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Case Report

Proptosis Secondary to Meningioma of the Sphenoid Wing

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Abstract

Objective: Presentation of a clinical case of a female patient with meningioma in the sphenoid wing with ophthalmological manifestations. Spheno-orbital meningiomas are rare intracranial tumors that arise in the sphenoid wing. These tumors can invade important neurovascular structures making radical resection difficult, and residual tumors often lead to recurrence.

Summary: 44-year-old female patient referred by a clinician for proptosis of 6 months of evolution. The ophthalmological examination presented proptosis in the right eye, with inferior projection, preserved ocular motility in 9 quadrants, with diplopia in extreme superior and extreme lateral gaze. Visual acuity 10/10 both eyes. Computerized visual field od altered oi normal. Normal eye fundus at the time of examination. Brain MRI is requested where a marked thickening and heterogeneous signal, predominantly hypotensive in T2, are reported. This alteration conditioned compression and displacement of the external rectum. Interconsultation with neurosurgery is performed and diagnostic and therapeutic resection is planned.

Pathology reports meningioma meningotheliomatous intraosseous grade 1 of the sphenoid wing.

On postoperative ophthalmological examination, the patient presented no proptosis, preserved motility in 9 quadrants without diplopia, VA 10/10, normal computerized visual field in both eyes.

At six months post-surgery, there are no imaging or clinical signs of recurrence.

Conclusion: The maximum safe resection with preservation of function continues to be the most important prognostic factor associated with lower recurrence rates and the performance at the right time of surgery reduces permanent ophthalmological alterations.

Keywords: *Sphenoid Meningioma; Proptosis; Visual Disability*

Introduction

Meningiomas are dural-based tumors that arise from meningothelial cells (Arachnoid cells of the "layer") [1]. They represent 37.6% of all primary brain tumors in adults, making them the most common type of intracranial tumor [2]. The incidence of meningioma increases with age, with a median age of diagnosis of 65 years [3]. Meningiomas are usually slow-growing and non-infiltrating, and their symptoms are variable and depend on their location. Plaque meningioma (EPM) is a rare type of meningioma defined by a sheet-shaped lesion that infiltrates the dura mater and sometimes invades the bone [4]. Most reported cases of EPM are WHO grade I, with very few cases being grade II or III [5].

Despite great advances in surgical procedures, meningioma plaque sphenoorbital remains a serious problem. MEP comprises between 2% and 9% of all meningiomas and up to 18% of sphenoid wing meningiomas [6]. It is 3 to 6 times more prevalent in women and usually appears in people between 40 and 50 years of age [7]. Plaque meningioma was first introduced by Cushing and Eisenhardt in 1938 to describe carpet-like tumor growth associated with hyperostosis, most commonly seen on the sphenoid ridge with orbital involvement [8]. Proptosis is the most common symptom of the disease, although symptoms such as headache, ptosis, diplopia, and unilateral visual impairment due to compression of the optic nerve by canal stenosis [9].

The diagnosis is made by computed tomography (CT) with coronal and axial slices, these scans are the best slices to observe bone hyperostosis. Also, the intradural region, optic canal, and superior orbital fissure (SOF) involvement can be examined using magnetic resonance imaging (MRI) [10].

Treatment of AEM includes observation with follow-up scans at intervals, microsurgery, radiosurgery, and/or radiation therapy. Given the increasing incidence of incidental discovery of these tumors while imaging is ordered for other reasons, most tumors are initially simply followed until there is: (1) documented growth and/or; (2) development of tumor site-specific symptoms. In the absence of a history of other systemic tumor involvement, the interval for imaging is two 6-month scans followed by annual imaging. In a patient with another systemic cancer, a shorter scan of 2 to 3 months is needed to help differentiate a benign from a more aggressive condition based on rapidity of growth [11].

Surgery is the best treatment for these lesions, used for the total resection of the tumor and the affected dura [12]; the goal of total removal of the tumor when feasible is the main therapeutic strategy for meningioma. The extent of the resection has a great impact on the rate of recurrence.

Conventional radiotherapy or radiosurgery has been introduced as an alternative or ancillary therapy in cases where total tumor resection is not possible. However, due to the proximity of the tumor to the optic nerve and the vital elements involved, there is a risk of considerable complications [13].

Clinical Case

44-year-old female patient referred by a clinician for proptosis of 6 months of evolution. The ophthalmological examination presented proptosis of the right eye with progressive growth, non-painful, with inferior projection, non-pulsatile. Preserved ocular motility in 9 quadrants, with diplopia in extreme superior and lateral gaze. Visual acuity 10/10 both eyes without correction. Computed visual field od peripheral scotomatal defect oi normal. Normal eye fundus at the time of examination. Brain MRI is requested where a marked thickening and heterogeneous signal, predominantly hypotensive in T2, are reported. This alteration conditioned compression and displacement of the external rectum. Contrast -enhanced CT reports bone destructuring with alteration of the right greater wing of the sphenoid, which presents a sclerotic lesion and bulging of the external orbital wall towards the medial side and irregular edges with a frayed appearance towards the rear. Said lesion measures 27 x 31 x 19 mm (Long x AP x Tr) and generates proptosis of the ipsilateral eyeball by displacement of the lateral rectus without invading it. It extends to the orbital extraconal fat and to the convexity of the right temporal lobe, showing marked soft tissue enhancement. It could correspond to a malignant neoformative process in the first place.

Interconsultation with neurosurgery is performed and diagnostic and therapeutic resection is planned.

It is performed under general anesthesia, supine position, with the head of the bed slightly above the heart, right pterional approach, bone platelet dissection and dura mater anchorage, it is observed in the orbit, sphenoid region with extension to the base of the skull, bleeding hyperostosis, An extensive resection is performed, involving the greater and lesser wings of the sphenoid up to SOF. The lateral

and superior walls of the orbit were then excised until the periorbita was completely exposed. The periorbita was explored, and infiltrated orbital fat was resected. The optic canal was decompressed to improve and preserve visual function.

cancellous bone is visualized in the pterional region, rupture of the dura mater by the tumor with intracranial invasion is observed, said invasion is dissected and resected with microsurgery and ultrasonic aspirator. The orbital cone, optic nerve and lateral wall of the orbit are decompressed. Titanium mesh is placed for cranial reconstruction. A forced duction test is performed to ensure that there is no impact from the orbital reconstruction on the orbital muscles.

Surgical resection of sphenoid wing meningioma with orbital invasion is technically demanding due to the difficult removal of hyperostosis with sufficient resection margin, reconstruction of the bone structure and dura mater, and preservation of important anatomical structures such as the optic nerve, the oculomotor nerve, the trigeminal nerve or the internal carotid artery [14].

According to several studies, the degree of preoperative impairment and the full extent of optic nerve decompression are the most important prognostic indicators of postoperative optic nerve function.

Pathology reports meningioma meningotheliomatous intraosseous grade 1 of the sphenoid wing.

Postoperative ophthalmological examination 15 days later, the patient presented no proptosis, preserved motility in 9 quadrants without diplopia, VA 10/10, normal computerized visual field in both eyes.

At six months post-surgery, there are no imaging or clinical signs of recurrence.

Discussion

Sphenoid wing meningiomas (SAMs) are one of the three most common tumors among intracranial meningiomas. They most commonly occur in the fourth decade of life and like meningiomas in other localities there is a female predominance. The most common symptoms at presentation are headache, seizures, and visual disturbances. The forms of plaque hyperostosis occur almost exclusively in women and present with painless unilateral proptosis, decreased visual acuity, diplopia, which can affect the aesthetic appearance and quality of life [15]. The clinical manifestations found in the literature were those presented by our patient.

Plaque hyperostosis tumors, also known as meningiomas sphenoorbital, are slow growing and most of the tumor burden is within the bone. These tumors require a more extensive fronto -temporal-orbital-zygomatic craniotomy with removal of hyperostotic bone medially to the foramen ovale and rotundum, optic canal, roof and lateral wall of the orbit, as well as infiltrated [16] periorbita.

The typical presentation is that of a middle-aged, female person with progressive and painless proptosis, as in the case presented by us. A dull periorbital pain is described. There is usually edema of the lower eyelid and complaints of double vision or blurred vision, diplopia was one of the reasons for consultation of our patient. For patients with prolonged compression of the optic nerve, there may be loss of optic nerve fibers with reduced visual acuity, impaired visual fields, altered color vision, and abnormal ocular coherence tomography (OCT), in our case the VA was normal but campimetry was affected on the side of proptosis. In extreme cases there may be problems with corneal keratitis due to exposure and visual acuity alteration, these symptoms did not occur in the case.

Mass meningioma is defined by its characteristic flat “carpet-like” growth along the bony contour. EPM can also be associated with hyperostosis as in the case of our patient. EPM arises more commonly in the sphenoid wing and orbital regions and, less frequently, along the cerebral convexity, the temporal bone and the foramen magnum, as we have already mentioned in our patient the condition was the sphenoid wing, which agrees with what was found in the literature review.

The clinical presentation of EPM depends on its location and spread.

Symptoms arise either due to direct neural compression and invasion by the tumor or due to bony hyperostosis that can narrow the foramina and fissures through which the neural structures pass. In EPM of the sphenoid wing, such neural compression can result in decreased visual acuity and visual field defects. Hyperostosis of the orbital bones can result in proptosis, some patients also complain of retrobulbar pressure, orbital pain, and headache [17]. The degree of proptosis can be measured by the ophthalmologist directly, and axial magnetic resonance imaging can help calculate the exophthalmos index, that is, the relationship between the position of the eyes on the affected side with that on the normal side. The distance of the eye position is measured from the lens to a horizontal line drawn between the posterior part of the frontozygomatic process at the same level. This index can be followed after treatment to assess response and look for redevelopment that might suggest a recurrent tumor. At the time of consultation, the patient already presented all the symptoms of compression and orbital occupation, this was the reason why surgical intervention was decided instead of controls with tomography and resonance, as recommended by some authors who emphasize the strategy of " wait and see" due to its slow growth rate 0.3 cm^3 [18].

EPM presents a diagnostic challenge due to its unusual radiological appearance. First-line imaging studies include computed tomography (CT) to delineate bony involvement and bony hyperostosis, which is seen in 13% to 49% of EPM [19]; and magnetic resonance imaging (MRI) to identify dural and intradural involvement. Suppressed T1-weighted MRI allows assessment of orbital and fat invasion with contrast. Hyperostosis is often seen in a periosteal pattern with irregularity on the surface of the structures involved and inward bulging of the lesion [20]. All these imaging signs were found in those performed on the patient.

Regarding the reconstruction of the walls of the orbit, we carry it out to avoid the possibility of post-surgical enophthalmos or exophthalmos.

The complications of EAM resection found were blindness, limitation of extraocular movement, facial numbness, cerebrospinal fluid leak and exposure of the cranial fixation plate with wound infection [13,21]. To date, our patient only reports hyperesthesia on the scalp.

Despite the locally invasive nature, most EPM cases are still classified as WHO grade I tumors due to a low proliferative index [22]. The pathology report of our patient was grade 1.

Conclusion

Maximum safe resection with preservation of function remains the most important prognostic factor associated with lower recurrence rates, and timely surgery decreases permanent ophthalmologic abnormalities.

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