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CASE STUDY

Extracranial meningioma of the paranasal sinuses

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KEYWORDS
Extracranial meningioma;
Paranasal sinuses;
Unusual localization;
Surgical approach

Abstract
Extracraneal meningiomas are infrequent and their localization in the paranasal sinuses is rare. We present the case of a patient with a primary meningioma of the paranasal sinuses. We describe the surgical approach and review the current literature.

PALABRAS CLAVE
Meningioma extracraneal;
Senos paranasales;
Localización inusual;
Abordaje quirúrgico

Meningioma primario extracraneal de senos paranasales

Resumen
Los meningiomas extracraneales son poco frecuentes y es rara su localización en los senos paranasales. Presentamos el caso de un paciente con un meningioma primario de senos paranasales. Describimos el abordaje quirúrgico realizado y revisamos la literatura actual al respecto.

Introduction

Meningiomas tend to settle at the level of the central nervous system; their extracranial location is very rare. We describe the case of a patient who presented a primary extracranial meningioma at the level of the paranasal sinuses. This unusual location forced us to use a combined surgical approach to achieve complete excision of the lesion. Subsequently, we reviewed the literature on this respect.

Case study

Forty-eight-year old male patient who attended consultation due to a right nostril respiratory failure of 3 months evolution. As prior pathological history of interest, he referred that at age 12 he was operated on due to a chromophobe pituitary adenoma that was accessed through a right temporal craniotomy; a right amaurosis and a left visual deficit from

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chiasmatic alteration as well as panhypopituitarism were left as sequelae.

CT and MRI showed a mass occupying almost the entirety of the right nostril, including maxillary, sphenoid and ethmoid sinuses, without affecting the dura mater or endocranial structures. This caused the erosion of the medial wall of the right maxillary sinus as well as orbital floor and the lamina papyracea. Biopsy carried out under endoscopic control reported a grade I transitional nasal meningioma with low mitotic activity. It was decided to perform surgery together with the Maxillofacial Surgery and Neurosurgery Services, using a combined approach path: coronal and right paralateronasal craniotomy (Figures 1-2). After medial maxillectomy, the excision of the tumour and the adjacent paranasal structures, we reconstructed the orbit and maxilla with titanium screws and No. 3 plates.

One year after the surgical intervention, the patient is free from disease.

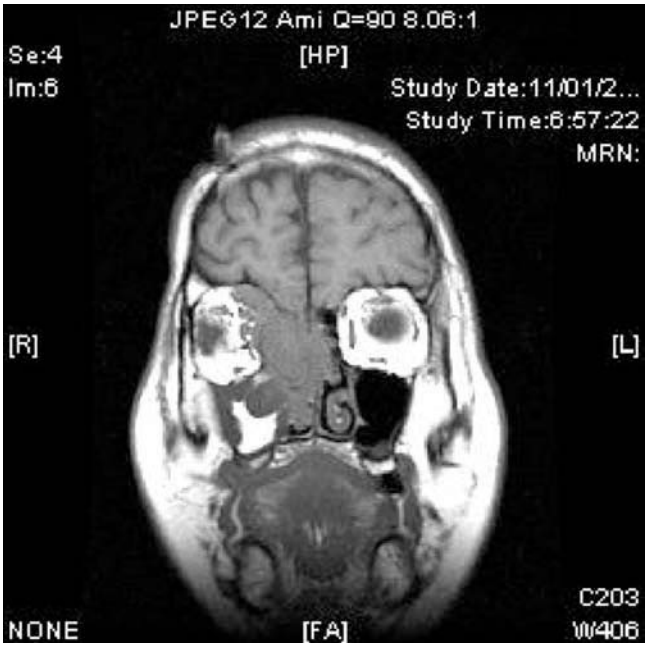

Figure 1 MRI. Involvement of the maxillary sinus, orbit and right nasal fossa by the meningioma.

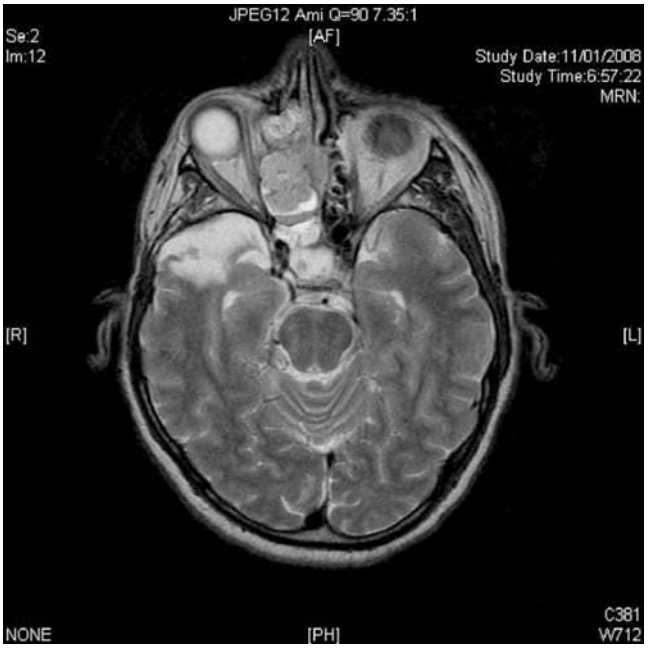

Figure 2 MRI. Axial section. Tumour occupation of the right ethmoid.

Discussion

Meningiomas are benign, slow-growing tumours, usually intracranial; they represent between 13% and 26% of all CNS tumours. They usually appear between the 4th and the 6th decade of life and are rare in the paediatric age, with a clear predominance in women (75% of cases).

Extracranial location accounts for 1% to 2% of all these tumours[2,3] and has been reported more frequently in men and in young individuals.[4]

They are considered primary when they originate independently and secondary when they show a direct communication with the intracranial region.[5] Most extracranial meningiomas are secondary, since up to 20% of the intracranial cases present extracranial extension.[6,7] Histologically, primary extracranial meningiomas do not differ from intracranial.

Most of these tumours are sporadic and the aetiology remains unclear.[5,8]

In very specific cases, it is possible to identify a possible cause, such as exposure to radiation, neurofibromatosis type II, abnormalities of chromosome 22, exposure to a virus of the adenovirus type (among others), hormonal factors, etc. There are several hypotheses about its origin, which derive from arachnoid cells of the cranial nerves[9] or ectopic meningocytes.[10]

In general, the most common signs and symptoms of paranasal sinus meningiomas may mimic cases of sinusitis with nasal obstruction, anosmia, facial pressure or pain, epistaxis and rhinorrhea.[11,12] Up to 10% of cases may remain asymptomatic even in advanced stages. Clinical examination should be comprehensive and include nasal endoscopy, which usually reveals a firm mass, pink to grey in colour, globular or lobulated but well circumscribed, with displacement but without infiltration of surrounding tissue.[13]

The differential diagnosis should include mucocele, olfactory neuroblastoma, carcinoma, haemangioma, sarcoma and angiofibroma.[14,15] Histology is therefore essential and four patterns are described for clinical purposes: syncytial (meningothelial), fibroblastic, transitional (as in our case) and angioblastic.[2,13] The majority of primary extracranial meningiomas of the paranasal sinuses are of the meningothelial type.[7]

Immunohistochemistry is helpful in confirming the diagnosis.[5,13] These tumours tend to show strong positivity towards vimentin and epithelial membrane antigen (EMA), as indeed occurred in our patient, and are focally positive for s-100, keratin and CEA.[5,16,17]

Both CT and MRI are essential in preoperative surgical planning.[2] Surgery is the only curative treatment; radiation therapy is therefore reserved as a palliative approach.[15,18-20]

Complete surgical resection of the lesion may be performed through endoscopy, open surgery or a combination of both.[21]

The recurrence rate varies according to histological subtypes, with worse prognosis for the angioblastic variant as resection is more difficult and recurrence is therefore more frequent.[13] The prognosis is generally good. In cases of tumour recurrence, it usually appears in the same location as the initial case, probably representing residual disease more than tumour recurrence.[14]

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