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A case report.

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Meningioma en plaque. A case report

Meningiomas as anatomo-pathological entities were first described in the sixteenth century, but it was Harvey Cusching who in 1922 proposed to identify them with the term "meningioma".
They are the most common intracranial tumors, occur between middle and late age and prefer female sex. The symptoms depend on their position with respect to the brain regions. These are mostly benign neoplasms, but there are atypical and even malignant forms. Plaque meningioma refers to a specific macroscopic aspect of the meningioma, due to diffuse dural involvement and usually with extracranial extension. We report here a case of plaque convex meningioma with typical grade I.

Key Words: En plaque, Meningioma, WHO Meningioma

Introduction

Meningiomas were fisrt described in the 16th century; Harvey Cushing coined the term "Meningioma" in the 1922.
Meningiomas are 15-30% of intracranial tumors, with a higher incidence in women (F:M = 2:1).
The highest incidence is in patients over 65 years, while they are rare in children.
The meningiomas originate from the cells of the arachnoid villi and, in order of frequency, they are localized to the convexity, to the cranial base and in the posterior cranial fossa [1].
According to the classification WHO 2007 we distinguish meningiomas WHO grade I, benign; Atypical Meningiomas WHO grade II, characterized by high mitotic activity; Anaplastic meningiomas WHO grade III, malignant.

En Plaque meningioma is a rare type of meningioma characterized by infiltrative nature, sheet-like growth and times invading the bone.
Meningiomas-en-plaque (MEP) comprise 2.5% of all meningiomas. While they typically arise in the sphenoid wing, convexity MEP are comparatively rare and are often confused with meningeal sarcoidosis, osteoma, tuberculoma, or fibrous dysplasia, with very little information published in the literature.
En plaque meningioma is an uncommon but distinct entity which shows carpet-like proliferation along the dura mater, in contrast to ordinary, massive meningiomas occurring as an intracranial mass with broad attachment to the dura mater. En plaque meningiomas typically develop in the spheno-orbital regions and manifest as ocular motor paresis, visual impairment, and proptosis, whereas those arising in the convexity are rare. Hyperostotic changes of the affected skull are thought to be characteristic findings of en plaque meningioma but may be confused with other pathological conditions such as fibrous dysplasia, osteoma, primary meningeal fibrosarcoma, and meningeal sarcoidosis [2,4,7]. The molecular genetics of meningiomas have recently been investigated but still far from a systematic understanding [1,8].

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Here we describe a case of a plaque meningioma of the convexity presented with peculiar extracranial extensions identified by histological examination [1,16].
We describe our surgical experience on the treatment of this rare type od Meningioma.

Case Report

We report our experience with a patient, aged 45, who came to our attention for persistent headache.
Neurological examination at the entrance showed no significative alterations.
The physical examination showed a swelling in the left frontal-temporal area of hard consistency.
Neuroradiological investigations (CT and MRI) showed the presence of a massive bone lesion at the level of the left sphenoid wing (Figs. 1, 2, 3).

The patient was then subjected to surgical intervention with excision of the lesion described, using a left frontal-temporal approach (Fig. 4).
The dura mater, also thickened, was then removed (Figs. 5-6).
Finally, cranioplasty was performed using Methyl methacrylate material (Fig. 7).
The histological examination of the lesion referred to Meningioma En Plaque (Fig. 10).
The neuroradiological follow-up at 3 years (CT and MRI with contrast medium) have not shown, to date, signs of recurrence of the lesion (Figs. 8-9).

Discussion

Meningioma En Plaque (MEP) is characterized by a remarkable disproportion between meningeal tumor and

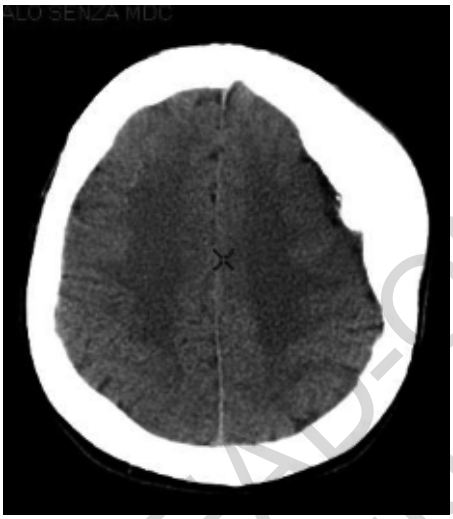

Fig. 1: Pre-operative axial CT scan A.

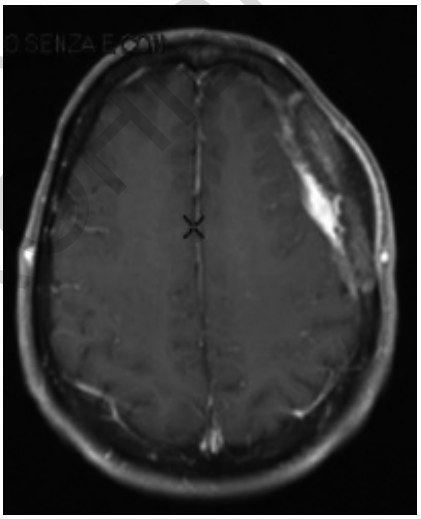

Fig. 3: Pre operative axial T1 MRI image.

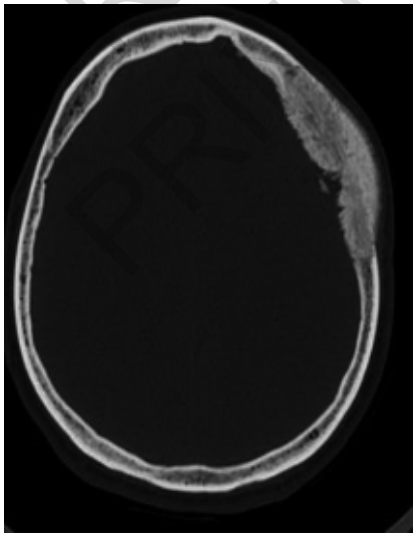

Fig. 2: Preopertive axial bone CT scanMa.

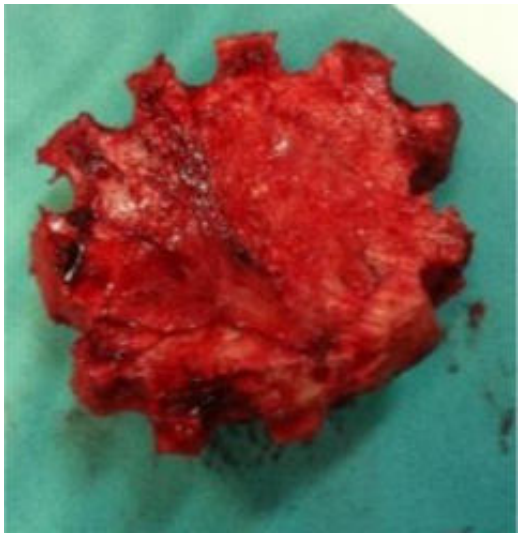

Fig. 4: Bone flap with more burr hole.

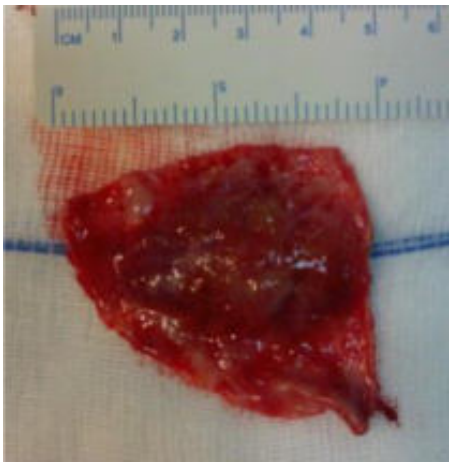

Fig. 5: Extension and infiltration of dura mater.

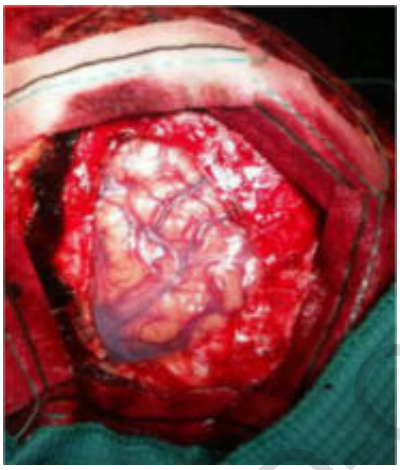

Fig. 6: brain after removal meningiomas.

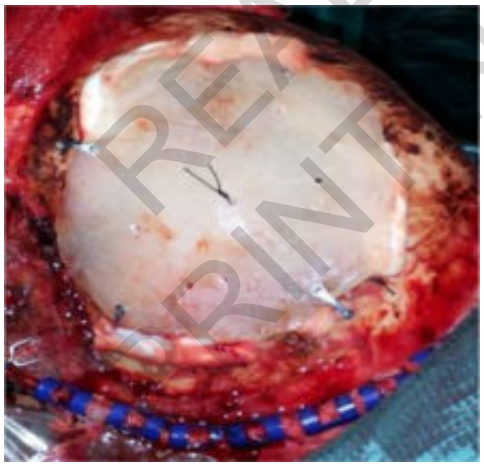

Fig. 7: cranioplastic with methyl methacrylate material.

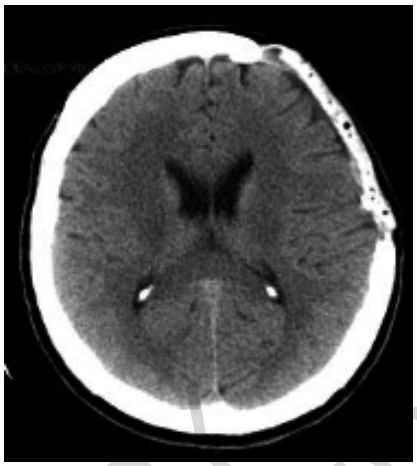

Fig. 8: post-operative axial CT scan.

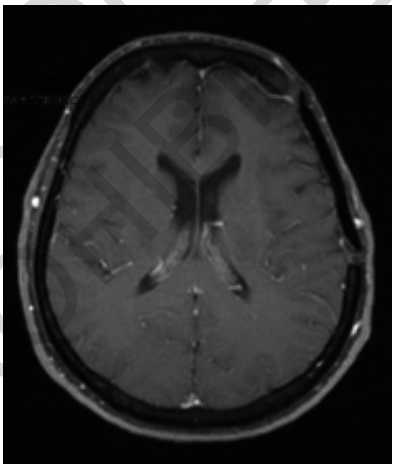

Fig. 9: T! post-operative MRI.

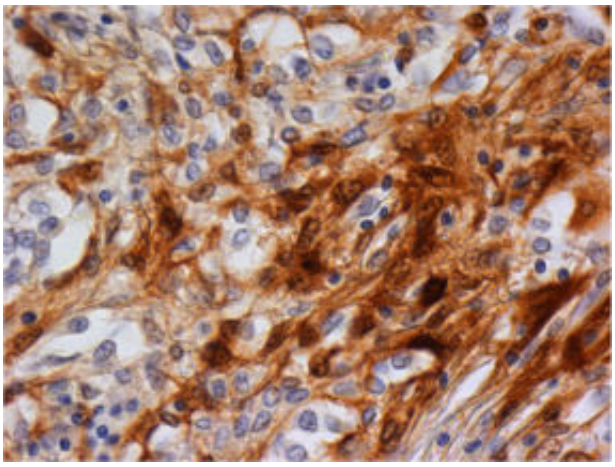

Fig. 10: ema X 400 immunohistochemistry.

bone reaction. The etiopathogenesis is still discussed.
This type of neoformation represents about 4% of intracranial meningiomas [2].
It occurs more often at the level of the sphenoid wing, with a thin extension on the inner surface of the hard and scarce intracranial expansion.

The extensive hyperostosis is evident, which can sometimes affect the sphenoid wing, the lateral orbit wall, the temporal bone and the external surface of the middle cranial fossa [3,4].
In addition to an accurate physical examination, the diagnosis is performed by CT and MRI.

Differential diagnosis involves osteoma-like lesions, fibrous dysplasia and osteoblastic metastases [5]. Surgery should be performed early.
The surgical approach must aim, as far as possible, at the total resection of macroscopically infiltrated bone [6]. Sometimes the therapeutic course, where necessary, can be completed by radiotherapy treatment.
The precise mechanism of how MEP induced hyperostosis is as yet unclear, though it is traditionally characterized by slow growth and sclerotic change in the overlying bone. The success in achieving total surgical resection of MEP with no tumor recurrence depends largely on its location and the extent of bone invasion.
The diffulty of complete resection of MEP is well documented in the literature for sphenoid wing MEP and is often acribed to extensive dural and calvarial involvement as well as the tumor's proximity to vital structures. Recurrence of MEP is common, reported to be around 25-50% for sphenoid wing MEP.
In the case of partial or subtotal resection, the current school of thougt is to treat residual tumor with radioterapy or conventional multibeam radiation.
The use of radiotehrapy to treat residual tumor is debated in the literature.
Some authors only recommend radiation treatment in the case of atypical ore malignant, MEP if there is dural or cavernous sinus invasion, or if follow-up imaging indicates recurrence [16].

Conclusion

Meningioma En Plaque is a rare but benign lesion. The molecular genetics of massive meningiomas have recently been investigated, while the genetic peculiarities of en plaque meningiomas are little known. Differences in the genetic background between these two entities are also not known. In the present case, en plaque and subcutaneous, massive meningiomas occurred simultaneously in the same patient. Furthermore, in contrast with the macroscopically intact inner table adjacent to the en plaque lesion, the outer table was eroded by the massive tumor overlying it. To date, we do not explain the underlying mechanism of them. Further investigation is needed to understand the biological behavior of meningiomas.
Total resection is the rule for the treatment of low-grade meningiomas. In the present case, en bloc tumour resection involving the affected dura mater, skull, and temporalis muscle was achieved. Meningiomas should be assumed as differential diagnosis of subcutaneous mass lesion even in the absence of an accompaniment of intracranial tumor growth. The best results in terms of complete resection of the pathology and clinical outcome are obtained when the diagnosis and treatment are carried out early. All the involved bone shoul be removed to prevent recurrence .

Riassunto

I meningiomi come entità anatomo-patologiche furono descritti per la prima volta nel XVI secolo, ma fu Harvey Cusching che nel 1922 propose di identificarli con il termine di "meningioma".
Sono i tumori intracranici più comuni, si manifestano tra la mezza e la tarda età e prediligono il sesso femminile. La sintomatologia è funzione della loro posizione rispetto alle regioni dell'encefalo. Si tratta per lo più di neoplasie benigne, ma esistono forme atipiche ed anche maligne. Il meningioma "a placca" si riferisce a un aspetto macroscopico specifico del meningioma, per coinvolgimento durale diffuso e solitamente con estensione extracranica.
Riportiamo qui un caso di meningioma convessità en placca con tipico grado I.

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