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Spinal Meningiomas: A Comprehensive Overview and Own Experience

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Abstract: Among intradural extramedullary tumours, neurinomas followed by meningiomas are the most common histological entities. Spinal meningiomas constitute only for 7.5–12.7 % of all meningiomas. More than $\frac{2}{3}$ of all spinal meningiomas are located in the thoracic spine. 86–95 % of the tumours are found purely intradurally.

Risk factors are ionizing radiation, genetic predisposition, and female gender.

For diagnosis, the methods of choice are magnetic resonance imaging scans, including T1- and T2-weighted images, with and without contrast agent. They show spheric contrast-enhancing tumours with their intradural and extra-

medullar localizations. The tumour 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 tumours). Distinct calcifications, which can be recognized in computed tomography, suggest a meningioma. Larger cystic areas rather indicate a neurinoma.

The treatment of choice is surgical removal using intraoperative ultrasound (for ideal tumour localization) and intraoperative neuromonitoring (for functional preservation). For surgery, we prefer the dorsal approach except for a few cases, sometimes laterally extended by partial resec-

tion of the vertebral joint or the head of rib in the area of the thoracic spine.

Postoperatively, the great majority of patients have excellent postoperative outcomes. In our own recent series between 1/2010 and 12/2012, 80 patients were operated on spinal tumours with 27 patients harbouring meningiomas (34 %). More than 90 % (26/27) showed clinical improvement with ameliorated gait pattern or even restarted walking without assistance. **Eur Assoc NeuroOncol Mag 2013; 3 (3): 118–21.**

Key words: spinal meningioma, spinal tumour

Introduction

Approximately $\frac{2}{3}$ of all intraspinal neoplasms are intradural extramedullary tumours. Among those, neurinomas followed by meningiomas are the most common histologic entities [1]. Spinal meningiomas occur less frequently than intracranial meningiomas. They constitute only for 7.5–12.7 % of all meningiomas [2].

Comparable to intracranial meningiomas their incidence is 2–3 times higher in women than in men. These lesions are a typical disease of the middle or older age [3, 4].

The purpose of this article is an update of this disease based on a literature review and our own experience. Additionally, we present our most recent own consecutive case series collected between 2010 and 2012.

Localization

More than $\frac{2}{3}$ 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 %) [4, 7]. In the latter, in 2 locations they are predominantly dumbbell tumours causing an enlargement of the intervertebral foramen.

Risk Factors

Most publications focus 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.

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 [24]. Additionally, neurofibromatosis type 2 with mutation of chromosome 22Q12 is an autosomally recessive, hereditary disease with elevated risk of developing meningiomas or schwannomas [25]. In a very recent publication, changes in the gene SMARCE1 could be identified in relation to an increased incidence of familial spinal meningiomas [26].

Gender

Women have a 2–3 times higher incidence of meningioma development. Additionally, the gender-related risk is slightly

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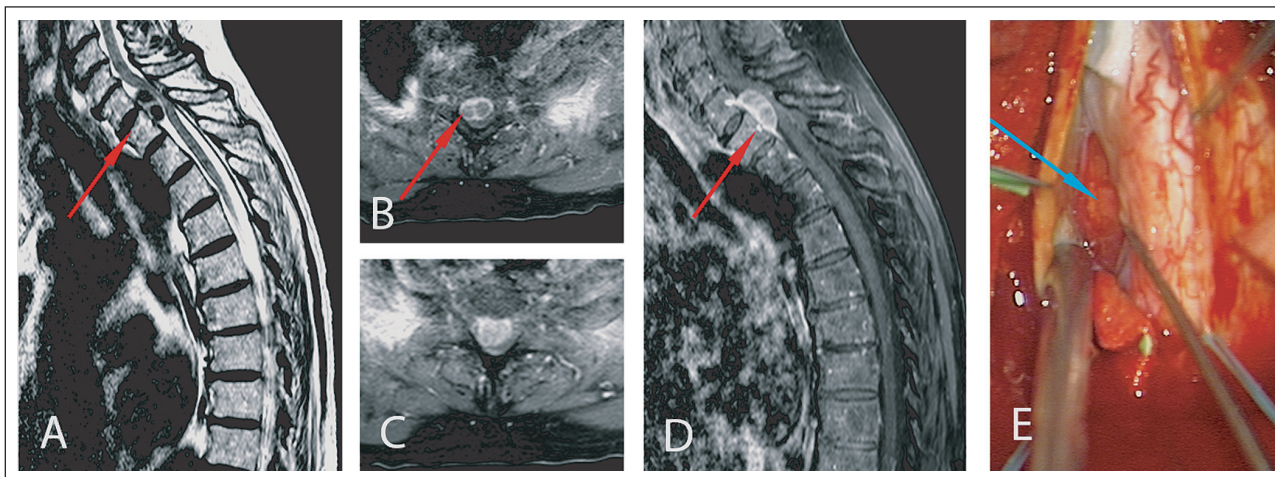


Figure 1. Pre- and intraoperative findings: (a) T2-weighted native sagittal MR image: the tumour can be nicely delineated (red arrow). (b, c) Axial T1-weighted contrast-enhanced sequence: inhomogeneous contrast enhancement is due to intratumoural calcifications (red arrow). (d) Sagittal contrast-enhanced T1-weighted image: again, the calcifications can be seen as contrast agent-free areas within the tumour (red arrow). (e) Intraoperative findings: the spinal cord is carefully mobilized under IOM. The tumour (blue arrow) can be seen at the left ventral dura.

higher in women who take contraceptives or receive hormone replacement therapy [27–29]. The coincidence of breast carcinoma and meningiomas has been observed for many years [30]. It may be due to a joint risk profile (age, genetics, environmental factors in interaction) [31, 32].

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 tumours, 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.

Management

Diagnosis and Operative Planning

The methods of choice are magnetic resonance imaging (MRI) scans, including T1- and T2-weighted images, with and without contrast agent (Figure 1). They show spheric contrast-enhancing tumours with their intradural and extramedullar localization [33]. The tumour 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 tumours). Distinct calcifications, which can only be recognized in computed tomography (CT), suggest a meningioma. Larger cystic areas rather indicate a neurinoma.

Cystic changes are very rare in spinal meningiomas in contrast to calcifications. The latter may influence the surgical

approach especially in ventrally positioned tumours. For this reason, we believe that it makes sense to perform a CT scan in ventrally positioned tumours to estimate the extent of calcifications prior to surgery. In central calcified tumours 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

We prefer the dorsal approach 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 tumour 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 tumours. The disadvantages are complications caused by the larger approach with vertebral body resection and the need for stabilization.

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 tumour we try to remove the vertebral arch 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 tumour. In ventrally positioned tumours, 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 tumours. Opening the dura directly at the tumour 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 tumour de-

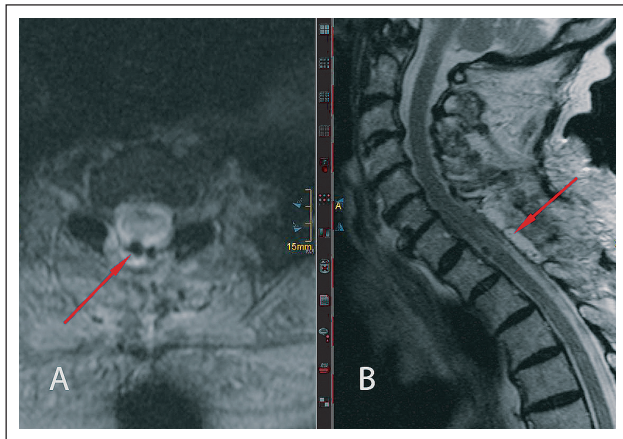


Figure 2. Postoperative MR scans: (a) T1-weighted contrast-enhanced MR images: the spinal cord (red arrow) is in normal location. No tumour remnants are visible. (b) T2-weighted native MR scan: the surgical dorsal approach and complete tumour removal are documented.

bulking is performed using bipolar coagulation and the cavitron ultrasonic surgical aspirator (CUSA) to reach the tumour matrix at the dura as fast as possible. After interruption of its blood supply the tumour can be better mobilized. This alleviates separation of the remaining avascular tumour 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 tumour removal.

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 [35].

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 our monitoring setting.

In general, tibial and median SEP are applied depending on the location of the pathology. Mostly surface values are derived but also subcortical components may be extrapolated.

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 [36–39].

IOM requires close cooperation of all groups involved in spinal tumour surgery [40]. 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.

Our Recent Case Series

Between 1/2010 and 12/2012, 80 patients were operated on spinal tumours with remarkable female prevalence (2,5:1) at our department.

The number of neurinomas (36 %) was nearly equivalent to meningiomas (34 %), followed by metastases (14 %) and other entities (16 %) like ependymoma, cavernoma, and compressive arachnoid cysts.

Postoperative Outcome

The great majority of patients have excellent postoperative outcomes (26/27 in our series). 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 tumour 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]. In our series, we had no case of epidural haematoma or fistula, but 2 cases of tumour recurrence.

Tumour Recurrence

The recurrence rate in spinal meningiomas is significantly lower than in intracranial meningiomas. As expected, total removal at first surgery is the key factor to avoid tumour recurrence. En plaque meningiomas, which often cannot be radically removed, show a significantly higher relapse rate. This is also the case in tumours with ventral matrix or in cases with severe calcifications of the tumour. Due to higher rates of subtotal tumour removal close to the arteria radicularis magna the recurrence rate in this location is also higher.

Concerning the treatment of tumour recurrence, reoperation is the first-line treatment option. The role of radiotherapy is still controversially discussed [3, 7, 33]. However, it is a possible treatment option in recurring tumours. During the years to come its significance will grow due to the development of new radiotherapeutic alternatives such as the proton beam radiation.

So far, we indicate radiotherapy only after recurrent surgery or in very old patients with an increased surgical risk profile. In cases of subtotal tumour removal, we closely follow the patients with special focus on further tumour growth (“wait and see”).

Discussion

Comparable to intracranial meningiomas, we find histological subtypes such as meningotheliomatous, hypoplastic, transitional, and psammomatous tumours.

The first 2 types are predominant in spinal meningiomas. Interestingly, the histological type does not seem to influence prognosis. Compared to intracranial meningiomas spinal tumours less frequently belong to WHO grades II and III [1–7]. Nevertheless, spinal meningioma represents an entity of its own.

Surgery is always the therapy of choice in spinal meningiomas. In the vast majority of patients, the operation results in significant improvement of the preoperative neurological deficits [2, 4–6, 33].

In rare tumours 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.

Concerning our surgical philosophy we prefer dorsal approaches whenever possible. Also, ventral tumours 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 tumours located exactly in the midline, the spinal cord may cover the tumour bilaterally. Only in these cases a ventral approach with vertebral body resection is necessary.

Conclusions

During the last 3 decades the prognosis of spinal meningiomas has improved for the following 3 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.

Conflict of Interest

No author has a conflict of interest related to this paper.

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