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CASE REPORT

THORACIC MENINGIOMA PRESENTING WITH PROGRESSIVE LOWER PARAPARESIS IN A FEMALE PATIENT IN HER 30'S

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Abstract. *The most common tumors located in the thoracic segment of the spinal cord are spinal meningiomas and schwannomas. Meningiomas arising from the sheaths of the spinal cord represent about 20% of all benign tumors of the spinal canal. Second place is occupied by metastases, which mainly affect bone structures and much less often – the spinal cord. Meningiomas are predominantly intradural extramedullary tumor formations, although some rare cases of extradural spinal meningiomas have been described in the literature. The World Health Organization divides these tumors into three grades according to their malignancy, which include 15 histological subtypes of meningiomas with a predominantly benign course of development. The psammomatous meningioma belongs to grade I with a benign course and its total extirpation leads to a good ten-year tumor control. Meningiomas are typically slow-growing tumors that commonly present clinically after the fifth decade of life, with a higher prevalence in female patients. The occurrence of thoracic meningiomas in younger individuals, particularly in the second and third decades is often associated with neurofibromatosis. In this report, we describe a female patient in her 30's with a one-year history of back pain radiating along the ribs, accompanied by progressive lower limb weakness and sensory disturbances over the past 30 days. The clinical complaints and the performed imaging diagnostics show the presence of an intradural, extramedullary tumor formation located at the level of TH5-TH6 vertebral projections and occupying more than 60% of the spinal canal. The treatment consists in total tumor extirpation by means of microneurosurgical technique and histological verification of the process. Subsequent radiotherapy for resected Simpson grade II spinal meningiomas and for psamoma variant of meningioma is not recommended. Clinical monitoring using magnetic resonance imaging is the gold standard for controlling tumor growth.*

Key words: *spinal meningiomas, spinal cord, inferior paraparesis, histology of meningiomas, thoracic spinal segment, intradural extramedullary tumors, neurofibromatosis*

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INTRODUCTION

Spinal meningiomas are rare tumors that are mainly located in the thoracic segment of the spine. They account for about 3% of all meningiomas of the central nervous system (CNS) and about 20% of benign tumors located in the region of the spinal cord [1, 2, 3]. These tumors originate from the sheaths of the spinal cord, where they form meningeal hyperplasia at the site of the dural tail. The gold standard in the diagnosis of meningiomas is magnetic resonance imaging (MRI), with the application of intravenous contrast, which shows the relation between the tumor and the spinal dura [1, 4, 7].

The epidemiological features of these tumors are mainly related to female sex, age over 50 years, genetic predisposition and the influence of radiation. Manifestation at an earlier age, as well as the combination of multiple meningiomas, or with bilateral schwannomas of the vestibular nerve, is associated with neurofibromatosis type 2 (NF2) [1, 4, 5]. The most common histological types localized in the thoracic segment of the spinal cord are psamoma meningiomas with predominance of calcifications. According to the classification of the World Health Organization (WHO), these tumors are of a benign nature and are classified as grade I. The localization of meningiomas of the spinal canal is most often in the posterior, posterolateral and lateral thoracic regions. Ventrally located memingyms are rare. The standard in their treatment is the surgical extirpation of the tumor by means of open microsurgical access, minimally invasive surgery or endoscopic treatment [1, 5, 7, 8].

In the present report, we describe a woman in her 30's with a clinical presentation of lower spastic paraparesis and conduction-type hypesthesia. An MRI of the thoracic spine supported the diagnosis of thoracic meningioma with spinal cord compression. The chosen operative approach, by means of micro-neurosurgical extirpation, helps to remove the tumor completely and to avoid postoperative surgical complications.

CLINICAL CASE DESCRIPTION

A woman in her 30's was admitted to the neurosurgery department due to complaints of back pain of an alarming nature, which started about a year ago. She repeatedly underwent treatment with analgesics, non-steroidal anti-inflammatory drugs and physiotherapy, however, without a lasting result. The pain was localized at the level of TH5, TH6 vertebral projections and radiated along the corresponding dermatomes, being more pronounced in the left thoracic half. The patient denies having any accompanying diseases and harmful habits such as smoking and radiation exposure.

Over the past 30 days, her symptoms have worsened, with increasing stiffness and numbness in her legs, indicative of impaired conductive-type sensation. The progressive weakness in her lower limbs led to several falls, necessitating the use of mobility aids. Upon admission, the neurological examination revealed a patient who was alert, oriented, and in clear consciousness, with no signs of meningeal or radicular irritation, and preserved cranial nerve function. Testing for latent paresis confirmed spastic paraparesis of grade II to III.

Increased tendon-periosteal reflexes for lower limbs, increased muscle tone, more pronounced for the left leg, impaired coordination for the legs, with suppressed surface sensation and preserved deep sensation, were found. Positive pathological Babinski reflex on the left was seen. The level of hepesthesia is conductive-type, distal to the fifth, sixth intercostal space and is bilaterally symmetrical and equally expressed. The function of the pelvic reservoirs and defecation are not impaired.

The performed diagnostic imaging included magnetic resonance imaging of the thoracolumbar spine (MRI), which demonstrated a space-occupying process localized at the level of TH5-TH6 vertebral projections. The tumor formation is located intradurally, extramedullary and occupies more than 60% of the volume of the spinal canal, which leads to pronounced medullary compression. Imaging interpretation includes T2, T1 and TIRM sequences in the sagittal, axial and coronal planes as well as the administration of intravenous contrast material, which achieves homogeneous filling of the tumor with contrast. The type of tumor, according to the imaging diagnostics, points to a meningioma localized in the thoracic region. Sagittal sections give good information on the localization of the tumor, and axial sections better define the location of the tumor relative to the spinal cord (the most common location in clinical practice is the dorsolateral). Figures 1, 2, and 3 illustrate the location of the tumor relative to the spinal cord, which can be used for preoperative planning of the operative access.

In the course of preoperative preparation, laboratory tests did not show any deviations from the norms. A chest X-ray was performed, which is part of the standard panel of investigations when performing neurosurgical operations under general anesthesia. Chest X-ray showed a symmetrical chest, non-en-

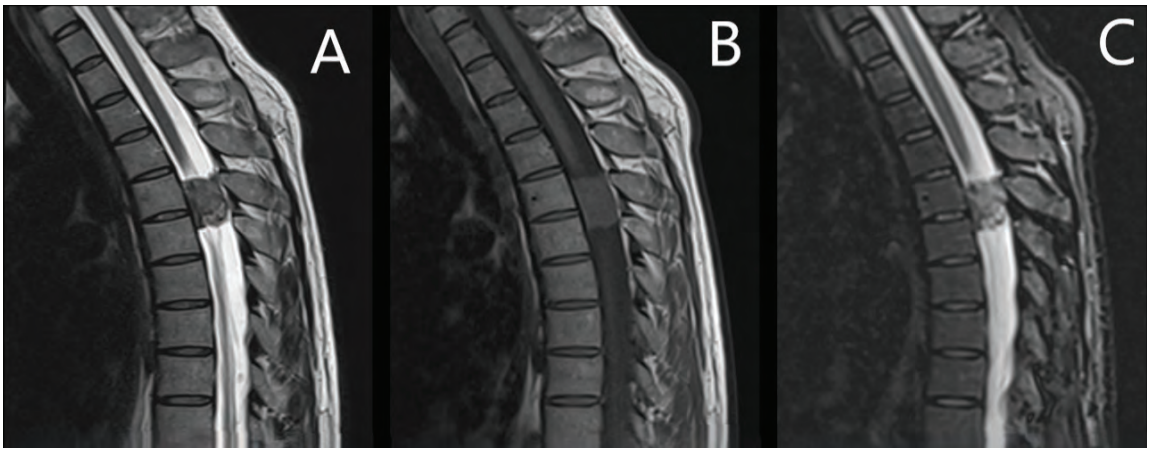

Fig. 1 (**A, B and C**). MRI of the thoracic spine shows a meningioma formation located at the level of TH5-TH6 vertebral projections. Panel A presents a sagittal T2 sequence showing a dorsolaterally located hyperdense tumor with a lobular structure. Panel B presents a sagittal T1 sequence in which the tumor has an isointense appearance to mildly hyperintense in places. Panel C presents a TIRM-sequence with a subthreshold magnetic signal of the adipose tissue and shows a hyperintense tumor mass

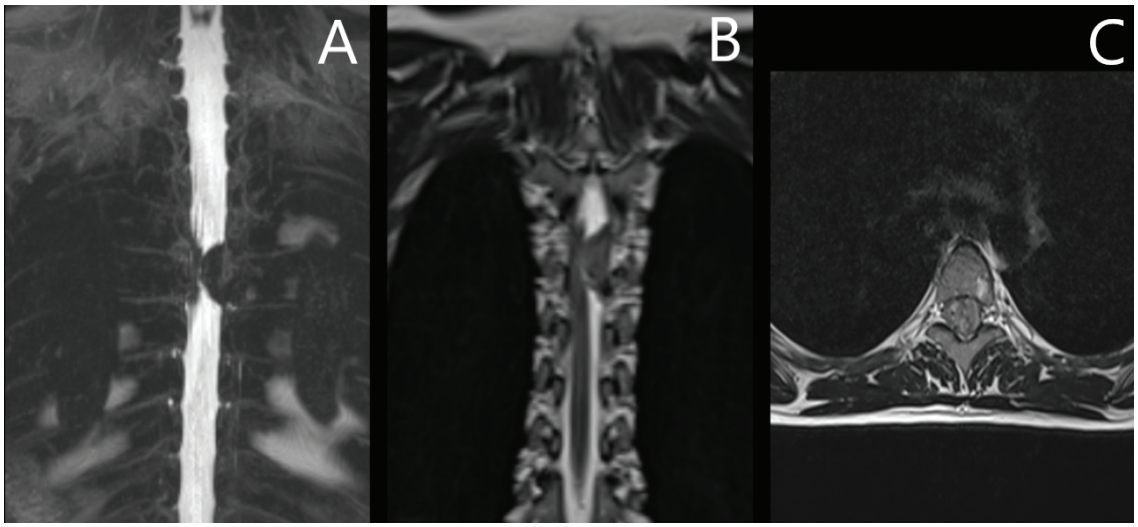

Fig. 2 (**A, B and C**). MRI of the thoracic spine in the coronal and axial planes. Panel A presents a virtual myelography showing the disturbed CSF flow. Panel B presents a coronal view of a T2 sequence showing the dorsolateral left-sided location of the meningioma. Panel C shows an axial section through the middle of the tumor, where a clear border between the meningioma and the spinal cord is difficult to identify

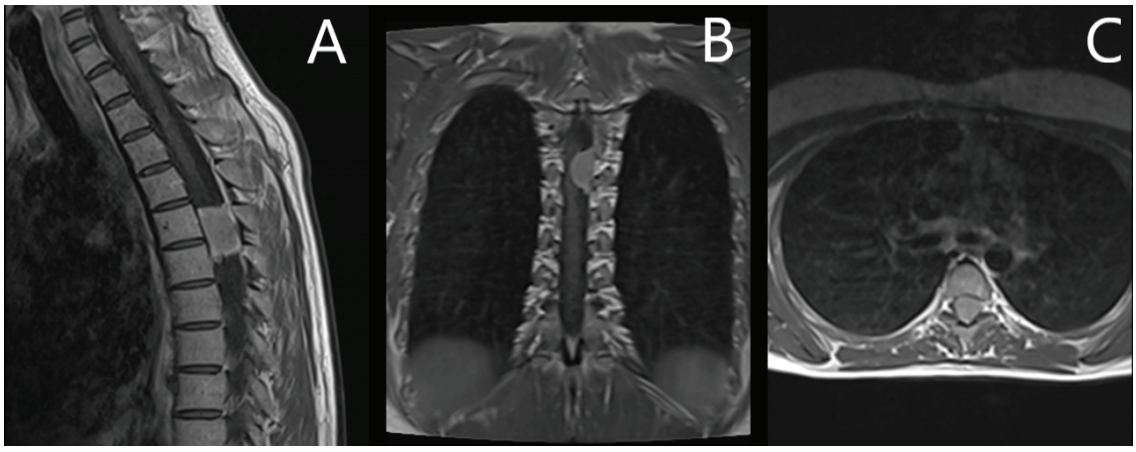

Fig. 3 (**A, B and C**). MRI of the thoracic spine with the application of gadolinium in a T1 sequence of sagittal, coronal and axial planes showing a homogeneously filled hyperintense tumor mass

larged hilar shadows, preserved lung parenchyma, diaphragms were normally configured. Unenlarged heart shadow was seen. In conclusion, there was no radiographic evidence of an active inflammatory process or infiltrative disease (Figure 4).

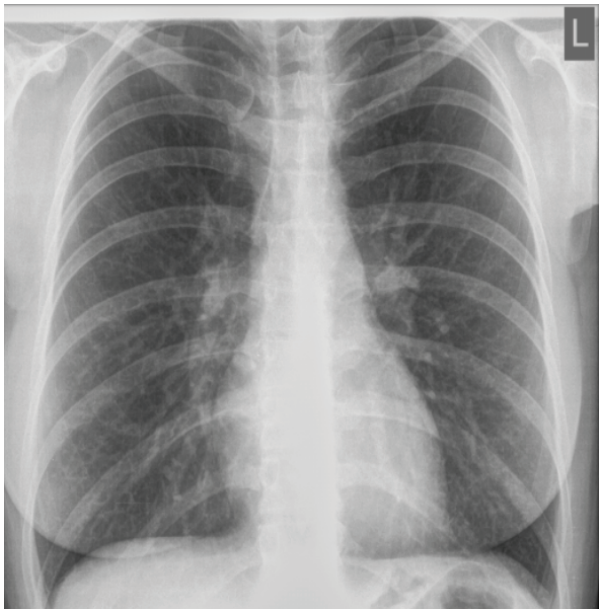

Fig. 4. X-ray of a lung with preserved parenchymal transparency and no evidence of infiltrative diseases

Surgical extirpation of tumors of meningeal origin is a major part of their treatment. Preoperatively, the level was verified by C-arm fluoroscopy and approached with a median dorsal incision at the level of TH5-TH6 vertebral projections. A laminectomy was performed on them, preserving the joint complexes (the spinous processes and arches of the TH5 and TH6 vertebrae were removed). In this way, no destabilization of the thoracic spine segment occurs, which is preferred in similar type of surgical interventions in our department. A median durotomy was performed along the course of the laminectomies, where an encapsulated tumor formation was encountered. Under microneurosurgical control, dissection of the arachnoid was carried out, medial enucleation of the tumor process was performed, which macroscopically appeared whitish-white with a granular structure. It is devascularized from its feeding vessels and in a good arachnoid plane it is removed from the adjacent neural structures totally with coagulation at its capture site (Simpson II) (Figure 5).

The morphological result shows the type of tumor most accurately. The psammomatous meningioma is classified as WHO grade I with a predominantly benign nature. Histologically, it is composed of numerous psamoma bodies adjacent to proliferated meningothelial cells. They are oval formations composed of concentrically arranged calcified collagen fibers. Occasionally, psammosomal bodies may confluent into large calcium masses, and more than half of the tumor component must be represented by them (Figure 6).

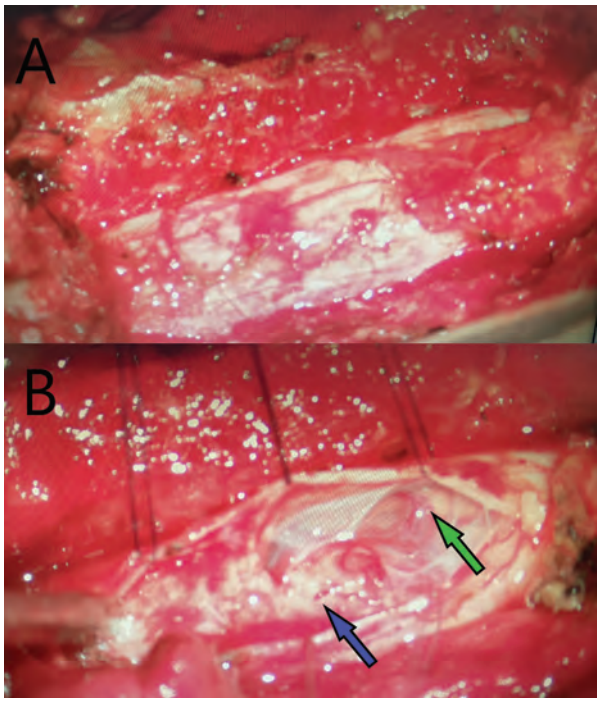

Fig. 5 (**A and B**). Intraoperative miroscopic images during tumor removal. Panel A presents an apparently intact but highly stressed dura mater spinalis. Panel B shows the tumor process (blue arrow) and the dislocated and compressed spinal cord, with arachnoid (green arrow)

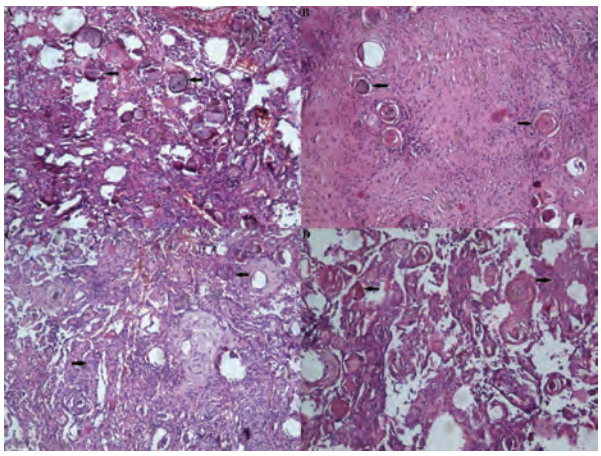

Fig. 6 (**A-D**) Tumor histology showing round, oval calcifications between meningothelial whorls (H & E stain; original magnification x 100)

Postoperatively, the patient recovered well, was verticalized and mobilized on the second postoperative day. Complications from the side of the surgical wound and liquorrhea were not observed. There was an improvement in the manifestations of hypesthesia, and

the motor neurologic deficit showed good trends of improvement. A month after the surgical intervention, the clinical examination showed a good recovery of the motor function – I degree lower paraparesis.

Oral consent for publication has been obtained from the patient.

DISCUSSION

Spinal meningiomas are significantly less common compared to their cranial location and, according to literature, account for about 1.2% to 3% of all meningiomas of the CNS [1, 2]. They can be located in different places along the spinal canal, being the most common in the thoracic region, followed by cervical localization and lumbar region. Clinical complaints are leading, as most often they start with thoracic pain (depending on tumor localization), followed by motor deficits due to medullary compression and ischemic myelopathy, combined with sensory changes.

In the clinical case that we report, the patient's symptoms developed in a manner consistent with the patterns described in the literature. With the advent of imaging, these processes have become significantly more recognizable, and the diagnosis of thoracic meningioma takes on average several months. In most clinical studies of large patient series, the diagnosis was made within 10 months [1, 2, 3, 7, 12]. Magnetic resonance imaging (MRI), in which meningiomas of the thoracic spine have an isointense to hyperintense signal in the T2 sequence and hypointense, and sometimes with intermediate intensity in the T1 sequences, is the main place in the diagnosis. Contrast enhancement contributes to an increase in the density of the tumor, which clearly delineates the boundaries of it.

Yeo et al. classified spinal meningiomas into four categories according to MRI images [1]. The so-called "dural tail" is visible, as is the location of the tumor relative to the spinal cord. Dorsal and dorsolateral location of the tumor is reported as the predominant part in most studies. The ventral location of the tumor in the thoracic segment is rare and predisposes to other types of operative approaches. In the literature, it is reported that ventral location of meningiomas is more characteristic of cervical localization. Meningiomas of the "en plaque" type have also been described, which grow on a broad base and occupy a large area of the spinal cord [1, 3, 4, 5, 7, 12].

The basis of the diagnosis is the histological examination of the tumor. In this way, it is determined to which grade the meningioma belongs and what are its histological features. WHO divides meningiomas into three grades according to the degree of their malignancy. Grade I and II meningiomas are benign in nature, and surgical treatment with total extirpation of the tumor and coagulation of its attachment site (Simpson grade II) or total extirpation of the tumor with excision of its attachment site along the dura (Simpson grade I) significantly reduces the risk from relapse [5, 7, 12, 13].

WHO grade III meningiomas have a tendency toward recurrence and malignant malignancy [1, 3]. The most common histological types that can be encountered in spinal or brain localization are: psammomatous, atypical meningioma, meningothelial, transitional type, metaplastic, and for the thoracic localization of the tumor, the most characteristic is the psammomatous variant with the accumulation of calcifications, as is also in the case reported by us. Psammomatous meningiomas should generally predominate due to their structure composed of psammoma bodies. In its progression, it is reported to probably begin as a transient meningioma and gradually undergo dystrophic calcification near the meningothelial cells over time.

With the development of molecular biology and genetics, determining the genetic status of the tumor has become an essential part of the complex treatment of CNS tumors, including meningiomas. Women after the fifth decade of life have a more serious predisposition to the manifestation of meningioma, but at an early age meningiomas occur in neurofibromatosis type 2 (NF2) [3, 8, 9 10, 11]. The NF2 gene is located on chromosome 22q12.2 and leads to the production of a specific protein called merlin, involved in the activation of several signaling pathways. Merlin protein is a tumor suppressor and its loss is characteristic of NF2 meningiomas. There is also a second group of patients in whom mutations in the NF2 gene are not detected, but other types of mutations are detected. AKT1 and TRAF7 have been found in WHO grade I meningiomas, and many other predisposing mutations in meningiomas have been reported. To establish a diagnosis of neurofibromatosis, key considerations include clinical criteria such as the presence of bilateral vestibular schwannomas, the occurrence of meningiomas at multiple sites within or outside the central nervous system, and supportive genetic tests. Although NF2 is generally a benign disorder, the localization of tumors can pose significant surgical challenges and may result in severe complications, including death [10, 11]. This disease has a family predisposition and can be inherited in generations. The most common accompanying tumors are: vestibular schwannomas, peripheral schwannomas (tumors located along the plexuses), meningiomas,

glial ependymoma, meningoangiomatosis, which occurs rarely, and glial microhamartomas [11, 14].

New trends in the treatment of NF2 focus on studies of focal adhesion kinase (FAK). Its inhibition has been shown to lead to NF2 gene loss and thereby achieve tumor control [15]. In our case, the patient is subject to systematic clinical observations through clinical examinations and MRI of the thoracic spine. The established criteria for our department are that imaging diagnostics and examination once a year lead to systemic control in case of tumor recurrence.

Spinal thoracic meningiomas are defined as predominantly benign tumors with slow local growth. Single cases of extradural meningiomas as well as intramedullary meningiomas have been described in the literature. Operative treatment remains the gold standard, with the goal of achieving total tumor resection. Dorsolaterally located meningiomas do not present a surgical challenge for their removal. A classic microneurosurgical approach or a minimally invasive neuroendoscopic approach can be used. We accept as a more convenient method of treatment the microneurosurgical approach by means of laminectomy with the application of intraoperative intravenous corticosteroids, as well as perioperatively. In the literature, there is no sufficient reliable evidence regarding their benefit, but it is believed that they reduce peritumoral edema and thus contribute to fewer iatrogenic injuries to the spinal cord [2, 4].

The use of intraoperative ultrasound is extremely helpful in tumor localization. In some cases, an intraoperative ultrasonic aspirator can be used, which has shown to reduce bleeding from the tumor. The application of intraoperative neuromonitoring, by means of somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs), has an important role in atypical localization of the tumor and adhesion to important neural structures that do not allow to be resected [2]. Radical surgical resection for WHO grade I and II tumors does not require adjuvant therapy or radiation therapy. In the presence of tumor recurrence, high degree of malignancy, subtotal resection or partial, NF2 and the presence of other tumors, the use of fractionated radiotherapy, stereotactic radiosurgery, tyrosine kinase inhibitors, brachytherapy, or a combination of the above methods is indicated. Fractionated radiation therapy is most commonly used, and in some types such as anaplastic meningioma, treatments blocking the expression of vascular epidermal growth factor (VEGF) are used [2, 3, 5, 9, 11].

Postoperative complications and improvements were determined by the Frankel scale, which examines neurological status. In our case, an improvement over it was achieved. Large clinical trials reported a 3% WHO grade I tumor recurrence and total tumor resection after a median period of 76.5 months [2].

CONCLUSIONS

Meningiomas of the thoracic spine region represent a rare and sometimes challenging area in neurosurgery and oncology management aspects. A thorough understanding of their epidemiology, clinical features and treatment options is essential for neurosurgeons. The appearance of these tumors at a young age may be an incidental finding or part of the neurofibromatosis disease syndrome. Treatment outcomes indicate that surgery is the most effective approach for thoracic spine meningiomas, achieving high rates of complete resection and excellent long-term prognosis.

Radiation therapy may be considered an alternative for inoperable or residual lesions, although its effectiveness is generally lower compared to surgical treatment. Operative guidelines are towards minimally invasive and endoscopic methods. Histological diagnosis is crucial to determine the subsequent treatment and the following clinical monitoring. Future research should aim to elucidate the molecular and genetic factors involved in the development of thoracic spine meningiomas and identify potential therapeutic targets for this rare and debilitating disease.

Informed Consent for a Clinical Case: *Written informed consent was obtained from the patient for the publication of this case report, including any accompanying images.*

Conflict of Interest Statement: *The authors declare no conflicts of interest related to this work.*

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Ethical statement: *This study has been performed in accordance with the ethical standards as laid down in the Declaration of Helsinki.*

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