

A Neglected Giant Cervical Intradural Extramedullary Tumor

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ABSTRACT

Primary spinal tumors are rarely seen, and representing 4-8% of all central nervous system tumors. Extradural tumors constitute approximately 40% of intraspinal tumors whereas intradural-extramedullary tumors make up the remaining approximately 60%. These tumors are classified as extradural, intradural, extramedullary, and intramedullary depending on their origin and anatomic location. Intradural extramedullary spinal tumors form schwannomas and meningiomas.

Spinal meningiomas are usually intradural extramedullary tumors. Meningiomas can be seen in epidural localization, extradural extension only, with nerve rostral invasion as, vertebral meningioma, or as multiple spinal meningioma.

Spinal meningiomas are the most common intradural-extramedullary tumors and are generally slow-growing. Spinal meningiomas may have different clinical manifestations depending on their location and size. Surgical treatment should be planned according to the size of the tumor in the clinical situation after diagnosis.

This study reports the case of a 57-year-old female patient presenting with a one-month history of increasing weakness in the lower extremities and numbness in the upper extremities. In addition, the report includes a literature review.

Keywords: giant cervical spinal tumor, meningioma, posterior stabilization, neglect

ÖZET

İhmal edilmiş bir dev servikal intradural ekstramedüller tümör

Primer spinal tümörler nadiren görülür ve tüm merkezi sinir sistemi tümörlerinin %4-8'ini temsil eder. Ekstradural tümörler, intraspinal tümörlerin yaklaşık %40'ını oluştururken, intradural ekstramedüller tümörler, intraspinal tümörlerin yaklaşık %60'ını oluşturur. Bu tümörler, orijinlerine ve anatomik yerlerine bağlı olarak, ekstradural, intradural, ekstramedüller ve intramedüller olarak sınıflandırılırlar. Intradural ekstramedüller spinal tümörleri, schwannomalar ve meningiomalar oluştururlar.

Spinal meningiomalar, genellikle intradural-ekstramedüller tümörlerdir. Meningiomalar, sadece epidural yerleşimli, ekstradural yayımlı, sinir kökü invazyon, vertebral meningioma, multipl spinal meningioma da görülür.

Spinal meningiomalar, en yaygın intradural ekstramedüller tümörler olup genellikle yavaş büyür. Spinal meningiomaların yeri ve büyüklüğüne bağlı olarak farklı klinik belirtileri olabilir. Teşhis sonrası klinik durum ve tümörün büyüklüğüne göre cerrahi tedavi planlanmalıdır.

Bu çalışma, alt ekstremitelerde güçsüzlük artışı ve üst ekstremitelerde uyuşukluk ile başvuran ve bir aylık öyküsü olan, 57 yaşındaki bir kadın hasta olgusunu bildirmektedir.

Anahtar kelimeler: dev servikal spinal tümör, meningioma, posterior stabilizasyon, ihmal

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Date of submission: February 9, 2017

Date of acceptance: February 20, 2017

Introduction

Intradural extramedullary (IDEM) tumors account for about 60% of intraspinal tumors (1). The most common type (25-45%) of IDEM tumors are meningiomas that originate from

arachnoid cap cells; they are often benign but may rarely be malignant (2,3).

IDEM tumors usually are of benign nature. However, they may lead to severe neurogenic deficits through compression of the medulla, given that they are located in an anatomically

narrow area (4). In line with the advancements in imaging techniques, particularly in magnetic resonance imaging (MRI), early diagnosis of IDEM tumors has minimized the delays in the treatment of the tumors, and complete healing can be achieved through total excision of the tumor using distinct approaches. However, treatment attempts for the symptoms caused by spinal tumors prior to the development of neurological symptoms vary between different departments (e.g. physical therapy, algology, and orthopedics). This delay in diagnosis causes the condition to become a late-stage tumor.

In this report, we present a case who underwent various treatments in other departments and was finally diagnosed in our department with a neglected IDEM spinal tumor completely occupying the cervical canal.

Case Report

History and Examination

A 57-year-old female patient presented with a two-year history of progressive weakness and numbness in the legs, difficulty in walking, and severe pain in the neck and back. Medical history revealed that the patient had undergone numerous conventional treatments in various parts of Van province but had achieved no relief of symptoms. Subsequently, the patient was given a course of medical treatment and physical exercise by several departments including physical therapy, orthopedics, and algology). She was seen first by physical therapy on November 3, 2015. The patient then attended the orthopedic clinic on

November 25, 2015. After the emergence of walking difficulty the patient was referred to our department on December 22, 2016. The patient's surgery was performed on December 23, 2016.

The patient had no history of chronic diseases including diabetes and hypertension.

Neurological examination revealed paraparesis in both lower extremities (motor strength: 3/5), hypoesthesia below the C6 dermatome, and increased reflex in the knees and ankle joints. The patient was walking with the support of a walking aid and had spasticity limited to the lower extremities. Cervical T2 sagittal and axial MRI revealed a contrast-enhanced IDEM tumor, lying along the C7 vertebral body, completely occupying the spinal canal, and causing severe cord compression (Figure 1).

Surgical Technique

The patient was operated at Yuzuncu Yil University, Faculty of Medicine, Department of Neurosurgery.

In the prone position, a hemilaminectomy of C5 and C7 and a total laminectomy of C6 were performed via posterior approach. The tumor was soft, partially hemorrhagic, and had a clear cleavage plane. The tumor was totally excised by using microdissection techniques. The dura mater was closed primarily. In order to prevent the development of kyphotic deformities, posterior stabilization was achieved with lateral mass screws inserted at the level of C3-C6-C7 and pedicle

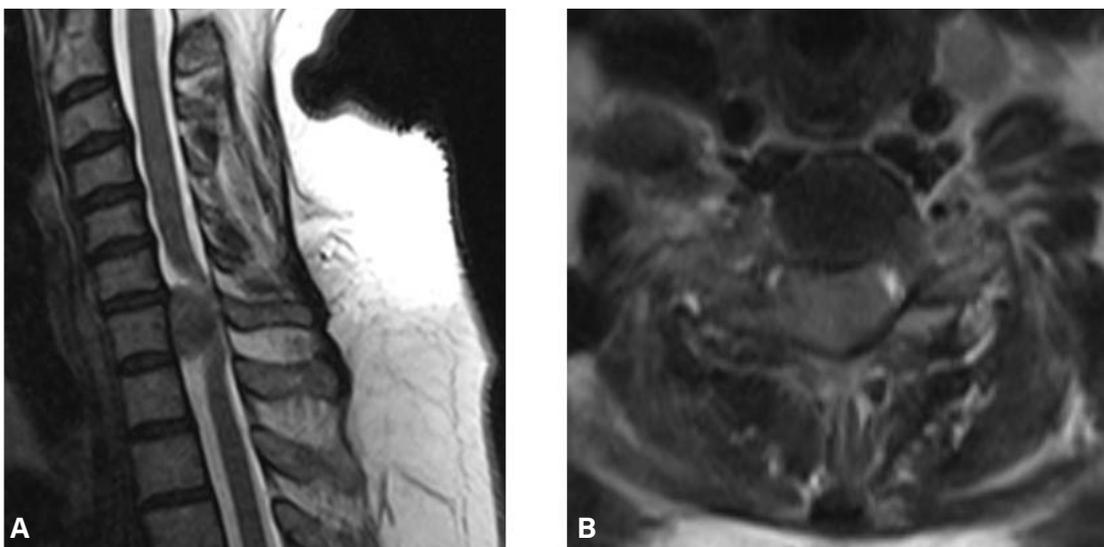


Figure 1: A) Preoperative cervical sagittal T2 MRI. B) Preoperative cervical axial T2 MRI.

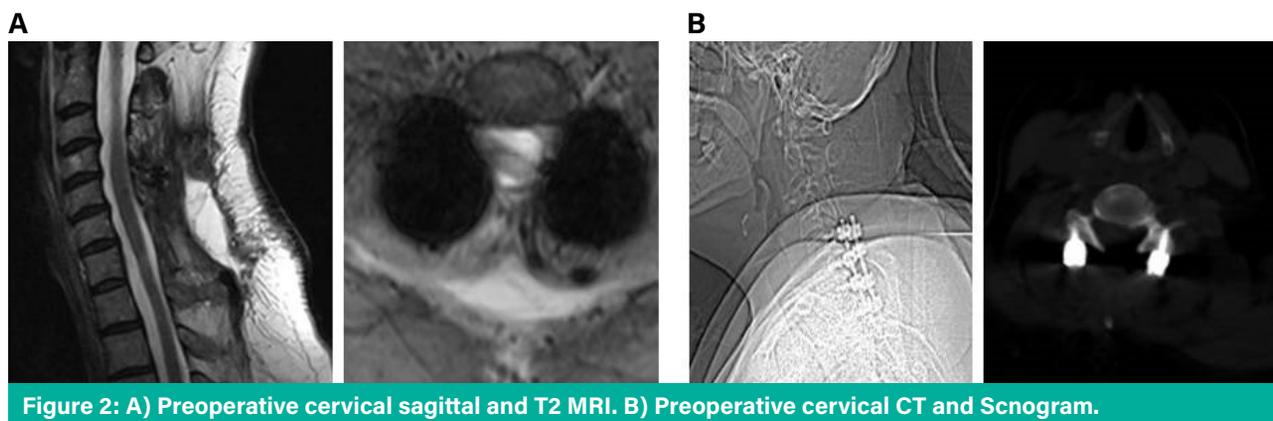


Figure 2: A) Preoperative cervical sagittal and T2 MRI. B) Preoperative cervical CT and Scnogram.

screw fixation at T1 (Figure 2). The tumor was compressing the spinal cord from the front. Since 80% of the vertebral stability is obtained from the vertebral corpus, an anterior approach, which is a more aggressive surgery, was not preferred. Posterior segmental instrumentation including lateral mass screw and pedicle screw was applied to the patient as a postoperative approach.

Follow-up

Following the surgery, the patient had 4/5 motor strength in the lower extremities and the spasticity in the lower extremities resolved. The patient was followed up in our clinic and the pathological examination indicated a psammomatous meningioma (Grade 1). The patient was referred to the physical therapy department for rehabilitation. All the symptoms resolved at six months postoperatively.

Discussion

Meningiomas originate from arachnoid cap cells and are usually of benign character. However, they may also be malignant, though rarely. Spinal meningiomas are often located in the IDEM space, but it is uncommon for a spinal meningioma to have nerve root invasion or an extradural or solitary epidural location (5,6). In the literature, a total of 17 cases of extradural meningioma have been reported by Frank et al. (6) Localization of our case was IDEM, and it was of benign character.

Spinal meningiomas may have different clinical manifestations depending on their location and size. Unless detected incidentally, these tumors are diagnosed when they cause severe neurological deficits, since they have a slow rate of growth. Among these deficits, motor and sensory deficits,

hyperreflexia, spasticity, and urinary and fecal incontinence can be seen in isolation or together (5). Spinal meningiomas were mostly thoracic (56%), and cervical (39%) (6,4). In our patient, the tumor was located in the cervical region.

In the absence of MRI, radiological diagnosis of spinal meningiomas can be achieved by direct radiography and computed tomography (CT). Direct radiography shows 25% of scoliosis and also displays bony erosion if the tumor is a late-stage tumor and determines interpedicular widening in the presence of invasion in interpedicular distances. CT is particularly useful for the detection of calcification in psammomatous meningiomas (7). With the advent of MRI, the establishment of the diagnosis of IDEM tumors has become relatively easier. We used cervical MRI for diagnosis but the patient was diagnosed late because no MRI had been used in the other centers.

The surgical approach to be used in the treatment of spinal meningiomas depends on the size and location of the tumor and whether the tumor is single or multiple. Approaches can be anterior or posterior. In the literature, a number of disadvantages have been reported for the anterior approach, including (I) limited view of the surgical site due to restricted accessibility, (II) high risk of BOS fistula after surgery, (III) excessive hemorrhage due to the extensive epidural venous plexus, (IV) prolonged operative time due to anterior stabilization with cage and plate during and after corpectomy, and risk of morbidity (8). By using the posterior approach, the tumor can be excised by laminectomy, hemilaminectomy, and costotransversectomy. However, when used in the tumor locations at the junction site and in cases that require multiple-level laminectomy, this approach may require posterior

stabilization due to the sagittal balance disorder that may occur in the late stage of the disease (9). In our patient, posterior stabilization was achieved with lateral mass screws and pedicle screws in order to prevent the development of sagittal balance disorders and cervical kyphosis in the cervicothoracic region. At six month postoperatively, no complications were detected in the patient. The spinal meningiomas located in the cervical and upper thoracic segments are often located at the anterior aspect of the spine, as seen in our patient. Although it is highly difficult in the posterior approach to remove a tumor without causing neurological deficits, this approach provided successful outcomes in our patient.

Conclusion

The patient's preoperative neurological state is the key factor affecting the outcome of the treatment of spinal tumors. In the case presented, the preoperative neurological deficits completely resolved after the total excision of the tumor. It is highly uncommon for a meningioma to completely occupy the

spinal canal in the cervical region. Early diagnosis of our patient was prevented due to attempts of treatments in other departments.

Since the IDEM tumors have a slow rate of progress, these tumors should be diagnosed by clinicians working in other departments before they become late-stage tumors, or such patients should be promptly referred to a neurosurgery clinic. The primary aim in the treatment of IDEM tumors is to achieve total excision of the tumor and to preserve or restore spinal stability.

Contribution Categories	Name of Author
Follow up of the case	A.A., M.A., M.E.A.
Literature review	A.A., F.K., M.E.A., M.A.
Manuscript writing	A.A., M.A., F.K.
Manuscript review and revision	A.A., F.K., M.E.A.

Conflict of Interest: Author declared no conflict of interest.

Financial Disclosure: Authors declared no financial support.

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