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Case report

Ethmoid tumor and oncogenic osteomalacia: Case report and review of the literature

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ABSTRACT

Introduction: Oncogenic osteomalacia is a very rare disease usually caused by a phosphaturic mesenchymal tumor, particularly the "mixed connective tissue type", secreting FGF-23 hormone.
Objective: The authors report a case of ethmoid tumor associated with oncogenic osteomalacia and discuss management based on a review of the literature.
Case summary: A 41-year-old woman with multiple fractures causing major disability was diagnosed with early-onset osteoporosis. CT scan followed by MRI, performed due to the concomitant presence of nasal obstruction, showed a right ethmoid tumor in contact with the dura mater and periorbital tissues, but with no signs of invasion. Endoscopic resection was performed with reconstruction of the defect of the cribriform plate by a nasoseptal flap. Nasal and bone symptoms subsequently resolved. Histological examination revealed a phosphaturic mesenchymal tumor.
Discussion: Twelve cases of mesenchymal tumor of the ethmoid sinus associated with oncogenic osteomalacia have been reported to date. FGF-23 assay and whole-body MRI with STIR sequence are useful for the diagnosis. A very favorable outcome is observed after surgical treatment in the majority of cases.

1. Introduction

Oncogenic osteomalacia is a very rare disease due to defective bone mineralisation. It is caused by a usually benign mesenchymal tumor secreting Fibroblast Growth Factor 23 (FGF-23) hormone. This hormone prevents phosphate reabsorption by inactivation of the sodium-phosphate pump in the proximal renal tubule [1], inducing renal phosphate leakage, associated with compensatory phosphate release by bone, resulting in bone demineralisation. The symptoms of osteomalacia, such as bone pain, muscle weakness, walking difficulties or fractures secondary to minimal trauma, are therefore nonspecific [1–2].

Oncogenic osteomalacia generally affects subjects over the age of forty, with no sex predominance [3]. The primary tumor, usually a phosphaturic mesenchymal tumor, mixed connective tissue variant (formerly called haemangiopericytoma), is situated in the head and neck in 27% of cases [1], predominantly involving the mandible and paranasal sinuses [3]. Standard treatment consists of surgical resection with adequate margins [1–2].

The objective of this study was to report a case of ethmoid tumor associated with oncogenic osteomalacia and discuss the management in the light of a review of the literature.

2. Case report

A 41-year-old woman was diagnosed with early-onset osteoporosis in a context of multiple microfractures of the ischium and femoral head, causing major disability and pain requiring step 3 analgesics. Contrast-enhanced CT scan of the facial bones, performed due to the concomitant presence of nasal obstruction, showed a polypoid, hypervascular obstructive mass of the right nasal cavity and ethmoid sinus with osteolysis of the lamina papyracea and the roof of the ethmoid sinus. This mass extended into the ipsilateral frontal recess with moderate invasion of the frontal sinus. Gadolinium-enhanced MRI of the paranasal sinuses visualized the intracranial tumor extension in contact with but not invading the dura mater. The tumor was in contact with the periorbital tissues, with no signs of intraorbital extension and no contralateral extension (Fig. 1 A and B).

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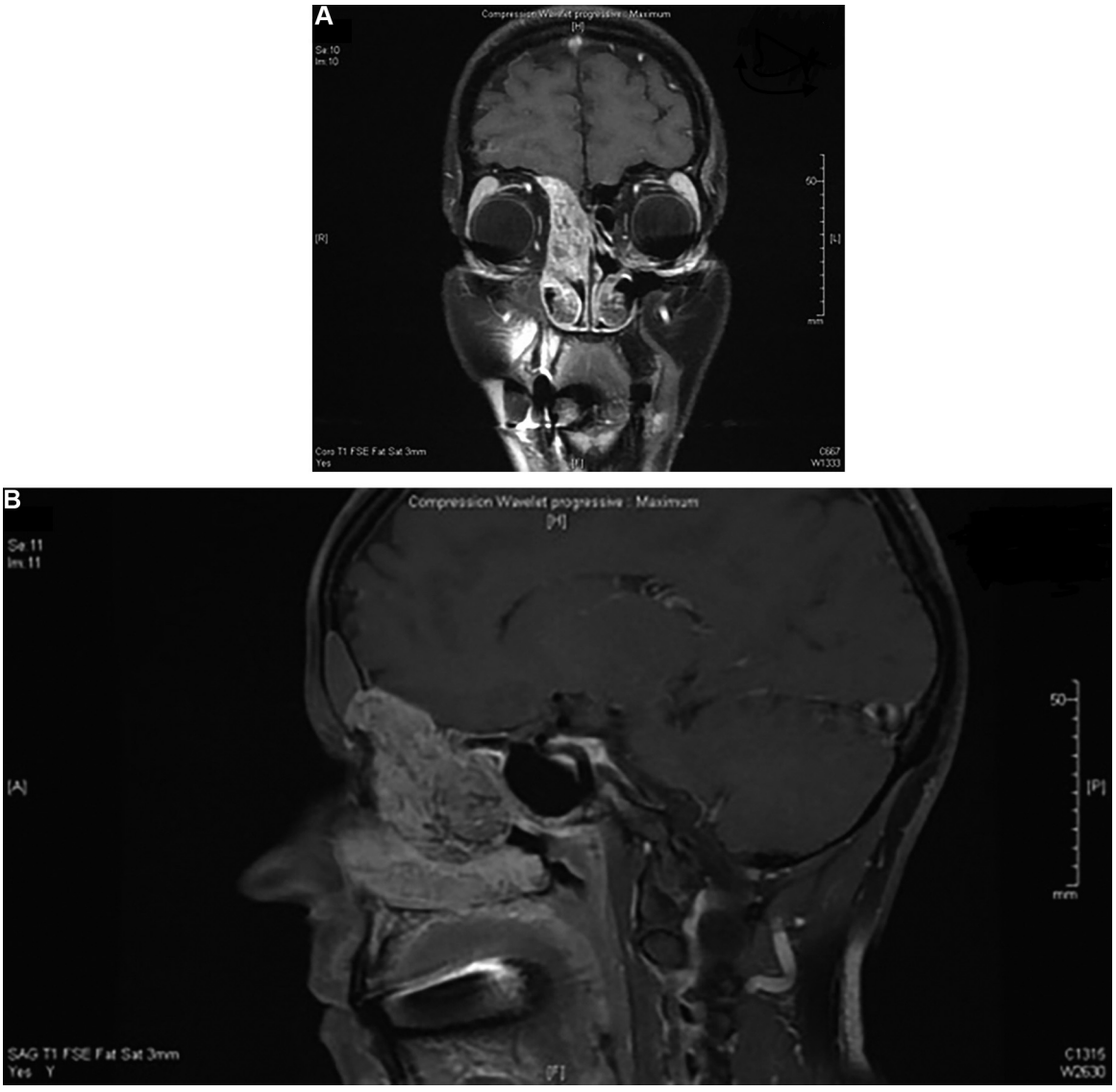

Fig. 1. A and B. Pretreatment MRI of the ethmoid tumor, coronal (A) and sagittal (B) sections, showing tumor extension in contact with the dura mater and lamina papyracea and the absence of contralateral extension.

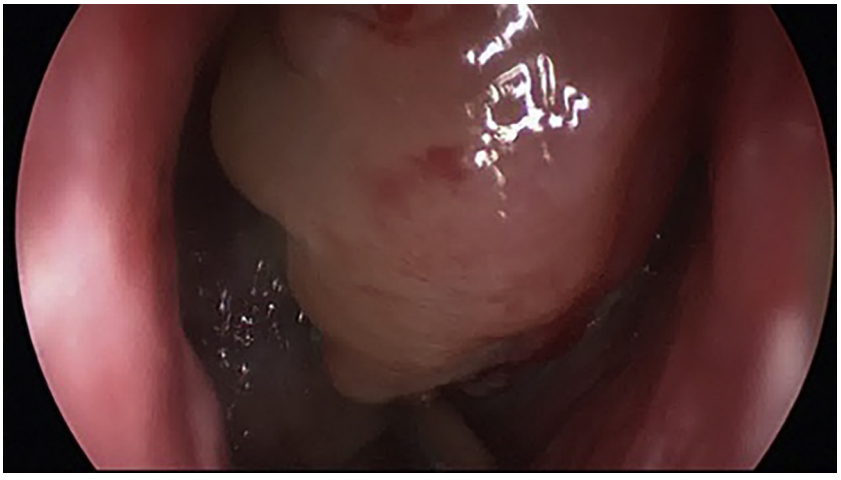

Fig. 2. Initial nasal endoscopy showing an atomic beige polyp in the right nasal cavity.

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