Fronto-orbital osteoma. Answer to the e-quiz ‘‘Unilateral exophthalmos in a 30-year-old man’’☆

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Case

Mr D.L., 30 years old, with no notable medical history, was hospitalized for right exophthalmos, which had been developing for a year and a half and had recently become painful. Clinical examination found irreducible non-axial right exophthalmos with inferior and posterior globe deviation, limitation of ocular mobility upward, and conjunctival congestion. MRI of the cerebrum and orbit was performed (Fig. 1).
What is your diagnosis?

On reading this case report, which of these diagnoses would you choose:
• meningioma of the frontal plate with orbital extension?
• enlarged frontal sinus?
• fronto-orbital osteoma?
• calcified intraorbital angioma?
• fibrous dysplasia of the orbital roof?

Diagnosis

Fronto-orbital osteoma.

Comments

The coronal MRI slices in T2 and in T1 with fat saturation (fat sat) after gadolinium injection (Fig. 2) showed a lobulate mass with two components: the larger part with a negligible signal, essentially black, and the smaller lobe with an intermediate signal in T1 and in T2 without enhancement after gadolinium injection. It began in the frontal sinus and extended into the orbit, exerting a mass effect on the eyeball, the optic nerve, and the upper right muscle. It extended at the top to the anterior cranial fossa, in displacing cerebral parenchyma without meningeal uptake of the gadolinium. This mass evokes a highly calcified process.
The patient underwent CT of the orbit (Fig. 3), which confirmed the presence of a multilobulate calcified process with two components: one dense and compact, and one with a ground-glass appearance. This process developed in the frontal sinus, extended to the anterior cranial fossa and to the orbit without any associated bone deformity. Osteoma was diagnosed. Another small osteoma of anterior right ethmoidal cells was also found (Fig. 3b).

Surgery was performed with a bifrontal approach and included a right frontal component. Fragmentation of the fronto-orbital process allowed complete excision. The pathology examination confirmed the diagnosis of mixed osteoma.

Discussion

Osteoma of the sinuses is a benign bone tumor that grows slowly and continuously and can occur at any age. Several forms exist: lobulate and pedunculated [1,2]. It is the most frequent benign tumor of the paranasal sinuses [3] and affects, in descending order of frequency, the frontal sinus, ethmoidal cells, and the maxillary sinus. The sphenoidal sinus is affected only rarely [1,4]. Often the orbital damage is only an extension from the frontal sinus or ethmoidal cells.

Fronto-orbital osteomas account for 0.4 to 5% of orbital tumors and from 0.6 to 2.5% of the causes of tumor-related exophthalmos [5].

Osteoma results from metaplasia of the connective tissue of the sinus mucosa. Three theories have been proposed to explain its onset: traumatic, infectious and embryonic. In the first, the trauma is responsible for the existence of osteoblastic activity greater than the osteoclastic activity. The second hypothesizes that infection stimulates osteogenesis. The embryonic theory postulates that this tumor develops at the borders of tissues of different embryonic origin, that is, it results from the ossification of fetal cartilage located at the junction of the frontal sinus and the ethmoidal cells, the former of bone membrane origin and the second of enchondral bone origin [1,4,5]. When several tumors are present, it is essential to look for Gardner syndrome [3,5,6].

Osteoma is thought to have three histologic types: an eburnated or ivory-like type, a more aggressive spongy type, and a mixed type [2,5—7]. Classically, sinus osteomas are asymptomatic. When they block the ostomeatal intersection, they can cause sinusitis. Depending on their size, their extension into the orbit causes various signs, including exophthalmos, diplopia, and limitation of ocular movements [1,2,5], all seen in our case.

Imaging, especially CT and MRI, makes possible not only their diagnosis but also the exhaustive staging that helps to choose the best surgical technique. In CT, osteoma, depending on its degree of mineralization, appears as a dense, polycyclic process, well defined compared with healthy bones, in contrast to the clarity of the sinuses and resembling the cortical bones in its ivory form or taking on a ground-glass appearance in its spongy form. These two aspects can coexist in the same lesion (mixed form), as in our case. MRI is less effective for studying these bone lesions: small compact osteomas might not be detected because they generate negligible signals and can thus be confused with intrasinus air [5,7]. At the same time, MRI is more accurate for the study of adjacent structures such as the optic nerve, the eyeball, and the orbit’s vascular structures and muscles.

The primary differential diagnoses for osteoma are meningioma of the frontal plate with orbital extension, an orbital focal spot of fibrous dysplasia, an ossifying fibroma, an osteoblast or Paget disease [3,5]. Small asymptomatic osteoma must be monitored annually, while bulky symptomatic osteoma must be excised surgically and completely, in view of the risks of neurological and orbital complications, on the one hand, and the risk of recurrence on the other. This resection may be performed in a single block or by fragmentation. The surgical approach depends on the osteoma stage, determined by the various imaging examinations [1,4—6].

Figure 3. Orbital CT. Right fronto-orbital process including a compact component (black arrow) and another component with a ground-glass appearance (empty arrow) exerting a mass effect on the eyeball (asterisk), the optic nerve (arrow head) and the upper right muscle. Note the presence of a small compact osteoma of right ethmoidal cells (red arrow). a and b: coronal reconstructions in the bone window; c: oblique sagittal reconstruction in the parenchymal window, including the optic nerve.
Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


