



Sinonasal Phosphaturic Mesenchymal Tumor with Intracranial Extension

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ABSTRACT

Objectives: Phosphaturic mesenchymal tumors are rare neoplasms that are known to cause renal phosphate wasting resulting in systemic bone demineralization and osteomalacia. These tumors are often discovered during the search for the cause of osteomalacia and diagnosis is often delayed. Only 17 previously published tumors were localized to the sinonasal area with even fewer reported to be locally destructive with intracranial destruction. Through a case report and literature review we discuss the clinical presentation, radiographic and histologic features, and treatment options for this rare tumor.

Methods: Case report.

Results: A 29-year-old male was transferred to Ronald Reagan Medical Center at UCLA with an extensive sinonasal mass found on imaging. The patient had a 10-year history of unexplained osteomalacia and lower extremity weakness. He presented to an outside hospital with a 1-month history of double vision and right-sided proptosis. Imaging demonstrated a large heterogeneous mass centered in the right sinonasal cavity with extension into the right orbit and destruction of the posterior table of the frontal sinus. The patient underwent anterior craniofacial resection of the tumor followed by adjuvant radiation therapy. Definitive histopathological diagnosis was consistent with phosphaturic mesenchymal tumor.

Conclusion: Phosphaturic mesenchymal tumor is a rare neoplasm in the sinonasal region that causes tumor induced osteomalacia. Early recognition is of utmost importance in order to minimize the debilitating consequences of long term osteomalacia as well as local tumor invasion. Although intracranial extension is rare, treatment with surgery followed by radiation may be warranted in order to maximize disease free survival.

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BACKGROUND

Phosphaturic mesenchymal tumors (PMT) are extremely rare tumors that are most commonly found in the extremities. Head and neck sites are the second most common location with over half of the cases occurring in the sinonasal cavity. There have been only seventeen cases of sinonasal PMT reported in the literature and intracranial extension is even more uncommon.

PMT is associated with a paraneoplastic syndrome caused by tumor cell production of fibroblast growth factor 23 (FGT-23), a hormone like substance that affects the proximal tubule of the kidneys. This results in renal phosphate wasting, mobilization of calcium and phosphate from the bone, and osteomalacia leading to pathologic fractures. Laboratory findings are significant for hypophosphatemia and hyperphosphaturia with normal serum calcium and parathyroid hormone. Patients are often diagnosed with PMT during workup for the cause of progressive weakness and bone pain, however, early detection is not common due to the nonspecific presenting symptoms and the slow growth of the tumor.

Microscopically these tumors consist of bland spindle cells with grungy calcifications, multinucleated giant cells in a chondromyxoid matrix, and minimal mitosis. Immunohistochemistry of PMT is typically positive for vimentin, and negative for S-100, CD68, desmin, CD34, and cytokeratin. Diagnosis is confirmed with molecular testing showing overexpression of FGT-23. Although imaging with CT and MRI cannot differentiate PMT from other sinonasal tumors, imaging is important to assess tumor extent and to aid in surgical planning. Octreotide scans have been shown to help in tumor localization.

Most cases of PMT behave as benign neoplasms with low risk of metastasis and recurrence. Therefore, traditional treatment has been surgical with wide local excision. Resolution of the paraneoplastic syndrome reportedly occurs following resection. However, there are rare case reports of recurrent and metastatic tumors requiring adjuvant therapy.

We present an unusual case of PMT of the sinonasal cavity with bony destruction and intracranial extension that underwent subtotal resection with adjuvant radiation.

METHODS

The medical records of a single patient treated at a tertiary academic center for sinonasal phosphaturic mesenchymal tumor were retrospectively reviewed.

RESULTS

A 29-year-old male presented to an outside hospital with a one-month history of gradually worsening right sided proptosis, blurry vision, headaches, and nasal obstruction. The patient also reported a 10-year history of progressive weakness and fatigue that had left him wheelchair bound. CT of the sinus revealed a large, mixed cystic and solid sinonasal mass measuring 5.7 x 3.2 x 8.3cm (Image 1). The mass centered in the right nasal cavity and extended into the right maxillary, ethmoid, and frontal sinuses as well as the right orbit. There was destruction of bony structures including the posterior wall of the right frontal sinus with extension into the intracranial space (Image 2). MRI was significant for mass effect on the right frontal lobe and dural enhancement in the right frontal region (Image 3).

The patient was transferred to Ronald Reagan UCLA Medical Center for further evaluation and management. Endoscopic examination revealed a large, friable mass filling the entire right nasal cavity. Biopsy of the mass was diagnosed as a low-grade spindle cell lesion, consistent with PMT. The histopathology demonstrated bland spindle cell proliferation with rare osteoclast-like giant cells and inconspicuous mitotic figures (Image 5,6).

The patient underwent a subtotal anterior craniofacial resection of the sinonasal tumor through a bifrontal craniotomy and lateral rhinotomy. Postoperative course was uncomplicated. Final pathology confirmed the diagnosis of phosphaturic mesenchymal tumor. The patient received postoperative radiation therapy with 54Gy in 30 fractions. He was recently seen for a 12-month follow-up visit. He had no evidence of recurrent disease on endoscopic exam or head and neck imaging (Image 4). He reported improved strength and he can now walk long distances with a cane.

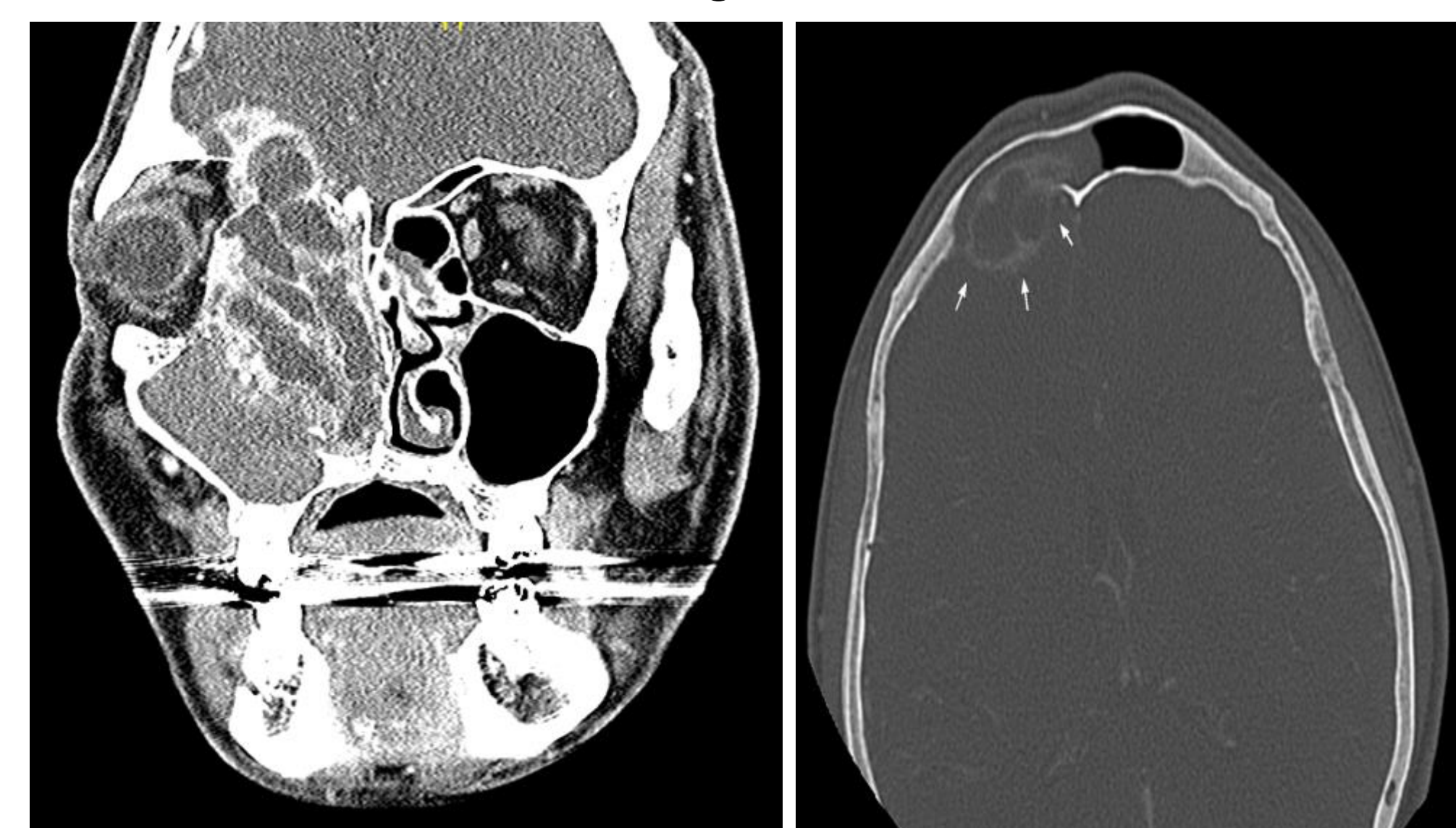


Image 1: Coronal CT sinus with large, multi-loculated right-sided sinonasal mass

Image 2: Axial CT with posterior frontal sinus wall invasion

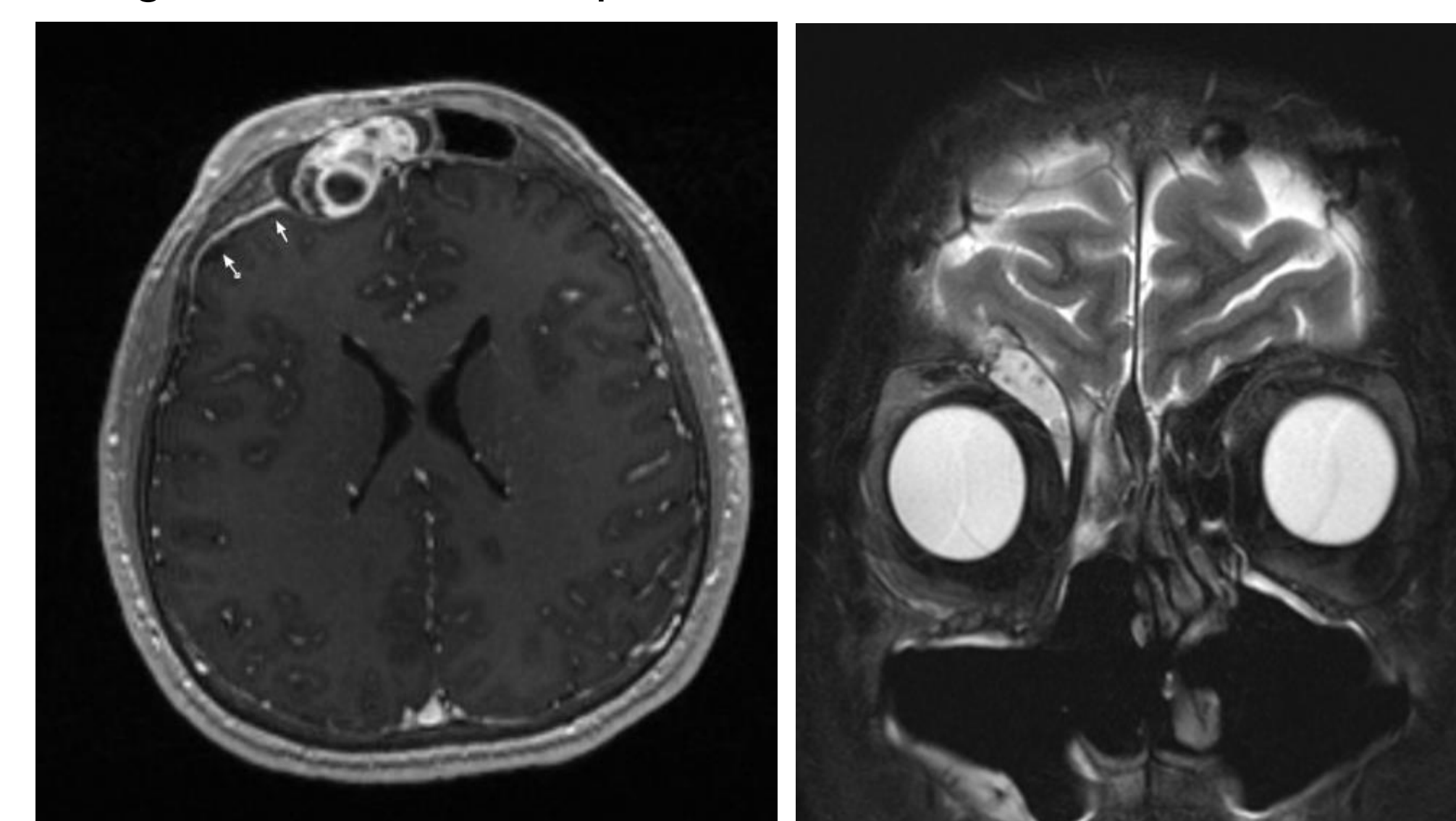


Image 3: T1 post-contrast axial MRI showing dural enhancement

Image 4: T2 fat suppressed coronal MRI with significantly decreased enhancement in paranasal sinuses

Image 5: Low-power magnification shows a spindle cell tumor with variable cellularity. H&E, 10X.

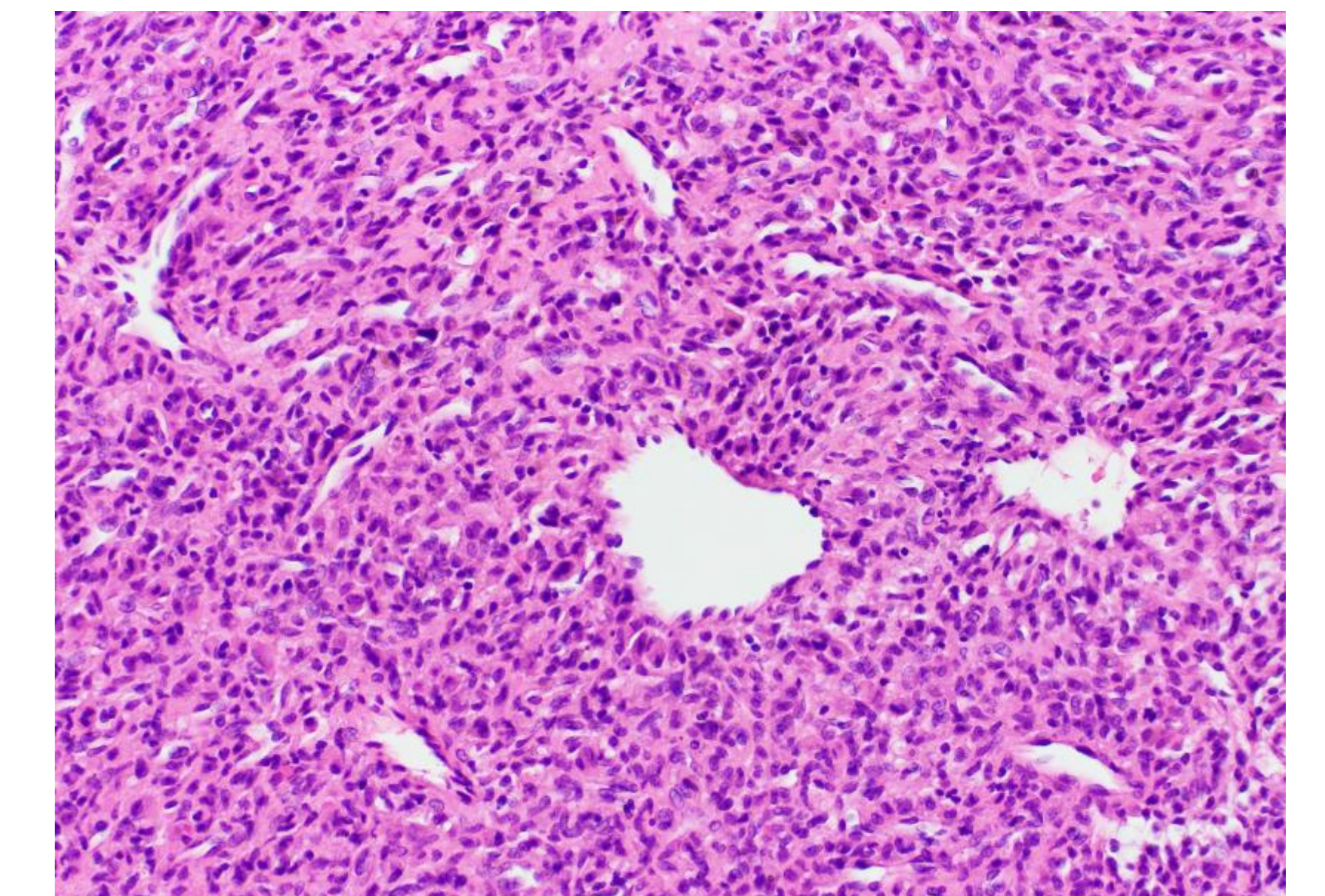
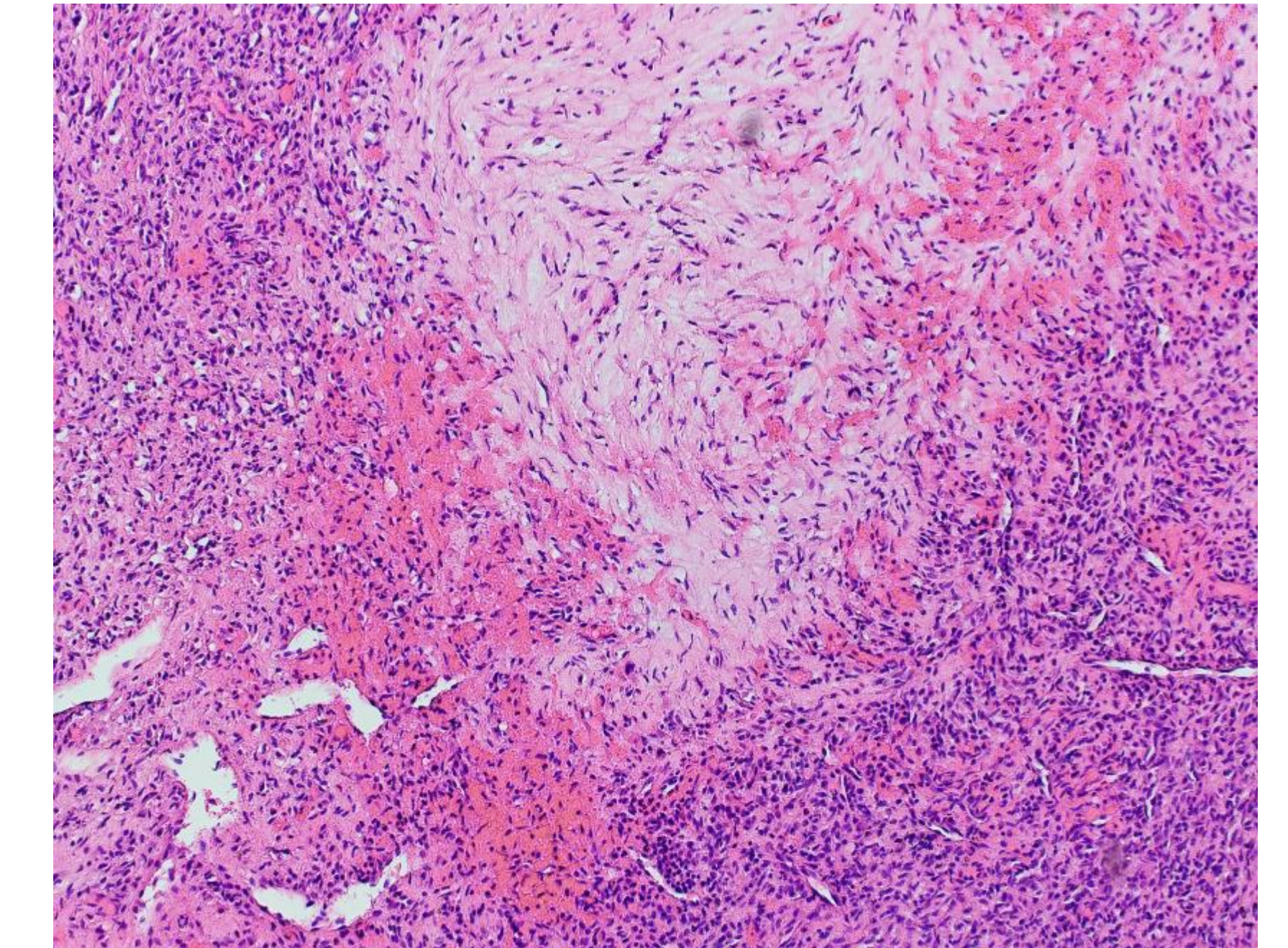


Image 6: Hypercellular areas show no specific pattern of growth and are composed of oval to spindle cells with vesicular chromatin, inconspicuous nucleoli and moderate amounts of eosinophilic cytoplasm. No significant pleomorphism or mitotic activity is present. H&E, 20X.



DISCUSSION

Phosphaturic mesenchymal tumor of the head and neck is an exceedingly rare tumor and diagnosis is often delayed. The majority of these tumors reported in the literature are slow growing, benign tumors that cause problems through local growth and paraneoplastic syndrome. The mainstay of therapy is complete surgical excision with negative margins. However, there are rare case reports of recurrent and metastatic tumors requiring adjuvant therapy.

The patient presented here had an extensive sinonasal tumor that was unique in its bony destruction and intracranial extension. Given the proximity of the tumor to the orbit and frontal lobe of the brain, the patient underwent a subtotal resection followed by adjuvant radiation therapy. This case highlights the importance of early detection in order to minimize the effects of long term osteomalacia and local tumor growth. Treatment with surgery followed by radiation therapy may be warranted in order to minimize morbidity and maximize disease free survival.

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