European Annals of Otorhinolaryngology, Head and Neck diseases 136 (2019) 335–336

Available online at
ScienceDirect
www.sciencedirect.com

Elsevier Masson France
EM|consulte
www.em-consulte.com/en

Letter to the Editor

Osteomalacia secondary to delayed diagnosis of a maxillary mesenchymal tumour

1. Case report

A 21 year-old woman consulted in 1999 for spontaneous left metatarsal fractures. Clinical work-up revealed hypophosphoraemia, phosphaturia and bone demineralisation on bone densitometry.

Three years later, the patient presented with stress fracture of the hip and deterioration of hypophosphoraemia despite vitamin D and calcium supplementation. The negative gastrointestinal work-up eliminated the hypothesis of malabsorption syndrome. The phosphorus reabsorption rate was decreased, confirming the renal origin of the hypophosphoraemia. The diagnosis of osteomalacia due to phosphate diabetes was then proposed, but no underlying cause was identified.

Genetic origin was excluded due to the absence of any notable history and there were no arguments in favour of tubulopathy such as Fanconi syndrome.

Overproduction of phosphatonin in the form of FGF23 by a secreting tumour appeared to be the most likely cause in view of the positive results of the first FGF23 assay. However, chest/abdomen/pelvis Computed Tomography (CT), positron emission tomography (PET)-CT and scintigraphy did not reveal any signs of a metabolically active tumour, and subsequent FGF23 assays were normal.

Isolated hypophosphoraemia due to excessive renal losses persisted for many years.

In 2015, FGF23 assay was again elevated, suggesting the hypothesis of an FGF23-secreting tumour. This time, PET-CT demonstrated an increased uptake in the maxilla. Facial MRI revealed a $14 \times 7 \times 8$ mm nodular mass of the palatine aspect of the left maxilla (Fig. 1). FGF23 assay was markedly elevated early in January 2018, justifying resection of this lesion (Fig. 2).

Histological examination of the operative specimen suggested a benign spindle cell phosphaturic mesenchymal tumour. Serum phosphorus had returned to normal on the 7th postoperative day and the patient was asymptomatic after one year of follow-up.

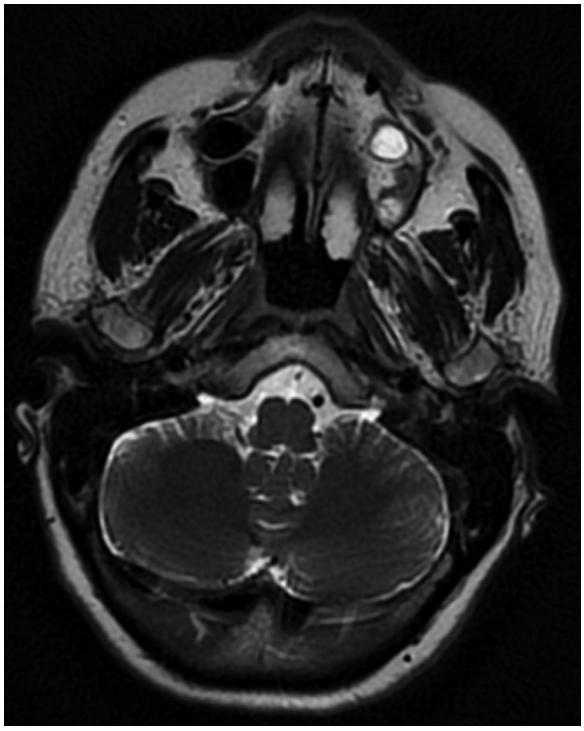

Fig. 1. MRI T2-weighted sequence, axial section, showing a high-intensity left maxillary lesion view.

2. Discussion

Oncogenic osteomalacia is a difficult diagnosis: localization of the FGF23-secreting tumour constitutes a major challenge in the management of these patients, as tumour resection allows complete cure [1].

The late onset of the mesenchymal tumour (20 years in the present case) and its painless nature often leads to delayed diagnosis [2]. There also does not appear to be any direct correlation between the size of the tumour, which is often small, and the magnitude of its clinical and laboratory consequences. The diverse sites of these tumours (brain, legs, facial bones) further complicates their detection [3].

In the presence of osteomalacia, it appears essential to start by eliminating the various differential diagnoses before considering a neoplastic origin [4,5]: congenital rickets, Fanconi syndrome, and other paraneoplastic syndromes of FGF23-secreting tumours: solid cancers (lung, prostate), haemangiomas and haematological malignancies. In the absence of any typical clinical features, it then appears reasonable to perform a number of complementary investigations. Indium-labelled octreotide has recently been shown to be useful in this setting [4,5]. This molecule has a high affinity for stomatostatin and stomatostatin receptors are overexpressed on the surface of phosphaturic mesenchymal tumour cells. Octreotide scintigraphy or octreoscan functional imaging are useful examinations for the detection of these tumours and repeated whole-body scans should always be performed before “blind” morphological imaging examinations in a context of phosphate diabetes [5].

https://doi.org/10.1016/j.anorl.2019.02.007

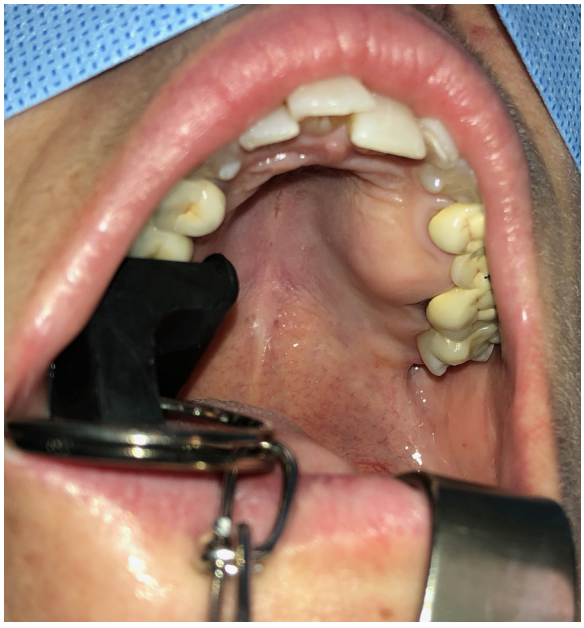

Fig. 2. Intraoperative view of the mesenchymal tumour of the palatine aspect of the maxilla over teeth 24 and 25.

Disclosure of interest

The authors declare that they have no competing interest.

References

[1] Chazal T, Khanine V, Lidove O, Godot S, Ziza J-M. Tumor-induced osteomalacia caused by a late-revealing phosphaturic mesenchymal tumor. Rev Med Interne 2017;38:412.

[2] Mori Y, Ogasawara T, Motoi T, et al. Tumor-induced osteomalacia associated with a maxillofacial tumor producing fibroblast growth factor 23: report of a case and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2010;109:63.

[3] Allevi F, Rabbiosi D, Mandalà M, Colletti G. Mesenchymal phosphaturic tumour: early detection of recurrence. BMJ Case Rep 2014.

[4] Sahnoune I, Tazi-Mezalek Z, Essaadouni L, et al. Oncogenic osteomalacia in a patient with hemangioma: a clinical diagnosis. Jt Bone Spine Rev Rhum 2006;73:1158.

[5] Pedrazzoli M, Colletti G, Ferrari M, Rossetti G, Moneghini L, Autelitano L. Mesenchymal phosphaturic neoplasm in the maxillary sinus: a case report. Int J Oral Maxillofac Surg 2010;39:32.

L. Nakhleh [a,*]
S. Zink [b]
J.M. Raguin [c]
T. Raguin [a]
[a] *HUS, service d'ORL et de chirurgie cervico-faciale, hôpital de Hautepierre, avenue Molière, 67000 Strasbourg, France*
[b] *HUS, service de chirurgie maxillo-faciale, nouvel hôpital civil, 1, place de l'hôpital, 67000 Strasbourg, France*
[c] *Cabinet du Dr J.M. Raguin, 13, rue Arthur-Bourdin, 25300 Pontarlier, France*

* Corresponding author.
E-mail address: Laura.nakhleh@chru-strasbourg.fr (L. Nakhleh)