Original Article

A Retrospective Study of Phosphaturic Mesenchymal Tumours in a Tertiary Care Center

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Abstract

Background: Phosphaturic mesenchymal tumour (PMT) is a rare neoplasm with a distinct clinical presentation. It overproduces fibroblastic growth factor 23 (FGF23), a peptide-like hormone that decreases renal tubular phosphate absorption and inhibits 1α-hydroxylase, reducing 1α,25-dihydroxy vitamin D3 levels. It also mobilizes calcium and phosphate from bones and suppresses osteoblastic activity, resulting in osteomalacia. Given its rarity and unique biochemical profile, early recognition is crucial. This study reviews 14 cases of PMT reported in our institution.
Methods: This study was conducted in the Department of Pathology, Amrita Institute of Medical Sciences, Kochi, Kerala and retrospectively identified fourteen cases reported as PMT from 2011–2020 from the institution's surgical pathology archives. Cases diagnosed histopathologically as PMT or those with osteomalacia and supportive DOTANOC scan or elevated FGF23 levels were included. Outcomes evaluated.
Result: Fourteen cases were analyzed. Eleven were confirmed as PMT histopathologically, and three were diagnosed clinically, based on Ga-68 DOTANOC scan and high FGF23 levels. One case from the soft tissue near pyriform fossa showed features of low grade malignancy with necrosis, apoptotic debris and increased mitosis.
Conclusion: The nonspecific presentation of PMT often delays diagnosis. Histopathology, serology, and detailed radiological imaging are valuable diagnostic tools. Complete surgical excision is curative, leading to resolution of osteomalacia, clinical symptoms, and abnormal laboratory parameters. Oncologists and pathologists should recognize this entity, as early diagnosis and treatment provide excellent outcomes.

Keywords: phosphaturic mesenchymal tumour; osteomalacia; FGF23; Ga-68 DOTANOC scan

Introduction

Phosphaturic mesenchymal tumours (PMTs) are distinct neoplasms of uncertain differentiation that clinically presents as oncogenic osteomalacia, a paraneoplastic syndrome [1, 2]. The neoplastic cells overproduce fibroblastic growth factor (FGF23), a peptide hormone-like substance that is a physiologic regulator of phosphate levels. FGF23 decreases renal tubular phosphate absorption and inhibits 1 α, hydroxylase which in turn reduces the levels of 1-α, 25-dihydroxy vitamin D3. It also mobilizes calcium and phosphate from bones, reduces the osteoblastic activity, leading to widespread osteomalacia. The characteristic hallmarks of this disorder are bone pain, muscle weakness, multiple fractures, elevated or inappropriately normal plasma FGF23, inappropriately normal or low 1,25-dihydroxy vitamin D, phosphaturic hypophosphatemia, and resistance to vitamin D supplementation [3]. Diagnosis is established by chronic hypophosphatemia due to renal phosphate wasting, increased FGF23 blood levels and decreased 1,25 hydroxy vitamin D. These tumours show equal gender preponderance and it affects adults between 30-40yrs though younger age group involvement has been reported [4]. They are

commonly located in the extremities followed by the head and neck region, of which sinonasal cavity is the most common site. They are rarely seen in the retroperitoneum, viscera and mediastinum [1, 2]. PMTs usually behaves benign; although malignant cases have been recorded.

Identifying this tumour is crucial, as it has infiltrative borders and wide local excision with negative margins is curative. A thorough work up is therefore required. Previous studies show fibronectin 1 (FN1)-FGFR1 fusion genes in 60

Materials and Methods

This study was conducted in the Department of Pathology, Amrita Institute of Medical Sciences, Kochi, Kerala. We retrospectively identified 14 PMT cases reported between 2011–2020 from the institution's surgical pathology archives. Cases diagnosed histopathologically or based on presentation of osteomalacia with DOTANOC scan detection or high FGF23 levels were included. Tumours were medically managed or surgically excised and histopathologically evaluated. Outcomes were documented.

Results

Of the 14 cases, 11 were histopathologically proven as PMT, 3 were treated as PMT based on Ga-68 DOTANOC scan and elevated FGF23 levels. Eleven patients were male and three female. Mean age was 45.9 $\pm$ 18.0 years. Four lesions occurred in the femur, four in the head and neck region and others in soft tissue, D4 vertebra and fibula. All presented with osteomalacia and had significant disabilities pertaining to tumour location. Joint pains were common. One elderly patient sustained ankle fracture.

All the patients had hypophosphatemia, mean serum phosphorus level was 0.86 $\pm$ 0.60 mg/dL. Alkaline phosphatase (ALP) level was raised, mean 306.2 $\pm$ 267.9 IU/L. Serum FGF23 levels were assessed in 12 cases. The mean FGF23 level was 1491.2.0 $\pm$ 3355.7 RU/m, indicating the values were either elevated or inappropriately normal for the degree of hypophosphatemia. Few cases Ga-68 DOTANOC scan done in 11 patients, all demonstrated somatostatin avid lesions. Representative images from six patients shown in (Figure 1A–F) was diagnostic of the tumour. 11 of the patients underwent surgical excision, 2 were medically managed, and 1 patient underwent radiofrequency ablation.

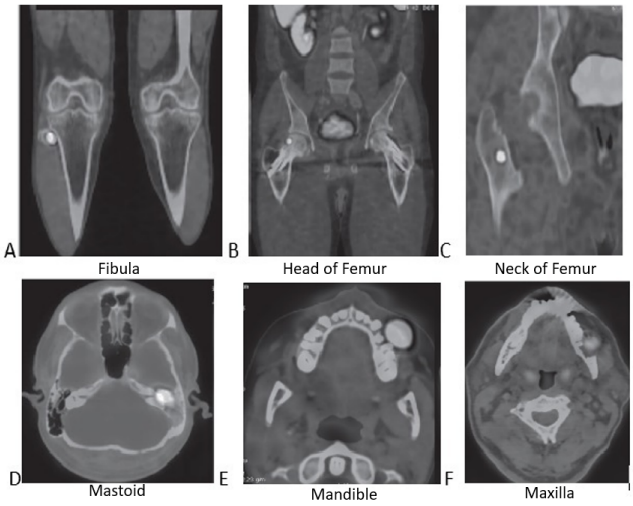

Figure 1: Ga-68 DOTANOC PET/CT of cases showing somatostatin avid lesions: A) fibula, B) head of femur, C) neck of femur, D) mastoid, E) mandible, F) maxilla.

Grossly tumours were circumscribed to ill-defined with infiltrative margins. Cut surface were grey white to grey brown, with some showing haemorrhagic areas. Histopathology showed variably cellular neoplasms composed of oval to spindle cells arranged in diffuse sheets, fascicles, cords (Figure 2) Vascularized connective tissue and areas of hyalinization in a hemangiopericytoma like pattern seen in 4 cases (Figure 3) and characteristic grungy calcification (Figure 4) was seen in 8 cases. Giant cells (Figure 5) were noted in 8 cases with myxoid and osteoid like matrix in the background. Mitoses was sparse and no necrosis or significant atypia in most cases. One case of soft tissue mass near pyriformis muscle demonstrated malignant features characterized by the presence of necrosis, apoptotic debris, and increased mitotic activity (4/10 HPF), without evidence of atypical mitoses. Immunohistochemistry was performed in five cases. Tumor cells showed diffuse vimentin positivity in four cases (naso-ethmoidal mass, left mastoid bone, nasal cavity mass, and right upper thigh soft tissue), while CD34, CD99, and SMA were consistently negative in the tested cases. One case (soft tissue near pyriformis muscle) demonstrated CD56 positivity, with SMA, CK, desmin, CD31, and CD34 negative, nonspecific S100 staining, and a

Ki-67 labeling index of 8–10%; LCA highlighted a few scattered large cells, while melanocytic, myeloid, and lymphoid markers were negative. In the thigh lesion, S100 showed focal faint positivity, whereas EMA and neuroendocrine markers (synaptophysin, chromogranin, NSE) were negative. Based on these findings; seven of the excised tumours were diagnosed as Phosphaturic mesenchymal tumour - mixed connective tissue variant (PMTMCT), one mandibular tumour was diagnosed as PMTMCT with ossifying fibroma like variant and one as PMT alone. The head of right fibula tumour was initially reported as spindle cell tumour of vascular origin but was later reclassified as PMT based on presence of osteomalacia and raised FGF23 levels. Another nasal cavity mass initially diagnosed as giant cell tumour on histopathology was later reclassified as PMT based on clinical features and raised FGF23 levels.

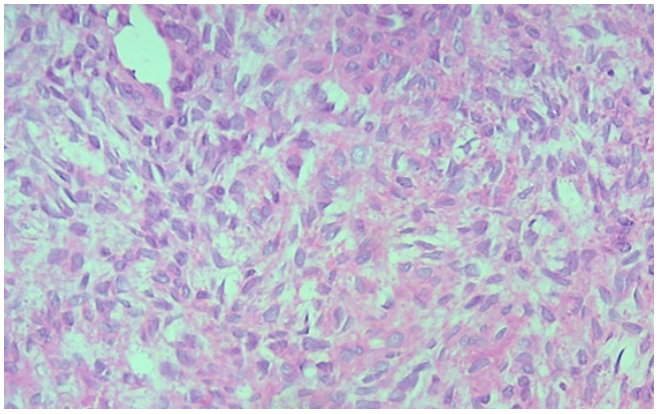

Figure 2: Spindle to stellate shaped cells [H & E, 40X].

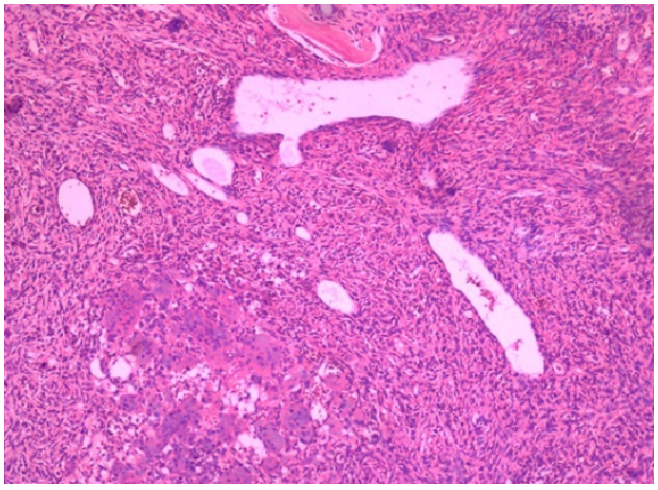

Figure 3: Staghorn vasculature in the tumour (demarcated by arrow) [H & E, 40X].

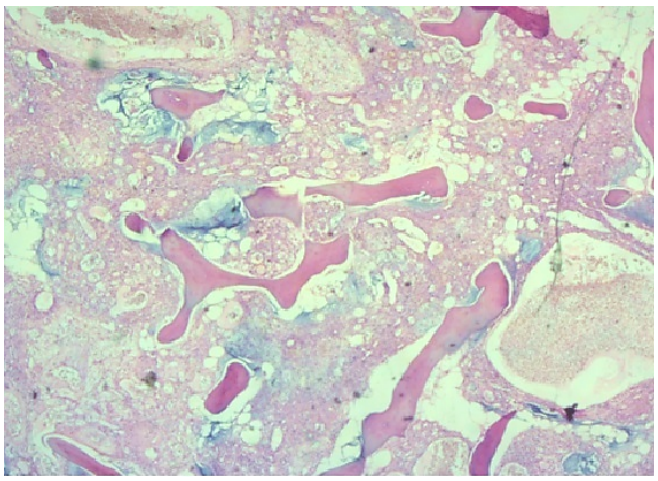

Figure 4: Characteristic grungy calcification (demarcated by arrow) [H & E, 10X].

Ten patients who were surgically managed and three medically managed patients on follow up are doing well with minimal

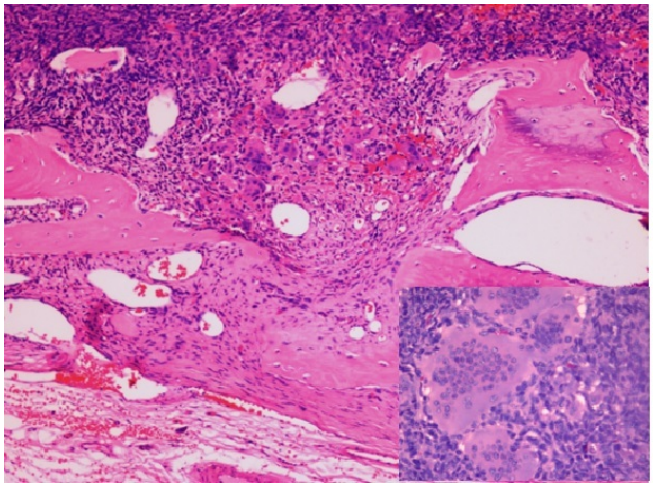

Figure 5: Areas with giant cells (inset) [H & E, 40X].

restrictions of daily activities. One case with a pyriformis muscle mass later developed pulmonary metastasis.

The clinical, histopathology and follow-up of all patients are summarised in Table 1.

Discussion

Tumour induced osteomalacia (TIO) is a rare acquired paraneoplastic syndrome characterized by low vitamin D levels, severe osteomalacia, bone pain and fractures. The pathological mechanism of PMTs is over-production of fibroblast growth factor 23 (FGF23).

In 1947, McCance first described TIO [8]. But the concept of TIO also known as oncogenic osteomalacia was first introduced by Prader and colleagues in early 1959 [9]. In 1972, Evans & Azzopardi [10] and Olefsky et al [11] identified distinctive lesions associated with TIO that were different from other known soft tissue and bone neoplasms. In 1987, Weidner and Santa Cruz [12] coined the term PMT.

PMTs often arise in the bone and soft tissue. The retroperitoneum, mediastinum and viscera are uncommon sites [1, 13]. In our case study, these tumours were seen mainly in the extremities (particularly femur), head and neck regions and the rest were seen in the soft tissue, vertebra and fibula. This tumour has male predilection and usually present in the fourth to fifth decades of life [14]. Most of our cases occurred in adults over 40 years, the mean age being 42 years, with extreme age presentations of 14 and 87 years. FGF23 overproduction in patients with PMT causes renal phosphate wasting which manifests as TIO presenting as bone pain, generalised weakness and pathological fractures [15], as was also seen in our cases. In a study done in 2004, by Folpe et al [24] analysed 32 cases and did a comprehensive review of literature of 106 cases it was found that 80% of mesenchymal tumours presenting as TIO were PMTs. The rest of the 20% included other tumours like Hemangiopericytoma, Giant cell tumour of bone (GCTB) and Osteosarcoma.

The combination of computed tomography (CT), magnetic resonance imaging (MRI), fluorodeoxyglucose positron emission tomography (FDG PET), RBC blood pool scan and 99mTc Sestamibi scan are the usual imaging modalities used to localize these tumours [16, 17, 18]. PMTs are generally smaller in size to locate on plain radiograph and CT scan, so further functional imaging is required. As PMTs express multiple surface somatostatin receptors, even occult tumours can be evaluated by Ga-68 DOTANOC and 99mTc Sestamibi scan. On CT scan these lesions typically appear osteolytic with narrow zone of transition and internal matrix. MRI shows T1 isointense, T2 hyperintense and solid enhancing with areas of dark T2 signal [3]. Larger masses on T2WI show increased signal intensity with multiple dark foci or vascular flow voids which correlate with high vascular proliferation of spindled stellate cells, arborizing capillaries, or the hemangiopericytoma-like appearance, found in histopathology [19]. A study done in the institution showed that Ga-68 DOTANOC PET/CT was an ideal initial imaging modality and could correctly identify these lesions in patients presenting with hypophosphatemic osteomalacia [20]. In our study, eleven patients were diagnosed as PMTs, based on the evidence on DOTANOC scan along with the low phosphorous and/or high FGF23 levels.

PMTs that occur in bone should be differentiated from brown tumour, giant cell tumour of bone, osteosarcoma, chondrosarcoma and fibrous dysplasia. However, only brown tumour presents with osteomalacia and the rest can be differentiated from PMT as they do not show features of osteomalacia. In soft tissue, PMTs are often misdiagnosed as hemangioma and fibroblastic tumours due to their vascular connective tissue and infiltrating nature of tumours respectively [21, 22]. In older adults presenting with multifocal lytic bone lesions, multiple myeloma is a key differential diagnosis given its higher incidence in this age group; however, metastatic carcinoma and other metabolic or marrow infiltrative disorders should also

be considered [23].

PMT being a rare tumour with its histologic heterogeneity and diversity are often misdiagnosed [24]. In 1987, Weidner and Santa Cruz [12] first termed these lesions as PMTs and subclassified them into four morphological groups. The largest group predominantly involving soft tissues is the mixed connective tissue type. The other variants that occur in the bone includes osteoblastoma-like tumours, non-ossifying fibroma-like tumours, and an ossifying fibroma-like tumour. Folpe et al.[24] on further study concluded that most PMTs belong to a single entity - mixed connective tissue type. Another PMT variant of mixed epithelial and connective tissue" (PMTMECT) tends to occur in head and neck region especially jaw.

Microscopically, they appear ill circumscribed due to infiltrating margins of tumour into adjacent tissue. The tumour cells are bland, spindled, stellate and sometimes primitive appearing cells in a background of richly vascularised stroma. These spindle cells are round to oval without any nuclear pleomorphism. The vessels show capillary hemangioma like areas with small arborizing vessels or a hemangiopericytoma pattern with hyalinised, branching vessels (staghorn vessels). A characteristic smudgy basophilic matrix is produced by the tumour cells which calcify in an unusual “grungy” or flocculent fashion producing “flower like crystals”. When the classic features like grungy calcification, vascularized connective tissue are absent, they make these cases difficult to diagnose based on morphology alone without biochemical and radiology supporting features. Hence complete work up is needed for a definitive diagnosis. The matrix may also have areas resembling primitive cartilage or osteoid, areas resembling soft tissue giant cell tumour, benign fibrous histiocytoma in soft tissue locations, giant cell tumour of bone, non-ossifying fibroma in bone and rarely aneurysmal bone cyst-like areas. Rarely this tumour shows woven bone formation at the center and periphery of tumour. Some tumours show myxoid change with an unusual trabecular pattern of arrangement of tumour cells. PMTMECT shows both mesenchymal and epithelial component. The epithelial component within these tumours form small, irregular nests arranged haphazardly throughout the tumour. The cytoplasm of the epithelial component was eosinophilic or clear and the nuclei were evenly distributed and unpolarized. Malignant PMTs have features like high nuclear grade, marked pleomorphism, high cellularity, necrosis, and elevated mitotic activity, resembling Undifferentiated pleomorphic sarcoma or Fibrosarcoma. In our study, one case of low-grade malignant phosphaturic mesenchymal tumor with features of necrosis, apoptotic debris and mitosis (4/10hpf), underwent surgical excision and subsequently developed pulmonary metastases during follow-up. In sinonasal locations a variable component of mature adipose tissue is also frequently present. Some PMTs show myxoid change [1, 9, 17]. PMT should be differentiated from many entities like Solitary fibrous tumour, Capillary hemangioma, Mesenchymal chondrosarcoma, when the tumour shows rich vascularized stroma and hemangiopericytoma like pattern. Sometimes the tumour shows myxoid change with trabecular pattern of epithelioid cells giving endocrine nature to tumour and resembling Paraganglioma or endocrine neoplasms. Presence of excess osteoclast like giant cells, fibrohistiocytic cells within the calcified matrix mimic Giant cell tumour of bone and soft tissue. Significant osteoid matrix within tumour resembles Osteosarcoma, Osteoblastoma, Ossifying fibroma of bone. In nasal cavity presence of many thick walled vessels, adipose tissue should be differentiated from Vascular malformation, Cavernous hemangioma, Angiofibroma and Adipocytic tumours.

In our study, IHC done for 5 cases. Four of these cases showed vimentin only phenotype as described by Folpe et al [3] in his review article. Cases with this phenotype show only vimentin positivity and all the other IHC markers CD34, CD99, SMA, S100, synaptophysin, chromogranin were negative. The tumour with malignant features, extensive IHC work up was done and this tumour showed strong immunopositivity for CD56 and all markers CD34, S100, desmin, pan CK, LCA, HMB45, MPO, CD3, CD20, Tdt were negative. Most PMTs express CD56, ERG, FGFR1, SATB2, and/or SSTR2A [2, 3, 19]. Now FGF23, FGFR1 immunohistochemistry is considered highly sensitive for these tumours. However, IHC has limited role in diagnosing these cases.

Only one case had a pre-operative biopsy and was initially misdiagnosed as neurogenic tumour due to faint S100 IHC positivity. Later, on excision, tumour showed osteoclast like giant cells admixed with hemorrhages, dilated blood vessels without any grungy calcification and IHC showed only vimentin strong positivity, S100 focal faint positivity while all other markers like NSE, synaptophysin, chromogranin was negative. Based on this morphology, case was diagnosed as giant cell tumour. On follow up this patient had raised FGF23 levels and therefore was clinically treated as PMT. Eleven cases were diagnosed as PMT on histomorphology correlating with clinical features, raised FGF23 and/or findings on DOTANOC scan. The other three cases were diagnosed as PMT based on levels of FGF23 and/or findings on DOTANOC scan alone.

Surgical excision of the tumour is the curative, though other modalities like radiofrequency ablation have also been found to be useful [25, 26]. This lowers the FGF23 levels and cures the tumour induced osteomalacia. Two of the patients in this study received phosphorus replacement therapy and Calcitriol tablets out of which one of them underwent excision later. One of the patients with a D4 lesion underwent D4 corpectomy and another with a right femoral head lesion underwent radiofrequency ablation. The rest of the eleven patients underwent surgical interventions.

Conclusion

Phosphaturic mesenchymal tumour (PMT) is a rare mesenchymal neoplasm with distinctive heterogeneous but recognizable histologic appearances. Clinically these patients present with gradual muscular weakness, bone pain, and pathological

fractures, identified as tumour induced osteomalacia (TIO) along with hypophosphatemia, hyperphosphaturia and high FGF23 levels. The nonspecific nature of this tumour, lack of clinical suspicion, failure to do serum phosphorous levels makes it diagnosis challenging at an early stage. Histopathologically, these tumours may be missed and may resemble other mesenchymal tumours. Histopathology, serology along with detailed radiological imaging like Ga-68 DOTANOC PET/CT are valuable tools for diagnosis. Complete surgical excision of the tumour is curative resulting in the resolution of the osteomalacia, clinical symptoms and biochemical abnormalities.

Therefore, PMT should be considered in all patients presenting with hypophosphatemic osteomalacia. Pathologist and oncologists must be aware of this entity avoiding misdiagnosis, as timely identification and treatment can cure the disease.

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Competing Interests: The authors declare that no conflict of interests exists.

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Table 1: Clinical, histopathology and follow-up of patients with phosphaturic mesenchymal tumours.

S.no	Age, Sex	Site	Symptoms	Osteo-malacia	Site	Histopathology	Our Diagnosis	Follow-up
1.	51/M	Left head of femur	Pain and weakness both hips and fracture head of femur	Present	Left head of femur	Fragments of bony trabeculae with an infiltrating pattern of spindle cells in sheets, fascicles. Chondromyxoid background, areas with osteoid like matrix, grungy calcification and hyalinised blood vessels. Mitosis rare.	Phosphaturic mesenchymal tumour - mixed connective tissue variant (PMT-MCT).	Doing well. Full functional capacity
2.	49y/M	Head of right fibula	Hypophosphatemia with hyperphosphaturia, decreased TMP-GFR Vitamin D deficiency with secondary hyperparathyroidism Osteoporosis of right femur and lumbar spine, osteopinea left femur.	Present	Head of right fibula	Spindle cells in sheets with intervening thin walled hyalinised vessels. Grungy calcification seen. No nuclear atypia or increased mitosis	Phosphaturic mesenchymal tumour - mixed connective tissue variant (PMT–MCT)	Doing well
3.	20y/M	Right proximal femur	Recurrent episodes of monoarthritis, in migratory fashion involving the both ankle and left knee	Present	Right proximal femur	Spindle cells in fascicles with intervening hemangiopericytoma like pattern of blood vessels and giant cells. Grungy calcification seen. Mitosis <1/10hpf.	Benign mesenchymal tumour – Mixed Connective Tissue Type (PMT–MCT)	Doing well. Full functional capacity
4.	54Y/M	Naso ethmoidal mass, right nasal cavity	H/o low back ache and pathological fracture of left hip, myopathy → oncogenic osteomalacia and mass in the right nostril	Present	Naso ethmoidal mass, right nasal cavity	Variably cellular neoplasm of oval to spindle cells arranged diffusely in a hemangiopericytic pattern. Intermixed histiocytes, lymphocytes and osteoclastic giant cells also noted with areas of haemorrhage. Focally, there is grungy matrix with ossification. No mitosis. IHC: Vimentin - Positive; CD34, CD99 and SMA – Negative	Phosphaturic mesenchymal tumour - mixed connective tissue variant (PMT–MCT)	Doing well
5.	48y/M	Left mastoid bone	Difficulty in walking, getting up from squatting position	Present	Left mastoid bone	Spindle cells in sheets in a chondromyxoid background with grungy calcification and staghorn blood vessels. Few osteoclast like giant cells noted. Mitosis sparse. IHC: Vimentin – Positive; S100 and CD34 - Negative	Benign mesenchymal tumour – mixed connective tissue type (PMT-MCT)	Incomplete resection followed by radiotherapy. Now on medical therapy
6.	14Y/F	Left maxilla	Pain and weakness, pain mostly in the knee and below the knee area	Present	Left maxilla	Moderately cellular infiltrative neoplasm of spindle cells in storiform and fascicular pattern. Stroma shows hyalinization and distinctive grungy calcification. Numerous osteoclast like giant cells and fibrohistiocytic areas seen. Mitosis 2/10 hpf. /no necrosis	Phosphaturic mesenchymal tumor - mixed connective tissue variant (PMT–MCT)	Doing well
7.	54/M	Left mandible	Generalised body bony pain for 6 months. Pain present in the lateral aspect of left leg increases while walking and while weight bearing.	Present	Left mandible	Fragments of bony trabeculae with an infiltrating pattern of spindle cells in fascicles. Woven bone rimmed by osteoblasts. Stromal hyalinisation seen. Occasional mitosis (1/10hpf)	Phosphaturic mesenchymal tumor mixed connective tissue (PMTMCT) with ossifying fibroma like variant	Doing well. Full functional capacity
8.	33y/M	Right upper thigh – soft tissue	Complaints of swelling over the back of the thigh	Present	Right upper thigh – soft tissue	Skin with a well circumscribed neoplasm in the dermis composed of cellular spindle cells arranged in diffuse sheets. Multinucleated giant cells and grungy calcification noted. Vessels exhibits hemangiopericytoma like pattern. Stroma is hyalinised at places & shows foci of myxoid, cartilaginous & osteoid areas. No mitosis / necrosis seen.	PMT	Doing well
9.	60y/M	Head of right fibula	c/o pain in both knees and weakness in lower limbs and associated with low back ache	Present	Head of right fibula	Infiltrating neoplasm in bone composed of spindle cells in fascicles and bundles. Staghorn like blood vessels. No mitosis.	Spindle cell tumor of vascular origin → reclassified as PMT	Doing well
10.	51y/M	Left nasal cavity	History of left nasal block and left sided epistaxis on and off for two years	Present	Left nasal cavity	Cellular neoplasm with infiltrative pattern of spindle cells intermixed with osteoclast like giant cells with intervening prominent blood vessels, hemorrhage in the stroma. IHC: S100- faint Positive; CK, CD34, NSE, CD99, Synaptophysin, chromogranin – Negative	Giant cell tumour → reclassified as PMT	Doing well
11.	49y/F	Soft tissue from pyriform muscle	History of left thigh pain, low back ache, generalized myalgia and weakness	Present	Soft tissue from pyriform muscle	Spindle cells in sheets. Hemangiopericytomatous like vasculature, Blood filled cystic areas & grungy calcified matrix is noted. Focal areas of necrosis & karyorrhectic debris noted. Increased mitotic figures (4/10/hpf) noted. No atypical mitotic figures seen. IHC: Vimentin, CD56 – Positive. SMA, CK – Negative, Ki67 - 8-10%, Desmin – Negative, S100 - Non-specific staining, CD31, CD34 - Negative LCA - Positive in few large cells HMB45, MPO, CD20, CD3 - Negative CD138, Tdt, Pax 5, CD68 – Negative	Low grade malignancy arising in a PMT	Bilateral pulmonary lung metastasis
12.	87Y/F	Ankle fracture, Weakness proximal muscles, multiple joint pains	Presented with weakness mostly involving proximal muscles, difficulty in self-mobilisation	Present	Ankle fracture, Weakness proximal muscles, multiple joint pains	High FGF23 levels, low phosphorous	PMT	Doing well
13.	41y/M	Head of right femur	Progressive weakness of right thigh muscles, back ache and hip pain aggravated by walking	Present	Head of right femur	No histopathology – diagnosed with high FGF23 and DOTANOC scan	PMT	Radiofrequency ablation of femoral head lesion - Partial recovery
14.	31Y/M	D4 vertebrae	Back pain, generalised body pain	Present	D4 vertebrae	D4 right sided [illegible] – detected with DOTANOC scan, low phosphorous and high FGF23	PMT	On medical treatment