

# SPINAL MENINGIOMAS: ILLUSTRATION AND EVALUATION OF OWN DATA AND REVIEW OF THE LITERATURE

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**Abstract:** Meningiomas are among the commonest primary extramedullary spinal neoplasms (Broager, 1953; Lombardi and Passerini, 1961) and they are most frequently encountered in the dorsal region (Sloof, Kernohan, and McCarty, 1964).

The most common histological entities among the spinal intradural extramedullary tumors are nerve sheath tumors (neurinomas) followed by meningiomas. The spinal group of meningiomas constitute approximately for 7.5–12.7 % of all meningiomas. More than 60 % of all spinal meningiomas are located in the thoracic spine. 86–95 % of the tumors are found intradurally.

Although the frequency of meningioma among primary spinal cord tumors is 25 %. multiple spinal cord meningiomas are extremely rare. Multiple spinal cord meningiomas have not been reported in the spinal cord meningioma series of A. Davis (45 patients), K. Katz (44 patients), Lombardi and Pasarini (71 patients) and by Haft and Shenkin (367 patients).<sup>(1-3,5,7,9).</sup>

Walter J. Levy and his colleagues and F. Carta et al. have reported respectively two multiple spinal cord meningiomas among patients who had spinal cord meningiomas. Single cases of multiple spinal meningiomas have been described by Rand, Rath et al. and Di Rocco et al. Only Rath and his colleagues reported multiple meningiomas, which were intradural thoracic and extradural cervical.<sup>(2, 4, 8,10, 14, 19, 22, 34, 45, 48, 55, 60-61)</sup>

Meningiomas usually arise within the dura; in some cases they can extend beyond it, but are rarely extradural in toto (Rasmussen, Kernohan, and Adson, 1940). When, however, this does occur; spinal meningiomas share with other predominantly extradural neoplasms, notably neurofibromas, a tendency to extend through adjacent intervertebral foramina into the thorax (Heuer, 1929; Naffziger and Brown, 1933). According to Bull (1953) and Gautier-Smith (1967), bony radiological changes are rare with meningiomas but common with neurofibromas, thus providing a valuable criterion for their differential diagnosis. The occurrence of multiple meningiomas in different neuraxial compartments is rather rare.<sup>(12-16, 19, 33, 45, 58-61).</sup>

Comparatively similar to cranial meningiomas, the main risk factors for the spinal group are ionizing radiation, genetic predisposition, and female gender.<sup>(4-9, 11-18, 22, 60)</sup>

CT scan without and with contrast and MRI scans, including T1 and T2-weighted images, with and without contrast are the usual diagnostic methods. They show spheric contrast-enhancing structures with extra- and intradural, intra- and extra-medullary localizations. The tumor matrix is typically in a lateral position. Sometimes it is difficult to differentiate meningiomas from neurinomas in the rare cases when meningiomas grow intra- and extradurally (dumbbell tumors). Distinct calcifications, which can be recognized in computed tomography, suggest a meningioma. Larger cystic areas rather indicate a neurinoma.<sup>(10, 19-23, 25-33, 56, 58-61)</sup>

The aimed treatment of spinal meningiomas is usually the total surgical excision of the tumor with assistance of neuronavigation, using of intraoperative ultrasound (for ideal tumor localization) and intraoperative neuromonitoring (for functional observation and preservation).

For the surgical intervention, a dorsal approach is preferred, except for few cases, in which sometimes lateral extension is indicated by partial resection of the facet joint or the head of rib in the region of the thoracic spine.

Postoperatively, the majority of patients have satisfactory outcomes. Advances in radiologic and surgical techniques such as computed tomography (CT), magnetic resonance imaging (MRI), intraoperative ultrasound, evoked responses, ultrasonic aspirators and lasers have brought about better clinical results. However, the meningioma may re-occur, especially as a result of incomplete resection. The goal of surgical treatment must be total resection, if possible. Total resection of a spinal meningioma is usually possible, but if the tumor is ventral to the cord and calcified, surgery becomes hazardous and may damage the cord. The operative and long-term mortality rates of spinal meningioma have decreased recently. <sup>(24, 34-44, 57, 59, 61)</sup>

The purpose of this article is an update of this disease based on a literature review and the own experience with case illustration. Additionally, the most recent own consecutive case series is collected and presented between 2008 and 2012.

**Keywords:** Spinal meningioma, spinal tumor, psammomatous bodies, dumbbell tumor.

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## I. INTRODUCTION

Meningiomas of the spinal axis have been identified from C1 to as distal as the sacrum. Their clinical presentation varies greatly based on their location.

Meningiomas, which are situated in the atlanto-axial region may present similarly to some meningiomas of the craniocervical junction, while some of the more distal spinal axis meningiomas are discovered as a result of chronic back pain. Surgical resection remains the mainstay of treatment, although advancements in radiosurgery have led to increased utilization as a primary or adjuvant therapy.

According to their anatomical location, spinal tumors are classified as extradural and intradural. Intradural tumors could be intramedullary or extramedullary.

Primary extradural tumors: Relatively uncommon, accounting for 19% in the series of Elseberg. Neurinomas, meningiomas and lipomas occur in the order of frequency. Rarely a secondary malignant tumour without involvement of bone can occur. Extradural cysts (congenital or acquired), non-specific granulomas and hydatid cysts, are the other extradural lesions.

Intramedullary tumors (14% in the series of Elseberg): Most of the intramedullary tumors are malignant and belong to glioma group. Astrocytomas and ependymomas are the common lesions in the compartment.

Intramedullary tumors may become partly extramedullary when they break through the confines of the cord. Epidermoid, dermoid, angiomas, AV malformations and hemangioblastomas are such lesions.

Intradural extramedullary tumors: In the spine, these are the commonest intradural tumors (67% in the Elseberg series). They are common in middle age group and most of them are benign in nature, most of them being nerve sheath tumors (neurinomas) and meningiomas. The early diagnosis and surgical removal and relieving pressure on the cord along with an intensive rehabilitation give excellent results with best outcome.

Because of presence of filum terminale, intradural extramedullary ependymomas can occur in the cauda equine region.

While the diagnosis of cord compression may be obvious in few cases, it is often delayed because of the variations in the presenting symptoms which may simulate other disease and the variability of the progress of the symptoms. Pesna et al found a median delay in diagnosis of 24 months (range 3 days to 24 years) amongst 57 patients, referred to them between 1978 and 1988. The symptoms and signs include those produced by the involvement of the nerve roots anterior and posterior, the cord segments and the long tracts, the motor, sensory, autonomic and other tracts. Some other signs related and pertaining to the spinal column may also become apparent.

Spinal meningiomas are tumors originating from arachnoid cap cells most commonly situated in the intradural extramedullary region [1, 2]. They represent a high proportion of all spinal cord tumors. Spinal meningiomas tend to predominate in the thoracic region, although they are described in the cervical, lumbar, and rarely the sacral area [3-5]. They pose varying surgical challenges based on their regional location, as well as their anterior/posterior orientation to the spinal cord and cauda equina.

The various locations of spinal axis meningiomas, as well as the different surgical approaches, and adjuvant therapies should be discussed in this study.

Approximately 2/3 of all intraspinal neoplasms are intradural extramedullar tumors. Among those, neurinomas followed by meningiomas are the most common histologic entities [1, 14, 20-34]. Spinal meningiomas occur less frequently than intracranial meningiomas. They constitute only for 7.5–12.7 % of all meningiomas [2, 4, 7-9, 13-19, 21-23, 26, 28, 30, 33, 35, 38-44, 47, 49, 51, 53, 57, 58-61].

Comparable to intracranial meningiomas, their incidence is 2–3 times higher in women than in men (Diagram 1). These lesions are a typical disease of the middle or older age [1-7, 9-14, 45-61].

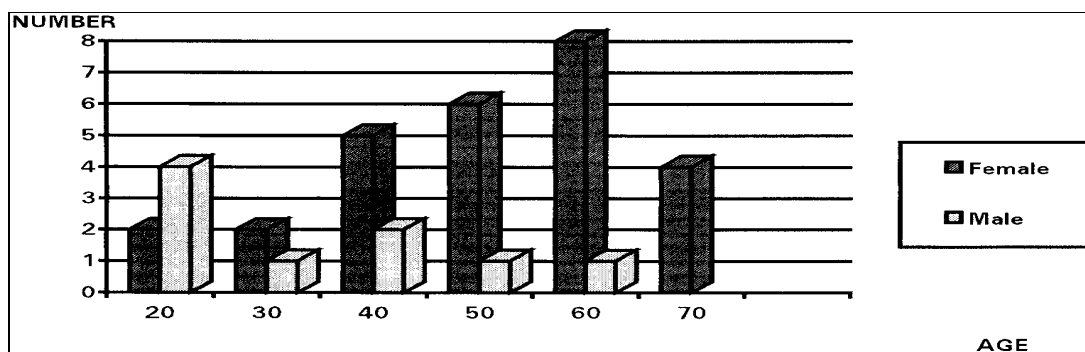


Diagram 1: Age and sex distribution of intraspinal tumors

Angiography also plays a critical role in surgical planning and may be utilized for preoperative embolization of hypervascular meningiomas. [3, 5, 7-12, 14-16, 22-29, 31, 33-36, 40, 42, 45-56, 58, 60].

## II. EPIDEMIOLOGY

Intradural extramedullary spinal cord tumors account for approximately two-thirds of all spinal cord tumors in adults [6].

Meningiomas, neurofibromas, and schwannomas are the most common type of tumor in this type of location [7,8]. Meningiomas represent about 40% of these tumors [6-22, 24, 44, 49-60]. The vast majority of spinal cord meningiomas are located in the thoracic region (Diagram 2 and Figures 1, 2, 3 and 4). [3-5, 9, 11-16, 18, 21-25, 29-33, 49, 51, 56, 58].

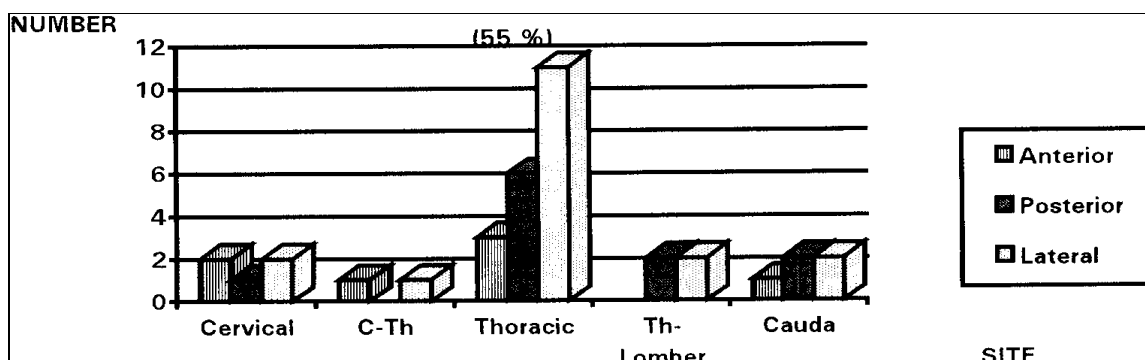


Diagram 2: The distribution of tumors.

Overall, spinal meningiomas account for about 7.5-12.7% of all meningiomas, occurring with less frequency than their intracranial counterpart. Most spinal cord meningiomas are located laterally. They typically arise from arachnoid cap cells

embedded in the dural root sleeve [6]. They can also arise from pial or dural fibroblasts. This may be accounted for by their mesodermal origin (13-18, 21-26, 29-33, 46-51, 53-58, 60-61).

This predominance of thoracic located meningiomas is seen only in females [10]. Spinal meningiomas occur about 2.5 times more often in females than males; with approximately 75-85% arising in women. The female preponderance is thought to arise from sex hormones or other receptor types common to women (10, 13, 15, 17, 19-23, 46, 49, 58-61).



**Fig. 1** (a) sagittal T1-weighted MRI showing homogeneous contrast uptake and broad contact to the ventral dura. (b) Sagittal CISS sequence and (c) axial CISS sequence revealing clear demarcation between tumor and myelon

Some authors believe that progesterone and estrogen receptors actually have opposing prognostic indications in regards to meningiomas. Pravdenkova et al. observed that the expression of progesterone receptors alone in meningiomas, signifies a more favorable and biological outcome [7-9, 11, 15-19, 24-28, 49, 55, 57-59]. They also found that either a lack of estrogen and progesterone receptors, or the presence of estrogen receptors in meningiomas, correlated with a more aggressive clinical behavior, progression, and recurrence. Hsu and Hedley Whyte also found that the presence of progesterone receptors, even in a small subgroup of tumor cells, indicated a more favorable prognostic value for meningiomas (8, 10, 12, 18, 20, 22, 26, 29, 31, 36-40).

An interesting epidemiological feature of spinal meningiomas, is that not only they are more common in women, but there is a sharp rise in postmenopausal females [9]. It has been postulated that these tumors are so grossly over-represented in postmenopausal females as a result of an association with osteoporosis. Previous authors have made suppositions that meningeal stretching or spinal meningeal trauma from direct contact with bone fragments of osteoporotic fractures, may in fact lead to a meningeal reparative proliferative process leading to tumor formation (5, 9, 11, 15, 19, 33, 40, 43-44, 50-53, 57, 59, 60).

Meningiomas may also be found in the region of the atlas and axis. The incidence of cervical meningiomas is about 14-27% [10]. They have a similar clinical presentation as foramen magnum meningiomas. The reason for this is that many cervicomedullary junction meningiomas expand to the high cervical region, just as many high cervical meningiomas expand rostrally through the foramen magnum [6, 8, 10-11, 14-18, 20, 23, 25, 28-30, 40]. Surgical treatment of meningiomas located in the high cervical region must be done with care to minimize bony removal in order to maintain mechanical stability [6, 8, 11].

The majority of spinal meningiomas are intradural, however a small percentage can be extradural, or both intradural and extradural. Although extremely rare, intradural, intramedullary spinal meningiomas have been described [13-16]. The incidence of lumbar spinal meningiomas is about 2-14% [10]. Sacral meningiomas have been described, although extremely rare (11, 17, 21, 29-33, 35, 37, 39, 41, 43-46, 49-51, 53-58).

### III. RADIOLOGICAL DIAGNOSIS

Cranial CT scan without and with contrast is very helpful, but MRI is the imaging modality of choice since it often delineates the characteristic dural origin of meningiomas. They are typically isointense or hypointense to gray matter on T1, and isointense or hyperintense on T2. Intraspinous meningiomas radiographically display avid homogenous

enhancement with contrast (Figure 1) <sup>[19]</sup>. Enhancement of the adjacent dura, or dural tail, is also characteristic of a meningioma <sup>[20]</sup>. CT scan will often demonstrate a calcified lesion in the spinal canal <sup>[19]</sup>. Radiographic evidence of a calcified spinal meningioma on plain x-rays is exceedingly rare, but some reports have quoted such a finding <sup>[21]</sup>. The dural attachment of spinal meningiomas is typically quite broad. However, similar to cranial meningiomas, dural tail in the MRI can be observed frequently. <sup>(18-24, 33-39, 41-46, 51-55, 57-61)</sup>

En plaque types of meningiomas are rare. However, partial circumferential calcification of the dura should raise the suspicion of this uncommon subtype of meningioma <sup>[22]</sup>. More commonly, spinal meningiomas are grossly smooth and fibrous, or fleshy with friable features [23]. There is usually a very well-defined epidural space in the spine. This is likely the main reason bony involvement is so rare in spinal meningiomas, as opposed to intracranial meningiomas. Also, unlike their intracranial counterparts, spinal meningiomas do not penetrate the pia. One reason this may be the case is that the manifestation of spinal cord compression and myelopathy are typically noticed by the patient “earlier” than the average intracranial meningioma. Therefore, surgical resection may be undertaken before penetration of the pial layer occurs <sup>[24, 27, 29, 33, 40]</sup>. This particular trait of pial preservation is also postulated in part by what has been described as an “intermediate leptomeningeal layer” <sup>[24]</sup>. This works in favor of a more complete and simplified surgical resection.



**Fig. 2** (a) T1-weighted MRI showing a round, well-demarcated tumor with intense contrast uptake. (b) Sagittal CISS sequences imaging the compression of the myelon by the well-confined tumor. (c) Contrast-enhanced, axial T1 images displaying the typical dumbbell shape

#### IV. CLINICAL PRESENTATION

Clinically, intraspinal meningiomas may have varying presentations depending on where they are located along the spinal axis.

Cranio-cervical junction and high cervical junction meningiomas typically present with some degree of myelopathic features and suboccipital pain. They may also cause atrophy of intrinsic hand muscles. Spinal tumors in general may cause a dull ache, and may also cause radicular symptoms if a nerve root is involved <sup>(12, 25, 38)</sup>.

**Symptoms:** At the beginning of the disease, mostly sensation disorders, a discrete spasticity of extremities, and gait disturbance are observed. Due to the slow growth tendency of these tumors, their symptoms remain often untypical for a long period of time. Because of the higher patient age (> 50 a) the altered gait pattern is often misinterpreted as ordinary joint pain. Due to these non-characteristic clues the correct diagnosis is often significantly delayed, especially in the most frequent location of the thoracic spine. Diagnosis remains unclear until the typical vesical and rectal disorders and progressive paraparesis emerge. With the help of magnetic resonance imaging spinal meningiomas are diagnosed earlier than several years ago.

Hydrocephalus is a rare sequelae of spinal meningiomas at any level, but is most common with meningiomas in the cervical region [26]. This is likely from increased protein production by the tumor, causing obstruction of CSF flow and absorption [26]. Corticospinal tracts are particularly vulnerable with thoracic tumors such as meningiomas. A clinical picture of thoracic myelopathy may ensue, displaying as long-tract signs. A foot drop may be an initial finding, with weakness of the tibialis anterior and extensor hallucis longus. Dorsally located midline thoracic meningiomas may cause a

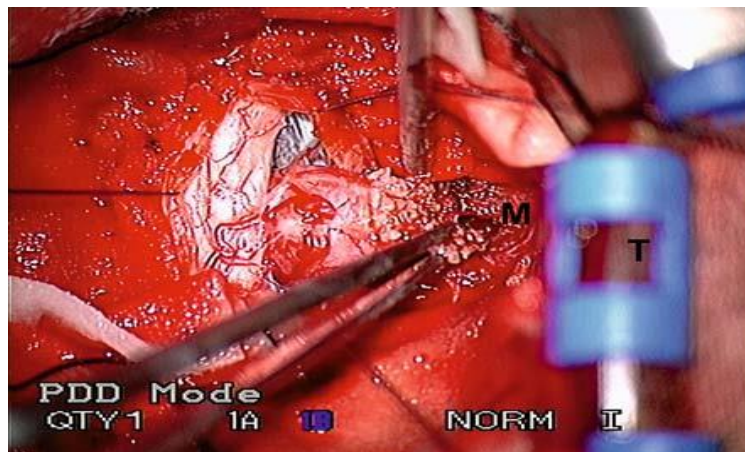


sensory ataxia from mass effect upon the posterior columns [6]. Bowel and bladder involvement may be a late manifestation. A common complaint of patients with extramedullary meningiomas is pain with recumbancy [3, 6, 9, 14, 18, 29-33, 35-37, 39-43].

## V. LOCALIZATION

More than 2/3 of all spinal meningiomas are located in the thoracic spine (67–84 %), followed by 14–27 % in the cervical spine and 2–14 % in the lumbar spine. Typically, they are found purely intradurally in 86–95 %. Only 5–14 % have an additional extradural part [3–7].

On rare occasions, spinal meningiomas occur completely extradurally (3–9 %). In the latter, in 2 locations they are predominantly dumbbell tumors (Figure 3) causing an enlargement of the intervertebral foramen. (3, 5, 9, 12, 23, 26, 28-33, 38, 40, 42, 44, 49, 51, 53, 56, 58, 61)



**Figure 3: Dumbbell schwannoma in the cervical spine.**

## VI. RISK FACTORS

Most publications were focussing on risk factors for the development of meningiomas in general [8]. Only a few papers refer directly to spinal meningiomas.

Ionizing Radiation Hiroshima and Nagasaki survivors showed an elevated risk of developing intracranial meningiomas. Their risk depended on their vicinity to the epicentre of the nuclear explosion [9–11]. Several US studies reported a significant correlation between X-ray dosage prior to the 20th year of life with the risk of meningioma development [12–14].

Also acute lymphoblastic leukaemia (ALL) patients showed an elevated risk of meningioma development after a latency of decades [15–20]. The latter lesions are more frequently multifocal, atypical, or malign [21, 22].

It is unclear whether this risk is caused by irradiation of the whole neuroaxis alone or whether additional factors such as chemotherapy are causative.

## VII. GENETICS

Changes or complete or partial loss of chromosome 22 may play a role in the development of meningiomas. Other changes in the gene loci are also associated with carcinogenesis and could play a role in the development of spinal meningiomas [23]. Contradictorily, Ketter et al documented a series of 23 spinal meningiomas, all of which showed a regular set of chromosomes or a monosomy 22 [13, 24, 32, 35-39].

### *Associations with neurofibromatosis Type 2 (NF2):*

The neurofibromin 2 gene (NF2) is located at the 12.2 region on chromosome 22. Additionally, neurofibromatosis type 2 with mutation of chromosome 22Q12 is an autosomally recessive, hereditary disease with elevated risk of developing meningiomas or schwannomas [23, 25-29, 31-33].

Loss of heterozygosity of the NF2 gene is the most common genetic abnormality found in meningiomas, and is responsible for 60% of sporadic as well as the majority of NF2 associated meningiomas. The NF2 gene codes for the protein Merlin. The function of Merlin is not well understood, however, it is thought to play a role in the 2 hit model of

tumor inactivation. Up to 60% of meningiomas have an associated NF2 gene mutation, which may serve as the initiating event of tumorigenesis, particularly those demonstrating a mesenchymal phenotype<sup>(16, 25, 27, 29)</sup>.

In a recent publication, changes in the gene SMARCE1 could be identified in relation to an increased incidence of familial spinal meningiomas<sup>(24, 26, 29)</sup>.

#### **Monosomy of 1p chromosome and loss of Alkaline Phosphatase (ALPL) function:**

The complete activity loss of ALPL function and the immunologically detected loss of ALPL protein in areas of meningiomas with monosomy 1p indicate a cytogenetically undetectable inactivation of the homologous allele.

Müller P et al. (1999) described deletion of chromosome 1p and loss of expression of ALPL, which indicates progression of meningiomas. The apparently homozygous loss of ALPL expression supports the notion that ALPL is a candidate tumor suppressor gene in meningiomas.

#### **Gender:**

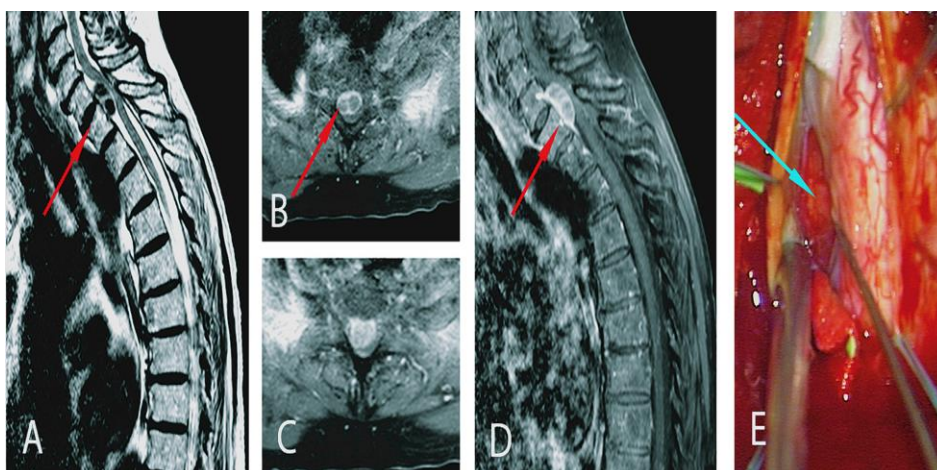
Women have a 2–3 times higher incidence of meningioma development. Additionally, the gender-related risk is slightly higher in women who take contraceptives or receive hormone replacement therapy<sup>[27–29, 33, 35, 39, 44, 50, 56, 58, 60–61]</sup>. The coincidence of breast carcinoma and meningiomas has been observed for many years<sup>[30]</sup>. It may be due to a joint risk profile (age, genetics, environmental factors in interaction)<sup>(14–29, 31, 32, 34–42)</sup>.

## VIII. MANAGEMENT

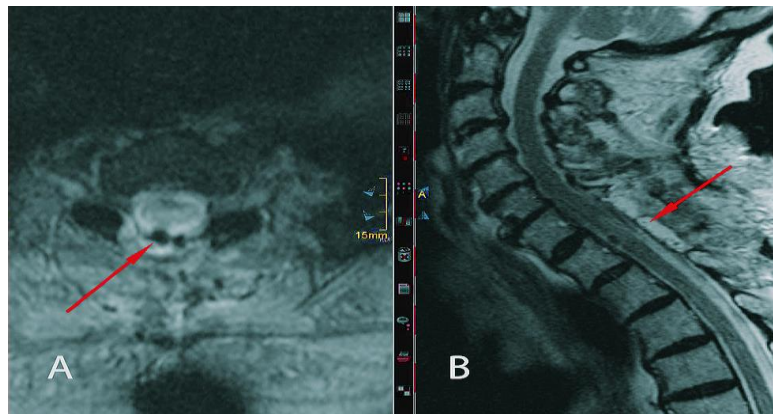
#### **Diagnosis and Operative Planning:**

The methods of choice in the diagnostics are CT scan without and with contrast and the magnetic resonance imaging (MRI) scans, including T1- and T2-weighted images, with and without contrast agent (Figure 2 and 3). They show spheric contrast-enhancing tumors with their intradural and extramedullar localization<sup>[31, 33, 35, 38, 41, 44]</sup>. The tumor matrix is in a lateral position in most of spinal meningiomas, more often dorsolateral than ventrolateral. Extensive growth and infiltration of the pia are significantly less frequently observed than in intracranial meningiomas.

It is sometimes difficult to differentiate meningiomas from neurinomas in the rare cases when meningiomas grow intra- and extradurally (dumbbell tumors). Distinct calcifications, which can only be recognized in computed tomography (CT), suggest a meningioma. Larger cystic areas rather indicate a neurinoma (Figures 4 and 5).



**Figure 4 (A-E): Pre- and intraoperative findings: (a) T2-weighted native sagittal MR image: the tumor can be nicely delineated (red arrow). (b, c) Axial T1-weighted contrast enhanced sequence: inhomogeneous contrast enhancement is due to intratumoral calcifications (red arrow). (d) Sagittal contrast-enhanced T1-weighted image: again, the calcifications can be seen as contrast agent-free areas within the tumor (red arrow). (e) Intraoperative findings: the spinal cord is carefully mobilized under IOM. The tumor (blue arrow) can be seen at the left ventral dura.**



**Figure 5: Postoperative MR scans: (a) T1-weighted contrast-enhanced MR images: the spinal cord (red arrow) is in normal location. No tumor remnants are visible. (b) T2-weighted native MR scan: the surgical dorsal approach and complete tumor removal are documented.**

Cystic changes are very rare in spinal meningiomas in contrast to calcifications. The latter may influence the surgical approach especially in ventrally positioned tumors. For this reason, It's believed, that it makes sense to perform a CT scan in ventrally positioned tumors to estimate the extent of calcifications prior to surgery. In central calcified tumors that are completely covered by the spinal cord total removal via a dorsal or dorsolateral approach is very difficult and may only be carried out at an elevated neurological risk.

#### ***Surgical Technique:***

The dorsal approach is preferred except for a few cases, sometimes laterally extended by partial resection of the vertebral joint or the head of rib in the area of the thoracic spine [2, 5, 33]. The rare ventral approach is discussed in the literature as an alternative mainly for purely ventral tumor locations completely covered by the spinal cord. The intention is to minimize manipulations at the spinal cord. The advantage of the ventral approach is a lower neurological risk for the spinal cord and better chance of radical removal in ventral tumors. The disadvantages are complications caused by the larger approach with vertebral body resection and the need for stabilization.

Traditionally, a posterior or posterolateral approach has been performed for cervical meningiomas, whether they are predominantly located dorsally, laterally, or even ventrally. Cervical meningiomas often can be a surgical challenge, as they are more often situated anterior to the spinal cord, as opposed to thoracic meningiomas which have a predilection for more posterior and lateral locations [23, 25, 27, 28-32, 35]. Klekamp and Samii reported on 130 spinal meningiomas, 27% of which were located in an anterior location [21, 23, 26, 28, 32, 36-44, 52, 54-57]. These were all resected via a posterior approach including a laminectomy with or without facet joint resection. No specific indications for cervical-based meningiomas was given in this series [14, 23]. Solero et al. reported on 174 spinal meningiomas and used a posterior approach for all posterior cervical meningiomas and all 7 of the reported anterior situated meningiomas [3, 5, 13, 19, 23, 25, 28-30, 34-37, 40]. In the majority of intraspinal meningiomas including those situated in the ventral cervical region, a posterior laminectomy gives adequate exposure [4, 6, 14, 16-20, 24, 26, 28, 30-34, 50].

#### ***Intraoperative Ultrasound:***

The application of intraoperative ultrasound [34] improves the precise localization and helps avoid unnecessarily large approaches with multi-level laminectomy. Depending on the longitudinal extension of the tumor, the removal of the vertebral arch was done usually only at one level. In many cases, surgery can be performed via an extended interlaminar approach with partial preservation of the vertebral arches. In younger patients, the vertebral arch should be preferably restored by laminoplasty, especially in the lumbar and cervical spine. The dura is opened paramedially vertically depending on the lateralization of the tumor. In ventrally positioned tumors, the incision can be enlarged laterally by way of a dural flap. Resection of the denticulate ligaments allows for a better view, especially of ventral tumors. Opening the dura directly at the tumor site instead of choosing the common median incision leaves the spinal cord mostly covered by the dura during surgery. This reduces the risk of spinal cord impingement in the slit-like area of the dural opening.



Comparable to intracranial meningiomas, spinal tumor debulking is performed using bipolar coagulation and the CUSA ultrasonic surgical aspirator (CUSA) to reach the tumor matrix at the dura as fast as possible. After interruption of its blood supply the tumor can be better mobilized. This alleviates separation of the remaining avascular tumor from the spinal cord. Postoperative MRI screening with contrast agent is mandatory within the first 72 hours (Figure 2) to reliably evaluate the extent of tumor removal.

Large ventrally located tumors may retract the spinal cord to some degree, and as a result, can still be approached posteriorly. To augment further anterior exposure through a posterior approach, subtle surgical maneuvers may be employed. Suturing of a noncritical dural root or a divided dentate ligament may be used as further retraction [6]. However, the posterior cervical approach still carries some degree of spinal cord manipulation to reach a ventrally-located meningioma. This may place the patient at higher risk of cord injury during the surgery. Dorsally-located cervical meningiomas can be approached via standard posterior cervical exposure. Although this is the approach of choice it may require stabilization if the patient becomes kyphotic over time [24, 28, 30]. The anterior cervical approach has also been used with success in resecting ventral cervical meningiomas [27, 29-32]. Advantages of an anterior approach to ventral cervical meningiomas include a large bony window for surgical exposure with direct visualization of the intradural pathology ventral to the spinal cord, extradural coagulation of anterior tumor feeders, and lack of necessity for cervical cord manipulation and retraction [27]. Lenelle et al. described an anterior cervical meningioma behind the C5 vertebral body that was resected with good neurological outcome, via a C5 corpectomy, while also taking the lower part of C4 and the upper part of C5 [32]. Sawa et al. reported a meningioma from the foramen magnum down to T2 [31]. This tumor was fully resected via a C3-T1 corpectomy also with a good neurological outcome. Banczerowski et al. described an anterior resection of C3 and C5 meningioma via corpectomies at those respective levels [29]. A significant disadvantage of the anterior cervical approach includes the necessity for a minimum of anterior cervical fixation. In cases of a three-level corpectomy, supplementary posterior fixation has been shown to increase biomechanical stability [33]. An additional disadvantage of an anterior approach is the potential for a CSF leak. Although this is certainly a risk with a posterior approach as well, with anterior approaches, there tends to be a fair amount of dead space between the dura and bone graft or cage [27]. In light of an anterior CSF leak, there is minimal to no tamponade effect due to this previously described dead space, and this can lead to post-operative complications related to the leak. This may be prevented by a watertight dural closure, application of fibrin glue, prophylactic lumbar drain, and filling the dead space with muscle fascia from a site such as the iliac crest [29, 34, 35]. Ventrally located cervical meningiomas with an eccentric component may be accessed via a hemilaminectomy and facetectomy. Ventral thoracic tumors may also be accessed via a costotransversectomy or lateral extracavitary approach [36]. Anterior transthoracic approaches are can be used for extradural thoracic lesions. This surgical technique is less frequently used for their intradural counterparts such as meningiomas. However, a transthoracic transvertebral approach has been described before for an anterior thoracic intradural calcified meningioma [37]. On rare occasion, a spinal meningioma may present as a dumbbell tumor, which has the appearance of a nerve sheath tumor. In order to resect this type of tumor, the nerve root at the level affected is typically sacrificed with minimal neurological sequelae [6, 38]. The exceedingly rare intramedullary meningioma has been treated using surgical cordectomy as the operative technique [39]. Although uncommon, meningiomas have been described in the lumbosacral spine [17, 40-43]. Overall, spinal meningiomas have some favorable features relative to intracranial meningiomas in regards to their resection. Typically, there is no bony involvement and major blood vessels are not usually in the path of surgical resection [24, 44]. In regards to the dural base of the meningioma, a surgeon may opt to coagulate the dura extensively, or may excise the involved dural component, with subsequent patch graft reconstruction. No significant differences in recurrence rates have been found with these two techniques [5]. Long-term recurrence rates after total resection range between 3 and 23% [4, 18, 38, 45]. Multiple spinal meningiomas are a relatively rare occurrence, especially outside the association with NF2 [44, 46-47]. Treatment options for multiple spinal meningiomas must be tailored to their locations and the symptoms they are causing, as well as the premorbid condition of the patient. Meningiomas causing significant myelopathic or radiculopathic features should be treated, as opposed to watchful waiting. Surgery should be the mainstay of treatment for symptomatic multiple spinal meningiomas, although radiation treatment may play a role as well [23, 25, 29, 33, 36, 38-41, 43, 45, 48, 51, 55, 57, 60-61].

#### ***Intraoperative Neuromonitoring:***

The use of intraoperative neurophysiological monitoring (IOM) is based on the observation that the function of neurological structures usually changes by a measurable value before it completely fails [33, 35, 37-39].

In contrast to laboratory tests, IOM is carried out in a “hostile environment” with permanent electric smog which may impair monitoring.

In the area of the spine, somatosensory evoked potentials (SEP) – monitoring ascending pathways –, dorsal column somatosensory system, and motor-evoked potential (MEP) – monitoring descending pathways and the corticospinal motor system – are applied and used in the monitoring setting. In general, tibial and median SEP are applied depending on the location of the pathology (Figure 6). Mostly surface values are derived but also subcortical components may be extrapolated.

	Cervical	Thoracic	Lumbosacral	Conus medullaris
SEP	++	++	(+)	++
EMG	++	-	++	++
MEP (ventral locations)	+	+	(+)	++
++, important; +, helpful; -, unnecessary				

**Figure 6: SEP, EMG and MEP in spinal meningiomas**

The use of SEPs alone for the monitoring of motor function is inadequate. For MEP, electrical transcranial stimulations are applied and EMGs are derived from the extremities <sup>(31, 34, 36-39, 42-48)</sup>.

Intra-operative monitoring (IOM) requires close cooperation of all groups involved in spinal tumor surgery <sup>[40]</sup>. Also anaesthesia has to be adapted to IOM, as temperature and blood pressure may influence the measured potentials. Additionally, “lost” electrodes may cause erroneously positive monitoring results and can therefore influence the surgical strategy and prolong operation time considerably.

**Radiation Therapy:**

The use of radiosurgery and adjunctive radiation therapy for the treatment of craniovertebral and spinal meningiomas is an evolving topic. Microdissection and resection of spinal meningiomas remains the gold standard for treatment <sup>[18, 49-52]</sup>. Adjunctive radiation therapy is considered primarily in cases of subtotal resection of recurrent meningiomas and/or when the risk of surgery is too high given the patient’s comorbidities or tumor location <sup>[49]</sup>. Frequently cited reasons for subtotal resection of meningiomas include an anterior tumor location (particularly in the thoracic area), recurrent tumors with arachnoid scarring and cord tethering, and en plaque and calcified meningiomas <sup>[18, 49-51,53]</sup>.

Because of the well documented success of primarily treating intracranial meningiomas with radiosurgery, it has been thought that radiation would be successful in spinal tumors of similar pathology <sup>[48, 51, 55, 59-60]</sup>. However, historically, primary radiosurgery treatment of spinal lesions has been problematic because conventional external beam radiotherapy relied on manual patient positioning and lacked the precision to deliver a large enough dose safely near the spinal cord <sup>[51,54]</sup>. Unlike intracranial systems that utilize the cranium as a rigidly immobilized reference to the lesion, early spinal systems used an invasive external frame system that was attached directly to the spine <sup>[51, 52, 54]</sup>. Subsequently, several frameless stereotactic radiosurgery techniques have been developed which utilize real time image tracking to achieve spatial accuracy of 1mm <sup>[51-54]</sup>. These systems allow high dose fractions of radiation to be delivered to the target tissue while sparing adjacent structures <sup>[51, 54]</sup>.

For conventional fractionated external beam therapy the dose tolerance for the spinal cord is thought to be 45 to 50 Gy delivered in 1.8 to 2 Gy fractions <sup>[50, 52]</sup>. Proton stereotactic radiosurgery has also been reported with similar promising results with the advantage of delivering significantly lower radiation to normal tissue <sup>[55]</sup>. Recent results of radiosurgery for spinal meningioma treatment have been promising.

The response rates seen with radiosurgery for spinal meningiomas appear to be comparable to responses rates for similar intracranial lesions <sup>[50, 53, 57, 59, 61]</sup>.

While most studies lack adequate long term follow-up necessary to establish the durability of the procedure, the majority of patients have been found to have stable or decreased tumor size on subsequent radiographic imaging<sup>[50]</sup>. In a study on foramen magnum meningiomas, Zenonos et al. found that 10 of 17 patients had symptomatic improvement with the remaining 7 patients remaining clinically stable at 6 months.

Dodd et al. showed in a study of 51 patients with benign intradural spinal lesions, that at one year follow-up, 61% of the patients had stable lesions and 39% had smaller lesions, with only 3 patients requiring surgery for worsening symptoms<sup>[43-46, 50-55, 60]</sup>. Gerszten et al. showed in a series of papers good long term radiographic control and symptomatic improvement for patients treated with both primary and adjunctive radiosurgery<sup>[51, 54]</sup>. Other authors have reported similar results<sup>[50, 53, 56, 58-61]</sup>. The technique seems to be effective in reversing neurologic deficits due to radiculopathies with better improvement for symptoms of pain and strength rather than sensory loss [50, 56]. Small asymptomatic tumors were more likely to have excellent regression responses and small symptomatic tumors were more likely to have symptomatic improvement<sup>[57]</sup>. Large tumors with significant brain stem compression and severe disability were not found to benefit from radiosurgery<sup>[57]</sup>. One of the major complications of radiation therapy is radiation induced myelopathy<sup>[50]</sup>. Delayed myelopathy has been of particular concern for radiosurgery of benign spinal lesions such as meningiomas. This concern stems from the fact that patient populations with these pathologically benign lesions have prolonged life expectancies compared to those with malignant lesions, and thus, myelopathy can be a burdensome side-effect to deal with on a chronic basis<sup>[51]</sup>. The reported incidence of radiation complications ranges from 0.2 to 5%<sup>[54]</sup>. Factors that have been associated with this complication include total dose and fraction size, length of irradiated spinal cord, and the duration of treatment<sup>[48, 50, 52, 57]</sup>.

Some authors have hypothesized that previous open surgery may predispose the spinal cord to subsequent radiation injury and have observed that the lower cervical levels and dorsal columns appear to be more susceptible<sup>[53]</sup>. Two types of radiation injury can occur. The first type is spinal cord edema, which may resolve spontaneously after completion of therapy, while the second is delayed myelopathy<sup>[54]</sup>. Delayed myelopathy typically occurs between 6 and 24 months<sup>[50, 52]</sup>. Diagnosis of cord damage can be seen on T2-weighted signal changes and have been reported to be treated with a combination of steroids, vitamin E and gabapentin<sup>[52]</sup>. Overall, radiation therapy for selected spinal meningiomas, is a plausible treatment option in the repertoire of the neurosurgeon.

### ***Spinal Angiography:***

Spinal angiography is often performed prior to surgical intervention in the case of lower thoracic meningiomas<sup>[58]</sup>. The artery of Adamkiewicz is the major provider of blood supply to the lumbar and sacral spinal cord. This is the largest known anterior segmental medullary artery. This artery is typically located between T8 and L3, and at T9 or T10 in about 50% of cases. It originates from the left side roughly 75% of the time<sup>[40, 45-50, 59-61]</sup>.

Unintentional interruption to this vessel during surgery could result in ischemia of the ventral horn of the spinal cord, in addition to the ventral commissura and the sympathetic centers of the intermediolateral region. This can subsequently manifest as distal paraplegia, pain and temperature sensory loss, and incontinence of bowel and bladder functions.

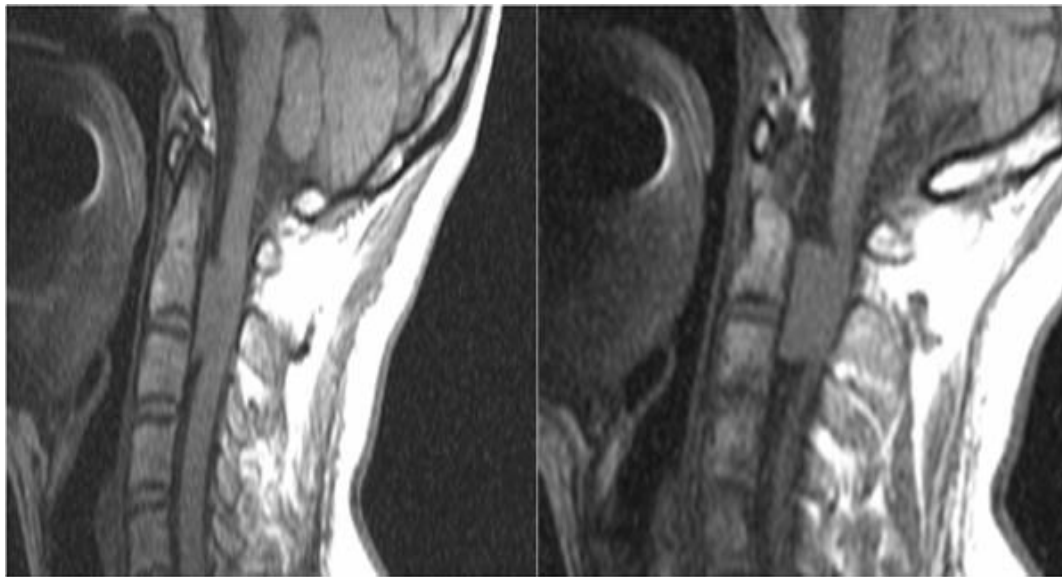
Pre-operative spinal angiography allows safe determination of the topography of the artery of Adamkiewicz. If a spinal meningioma is located at the same level as this artery, a contralateral surgical approach, or other selective surgical technique may be employed to avoid damaging this important vessel [59]. Spinal angiography can also be used pre-operatively as a means of embolization. Some spinal meningiomas are hypervascular, and resection of such lesions can become hazardous or may incite termination of the procedure due to excessive intra-operative hemorrhage<sup>[60]</sup>. The goal of this pre-surgical maneuver is to decrease the vascularity of the tumor, by embolizing artery-to-artery deep capillary tumor bed feeders<sup>[55, 59, 61]</sup>.

Pre-operative spinal meningioma embolization has the ability to facilitate surgical resection by reduction of intraoperative hemorrhage, shrinking the tumor, and giving the surgeon an unimpeded surgical field. These benefits can augment a more complete tumor resection.

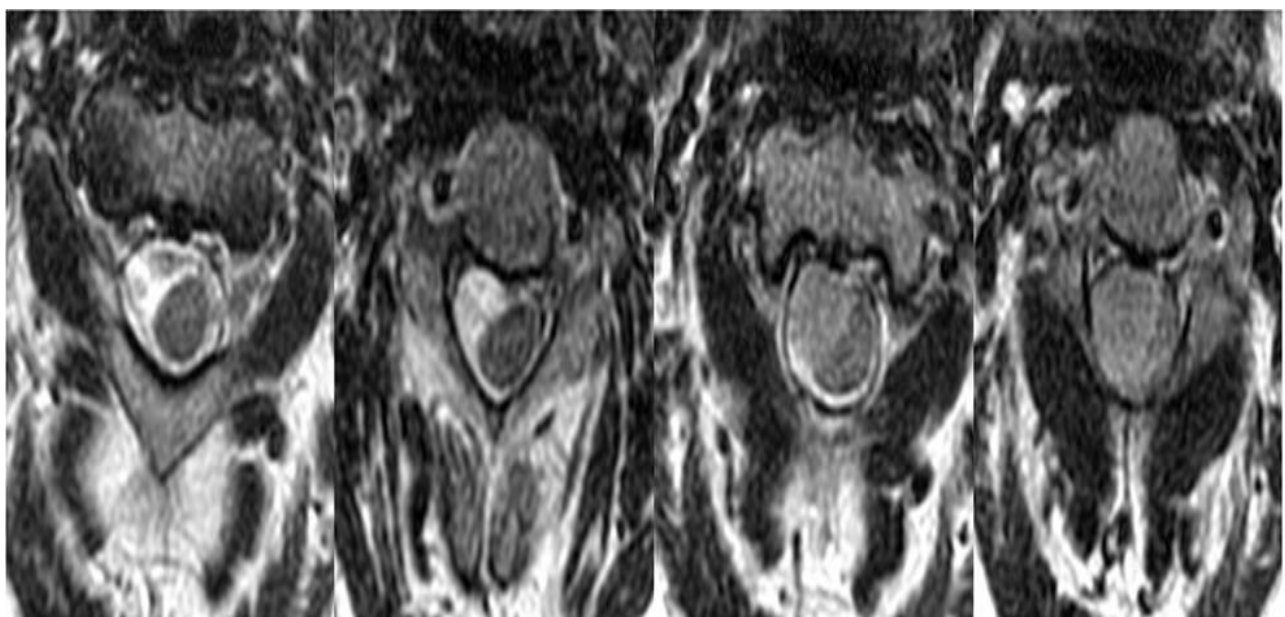
**IX. CASE ILLUSTRATION**

Female patient, 37 years old; otherwise there was no history of serious diseases. Onset of weakness and numbness was gradually in the right limbs. (Figures 7-10).

On presentation she was not able to walk, power grade in the right upper limb 3/5 and in both lower limbs 2/5. Postoperatively, the patient recovered very well. Diagnosis was cervical meningioma WHI grade I. Early neuro-rehabilitation was performed. After 6 months, the last follow up examination was done, in which the patient was able to walk without weakness in the upper limbs and with minimal weakness in the lower limbs (power grade 4+/5).

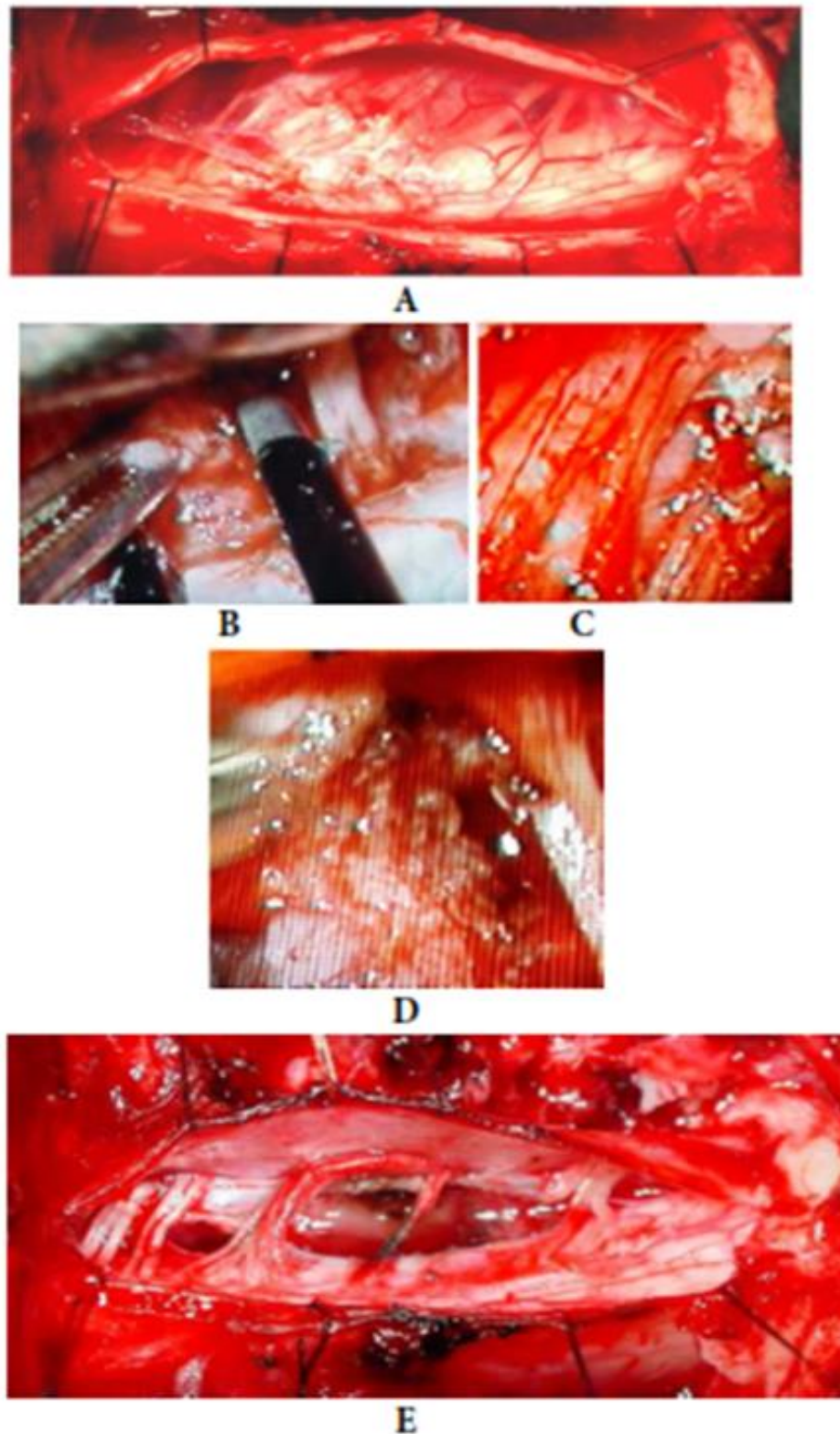


**Figure 7 - Preoperative MRI: sagittal T1w sequences without and with paramagnetic contrast media reveals the tislular nature and the homogenous enhancement of the tumor**



**Figure 8 - Preoperative MRI: Axial T2w sequences at the tumor level**





**Figure 9: Intraoperative photographs: A right antero-laterally placed, cervical meningioma, displacing the spinal cord laterally and anteriorly. Dura was opened, tacked laterally using 4-0 sutures and arachnoid was opened, dissected off the tumor and adjacent cord, B-D intraoperative photographs during tumor removal, E postoperative photography taken after removal of a cervical meningioma, without cutting right cervical roots.**



**Figure 10: Postoperative MRI: A sagittal T2 weighted image with no enhancement suggestive for local spinal cord oedema; B sagittal T1 image: a local postoperative edema.**

#### X. THE OWN CASE SERIES

The aim of this study was to analyze the clinical presentation, imageology, resectability, to know the incidence of different types of tumors in intradural extramedullary compartment and to study the surgical outcome. (Tables 1-3)

##### **METHODS:**

This was a retrospective study of 80 patients of intradural extramedullary tumors that were treated at the unit of neurosurgery, department of surgery, Barmherzige Brüder Hospital from February 2008 to October 2012 with remarkable female prevalence (57 female, 23 male; 2.5: 1). Age of patients was between 16 and 79 years. Mean age was 38.5 years.

The youngest patient in this series was a 19 year old woman and the oldest operated patient was a 79 years old female patient.

The majority of the patients were in 30-40 years age group: particularly there were 31 patients (38.7 %).

##### **RESULTS:**

The number of neurinomas / schwannomas (36 %) was nearly equivalent to meningiomas (35%), followed by metastases (14 %) and other entities (15 %) such as ependymoma, cavernoma, and arachnoid cysts.

Meningiomas were more common in females (72 %) whereas neurofibroma has shown male predominance (74.%).

##### **CLINICAL EVALUATION:**

The clinical presentation, imageology, resectability, histopathology, surgical outcome were studied.

The patients were thoroughly evaluated to assess the symptoms and signs with particular stress on motor and sensory deficits.

**Imageological evaluation:**

The patients were investigated with plain spinal radiography to note the change in spines and their joints.

All the patients were evaluated with MRI of the whole neuro axis for a better delineation of the lesion and to know its relations with cord and to know intrinsic cord changes due to tumor compression. Additional aim was to detect any other lesions in the whole neuronal axis (cranial and spinal).

**Surgical treatment:**

The cases were totally treated surgically by posterior or posterolateral approaches. Outcome and complications were evaluated.

**Follow up:**

All the patients were followed up regularly and the results were analysed. Ambulatory status was classified on admission by using Nurick – grading scheme. (See information board 1)

<b>Nurick – grading scheme:</b>
1. Normal walk.
2. Slight difficulty in walking.
3. Disability limiting normal walk.
4. Required assistance in walk.
5. Bed ridden.

**Information board 1: Nurick – grading scheme**

**Table 1: Location of tumours compared with other studies.**

Study	Location in percentage		
	Cervical	Thoracic	Lumbar
Lavy et al 1982 <sup>8</sup>	17	75	7
Solero et al 1989 <sup>13</sup>	15	83	2
Roux et al 1996 <sup>11</sup>	18	80	2
King et al 1998 <sup>14</sup>	14	84	2
Kleokamp J et al 1999 <sup>15</sup>	27	67	6
N Gottfried et al 2003 <sup>12</sup>	16	76	8
Present study	18	64	18

**Table 2: Presenting symptoms compared with other studies.**

Study	Pain	Weak-ness	Sensory	Sphincter Dysfunction
Lavy at al 1982 <sup>8</sup>	77*	66	74	40
Solero et al 1989 <sup>13</sup>	53	92	61	50
Roux et al 1996 <sup>11</sup>	72	80	67	67
King et al 1998 <sup>14</sup>	23	-	28	61
Kleokamp J et al 1999 <sup>15</sup>	50	16	6	6
N Gottfried et al 2003 <sup>12</sup>	68	64	84	48
Present study	89	100	89	44

Study	Outcome percentage			Ambulatory percentage	
	Improved	Stable	Deteriorated	Pre-operative	Post-operative
Lavy et al 1982 <sup>8</sup>	83	17	0	70	76
Solero et al 1989 <sup>13</sup>	53	33	10	70	92
Roux et al 1996 <sup>11</sup>	83	13	2	67	94
King et al 1998 <sup>14</sup>	95	1	4	74	97
Kleokamp J et al 1999 <sup>15</sup>	-	-	-	74	94
N Gottfried et al 2003 <sup>12</sup>	92	0	8	68	96
Present study	78	22	0	66	100

**Table 3: Functional outcome after surgery for spinal meningiomas.**

**Evaluation of the results of the own data:**

The incidence of nerve sheath tumor reported was 36% and that of meningioma 35% of all intradural extramedullary tumors. Majority of the nerve sheath tumors were present in 3rd decade and majority of meningiomas were present in 2nd and 3rd decades with female’s preponderance. Thoracic spine was common site of occurrence which is corresponding with the literature reported by Mc Cormick and Ramamurthy et al. (Information board 2)

Grade	Definition
0	No deficit
1	Mild focal deficit not significantly affecting function of involved limb; mild spasticity or reflex abnormality; normal gait
2	Presence of sensorimotor deficit affecting function of involved limb; mild to moderate gait difficulty; severe pain or dysesthetic syndrome impairing patient’s quality of life; still functions and ambulates independently
3	More severe neurological deficit; requires cane/brace for ambulation; may or may not function independently
4	Severe deficit; major sensorimotor deficit of more than one limb; patient functionally fully dependent

**Information board 2: Mc Cormick Score**

**Spinal nerve sheath tumors:**

The benign nerve sheath tumors, neurinomas, were the commonest intraspinal tumor in the present series of 29 total cases (36%). The incidence of nerve sheath tumors was reported as 25% in the series reported by Levy et al and McCormick et al. Majority of the tumors were seen in middle age group from 35-55 years in this series. Levy et al found that males were affected more commonly than females, which is corresponding with this series.

Neurofibromas occur frequently in the thoracic region, the rest being almost equally distributed between the cervical and lumbosacral regions. In this series 64% of tumors were located in thoracic region, 18% in cervical, 18% in lumbar region which is 7:2:2 ratios. Majority of the tumors (80%) were located posteriorly or posterolaterally to the spinal cord,



ensuring a greater percentage of surgical success without complications. The majority tumors (80%) were intradural and 10-15% extended through the dural root sleeve as dumbbell tumors occupied both intra and extradural compartments. The incidence in this series corresponds with that 75% and 10-15% of McCormick series report.

The giant neurofibromas in this series were 18% as type 5 which corresponds with series reported by Ramamurthy et al who reported it to be 10.9%.

The mean duration of presenting symptoms in this series was of 2 years and 2 months. Symptoms were back pain (89%), tingling and numbness (89%), weakness of limbs (100%), sphincter dysfunction 44, bladder disturbances (27%). Hyper-reflexia and severe spasticity noted in 72% was documented on initial neurological examination. Radicular pain and sensory symptoms were corresponding with series of Levy et al. corresponding intermittently with higher incidence of weakness and bladder disturbance.

Ambulatory status was classified on admission by using Nurick grading scheme. Majority of the patients (91%) were ambulatory on admission and 72% exhibited Nurick grade 1 to 3 status and 27% of patients were disabled with grades 4 or 5. One patient was bed ridden. All the patients were investigated with radiography of spine. The radiological changes found were flattening of pedicle, loss of pedicle shadow and enlargement of inter vertebral foramen were observed in 30% of cases. All the patients were investigated with MRI spine, and it was investigation of choice in this series.

#### **Surgical outcome:**

Surgery was indicated in all patients in this series and complete excision of the lesion was achieved in 99% of cases. This corresponds with results of Levy et al and Lot G et al who in their study achieved complete excision of lesion in 98% of cases. The rate of nerve root preservation in this series was 98%, without any persisting deficits after sacrificing the nerve root in 2 cases. There was no mortality in this series. No postoperative permanent / persistent neurological deterioration was noted.

#### **Functional outcome:**

In the immediate post-operative period on day 1 improvement in spasticity was seen in 90% of cases, and 85% had complete pain relief on follow up. 80% of patients had normal sensation that had prior sensory loss and 50% of patients who had grade II motor deficits pre operatively improved to normal, 10% of pre-operative patients with grade III deficit improved to grade II on 6 months follow up.

Bladder function improved in 90% of patients on follow up for 6 months, and 9% patients with grade V pre operatively had improved to grade IV in 12 months follow up. No case of postoperative deterioration was noted. The functional outcome result in the present series corresponds with the series reported by Levy et al.

#### **Meningioma:**

The mean age of the patients in meningioma cases was 36 years. In a study by Mayo Clinic they found that 37.5% of their patients belonged to less than 50 years of age. In the present study, the majority of the patients of meningioma were female (72 %), which is more or less corresponding to 87.5% of Mayo Clinic study.

The presenting symptoms of meningioma cases in the present study were pain in 89%, sensory disturbance (89%); weakness (100%), bladder dysfunction in 44% was corresponding to Roux et al series with 72%, 61%, 80% and 37% respectively. Gait normality was seen in all 9 (100%) of all patients in this series with 3(33%) were non-ambulatory which is corresponding with 32% in a series by O.N. Gottfried et al. <sup>(09, 11-12, 14-18, 22-26, 30, 34, 36, 39, 41, 44-46, 48-50, 57).</sup>

Majority of tumors (89%) were located in thoracic region, which is corresponding with 80% in a series of Mayo Clinic, and 83% in a study by Roux et al. <sup>(09-18, 22-27, 34-39, 44, 47, 52, 55).</sup>

In the present study, the sensory changes, gait abnormality, weakness was seen in majority of cases due to late presentation and large tumors in comparison to the Mayo Clinic study. This can be attributed to a random distribution of population due to availability of facilities, socio economic status of patients.

Spinal meningiomas are predominantly histologically benign tumors of the meningothelial and psammomatous variety. Total resection is the primary objectives of treatment. The key to successful spinal meningioma resection is judicious planning of the surgical corridor. To access to the tumor bulk, but equally important, to the tumor margins were noted.

Excision of dural margin in contrast to cauterization was associated with a lower recurrence rate. In this series all patients underwent posterior laminectomy, in case the tumors were located anteriorly the laminectomy was extended laterally towards the articular process to provide sufficient exposure and avoid pressure on spinal cord. Operating microscope was used. The goal of surgery was to minimize displacement of the spinal cord by undertaking an appropriately wide exposure, making the tumor and its dural attachment accessible. After dural opening a plane was developed. The tumor was then internally debulked and was removed from its dural attachment. Dural graft was used in rare cases; only 5 patients.

Total excision was achieved in 95% of cases in comparison with other series of 93% by Roux et al and King et al with 99% and 99% in Gottfried et al.

Overall functional outcome was excellent in this series and no deterioration seen: 78% of the patients were improved; 22% were stable, respectively.

The results are comparable King et al study and Gottfried et al study. In this series pre-operative ambulatory patients were 66%, non ambulatory 33% compared to King et al 74%. Postoperatively in this series 100% became ambulatory in 2 months period which is corresponding with 97% in King et al 96% in Gottfried et al 94% in Roux et al series.

Bladder dysfunction was 44% pre operatively in this series exhibited normal function after surgery in a period of 1 month, which corresponds with 95% cases in King et al study.

Mortality was nil in this series. The series results were corresponding with 0% mortality of Roux et al and O.N. Gottfried et al series.

In present series, histopathology examinations of the intraspinal meningiomas had shown 44% meningothelial and 56% psammomatous meningiomas (Table 4).

Study	Meningothelial	Psammomatous
Roux et al <sup>11</sup>	44%	20%
Present study	44%	56%

**Table 4: Histopathology of intraspinal meningiomas**

**Postoperative Outcome:**

The great majority of patients have excellent postoperative outcomes. More than 90 % show clinical improvement with ameliorated gait pattern or even restart walking without assistance [2-5, 7, 33]. Possible reasons for rare clinical deterioration are manipulations of the spinal cord, considerably sudden extension, or ischemia due to a vascular lesion.

A vascular disorder may occur mainly in meningiomas at the thoracolumbar region in proximity to the arteria radicularis magna. In such cases, it makes sense to refrain from radical resection and leave small tumor parts adhering to the artery untouched.

Rare complications requiring revision surgery include epidural haematomas (2-5 %) and cerebral spinal fluid fistulas (< 1 %) (2, 4-6, 8).

In the present series, no single case of epidural haematoma or fistula was detected, but 2 cases of tumor recurrence.

**Tumor Recurrence:**

The recurrence rate in spinal meningiomas is significantly lower than in intracranial meningiomas. In the present study, 2 tumor recurrences were identified and operated, which were meningiomas.

In spinal meningiomas, as expected, total removal at first surgery is the key factor to avoid tumor recurrence. En plaque meningiomas, which often cannot be radically removed, show a significantly higher relapse rate. This is also the case in tumors with ventral matrix or in cases with severe calcifications of the tumor.

Due to higher rates of subtotal tumor removal close to the arteria radicularis magna the recurrence rate in this location is also higher.

Concerning the treatment of tumor recurrence, reoperation is the first-line treatment option. The role of radiotherapy is still controversially discussed<sup>[3, 7, 33]</sup>. However, it is a possible treatment option in recurring tumors.

During the years, in order to reach its significance, it will grow due to the development of new radio-therapeutic alternatives such as the proton beam radiation.

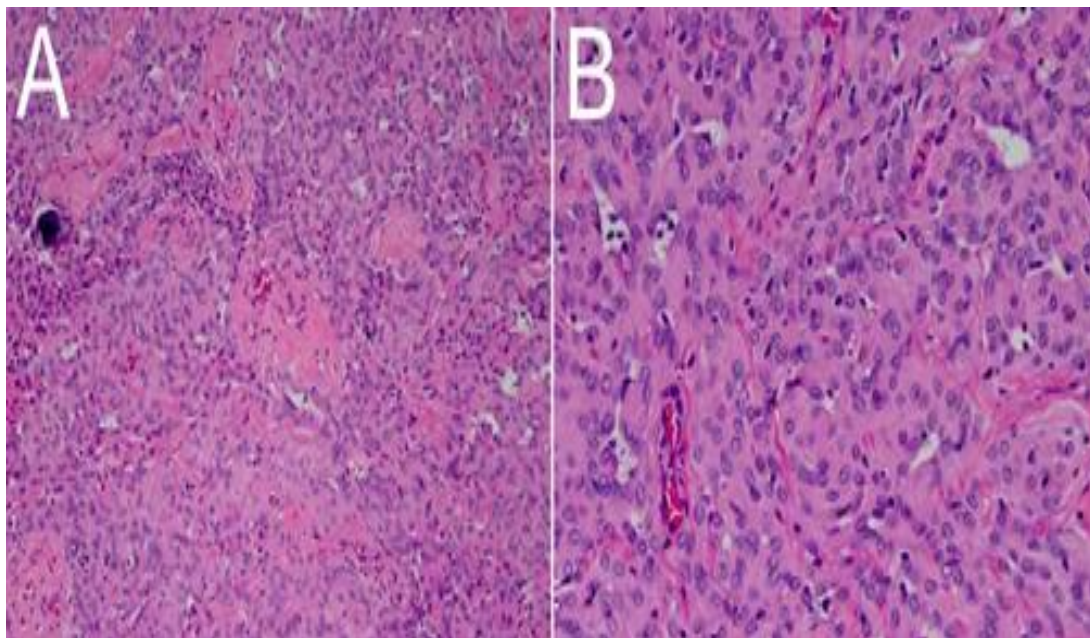
So far, indication of radiotherapy was only after recurrent surgery or in very old patients with an increased surgical risk profile. In two cases, a very tiny rim of tumor was remaining as it was tightly attached to the spinal cord. The two particular patients have undergone postoperatively a radiation treatment and were improved in the follow up examinations.

In cases of 3 subtotal tumors removal, it was the management with closely follow-up of the patients with special focus on further tumor growth (“wait and see”).

#### **Histopathology:**

Comparable to entities of intracranial meningiomas, the most common histological subtypes are meningotheiomatous, hypoplastic, transitional, and psammomatous tumors.

The first 2 types are predominant in spinal meningiomas. Interestingly, the histological type does not seem to influence prognosis. Compared to intracranial meningiomas, spinal tumors less frequently belong to WHO grades II and III<sup>[1-7]</sup>. Nevertheless, spinal meningioma represents an entity of its own. (Figures 11-13).



**Figure 11: 10X (A) and 20X (B) magnification. Hematoxylin and eosin stain shows monotonous cells with bland histology and poorly defined cell borders. Inconspicuous nucleoli are seen. The cells have round to oval nuclei and rare nuclear pseudoinclusions. Mitotic figures are not present. Whorls and a single calcification (Psammoma body) are present.**



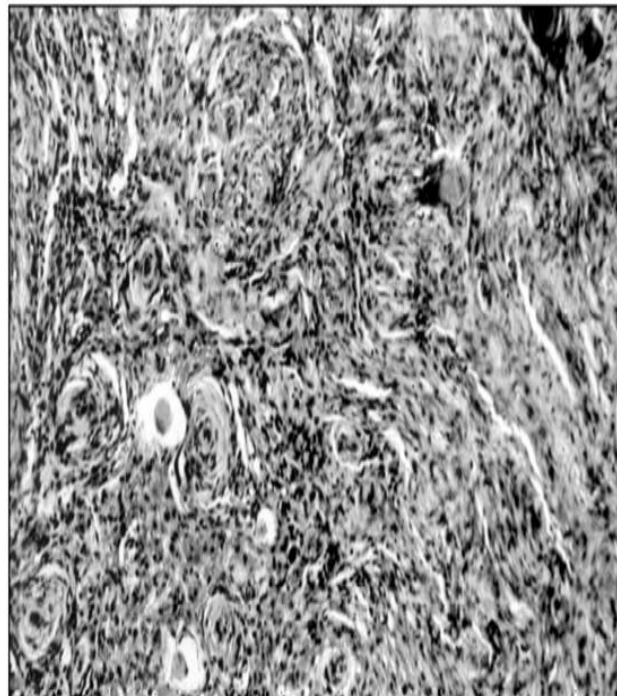


Figure 12: Characteristic pathological pattern of a psammomatous meningioma.

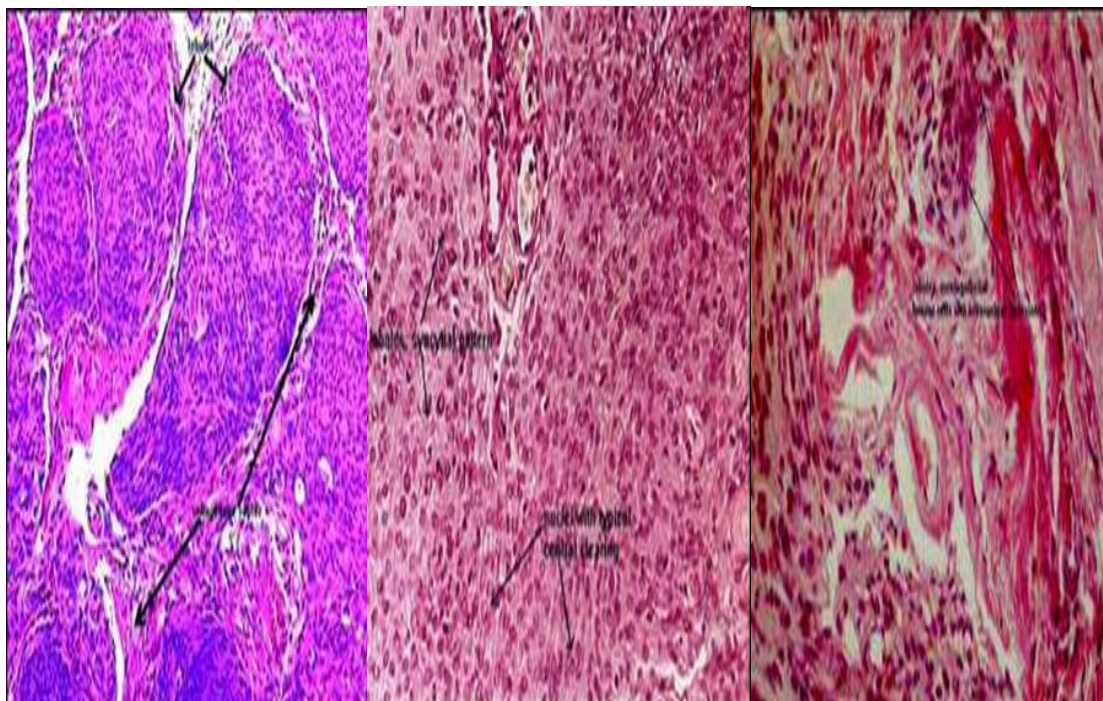


Figure 13: Meningothelial meningioma microscopy photographs: A.H&E staining, X 100: tumor cells form lobules surrounded by thin collagenous septae B.H&E staining, X 200: solid areas of cells with poorly defined cell membranes (syncytial appearance); like normal arachnoid, tumor cells are uniform, with oval or round nuclei with pale central clearing (slight tendency for the chromatin to be margined at the periphery) C.H&E staining, X 200: perilobular collagen and reticulin are variable; usually nodular vascular thickening are seen.



## XI. DISCUSSION

Spinal cord meningiomas represent a relatively high proportion of spinal cord tumors. Their location is most typically in the thoracic and cervical regions, however the less common lumbar and sacral meningiomas are described. Posterior or posterolateral approaches are most commonly employed for both ventral and dorsal spinal meningiomas, although anterior cervical and anterior transthoracic approaches are warranted for specific situations.

Nerve Sheath tumors and meningiomas are the most common in the group of intradural extramedullary spinal lesions and complete excision was possible in almost all cases of the presented series.

Surgery is always the therapy of choice in spinal tumors and spinal meningiomas, respectively. In the vast majority of patients, the operation results were associated with significant improvement of the preoperative manifestation of the neurological deficits (2, 4-6, 33, 35-39, 44, 49, 51-55).

Patient have usually a good benefit through the surgical intervention, in spite of poor pre-operative neurological status of the majority of the patients, as it was reported with similar results in this presented series and in other studies.

In rare tumors exclusively located ventrally or in close proximity to the arteria radicularis magna, the risk of complete removal has to be evaluated against the preservation of function on a case-by-case basis. In these patients, age plays an important role for the decision.

Radiation treatment can contribute to the management of some cases with spinal tumors. Indication of radiotherapy is mostly after recurrent surgery or in very old patients with an increased surgical risk profile. In some cases with subtotal excision of the tumor, the postoperative radiation treatment is helpful.

A dorsal approach is preferred whenever possible. Also, ventral tumors normally displace the spinal cord and thus create enough space for surgical manipulation using the dorsal or dorsolateral approach. In very rare cases of ventral tumors located exactly in the midline, the spinal cord may cover the tumor bilaterally. Only in these cases a ventral approach with vertebral body resection is necessary.

## XII. CONCLUSION

Approximately 2/3 of all intraspinal neoplasms are intradural extramedullar tumors. Among those, neurinomas followed by meningiomas are the most common histologic entities. Spinal meningiomas occur less frequently than intracranial meningiomas. They constitute only for 7.5–12.7 % of all meningiomas.

Prognosis of intraspinal-intradural extramedullary tumors is usually good, in spite of poor neurological status at the time of presentation.

Surgical resection remains the mainstay of treatment, although advancements in radiosurgery have led to increased utilization as a primary or adjuvant therapy. Angiography also plays a critical role in surgical planning and may be utilized for preoperative embolization of hypervascular meningiomas.

Adjuvant radiation treatment for spinal meningiomas is becoming an important treatment modality in cases where there is incomplete surgical resection. When hypervascular meningiomas are suspected based on standard imaging and angiography, pre-operative embolization can be utilized to aid the surgeon with a safer resection.

During the last 3 decades the prognosis of spinal meningiomas has improved for the following three reasons:

- Significantly earlier diagnosis because of magnetic resonance imaging and, consequently, better neurological status at the time of surgery.
- Reduction of surgical trauma and improvement of functional outcomes due to improved localization with the help of intraoperative ultrasound and the use of CUSA dissection and intraoperative neuromonitoring techniques.
- Avoidance of secondary defects (instabilities and postoperative deformities after years) with the help of laminoplasty when applying the dorsal approach and improved stabilizing techniques including spinal body replacement when applying the ventral approach.

### XIII. REFERENCES

- [1] Mawrin C, Perry A (2010) Pathological classification and molecular genetics of meningiomas. *J Neurooncol* 99: 379-391.
- [2] McLendon RE, Rosenblum MK, Bigner DD (2006) Russell and Rubinstein's Pathology of Tumors of the Nervous System. CRC Press.
- [3] Roux FX, Nataf F, Pinaudeau M, Borne G, Devaux B, et al. (1996) Intraspinal meningiomas: review of 54 cases with discussion of poor prognosis factors and modern therapeutic management. *Surg Neurol* 46: 458-463.
- [4] Levy WJ Jr, Bay J, Dohn D (1982) Spinal cord meningioma. *J Neurosurg* 57: 804-812.
- [5] Solero CL, Fornari M, Giombini S, Lasio G, Oliveri G, et al. (1989) Spinal meningiomas: review of 174 operated cases. *Neurosurgery* 25: 153-160.
- [6] Winn HR (2011) Youmans Neurological Surgery. Elsevier Health Sciences.
- [7] Bucci MN, Mc Gillicuddy JE, Taren JA, Hoff JT (1990) Management of anteriorly located C1-C2 neurofibromata. *Surg Neurol* 33: 15-18.
- [8] Hu HP, Huang QL (1992) Signal intensity correlation of MRI with pathological findings in spinal neurinomas. *Neuroradiology* 34: 98-102.
- [9] Preston-Martin S, Monroe K, Lee PJ, Bernstein L, Kelsey J, et al. (1995) Spinal meningiomas in women in Los Angeles County: investigation of an etiological hypothesis. *Cancer Epidemiol Biomarkers Prev* 4: 333-339.
- [10] Lee JH (2008) Meningiomas: Diagnosis, Treatment, and Outcome. Springer.
- [11] Pravdenkova S, Al-Mefty O, Sawyer J, Husain M (2006) Progesterone and estrogen receptors: opposing prognostic indicators in meningiomas. *J Neurosurg* 105: 163-173.
- [12] Hsu DW, Efid JT, Hedley-Whyte ET (1997) Progesterone and estrogen receptors in meningiomas: prognostic considerations. *J Neurosurg* 86: 1131-120.
- [13] Hafiz MG, Rahman MR, Yeamin MB (2013) Intradural intramedullary spinal cord meningioma in a seven years old female child. *Mymensingh Med J* 22: 180-185.
- [14] Moriuchi S, Nakagawa H, Yamada M, Kadota T (1996) Intramedullary spinal cord meningioma--a case report. *Neurol Med Chir (Tokyo)* 36: 888-892.
- [15] Sahni D, Harrop JS, Kalfas IH, Vaccaro AR, Weingarten D (2008) Exophytic intramedullary meningioma of the cervical spinal cord. *J Clin Neurosci* 15: 1176-1179.
- [16] Yuan D, Liu D, Yuan XR, Xi J, Ding XP (2013) Intramedullary thoracic spinal cord meningioma: a rare case report and review of the literature. *J Neurol Surg A Cent Eur Neurosurg* 74 Suppl 1: e136-139.
- [17] Teo M, Zrinzo L, King A, Aspoas AR, David KM (2010) Giant extradural sacral meningioma. *Acta Neurochir (Wien)* 152: 485-488.
- [18] Gezen F, Kahraman S, Canakci Z, Bedük A (2000) Review of 36 cases of spinal cord meningioma. *Spine (Phila Pa 1976)* 25: 727-731.
- [19] Weber DC, Lovblad KO, Rogers L (2010) New pathology classification, imagery techniques and prospective trials for meningiomas: the future looks bright. *Curr Opin Neurol* 23: 563-570.
- [20] Quekel LG, Versteeg CW (1995) The "dural tail sign" in MRI of spinal meningiomas. *J Comput Assist Tomogr* 19: 890-892.
- [21] Doita M, Harada T, Nishida K, Marui T, Kurosaka M, et al. (2001) Recurrent calcified spinal meningioma detected by plain radiograph. *Spine (Phila Pa 1976)* 26: E249-252.

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- [22] Gamache FW Jr, Wang JC, Deck M, Heise C (2001) Unusual appearance of an en plaque meningioma of the cervical spinal canal. A case report and literature review. *Spine (Phila Pa 1976)* 26: E87-89.
- [23] Klekamp J, Samii M (1999) Surgical results for spinal meningiomas. *Surg Neurol* 52: 552-562.
- [24] Salpietro FM, Alafaci C, Lucerna S, Iacopino DG, Tomasello F (1997) Do spinal meningiomas penetrate the pial layer? Correlation between magnetic resonance imaging and microsurgical findings and intracranial tumor interfaces. *Neurosurgery* 41: 254-257.
- [25] Stein BM, Leeds NE, Taveras JM, Pool JL (1963) Meningiomas of the Foramen Magnum. *J Neurosurg* 20: 740-751.
- [26] Feldmann E, Bromfield E, Navia B, Pasternak GW, Posner JB (1986) Hydrocephalic dementia and spinal cord tumor. Report of a case and review of the literature. *Arch Neurol* 43: 714-718.
- [27] Payer M (2005) The anterior approach to anterior cervical meningiomas: review illustrated by a case. *Acta Neurochir (Wien)* 147: 555-560.
- [28] Mayer M, Meier O, Auffarth A, Koller H (2013) Cervical laminectomy and instrumented lateral mass fusion: techniques, pearls and pitfalls. *Eur Spine J*.
- [29] Banczerowski P, Lipóth L, Vajda J, Veres R (2003) Surgery of ventral intradural midline cervical spinal pathologies via anterior cervical approach: our experience. *Ideggyogy Sz* 56: 115-118.
- [30] Giroux JC, Nohra C (1978) Anterior approach for removal of a cervical intradural tumor: case report and technical note. *Neurosurgery* 2: 128-130.
- [31] Sawa H, Tamaki N, Kurata H, Nagashima T (1993) Complete resection of a spinal meningioma extending from the foramen magnum to the second thoracic vertebral body via the anterior approach: case report. *Neurosurgery* 33: 1095-1098.
- [32] Lenelle J, Born JD, Collignon J (1986) [Ablation by the anterior transcorporeal approach of an ante-spinal cord cervical meningioma. Apropos of a case]. *Neurochirurgie* 32: 262-265.
- [33] Singh K, Vaccaro AR, Kim J, Lorenz EP, Lim TH, et al. (2003) Biomechanical comparison of cervical spine reconstructive techniques after a multilevel corpectomy of the cervical spine. *Spine (Phila Pa 1976)* 28: 2352-2358.
- [34] Ijiri K, Hida K, Yano S, Iwasaki Y (2009) Ventrally located cervical intramedullary cavernous angioma: selection of posterior and anterior approaches: case report. *Neurol Med Chir (Tokyo)* 49: 474-477.
- [35] O'Toole JE, McCormick PC (2003) Midline ventral intradural schwannoma of the cervical spinal cord resected via anterior corpectomy with reconstruction: technical case report and review of the literature. *Neurosurgery* 52: 1482-1485.
- [36] Steck JC, Dietze DD, Fessler RG (1994) Posterolateral approach to intradural extramedullary thoracic tumors. *J Neurosurg* 81: 202-205.
- [37] Jenny B, Rilliet B, May D, Pizzolato GP (2002) [Transthoracic transvertebral approach for resection of an anteriorly located, calcified meningioma. Case report]. *Neurochirurgie* 48: 49-52.
- [38] Kim MS, Eun JP, Park JS (2011) A dumbbell-shaped meningioma mimicking a schwannoma in the thoracic spine. *J Korean Neurosurg Soc* 50: 264-267.
- [39] Raza SM, Anderson WS, Eberhart CG, Wolinsky JP, Gokaslan ZL (2005) The application of surgical corpectomy in the management of an intramedullary extramedullary atypical meningioma: case report and literature review. *J Spinal Disord Tech* 18: 449-454.
- [40] Rutherford SA, Linton KM, Durnian JM, Cowie RA (2006) Epidural meningioma of the sacral canal. Case report. *J Neurosurg Spine* 4: 71-74.

- [41] Lista-Martínez O, Santín-Amo JM, Facal-Varela S, Rossi-Mautone E, RivasLópez LA, et al. (2012) [Multifocal clear cell meningioma in the sacral and lumbar spine. Case report and literature review]. *Neurocir Astur* 23: 259-263.
- [42] Bhadra AK, Casey AT, Saifuddin A, Briggs TW (2007) Primary atypical sacral meningioma--not always benign. *Skeletal Radiol* 36 Suppl 1: S91-94.
- [43] Toliás CM, Beale DJ, Sakas DE (1997) Giant lumbar meningioma: a common tumor in an unusual location. *Neuroradiology* 39: 276-277.
- [44] McCormick PC (1994) Anatomic principles of intradural spinal surgery. *Clin Neurosurg* 41: 204-223.
- [45] Guidetti B, Spallone A (1988) Benign extramedullary tumors of the foramen magnum. *Adv Tech Stand Neurosurg* 16: 83-120.
- [46] Petit H, Jomin M, Jeulin-Gaillard B, Warot P (1980) [Multiple "non-associated" spinal meningiomas]. *Lille Med* 25: 346-349.
- [47] Tsuji N, Nishiura I, Koyama T (1986) Extradural multiple spinal meningioma. Literature review a case report. *Neurochirurgia (Stuttg)* 29: 124-127.
- [48] Lloyd SK, Evans DG (2013) Neurofibromatosis type 2 (NF2): diagnosis and management. *Handb Clin Neurol* 115: 957-967.
- [49] Gottfried ON, Gluf W, Quinones-Hinojosa A, Kan P, Schmidt MH (2003) Spinal meningiomas: surgical management and outcome. *Neurosurg Focus* 14: e2.
- [50] Dodd RL, Ryu MR, Kamnerdsupaphon P, Gibbs IC, Chang SD Jr, et al. (2006) CyberKnife radiosurgery for benign intradural extramedullary spinal tumors. *Neurosurgery* 58: 674-685.
- [51] Gerszten PC, Ozhasoglu C, Burton SA, Vogel WJ, Atkins BA, et al. (2003) CyberKnife frameless single-fraction stereotactic radiosurgery for benign tumors of the spine. *Neurosurg Focus* 2003 14: e16.
- [52] Gerszten PC, Burton SA, Ozhasoglu C, McCue KJ, Quinn AE (2008) Radiosurgery for benign intradural spinal tumors. *Neurosurgery* 62: 887-895.
- [53] Kufeld M, Wowra B, Muacevic A, Zausinger S, Tonn JC (2012) Radiosurgery of spinal meningiomas and schwannomas. *Technol Cancer Res Treat* 11: 27-34.
- [54] Ryu SI, Chang SD, Kim DH, Murphy MJ, Le QT, et al. (2001) Image-guided hypo-fractionated stereotactic radiosurgery to spinal lesions. *Neurosurgery* 49: 838-846.
- [55] Halasz LM, Bussière MR, Dennis ER, Niemierko A, Chapman PH, et al. (2011) Proton stereotactic radiosurgery for the treatment of benign meningiomas. *Int J Radiat Oncol Biol Phys* 81: 1428-1435.
- [56] Hsu W, Nguyen T, Kleinberg L, Ford EC, Rigamonti D, et al. (2010) Stereotactic radiosurgery for spine tumors: review of current literature. *Stereotact Funct Neurosurg* 88: 315-321.
- [57] Samii PD med M, M.d MA (1992) Meningiomas Involving the Lower Clivus and the Foramen Magnum (Craniospinal Meningiomas) [Internet]; in : *Surgery of Skull Base Meningiomas*. Springer Berlin Heidelberg.
- [58] <http://emedicine.medscape.com/article/341870-overview>
- [59] Charles YP, Barbe B, Beaujeux R, Boujan F, Steib JP (2011) Relevance of the anatomical location of the Adamkiewicz artery in spine surgery. *Surg Radiol Anat* 33: 3-9.
- [60] Shi HB, Suh DC, Lee HK, Lim SM, Kim DH, et al. (1999) Preoperative transarterial embolization of spinal tumor: embolization techniques and results. *AJNR Am J Neuroradiol* 20: 2009-2015.
- [61] Gore P, Theodore N, Brasiliense L, Kim LJ, Garrett M, et al. (2008) The utility of onyx for preoperative embolization of cranial and spinal tumors. *Neurosurgery* 62: 1204-1211.