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Case report

Phosphaturic mesenchymal tumor of the nasal cavity and paranasal sinuses: A clinical curiosity presenting a diagnostic challenge

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ABSTRACT

Phosphaturic mesenchymal tumor (PMT) is a rare mesenchymal neoplasm associated with tumor-induced osteomalacia (TIO) and elevated serum FGF-23. Common in extremities, PMT rarely occurs in sinonasal region. We report a series of sinonasal PMT diagnosed at our institute over a 6-year period.

Six cases of sinonasal PMT were identified during this period, of which five presented with features of TIO. Median age of patients was 45.5 years. All six tumors were composed of stellate to spindled cells, with prominent staghorn vasculature in four cases. Typical smudgy matrix was seen in all cases, but only focally; grungy calcification was absent.

Accurate diagnosis of PMTs is imperative, as complete excision leads to dramatic resolution of TIO symptoms. Lack of knowledge of this entity prevents clinicians from ordering relevant investigations. Absence of specific morphological features, like grungy calcification, and presentation at atypical locations makes the diagnosis challenging. Awareness of this entity is essential in order to suspect PMT in patients presenting with a soft tissue mass and features of TIO, however unusual the location may be.

1. Introduction

Mesenchymal tumors in the sinonasal region are rare and include a variety of diverse entities, such as sinonasal hemangiopericytoma, nasopharyngeal angiofibroma, rhabdomyosarcoma, and a recently described entity, phosphaturic mesenchymal tumor (PMT). PMT is a rare bone and soft tissue tumor, associated with progressive hypophosphatemia, phosphaturia and tumor-induced osteomalacia (TIO) i.e. systemic demineralization of bone caused by and which may be cured by resection of a tumor [1]. Patients typically present with symptoms of gradually progressive weakness, bone pains and pathological fractures [2]. The term "phosphaturic mesenchymal tumor, mixed connective tissue variant" (PMTMCT) was coined by Weidner et al in 1987, following which Folpe described the clinical and histopathological features of 32 cases of this tumor in detail [1,3]. Most frequently seen in the extremities, PMT rarely occurs in the sinonasal region, with only 2 of 32 of Folpe et al.'s cases being located here [1]. Due to its rarity in the sinonasal location, most ENT surgeons are unacquainted with this neoplasm. In addition, the non-specific symptoms at presentation and the wide spectrum of histological features further serve to confound the surgeon, radiologist, as well as the pathologist. We report six cases of this unusual tumor with detailed clinicopathological findings, in order to illustrate the obstacles to its diagnosis, as experienced at a tertiary referral centre.

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2. Methods

All cases diagnosed between 2010 and 2016 as PMT at our centre were retrieved from the departmental archives. Clinical details, radiology (radiographs, MRI, bone scan, DOTANOC PET/CT), laboratory data (serum phosphate, calcium, alkaline phosphatase, and fibroblast growth factor 23 (FGF-23) levels), details of surgical procedures and post-operative course were reassessed by retrospective review of electronic medical records database (Table 1). Hematoxylin-and-eosin stained slides as well as immunohistochemistry performed for diagnosis were reviewed. Approval was obtained from the Institutional Ethics Committee to conduct this study.

3. Results

Six cases of PMT involving the sinonasal region were identified from our records, including one previously reported case [4]. Clinical details of the patients as well as results of investigations are summarized in Table 1. All patients were adults, with a median age of 45.5 years (range: 34–62 years; mean: 46 years), including 4 males and 2 females.

Imaging findings are shown in Fig. 1.

On histopathological examination (Table 2), all cases showed a mesenchymal neoplasm composed of haphazardly arranged stellate to spindled cells with indistinct cell borders, moderate amount of cytoplasm and ovoid normochromatic

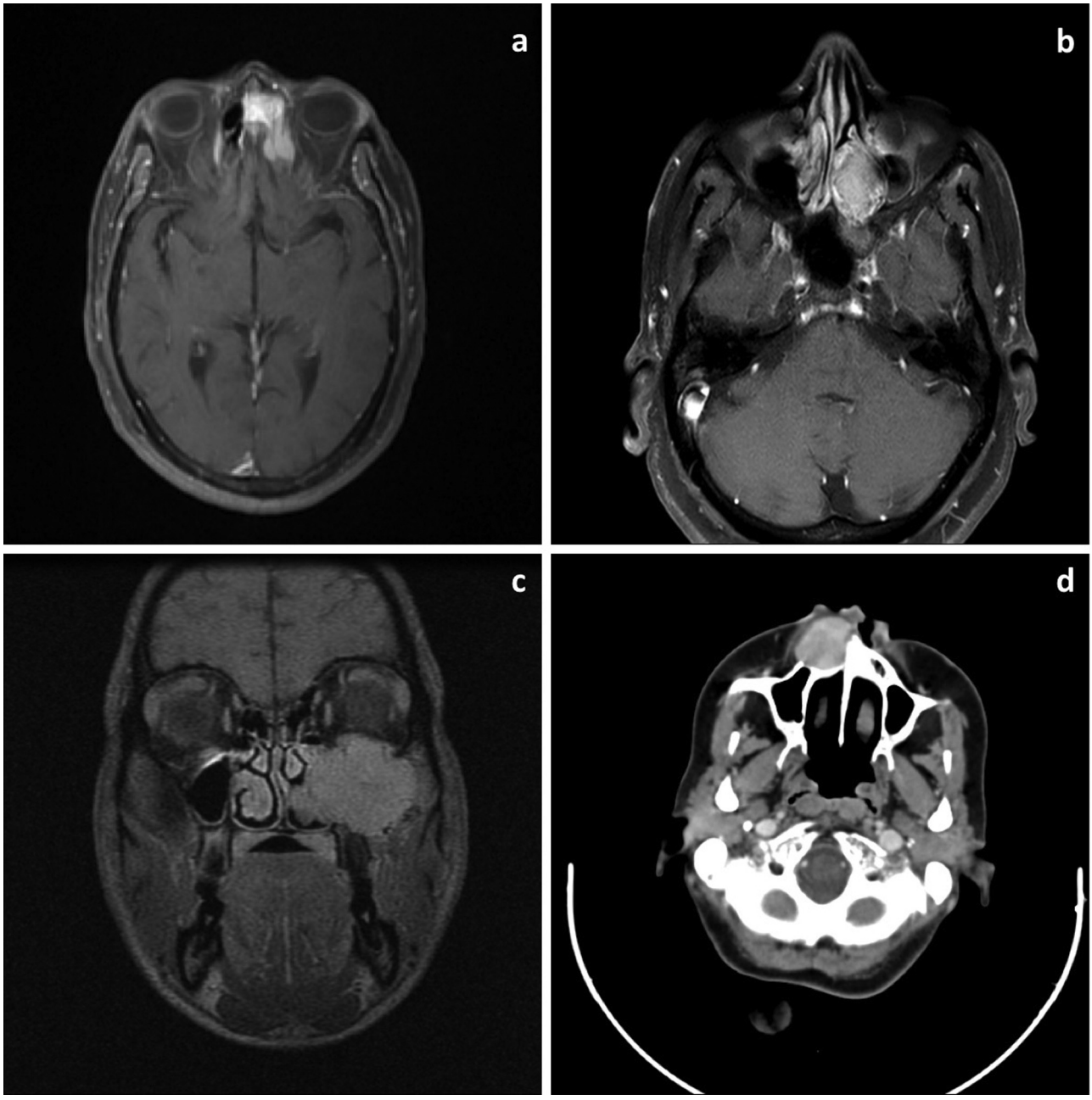

Fig 1. Axial post-contrast T1W MR images from cases 1 (a) and 3 (b) show enhancing lesions in left ethmoid air cells; coronal T1W MR image from case 5 shows a large, expansile mass filling the left maxillary sinus and extending into the nasal cavity (c); Axial CECT from case 6 shows a moderately enhancing mass in relation to the right upper alveolus, causing bony erosion (d).

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