Article

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Intracranial – extracranial meningioma: a case report of a rare entity

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Abstract: Meningiomas are common benign intracranial tumors. However, intracranial meningiomas with extracranial extension are exceedingly rare lesions with only a few reported cases in the literature. We report a case of an 86 years old female patient presented with enlarging mass over left parietal region for last 3 years. Besides cosmetic, she had no complaints. Head CT scan with bone window showed a large tumor consisting of intracranial and extracranial part, suggestive for meningioma. Patient was operated, and both parts of tumor were removed. Taking into account surgery indication and patient's age, bone flap was returned. The postoperative course was uneventful and patient fully recovered. In the case of epicranial tumor attached to the underlying bone, meningioma should be excluded.

Key words: Meningioma, Intracranial – extracranial meningioma, Epicranial tumor

Introduction

Meningiomas accounts for about 20-30% of all intracranial tumors. However, intracranial meningeomas with extracranial extension are exceedingly uncommon lesions with only a few cases reported in literature (1). We report a case of an 86 years old female patient with large intracranial meningioma with extracranial extension.

Case report

A 86 years old female patient referred to our neurosurgery department due to progressive epicranial growth over left parietal region for last 3 years. She had only complaint on cosmetic appearance. Patient neurological examination was normal. Local status revealed a hard growth over left parietal region with intact overlying skin. Patient had a positive history of treated hypertension. Brain CT scan with bone window revealed a large tumor

consisting of intracranial and extracranial parts involving left parietal region (figure 1). The tumor was invading the brain tissue, but there was no significant edema. Patient underwent operation and first the extracranial part was removed, after which parietal craniotomy was performed. Dura with overlying tumor was cropped and subdural part of tumor was separated from the underlying brain parenchyma. Part of tumor attached to bone flap was trimmed and taking account indication of surgery, as well as patient's age, bone flap was returned to place (figure 2). Histopathological study of the removed tumor confirmed the diagnosis of meningioma (WHO gradus I). The postoperative course was uneventful, and patient fully recovered. The control CT scan showed perfect position of bone flap, without signs of tumor rest (figure 3). Patient was discharged on 10th day after surgery. Follow up after 6 months was within normal expectations.

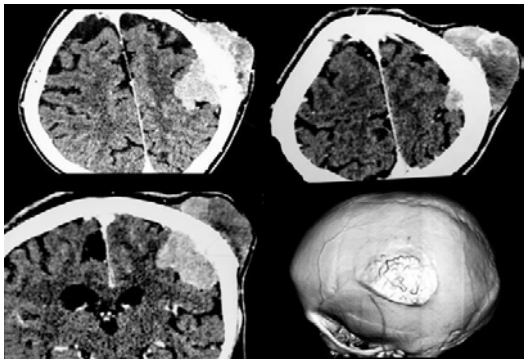

Figure 1 - Brain CT scan with bone window showing a large tumor consisting of intracranial and extracranial parts involving left parietal region

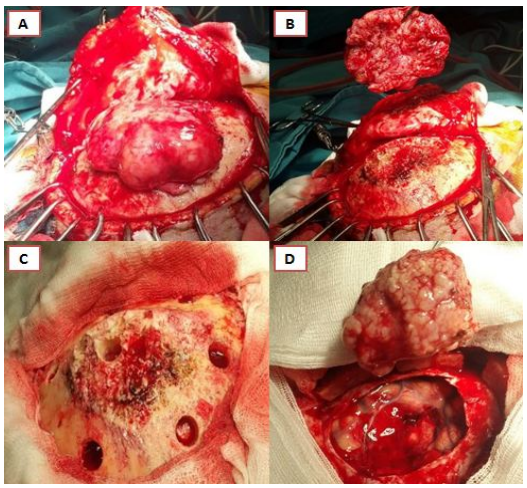

Figure 2 - Intaroperative view showing: (A) extracranial part of the tumor, (B) resection of extracranial part of the tumor, (C) bone flap after removal of extracranial tumor part, (D) resection of dura with intracranial (subdural) part of tumor

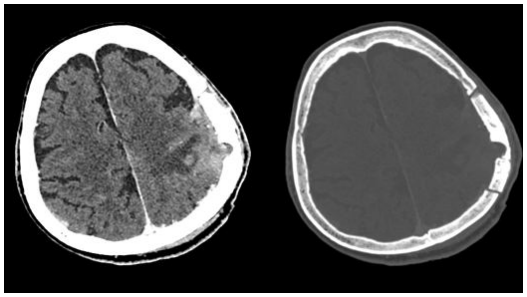

Figure 3 - The control brain CT scan

Discussion

Meningiomas account for about 20% to 30% of all intracranial tumors (1). The incidence rate of meningiomas is about 2.6/100,000 populations, being more frequent in female patients. Meningiomas incidence increases with age. Meningiomas typically have benign histological findings (5). This is in accordance with our case, since we presented a female patient with large meningioma which showed benign histopathological finding.

However, meningiomas with extracranial extension are rare lesions, with only a few cases reported in literature (6, 7).

According to study of Darweesh Al-khawaja et al, the locations of the extracranial meningiomas were: parietal region in 21 cases (31.8%), frontal region in 16 cases (24.2%), temporal area in seven cases (10.6%), occipital area in four cases (6.1%), frontoparietal in 11 cases (16.7%), frontotemporal in three cases (4.5%), temporoparietal in two cases (3.0%), and diffuse in two cases (3.0%) (2). Our patient had most usual site of tumor origin because tumor was located in the left parietal region.

Typically symptoms of meningioma are not specific, and usually are associated with seizures, headache, paresis, personality changes and visual impairment. The diagnosis is made through brain CT or MRI scan (3). Our patient only had complaint on cosmetic appearance, and her neurological examination was normal. In the case of progressive epicranial tumor growth, clinical exam and neuro-radiological evaluation are necessary even in the absence of neurological deficit or severe clinical symptoms.

As mentioned above, extracranial meningiomas are rare neoplasm's, accounting for less than 2% of all meningiomas, but are most often found in the head. The relative rarity, in addition to inconsistencies in nomenclature, limits the study of these tumors. Historically, they have been referred to as extracranial, extradural, extraneuraxial, and ectopic neoplasm's arising outside of the central nervous system. The most widely used classification of extracranial meningiomas was proposed by Hoye et al. (Table 1) (4).

TABLE 1

The most widely used classification of extracranial meningiomas

HOYE CLASSIFICATION SYSTEM FOR EXTRACRANIAL MENINGIOMAS DELINEATED BASED ON SITE OF ORIGIN (4)	
Type A	extracranial extension of meningioma with intracranial origin (secondary)
Type B	extracranial extension of meningioma arising from skull base foramen (primary)
Type C	ectopic tumor without connection to foramen, cranial nerve, or intracranial extension (primary)
Type D	extracranial metastasis with documented intracranial lesion (secondary)

In this case, the method of elimination led as to the conclusion that our patient had type A extracranial meningioma. Also, the tumor was firmly attached to the dura, with a large base, and thus probable origin of tumor was from this site. Taking into account indication of surgery (cosmetic appearance), patients age, as well as assumption that tumor originate from underlining dura, bone flap was returned to place due to better cosmetic appearance without much fear of tumor recurrence from the arachnoid cap cells which can be trapped in the bone flap or cranial sutures. Since follow up after 6 months was within normal expectations without signs of tumor recurrence, our hypothesis that patient had

type A extradural meningioma (extracranial extension of meningioma with intracranial origin) was probably right, but since this neoplasms are slowly growing tumors, further checkups are required.

In the case of extracranial meningiomas, surgical resection is the treatment of choice. If there is doubt about complete resection, the lesion should be followed with appropriate neuroimaging studies. In the case of recurrence, inoperability because of medical comorbidities, and in the case of incomplete resection, radiotherapy is recommended. Also, in all cases of WHO grade II and III lesions, adjuvant radiotherapy is suggested (2). In our patient gross total resection was achieved without need for adjuvant radiotherapy.

In the case of epicranial tumor growth, especially if attached to the underlying bone, meningioma should be excluded even in the absence of severe symptoms or neurological deficit. Surgical resection is the treatment of choice with adjuvant radiotherapy in some cases.

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