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Cervico-Occipital Meningioma in a 5-Year-Old Child: A Case Report

5 Yaşındaki Bir Çocukta Serviko-Oksipital Meningiom: Bir Olgu Sunumu

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ABSTRACT

AIM: Childhood meningiomas are scarce in clinical practice with an incidence ranging from 0.4 to 4.6% of all pediatric central nervous system (CNS) tumors. Cervico-occipital meningiomas account for 3.7% of childhood meningiomas and are slightly more frequent in male.

RESULTS: A 5-year-old female presented with febrile posterior cervico-occipital pain for 3 weeks. She was diagnosed with meningitis and treated for a similar period with adapted antibiotics. The pain persisted even after treatment. Magnetic resonance imaging revealed an enhancing subdural extra medullary mass of the cervico occipital junction, developing around the left vertebral artery. The characteristics of the lesion were strongly suggestive of a neuroma. Surgical removal of the tumor aiming the decompression of the spinal cord and nerve roots was performed with a surprising discovery: The tumor was tightly attached to the dura at the entry of the left vertebral artery. The resection was total and only a thin part close to the artery was left. The pathological findings confirmed the diagnosis of meningothelial meningioma.

CONCLUSION: Meningioma should be considered in the differential diagnosis of contrast enhancing subdural extra medullary lesions of the cervico-occipital junction in children.

KEYWORDS: Pediatric neurosurgery, MRI, Tumours, Child

ÖZ

AMAÇ: Çocukluk çağı meningiomlarına klinik uygulamada nadir rastlanır ve tüm pediatrik merkezi sinir sistemi (MSS) tümörlerinin %0,4 ila %4,6'sını oluştururlar. Serviko-oksipital meningiomlar çocukluk çağı meningiomlarının %3,7'sini oluşturur ve erkeklerde biraz daha sıktırlar.

BULGULAR: 5 yaşında bir kız çocuğu 3 haftadır mevcut olan febril posterior serviko-oksipital ağrıyla geldi. Menenjit tanısı kondu ve uygun antibiyotiklerle benzer bir süre tedavi edildi. Ağrı tedaviden sonra da devam etti. Manyetik rezonans görüntüleme sol vertebral arterin hemen çevresinde serviko oksipital bileşkede kontrast tutan bir subdural ekstramedüller kitle ortaya koydu. Lezyonun özellikleri kuvvetle bir nöroma düşündürüyordu. Omuriliğin ve sinir köklerinin dekompresyonu amacıyla tümörün cerrahi olarak çıkartılması gerçekleştirildi ve şaşırtıcı bir iyileşme oldu: Tümör sol vertebral arter girişinde duraya sıkı şekilde tutunmuştu. Rezeksiyon totaldi ve sadece artere yakın ince bir kısım bırakıldı. Patolojik bulgular meningotelyal menenjiyom tanısını doğruladı.

SONUÇ: Çocuklarda serviko-oksipital bileşkenin kontrast tutan subdural ekstra medüller lezyonlarının ayırıcı tanısında meningiom dikkate alınmalıdır.

ANAHTAR SÖZCÜKLER: Pediatrik nöroşirürji, MRG, Tümörler, Çocuk

INTRODUCTION

In a series of studies in the literature, the incidence of meningiomas range from 0.4 to 4.6% of all pediatric central nervous system tumors and pediatric forms represent approximately 1.5 to 2% of all meningiomas with a slight male predominance (2, 3).

Infantile meningiomas are exceedingly rare and the 2010 Central Brain Tumor Registry of the United States statistical report diagnosed only 84 cases occurring in the first decade of life whereas in adults 53.170 meningiomas were recorded.

Tumors of the cervico-occipital junction are classified as either extradural or intradural. Intradural tumors are further divided into intra medullary or extra medullary. The commonest intradural extra medullary tumors are schwannomas and neurofibromas. Meningiomas are infrequent in this location and account for only 3.7 % of all childhood meningiomas according to a recent study (2).

CASE REPORT

A 5-year-old girl with no previous history of trauma presented with febrile posterior cervico-occipital pain for three weeks. The child was diagnosed as having meningitis after positive lumbar puncture and was then treated with adapted antibiotics for a similar period.

The persistence of the cervico-occipital pain was astonishing. Brain Computed tomography was normal and Magnetic Resonance (MR) Imaging of neuraxes revealed an enhanced subdural extra medullary mass developed from the posterior cerebellar fossa to the third cervical vertebra. The lesion

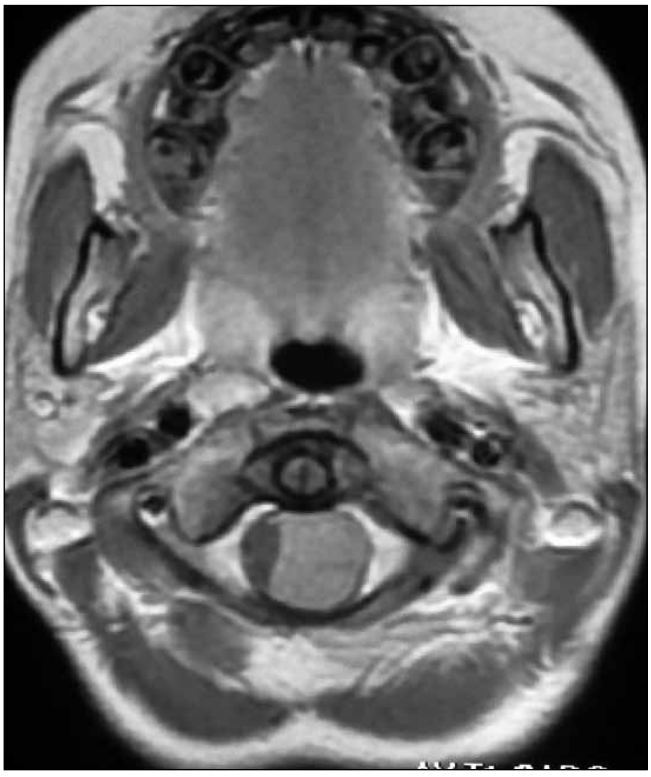

Figure 1: Preoperative contrast-enhanced axial T1-weighted MR image shows important tumor enhancement and cerebrospinal tract compression.

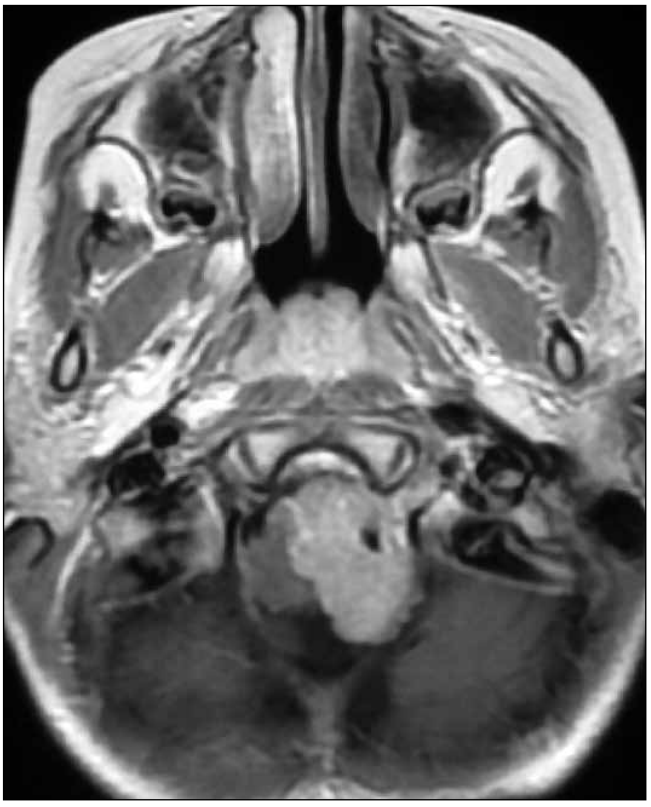

Figure 2: Preoperative contrast-enhanced axial T1-weighted MR image shows envelopment of the left vertebral artery.

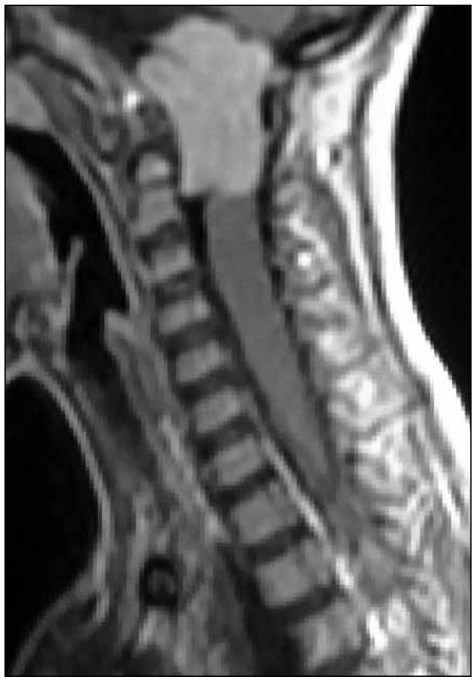

Figure 3: Preoperative contrast-enhanced sagittal T1-weighted MR image shows limits of the tumor to the cervico-occipital junction and mass effect.

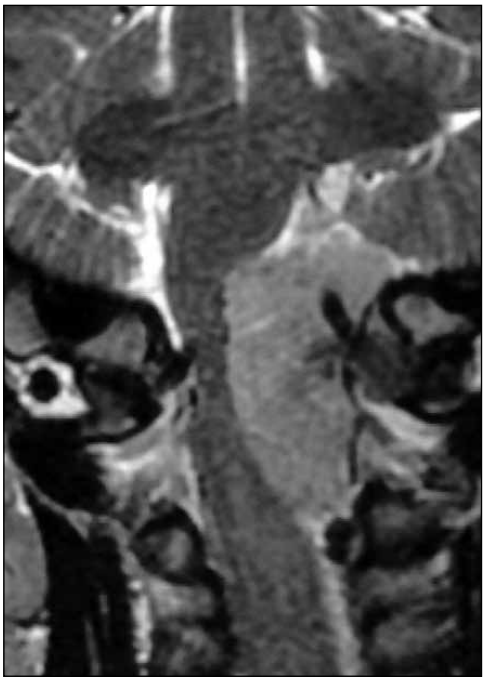

Figure 4: Preoperative coronal T2-weighted MR image shows slightly hyper intense tumor with important cerebrospinal tract compression.

enveloped the left vertebral artery that was slightly thickened and was responsible for a significant compression of the cerebrospinal tract (Figure 1, 2, 3, 4).

On physical examination, the patient had full muscle strength; she had no sensory deficit, pathologic reflexes, or long tract sign. The cranial nerves examination was also normal.

The patient was operated on quickly. During the surgery we opted for a right "park bench" surgical positioning. Then a partial left laminectomy of C1 and C2 associated to a 1.5 cm high opening of the foramen magnum was conducted. An inversed "L" dura incision exposed well the tumor that was in a close relation with several neural rootlets and small medullary arteries. The microsurgical resection was subtotal and we surprisingly found that the tumor was strongly attached to the dura at the entry hole of the left vertebral artery. Only a very thin tumor tissue was left there in order to avoid any accidental injury of the artery.

Immediately after surgery the patient noted a significant improvement of her pain and at the most recent follow up (two years after surgery) she was symptom free with no radiological recurrence.

DISCUSSION

Meningiomas are most commonly encountered after the second decade of life. However, they may occur at any age or even during fetal development. A recent report from the Central Brain Tumor Registry of the United States reports that only 2.5 % of all primary pediatric central nervous system tumors were meningiomas (3) and in this sub group the location of the tumor in the cervico-occipital region is far from being frequent and accounts for only 3.7% of all cases according to a recent study (1).

The incidence of meningiomas increases with age and more cases are reported in the second decade compared to the first. Another standpoint is that in contrast to adult meningiomas, there is no female preponderance among pediatric cases (1, 3).

Childhood meningiomas are characteristically known to have nonspecific symptoms and diagnosis is often difficult. The elasticity of the skull and non-cooperation among children compounds the problem. In the case of meningiomas developing within the cervico occipital junction, signs of increased intra cranial pressure are the most frequent (3).

In our case the child noted only cervico-occipital chronic pain that was not responsive to treatment.

Literature review regarding meningiomas of the cervico-occipital junction in children is poor, mainly because of the rarity of meningiomas in pediatric population first and of the location second.

Children who present with intradural extra medullary enhanced lesions located at the cervico-occipital junction can be expected to have schwannomas or neurofibromas because of their frequency and the diagnosis of meningioma is considered in the differential.

MR characteristics of pediatric meningiomas are similar to adult meningiomas. On MR imaging, the tumors are usually isointense to hypo intense on T1, iso to hyper intense on T2 and exhibit important enhancement (3).

The presence of a dural tail sign on MR is not obvious in all examples of pediatric meningiomas that make the radiological differential diagnosis with schwannomas difficult.

In our case the radiological features were strongly suggestive of a schwannoma.

Aggressive surgical treatment carried out in staged operations when necessary, is the treatment of choice in most cases, allowing complete excision of the tumor in about 70-80 % of cases.

Although there has been great development of the anatomical knowledge for this region, several controversies still exist regarding aspects of the operation (5).

Concerning the operative positioning of the patient, the sitting position should decompress the neuraxes upon opening of the posterior bone and dura.

Rich venous plexus is present around the cervical muscles, vertebral artery, and occipital bone leading to a potential risk of air embolism in the sitting position but not in the lateral oblique position.

In our case we opted for a right "park bench" position and the enlarged unilateral approach was sufficient to give us full view of the tumor.

The relationship of the meningioma and vertebral artery is important for surgical strategies and prognosis.

The MR Imaging showed a full encasement of the left vertebral artery by the tumor and it was clear that the protection of this artery as well as the lower cranial nerves is of great importance. During the surgery we performed a progressive microsurgical tumor excision and we only left a small portion strongly attached to the dura at the entry hole of the vertebral artery. This is in our opinion wise behavior in order to avoid any damage to the artery.

Better histological delineation has allowed the identification of highly aggressive meningeal neoplasms.

Most pediatric meningiomas are WHO grade I (80.6 %) with WHO grade II accounting for 10.4% and III for 8.1% (2).

In our case the pathological study confirmed the diagnosis of WHO grade I meningioma that was predicted according to the peroperative findings.

Post-operative management is as important as surgery. Recently, Thuijs et al. (4) found that that the biological behavior of pediatric meningiomas is more aggressive than that of its adult counterparts but guidelines for radiotherapy in children are limited.

There has been no surgical or neurological complication recorded immediately after surgery and, at last follow up (two

years after surgery), the patient was symptom-free with no radiological recurrence.

CONCLUSION

Meningiomas of the cervico-occipital region in childhood are relatively scarce with an unspecific clinical presentation. These tumors should be considered in the differential diagnosis of an intradural extra medullary tumor.

Surgery remains the gold standard with an increased safety and a high rate of complete removal especially with a unilateral approach in this particular region and at that age.

There is no sufficient evidence for the use of any post-operative radiotherapy in case of tumor remnants.

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