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Case Report

Pathology Section

Plexiform Fibrohistiocytic Tumour at a Rare Site: Frontal Region of Head with underlying Frontal Bone Invasion

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ABSTRACT

Plexiform Fibrohistiocytic Tumour (PFHT) is an intermediate malignant tumour of soft tissue. It is a rarely metastasising dermal subcutaneous neoplasm. It affects children and young adults. The age range of presentation is from birth to age 77 years. The tumour is most commonly located on the upper extremities, whereas the involvement of the frontal region is rare. The differential diagnosis includes benign fibrous histiocytoma, fibromatosis, giant cell tumour and neurofibroma. Plexiform fibrohistiocytic tumour is a slow-growing mass and which is considered a low grade malignant tumour. Treatment is simple excision. It can exhibit aggressive behaviour and rate of local recurrence is 12-38%. A 45-year-old female was presented with a solid mass on the frontal region of the head which was diagnosed as a plexiform fibrohistiocytic tumour with underlying frontal bone invasion on histopathological examination of an excised mass in the Pathology Department. Plexiform Fibrohistiocytic Tumour (PFHT) is a mesenchymal neoplasm that shows a multinodular growth pattern and which is composed of fibroblasts, histiocyte like cells and multinucleated osteoclast like giant cells. Clinically, it is usually a slow-growing mass with frequent local recurrence and rare regional lymphatic and systemic metastasis.

Keywords: Histocyte like cells, Low grade tumours, Mesenchymal tumour, Osteoclast like giant cells

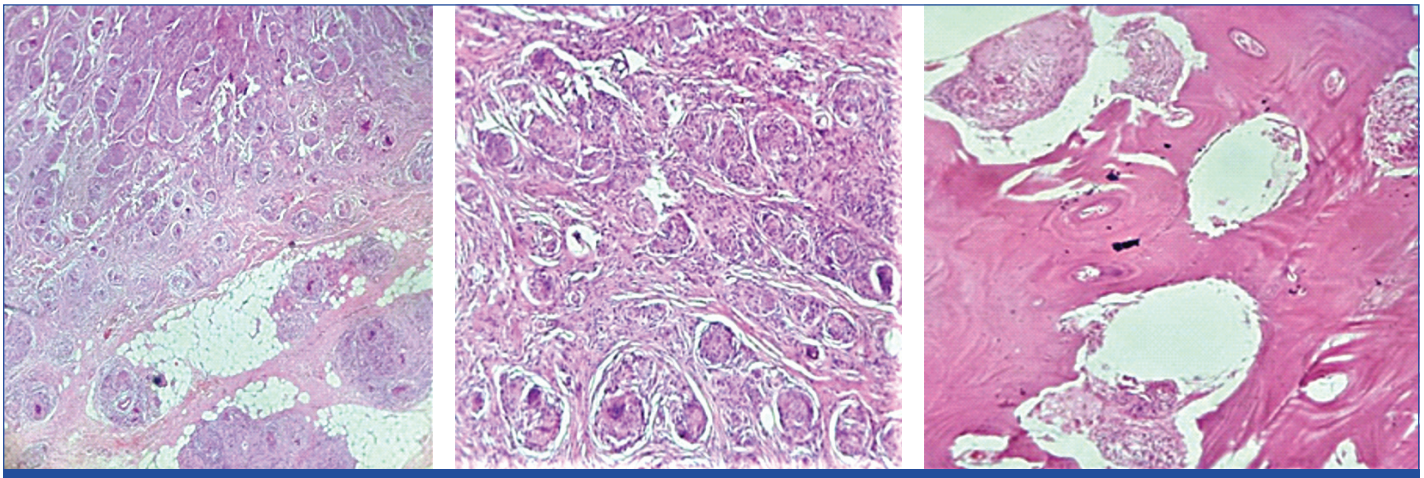
CASE REPORT

A 45-year-old female presented with a painful swelling on the frontal region of the head for one year. It was gradual in onset, progressive in nature and painless to start with, now became painful. On clinical examination, the swelling was 6×4 cm. The overlying skin was reddened. Surgical excision of the swelling in multiple pieces was done and sent to the Pathology Department in 10% buffered formalin solution.

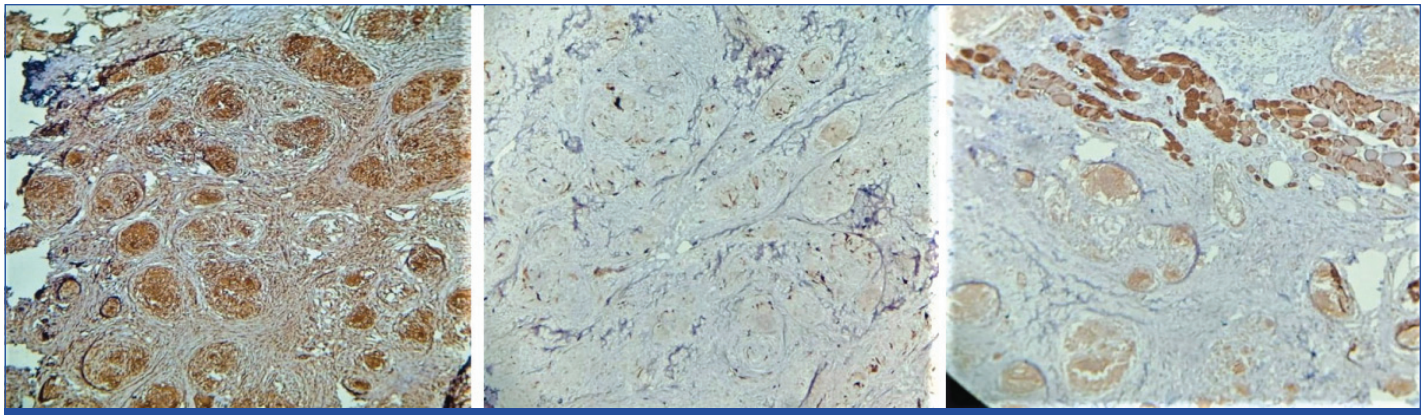
Grossly received multiple partial skins covered grey white soft tissue pieces along with bony pieces collectively measuring 5×4×4 cm. The cut section was grey white to grey tan. Routine processing was done followed by Haematoxylin and Eosin (H&E) staining. Slides were examined under a light microscope in which multiple section shows stratified squamous lining epithelium exhibiting focal thinning. The subepithelium shows a tumour arranged in small to medium sized whirling nodules extending from dermis to subcutaneous tissue. The tumour was composed of mononuclear histiocytoid cells, spindle cells and osteoclast like multinucleated giant cells [Table/Fig-1-3]. Intervening stroma was infiltrated by mild to moderate lymphomononuclear infiltrate. Adnexal structures were spared and underlying bony invasion was appreciated. Impression of PFHT mixed pattern with bone invasion were send for Immunohistochemistry (IHC). IHC results revealed positive strong Vimentin [Table/Fig-4]. S100 and desmin revealed negative expression [Table/Fig-5,6]. The postoperative follow-up of the patient could not be done.

DISCUSSION

Plexiform Fibrohistiocytic Tumour (PFHT) is a rare mesenchymal neoplasm of low malignant potential [1]. It usually affects children and young adults, with ages ranging from birth to age 77 years [2]. The tumour usually involves the upper extremities, followed by the lower extremity and trunk and head and neck is the least common site of involvement which is a rarely metastasising dermal subcutaneous neoplasm that presents as a multinodular growth pattern. Here, we present a case of PFHT with a rare location of the frontal region of the head. It was first described by Enzinger and Zhang in 1988 [2]. The PFHT is uncomman, and has been described in a smaller percentage of cases [3]. The PFHT is classified as so called fibrohistiocytic tumours of soft tissue. It has been associated



[Table/Fig-1]: Shows a tumour arranged in a plexiform pattern involving the subcutaneous adipose (H&E stain, 100x). **[Table/Fig-2]:** Higher magnification shows plexiform nodules with many osteoclast-like giant cells (H&E stain, 100x). **[Table/Fig-3]:** Tumour cells invading the frontal bone (H&E stain, 100x). (Images from left to right)



[Table/Fig-4]: Strong vimentin positive cytoplasmic expression in plexiform nodules (H&E stain, 100x). **[Table/Fig-5]:** IHC- S100 showed negative cytoplasmic expression (H&E stain, 100x). **[Table/Fig-6]:** IHC- Desmin showed negative expression in plexiform nodules with desmin positive smooth muscle bundles (H&E stain, 100x). (Images from left to right)

with low recurrence [4]. Local recurrence can occur in two to three years [5]. Only in 6% of cases, it has been metastasised to regional lymph nodes [2]. It has a good prognosis if it is completely excised. Clinically, the incidence of this tumour is higher in females and mainly affects children and adolescents, the mean age is reported to be roughly 14.5 years, it is rare after 30 years [6]. The PFHT is a classified in soft tissue tumours. There is only one reported case with bone invasion which was characterised by small poorly delineated, painless dermal and subcutaneous mass that slowly enlarges over months to years [2]. Cytologically, the tumour is characterised by multiple nodules of plexiform distribution as well as present in the deep dermis and in sub-dermal region of adipose tissue, containing mononuclear histocyte and multinucleated osteoclast like cells which are known to be proliferation of plexiform and we can observe bundles of fibroblast-like spindle cells surrounding these nodules. There are three different growth patterns:

1) Fibroblast type, spindled and infiltrating, often with lymphocytic inflammation, 2) Histiocytoid type, epithelioid with cannonball nodularity and osteoclast type giant cells, particularly in areas of haemorrhage; and 3) Mixed pattern. The predominant giant cell type is often accompanied by the destruction of the bone. It is rare to see myxoid changes, pleomorphism and cellular atypia.

The present case reported a mixed pattern- Plexiform fibrohistiocytic tumour with mitotic activity which shows underlying frontal bone invasion at the same time.

Immunohistochemically, the vast majority of tumour cells show a consistent immunophenotype including the histiocytoid cells positive for CD68, and the fibroblastic cells stain for Vimentin and Smooth muscle Actin (SMA) while negative expression of Desmin, S-100, AE1/AE3, CD34, Bcl-2, HMB-45, EMA, ERG, Caldesmon, β-catenin. A clonal t(16;17) was reported in a single case [7].

CONCLUSION(S)

The PFHT is an uncommon mesenchymal neoplasm fibrohistiocytic tumour of soft tissue and has been classified as so called fibrohistiocytic tumours. Involvement of the head and neck region with underlying bone involvement is rare. Extensive surgical resection is the mainstay of treatment. There is a low recurrence rate and distant metastases and hence long term follow-up is necessary.

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