“Sinonasal Phosphaturic Mesenchymal Tumor With Orbital and Skull Base Invasion: a Case Report”

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Abstract

Introduction: Phosphaturic mesenchymal tumor (PMT) is a rare neoplasm characterized by overproduction of fibroblast growth factor 23 (FGF23), causing oncogenic osteomalacia through renal phosphate wasting. Although most commonly found in the extremities, sinonasal and skull base involvement is clinically significant. Patients typically present with bone pain, fractures, and hypophosphatemia. CT, DOTATATE PET-CT, and MRI are essential for localization and surgical planning, while histology demonstrating spindle cells and osteoclast-like giant cells with FGF23 expression confirms the diagnosis. Complete surgical resection is curative and leads to normalization of phosphate levels.

Case Presentation: A 67-year-old man with osteomalacia presented with six months of progressive right orbital swelling and visual discomfort. Examination showed right exophthalmos. Imaging revealed a 9.2 × 5 × 4 cm mass involving the right ethmoid sinus, nasal cavity, frontal sinus, and orbit, with extension to the anterior cranial fossa but no cribriform plate involvement. Nasal endoscopy demonstrated near-total obstruction, and biopsy was performed.

Discussion and Conclusion: Sinonasal PMTs are rare and may initially mimic ophthalmologic conditions, delaying diagnosis. Imaging is critical to assess orbital and skull base extension and to guide multidisciplinary management. Histopathology confirms the diagnosis. Complete surgical resection remains the treatment of choice, though combined approaches may be required in cases with orbital or intracranial involvement. PMT should be considered in sinonasal lesions with orbital manifestations, particularly in patients with osteomalacia.

Introduction

- PMT is a rare mesenchymal neoplasm characterized by excess FGF23 production, causing oncogenic osteomalacia due to renal phosphate wasting and impaired vitamin D activation [1-5].
- Most commonly arises in the extremities, followed by the nasal cavity, paranasal sinuses, and skull base, making it relevant in otolaryngology [6-8].
- Diagnosis is suggested by bone pain, pathological fractures, and persistent hypophosphatemia.
- Imaging (CT, PET-CT DOTATATE, MRI) is essential for tumor localization, extent evaluation, and surgical planning [6-9].
- Histology showing spindle cells and osteoclast-like giant cells with FGF23 expression confirms the diagnosis [12].
- Surgical management is complex due to proximity to critical neurovascular structures, requiring a multidisciplinary approach [9-11].
- Complete surgical resection is the standard curative treatment.
- The endoscopic endonasal approach is preferred for sinonasal-confined lesions due to lower morbidity and high resection rates [10,11].
- Tumors with skull base or intracranial extension require open or combined approaches with otolaryngology, neurosurgery, and radiology collaboration [10,13].
- Skull base reconstruction is necessary after resection to prevent complications such as CSF leak and infection [13,14].

Case Presentation

- **Patient:** 67-year-old man with a history of osteomalacia and high blood pressure, undergoing treatment with calcitriol, calcium/vitamin D supplements, atorvastatin, Idena, Disgren, dutasteride (Avodart), Prestat, and Sumer. History of tonsillectomy. Denies allergies and tobacco use.
- **Chief complaint:** Progressive enlargement of the right orbit and visual discomfort for six months. Initially evaluated by ophthalmology on two occasions, who indicated the use of optical lenses, with no symptomatic improvement.
- **Physical examination:** Right exophthalmos, without nasal obstruction, snoring, or nocturnal congestion.
- **Imaging:** CT and MRI demonstrated a large solid lesion (9.2 × 5 × 4 cm) involving the right ethmoid cells and nasal cavity, affecting the superior and middle turbinates and the nasal septum, with extension to the contralateral anterior frontoethmoidal region and right frontal sinus. There was destruction of the medial wall of the right maxillary sinus with partial cavity occupation and secondary mucosal retention, as well as exudate in the posterior ethmoid cells due to drainage obstruction.
  Extensive invasion of the right orbit was evident, with destruction of the medial wall and partial erosion of the superior and inferior walls, causing exophthalmos and laterocaudal displacement of the globe, extraocular muscles, and optic nerve. Cranial extension to the anterior cranial fossa was observed, with frontobasal dural enhancement but no cribriform plate involvement.
  Posterior extension reached the choana with partial involvement of the superior sphenopalatine fossa. The tumor contained partially organized calcifications and cystic or necrotic areas. A small, well-defined 10 mm nodular lesion was also noted in the left sphenoid wing, without contrast enhancement or aggressive features.
  Nasal fibroscopy showed near-complete obstruction of the right nasal fossa by a mass.
- Endoscopic biopsy confirmed a low-grade spindle cell neoplasm consistent with phosphaturic mesenchymal tumor.

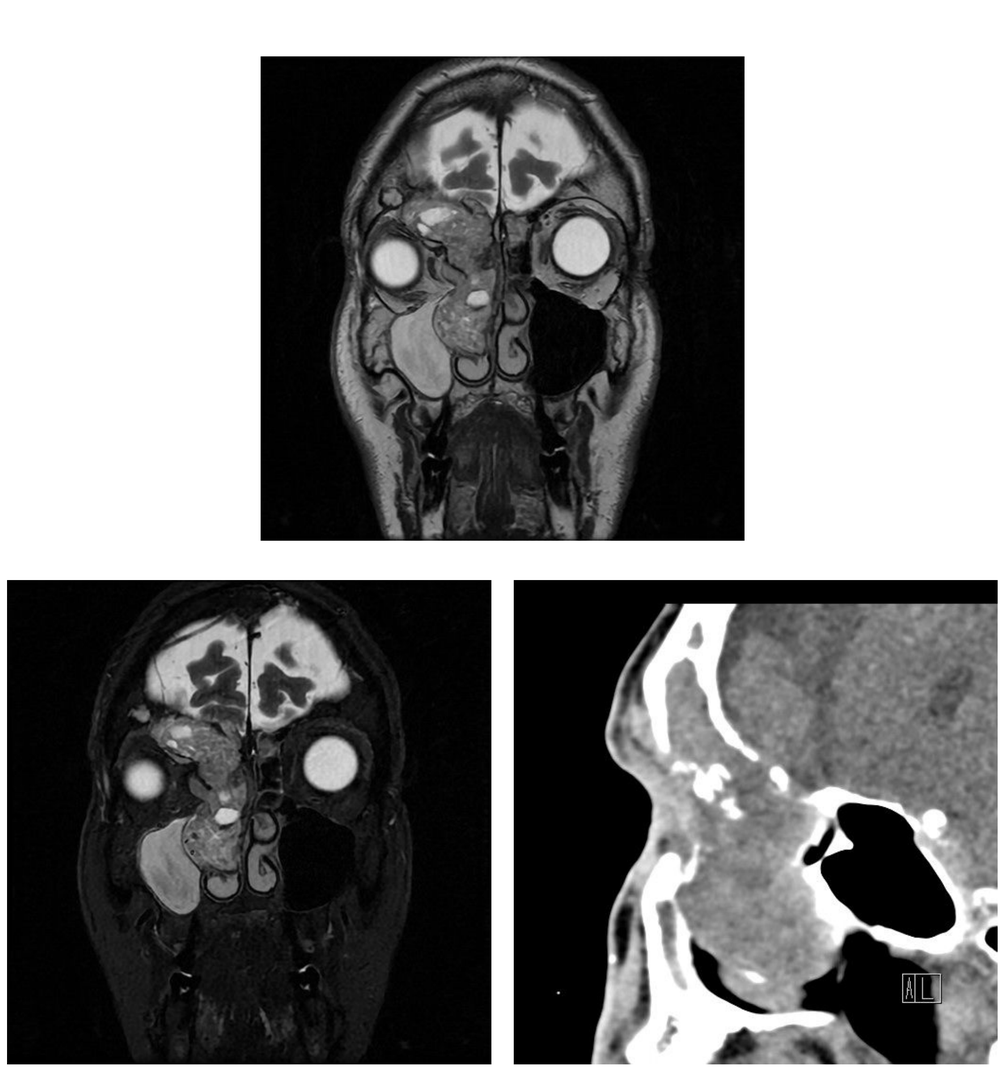

Figure 1. CT orbits MRI of the patient after first consult with ENT practitioner

Discussion

- PMT is a rare neoplasm causing oncogenic osteomalacia through excessive FGF23 secretion.
- Although most common in the extremities, sinonasal and skull base involvement is uncommon but clinically significant due to proximity to critical neurovascular structures and risk of diagnostic delay.
- In this case, orbital symptoms led to initial ophthalmologic evaluation; concurrent osteomalacia increased suspicion for PMT.
- CT and MRI defined tumor extent, revealing orbital invasion and anterior skull base extension, guiding multidisciplinary planning.
- Endoscopic biopsy showed a low-grade spindle cell neoplasm consistent with PMT.
- FGF23 overexpression on immunohistochemistry confirmed the diagnosis.
- Complete surgical resection is the treatment of choice, leading to rapid normalization of phosphate levels and resolution of osteomalacia.
- Endoscopic endonasal resection is preferred for most sinonasal cases; combined approaches may be required with orbital or intracranial extension to ensure adequate oncologic control.

Conclusions

Although rare, mesenchymal phosphate tumors of the sinonasal tract and skull base should be considered in the differential diagnosis of patients with unexplained orbital symptoms and osteomalacia. Comprehensive evaluation with advanced imaging, histopathological confirmation with immunohistochemistry for FGF23, and complete surgical resection form the basis of management. Multidisciplinary collaboration is essential to achieve curative results and preserve function in these complex cases.

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