Images in Otorhinolaryngology

Symptomatic Frontal Pneumoencephaly Secondary to Fibrous Dysplasia of the Sinus

Neumoencéfalo frontal sintomático secundario a displasia fibrosa sinusal

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The patient was a 38-year-old male, suffering from headache of 2 months’ evolution, refractory to medical treatment. On clinical examination, he was conscious, with bradypsychia and without focal neurological deficits. In the following days, he showed slow neurological deterioration, with ideational slowness, motor aphasia, agraphia, acalculia and worsening of the headache.

He underwent a brain CT scan, which revealed significant pneumocephalus in the left frontal area, with occupation of both frontal sinuses by a lesion (Fig. 1).

Figure 1
We performed a bifrontal craniotomy, observing occupation of both frontal sinuses by a bone mass that invaded the posterior wall of the sinus and caused an incisive spike set into the dura mater. We proceeded to mill the fibro-osseous lesion, removing the mucosa of both sinuses and of the nasofrontal duct. Subsequently, we excluded them with a bone substitute material, temporal muscle and fibrin glue (Fig. 2). The outcome was favourable, progressing until the patient was asymptomatic.

The isolated appearance of a bone lesion defines monostotic fibrous dysplasia (75%-80% of cases); however, McCune-Albright syndrome has to be ruled out through a clinical and endocrinological study and a bone scintigraphy scan showing other locations.

The diagnosis of suspicion is provided by iconography (Rx and CT). In the latter, we can distinguish three radiographic patterns: pagetoid (the most common), sclerotic, and cystic.

Definitive diagnosis is provided by the histological study. As long as it remains asymptomatic, we should adopt an expectant attitude. Surgical treatment should only take place as a response to the symptoms, and should involve complete removal of the lesion and the release of the structures involved.

There is the possibility of malignant transformation to osteosarcoma. Therefore, and due to its high tendency to recurrence, the evolution of these patients should be followed up.