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Case Report

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Phosphaturic Mesenchymal Tumor of the Anterior Ethmoid Sinus: A Rare Cause of Tumor-Induced Osteomalacia

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Abstract

Phosphaturic mesenchymal tumors (PMTs) are rare, benign neoplasms associated with tumor-induced osteomalacia (TIO). These tumors secrete fibroblast growth factor 23 (FGF23) which causes renal phosphate wasting, hypophosphatemia and skeletal demineralization. We report the case of a 52-year-old male who presented with multiple pathological fractures and hypophosphatemia. Imaging revealed a soft tissue lesion in the right anterior ethmoid sinus. The lesion was excised endoscopically. and histopathology confirmed the diagnosis of PMT. Postoperative normalization of phosphate levels and symptom improvement were observed. This case highlights the significance of prompt diagnosis and surgical management of sinonasal PMTs to prevent long-term complications

Keywords: Phosphaturic Mesenchymal Tumor, Tumor-Induced Osteomalacia, Hypophosphatemia, Sinonasal Neoplasms

INTRODUCTION

Phosphaturic mesenchymal tumor (PMT) is a rare, benign neoplasm often implicated in tumor-induced osteomalacia (TIO).[1] PMT is characterized by the production of fibroblast growth factor 23 (FGF23) which is a hormone that inhibits renal tubular phosphate reabsorption and decreases calcitriol production.

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These changes result in in hypophosphatemia and skeletal demineralization. Although most cases involve bones and soft tissues of the lower extremities head and neck locations represent approximately 10% of cases.[2]

The pathophysiology of PMT involves secretion of FGF23 which downregulates the sodium-phosphate co-transporters in renal tubules leading to renal phosphate wasting and hypophosphatemia. The resultant skeletal demineralization predisposes to fractures and bone pain. The lesion is histologically diverse and comprise of spindle cells in a myxoid or chondromyxoid matrix with low mitotic activity and vascular pattern.[3]

Clinically PMT presents with bone pain, muscle weakness and pathological fractures. Patients often experience prolonged symptoms before a diagnosis is made due to the rarity of the condition and its nonspecific presentation. Imaging studies, including CT and MRI, assist in localizing the tumor particularly in challenging anatomical regions such as the ethmoid sinus. Confirmation is achieved via biopsy and immunohistochemistry demonstrating spindle-shaped neoplastic cells expressing FGF23.[4]

In this case we describe a rare presentation of PMT localized to the anterior ethmoid sinus which was diagnosed following evaluation for hypophosphatemia and pathological fractures. He underwent functional endoscopic sinus surgery and the tumor was excised leading to biochemical normalization of serum phosphate levels and clinical improvement.

CASE REPORT

A 52-year-old male presented to the orthopedic department with complaints of generalized bone pain, difficulty ambulating, and multiple pathological fractures over the past year. His symptoms were insidious in onset and progressively worsened, causing significant disability. Initial laboratory evaluation revealed hypophosphatemia (serum phosphorus 1.8 mg/dL) with elevated alkaline phosphatase levels and normal calcium levels. Vitamin D levels were within normal limits.

Endocrinology consultation raised suspicion of tumor-induced osteomalacia (TIO), and further investigations revealed elevated serum FGF23 levels. Imaging studies, including a CT scan of the paranasal sinuses (PNS), identified a well-defined soft tissue lesion in the right anterior ethmoid sinus measuring 2.3 × 2.2 × 1.1 cm. The lesion caused mild scalloping of adjacent bone but without evidence of bony erosion (Figure 1).

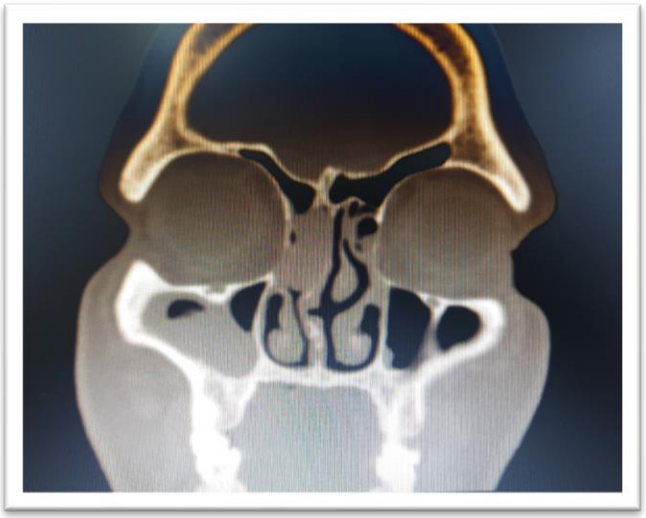

Figure 1 : Well-defined soft tissue lesion in the right anterior ethmoid sinus.

The patient underwent diagnostic nasal endoscopy (DNE) and subsequent excision of the lesion under general anesthesia. Gross examination of the excised specimen showed multiple fragments of greyish-white tissue measuring 3.5 × 2.2 × 0.6 cm. Microscopy revealed a neoplasm composed of spindle-shaped cells arranged in fascicles with elongated nuclei, distinct nucleoli, and moderate cytoplasm. Numerous osteoclast-like multinucleated giant cells and extensive vascularization with a branching "staghorn" pattern were noted. Focal areas of hemorrhage and myxoid change were present.

Postoperatively, the patient experienced normalization of serum phosphate levels within two weeks, and his symptoms improved significantly. Histopathology confirmed the diagnosis of PMT, and no recurrence was noted on follow-up at three months (Table 1).

Laboratory Investigations	Preoperative	Postoperative
S. Phosphate (mg/dL)	1.8	3.4
S. Calcium (mg/dL)	9.2	9.4
Serum FGF23 (pg/mL)	Elevated	Normal
Alkaline Phosphatase (IU/L)	480	220

Table 1 :- Lab Investigations in studied case.

DISCUSSION

Phosphaturic mesenchymal tumors are rare and often associated with TIO, a paraneoplastic syndrome caused by overproduction of FGF23.[5] Since its first description fewer than 500 cases of PMT have been reported worldwide. Head and neck involvement is even less common, with the ethmoid sinus being a rare location.[6]

In our case, the patient presented with classical features of TIO, including hypophosphatemia, multiple fractures, and muscle weakness. The ethmoid sinus lesion, confirmed by CT imaging and biopsy was consistent with previously reported cases. For example, similar cases by Folpe et al[7] and Argersinger DP et al[8] described sinonasal PMTs presenting with prolonged, nonspecific symptoms before eventual localization and successful excision.

Histopathological findings of PMT are characteristic and include spindle cells in a myxoid matrix, vascularization with "staghorn" patterns, and the presence of multinucleated giant cells. Immunohistochemical staining for FGF23 is a key diagnostic tool. Notably, surgical excision is curative in most cases, leading to rapid normalization of biochemical parameters and symptom resolution.[9]

PMTs in the sinonasal region present unique challenges due to their anatomical location and the proximity to critical structures. Endoscopic excision is the preferred approach, offering minimal morbidity while ensuring complete resection. Long-term follow-up is essential to monitor for recurrence.[10]

Our case highlights the importance of considering PMT in patients presenting with unexplained hypophosphatemia and pathological fractures. Prompt localization and surgical intervention can prevent further complications and improve quality of life.

CONCLUSION

Phosphaturic mesenchymal tumor of the ethmoid sinus is a rare entity associated with tumor-induced osteomalacia. Early recognition, imaging, and surgical excision are crucial for effective management and symptom resolution. This case underscores the need for heightened awareness of PMT as a differential diagnosis in hypophosphatemia.

Conflict Of Interest
None
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None

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