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CASE STUDY

Oncogenic osteomalacia due to phosphaturic mesenchymal tumour in infratemporal fossa

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KEYWORDS
Oncogenic osteomalacia;
Phosphaturic tumour;
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Abstract
Oncogenic osteomalacia is an uncommon syndrome characterized by phosphaturic tumours that produce mineral metabolism abnormalities. Head and neck is the second most frequent location of these tumours. We describe a case of a phosphaturic mesenchymal tumour in the infratemporal fossa that caused oncogenic osteomalacia, resolved by means of surgical excision.

PALABRAS CLAVE
Osteomalacia oncogénica;
Tumor fosfatúrico;
Neoplasia de cabeza y cuello

Osteomalacia oncogénica por tumor mesenquimal fosfatúrico en fosa infratemporal

Resumen
La osteomalacia oncogénica es un síndrome infrecuente que se caracteriza por alteraciones en el metabolismo mineral producidas por la presencia de tumores fosfatúricos. La región de cabeza y cuello es la segunda localización más frecuente. Presentamos un caso de tumor mesenquimal fosfatúrico de fosa infratemporal que producía una osteomalacia oncogénica que se resolvió con la exéresis quirúrgica del mismo.

Case study

Female, 64 years old, presenting a case of rapidly developing osteoarthritis in 2006. From the trauma service, she was referred to rheumatology where, in June 2007, she was diagnosed with hypophosphatemic osteomalacia from hyperphosphaturia and was treated with monosodium phosphate and vitamin D. The initial extension study to rule out oncogenic osteomalacia by PET did not show any pathological uptake. In July 2008, the study was repeated (this time with MRI) due to the persistence of pathological parameters, despite having increased the doses of phosphate and vitamin D. The MRI revealed a nodule in the lower right infratemporal fossa in close contact with the posterior and inferior wall of the maxillary sinus and

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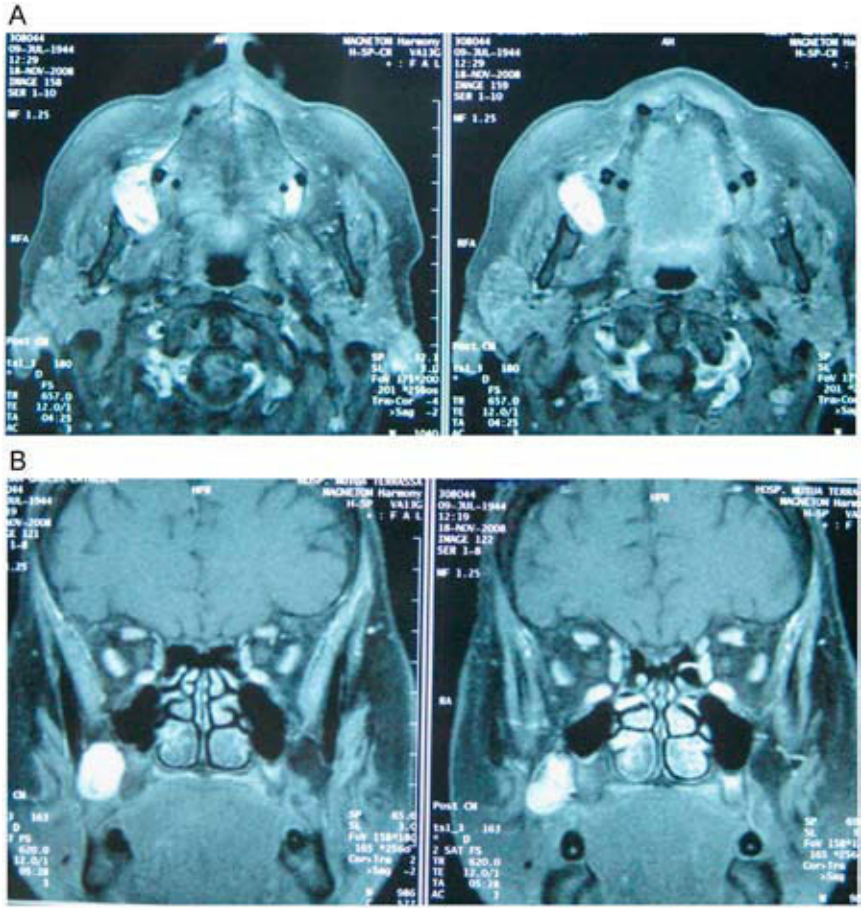

Figure MRI image of nodular lesions in the bottom right infratemporal fossa. Contacting with the maxillary sinus and pterygoid and masseteric muscles.

in contact with the pterygoid and masseteric musculature (Figure). The case was presented to the Committee of Head and Neck Oncology, and a mesenchymal tumour was postulated as a possible cause of the hypophosphatemic osteomalacia. Surgical excision was selected as a treatment. In December 2008, a highly vascular lesion was resected using a transoral approach and assisted by CO_2 laser; coagulation with bipolar radiofrequency clamp was very useful in the operation. The postoperative course was uneventful. The histopathology report informed of a solid, richly vascularised tumour, with a hemangiopericytoma-like pattern and extensive haemorrhage areas. Cell density was variable, predominantly moderate density areas alternating with hypocellular. The cell nuclei were rounded, oval or elongated, of monomorphic appearance, with no signs of atypia; the nuclei showed some isolated mitosis and eosinophilic cytoplasm with poorly defined edges. The cells were arranged in a diffuse, focally bundled pattern, with small microcystic spaces and focal marrow infiltration. We observed some isolated small foci of chondroid appearance. The definitive diagnosis was phosphaturic mesenchymal tumour with resection margins free of tumour. At 2 weeks after surgery, the laboratory parameters were normal. Control at 6 months after surgery continued to show normal laboratory values and no signs of recurrence

Discussion

Oncogenic osteomalacia is a rare disease characterised by a clinical-pathological syndrome consisting of hypophosphatemia from hyperphosphaturia, low plasma levels of 1.25-dihydroxyvitamin D and normal levels of serum calcium, parathyroid hormone and 25-hydroxyvitamin D, which are all associated with a normally benign tumour of mesenchymal origin. It was first described in 1947 by McCance.[1] About 170 cases have been published since then, although it was not until 2001 when the pathophysiology of the disease was presented. Shimada et al[2] identified fibroblast growth factor 23 (FGF23) as the causative factor. This growth factor is a potent regulator of vitamin D metabolism and phosphate homeostasis, which suppresses phosphate reabsorption and 1.25-dihydroxyvitamin synthesis.[2,3] In oncogenic osteomalacia, aetiological diagnosis is carried out years after the diagnosis of osteomalacia in more than 60% of cases. Most of these tumours appear in bone or soft tissue, with limbs being the main location. The head and neck region is the second location in frequency and appears in 27% of cases.[4] As for the location of the head and neck, the review published by Gonzalez et al[5] in 1998 showed 21 cases in the head and neck area, of which 57% were sinonasal and 20% were mandibular.

With regard to treatment, excision with free margins has shown a rapid correction of laboratory parameters. Radiation and chemotherapy do not offer good results, given the important vascular component of these tumours. In our case, the main difficulty was in the location, which required an external approach, too aggressive given that this was a benign disease. As the tumour was located in the lower end of the infratemporal fossa, we tried to approach it from the oral cavity, at the last upper molar level. This approach was sufficient for excision, given the small size of the lesion. The CO_2 laser facilitated dissection in a deep, narrow surgical field, although the significant tumour vascularization and consequent abundant surgical bleeding made its use difficult. There were no problems for wound closure, which was performed with direct suture of the mucosa. We believe the tumour probably originated in the bone wall of the maxillary sinus itself, given the presence of focal bone infiltration.

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