

Phosphaturic Mesenchymal Tumour of the Sinonasal Area Presenting As Oncogenic Osteomalacia: A Rare Presentation

Anju Anna Abraham, M.D¹, Vishnu Hari, M.D¹, Anitha Mathews, M.D², Sreejith G Nair, D.M¹, Suggeth M Thambi, M.D¹, Rony Benson, M.D^{1*}

¹Department of Medical Oncology, Regional Cancer Centre, Thiruvananthapuram 695011, India

²Department of Pathology, Regional Cancer Centre, Thiruvananthapuram 695011, India

*Corresponding author

Rony Benson

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Abstract: Phosphaturic mesenchymal tumours are rare benign tumours most commonly seen in extremities. Phosphaturic mesenchymal tumour of the sinonasal area is extremely rare. These tumors secrete a peptide-like hormone, fibroblast growth factor, resulting in urinary loss of phosphates and thus presents with oncogenic osteomalacia. Here we present a case of a 59 year old lady with sinonasal phosphaturic mesenchymal tumour and tumour induced osteomalacia.

Keywords: Phosphaturic, mesenchymal, tumours, phosphates, osteomalacia.

INTRODUCTION

Phosphaturic mesenchymal tumour is a rare benign neoplasm. Soft tissues of extremities are the predominant location of this tumour. Head and neck is a rare site for Phosphaturic mesenchymal tumour with Sinonasal region an even rarer location for the tumour. Irrespective of site of the tumor these tumors are peculiar due to the secretion of fibroblast growth factor, resulting in urinary oncogenic osteomalacia. Here we report the case of a 59 year old lady who presented to us with nasal obstructive symptoms that turned out to be a phosphaturic mesenchymal tumour with oncogenic osteomalacia.

CASE REPORT

A 59 year old lady presented with history of nasal obstruction, on and off rhinorrhoea and anosmia of 6 months duration.

On examination there was mass seen filling the left nasal cavity with no active bleeding. Contrast enhanced computed tomography [CT] of the paranasal sinuses and neck showed a 5 x 2.2 x 4.5 cm mass in left nasal cavity causing erosion of nasal septum, middle turbinate, maxillary sinus and sphenoid sinus (Figure-1a).

There was erosion of medial wall of left orbit and floor of anterior cranial fossa region of cribriform plate/olfactory bulb with minimal intracranial extension. Magnetic resonance imaging [MRI] of the face and neck showed a 4.6 x 6 x 2.3 cm lesion in left ethmoid sinus projecting to nasopharynx and a 6 x 6 x 7 cm contrast enhanced dural based lesion in left temporal region with differential diagnosis of metastasis/meningioma (Figure-1b). CT Thorax was done for possible primary/mets and was found to be normal. Biopsy from the lesion was done which showed a cellular spindle cell neoplasm. Immunohistochemistry revealed it to be Bcl2 weak positive, smooth muscle antigen (SMA) positive and S100 focal positive with MIB 12-17%. The initial differential diagnosis was biphenotypic low grade synovial sarcoma.

As a complete resection was doubtful, patient was treated with 3 cycles of Ifosphamide and Adriamycin and planned to reassess after 3 cycles for surgery. Post chemotherapy MRI showed a 7 x 4 x 3 cm lesion in left nasal cavity with intracranial and nasopharyngeal extension-stable disease compared with previous MRI. Patient underwent endoscopic resection of the nasal mass histopathology of which showed low grade spindle cell tumour with hemangiopericytomatous vasculature, which was negative for Cytokeratin, EMA, Desmin, Synaptophysin, CD34, S100, STAT6 and Nuclear INI1 expression in all tumour cells [Figure 2]. One of the differential diagnoses was phosphaturic mesenchymal tumour. Postoperative MRI showed residual mass at base of anterior cranial fossa, 30 x 8 mm and stable dural lesion in left temporal region. Nodular thickening in left nasal cavity suspicious for residual disease. Serum biochemistries showed serum calcium of 8.9 mg/dl (8.4-10.2), serum phosphorous of 2.4 mg/dl (2.5-4.5), and serum alkaline phosphatase of 267 IU (38-126). Serum vitamin D was 7.1 ng/ml, serum parathormone was 101 pg/ml, and spot urine phosphorous was 208 mg/l. DEXA scan taken showed T

score of -3.1 at L1-L4 and -2.1 at left femur suggestive of osteomalacia and the diagnosis of phosphaturic mesenchymal tumour was confirmed. Positron emission tomography (PET CT) scan done showed enhancing mass soft tissue uptake in base of anterior cranial fossa with SUV of 5.21 with multiple ribs with SUV of 4.81.

Since further resections not possible, she is kept on follow up and patient was started on calcitriol and phosphorus supplementation. She has completed 10 months of follow up now and is asymptomatic with normal serum and urine phosphorous.

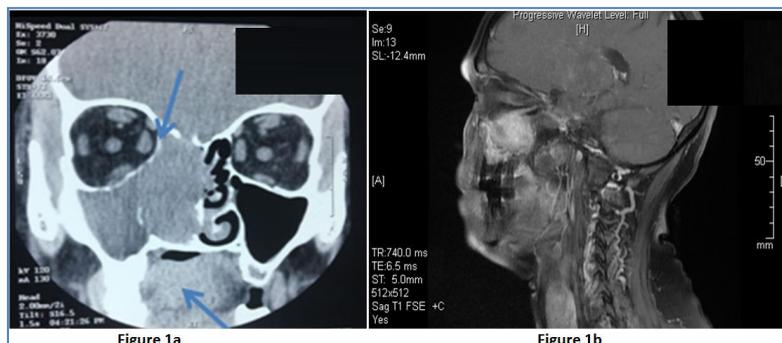


Fig-1: Contrast enhanced computed tomography of the paranasal sinuses and neck showing a 5 x 2.2 x 4.5 cm mass in left nasal cavity with erosion of left orbit and floor of anterior cranial fossa [Figure 1a] and Magnetic resonance imaging showing the tumor in the same location

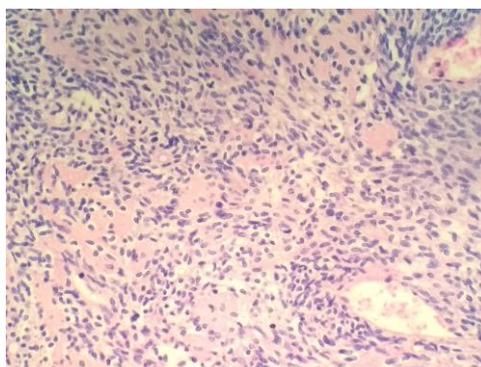


Fig-2: Photomicrograph showing low grade spindle cell tumour with hemangiopericytomatous vasculature

DISCUSSION

Named for its polymorphous histology, Weidner and Santa Cruz coined the term Phosphaturic mesenchymal tumour (PMT) in 1987. Phosphaturic mesenchymal tumour is a rare neoplasm of uncertain histogenesis that has been linked to tumour induced osteomalacia. Since 1959 to date around 300 cases have been reported [1].

The genesis of this benign tumour is largely unknown. Characteristic feature is production of phosphatonins like fibroblast growth factor (FGF-23) which cause renal phosphate wasting and induce oncogenic osteomalacia. Most common location of these tumors is extremities and appendicular skeleton which accounts for 95% of the cases. Head and neck is extremely rare for this tumor. Of the head and neck mesenchymal tumour 50% occurs in sinonasal area [2]. Morphological types include (1) Primitive appearing mixed connective tissue tumour, (2) osteoblastoma like tumour, (3) nonossifying fibroma like tumour (4) ossifying fibroma like tumour. Most frequently seen pattern is primitive appearing mixed connective tissue

tumour. It is often seen in soft tissues. Other 3 groups occur in bone.

Oncogenic osteomalacia is characterised by bone pain and pseudo-fractures. Usual etiological factors are benign mesenchymal or mixed connective tissue tumour usually phosphaturic mesenchymal tumour. There are also reports of association of oncogenic osteomalacia with haemangiopericytoma and malignant mesenchymal tumours like osteosarcoma and fibrosarcoma. Biochemical abnormalities include Hypophosphataemia, Hyperphosphaturia, decreased tubular phosphate reabsorption, Increased serum alkaline phosphatase in the presence of normal calcium, and vitamin D with a normal or slightly elevated serum Parath hormone (PTH). The serum FGF23 level will be very high. When the causative tumor was removed, there will be rapid resolution of the patient's biochemical abnormalities [3].

Complete excision of tumour with wide negative margins is curative. The role of chemotherapy and radiotherapy are not well defined due to the rarity

of this tumor. Radiofrequency ablation may be used when surgery is not possible [4, 5]. Patients who are unable to undergo surgery can receive vitamin D and phosphorus supplements. Postoperatively, FGF-23 levels fall off drastically and serum phosphate and 1, 25-dihydroxy vitamin D levels return to normal within days of surgery. Long-term skeletal changes reverse within months of surgery. Postoperative FGF-23 levels also may be used to determine the adequacy of tumor resection as persistent metabolic derangements can predict incomplete excision or lesion recurrence.

CONCLUSION

Sinonasal phosphaturic mesenchymal tumour is an extremely rare benign neoplasm which can cause oncogenic osteomalacia. Surgical resection with negative margins is the treatment of choice.

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