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PLEOMORPHIC HYALINISING ANGIECTACTIC TUMOUR WITH EARLY PHAT CHANGES – A CASE REPORT

Histopathology

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ABSTRACT

Pleomorphic hyalinising angiectactic tumour is an extremely rare low grade soft tissue neoplasm composed of spindled or pleomorphic neoplastic stroma containing hemosiderin, partially thrombosed ectactic vessels with circumferential hyalinization and variable inflammatory component. Occasionally different areas show myxoid spindle cell component coexist with PHAT ,provisionally termed as early PHAT(5).It represents as a precursor lesion if both PHAT and early PHAT coexist and appears identical to the lesion called hemosiderotic fibrohistiocytic lipomatous lesion. PHAT should be considered as intermediate grade malignancy with higher incidence of local recurrence. Hence diagnosis of this entity is needed to reduce the misdiagnosis of this lesion.(1)

KEYWORDS

Pleomorphic Hyalinising Angiectactic Tumour, Hemosiderotic Fibrohistiocytic Lipomatous Lesion.

INTRODUCTION

Pleomorphic hyalinising angiectactic tumour (PHAT) is a rare low grade tumour with locally aggressive behaviour with higher incidence of recurrence. It was first described by Smith et al in 1996. Since then about 100 cases have been reported including analysis of 41 cases by Folpe et al in 2004, largest among them.(1,2) It is now considered as neoplasm of uncertain behaviour of soft tissue under 2020 WHO classification of Soft tissue and Bone tumours. It usually affects patients from 10 to 79 years with slight female predisposition. It develops in subcutaneous tissue of the lower extremities mainly ankle and foot. Also seen as deep seated lesion in buttock, perineum, head and neck, viscera and arm.(1,3) Rarely seen in the skeletal muscle. Literature suggest morphological and genetical overlap with Hemosiderotic fibrolipomatous tumour(HFLT). Here we report a case of 32-year-old patient diagnosed as PHAT on excision biopsy with early PHAT features.

Case Report

A 32- year- old female, presented with a swelling in the medial aspect of right thigh for 6 months. No associated pain or functional disability. No increase in size. USG shows heterogeneously hypoechoic lesion with internal punctate hyperechogenic areas in the middle third of anteromedial aspect of right thigh, involving subcutaneous plane – Lipoma/ Lymphnode/neurofibroma. Patient underwent wide local excision from the local hospital. It was diagnosed as Benign Angiomyxoid soft tissue lesion and was sent to our hospital for further evaluation and definite diagnosis.

In our hospital we received the slides and blocks,sections were prepared from the blocks. Routine staining and immunohistochemical staining with CD34,Desmin,vimentin,SMA and S100 were done.

Microscopically, there is clusters of thin walled ectactic vessels lined by endothelium with a thick eosinophilic fibrin deposits surrounded by lamellated collagen(Figure1 A,B). Intravascular thrombi and hemosiderin pigment deposition noted. The surrounding neoplastic stroma shows spindle ,epitheloid to pleomorphic cells randomly distributed between the blood vessels embedded in stroma containing inflammatory cells(Figure 2A,B). Periphery shows early PHAT features like adipose tissue, myxoid stroma, bland fascicles of spindle cells with intracytoplasmic hemosiderin pigment and chronic inflammatory cells(Figure 3). Immunohistochemical studies were performed, the tumour cells were positive for CD34 and Vimentin (Figure 4), while it was negative for Desmin, S100 and CK (Figure 5).

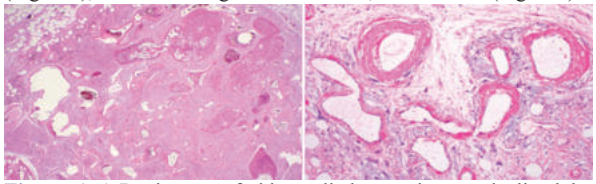


Figure 1 A,B clusters of thin walled ectactic vessels lined by endothelium with a thick eosinophilic fibrin deposits surrounded by lamellated collagen.

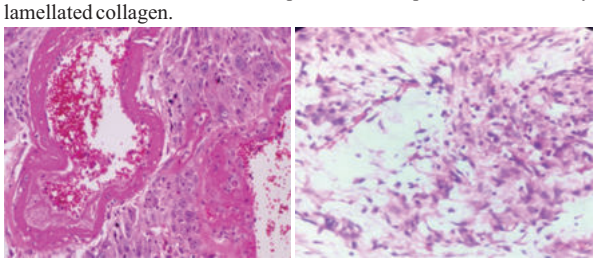


Figure 2 A,B Stroma shows spindle ,epitheloid to pleomorphic cells randomly distributed between the blood vessels embedded in stroma containing inflammatory cells

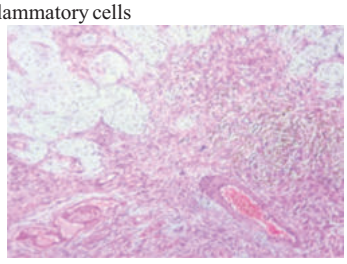


Figure 3.Early PHAT features like adipose tissue, myxoid stroma, bland fascicles of spindle cells with intracytoplasmic hemosiderin pigment and chronic inflammatory cells

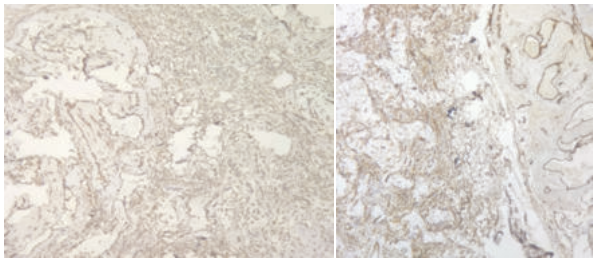
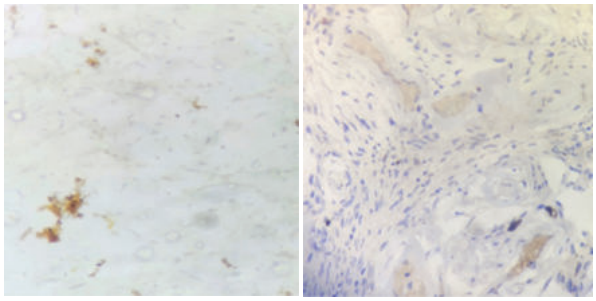


Figure 4 A,B IHC showing Vimentin and CD34 positivity .



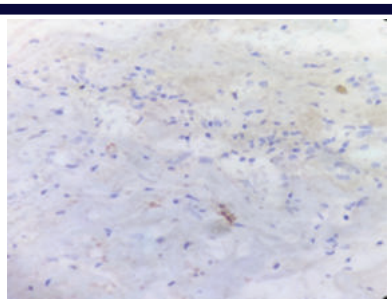


Figure 5 A,B,C. IHC negative for S100,Desmin and CK.

At the follow ups of 6 months and 12 months, the patient showed no local recurrence.

DISCUSSION

PHAT is a rare soft tissue tumour mostly seen in adults with slight female predisposition and occur predominantly in the lower extremities, first described by Smith et al(3).Other locations were perineum, buttock, arms, head and neck .The treatment modality includes wide excision with tumour free margins. The recurrence rate is about 30 -40 % when it is not possible for wide excision. Eventhough distant metastasis is not reported in literature.(5)

In literature most cases presents as lobulated appearance with myxoid and cystic areas with grey brown cut surface. Many of them having pseudoinfiltrative margins. Histopathologically characterised by clusters of thin walled ectactic vessels lined by endothelium with a thick eosinophilic fibrin deposits surrounded by lamellated collagen. Intravascular thrombi and hemosiderin pigment deposition noted. The surrounding neoplastic stroma shows spindle ,epitheloid to pleomorphic cells randomly distributed between the blood vessels embedded in stroma containing inflammatory cells. Immunohistochemically, PHAT is strongly positive for CD34, Vimentin and sometimes CD99, negative for S100, Desmin ,CK ,EMA and CD31.In our case also tumour cells are positive for CD34 and Vimentin. Many studies like by Folpe et al mention about the early PHAT features like mature adipose tissue, myxoid spindle cell component, hemosiderin pigment in the periphery of tumour which had an overlap with HFLP which was observed in our case also.(7)

The morphological features of PHAT points out differential diagnosis which includes Ancient Schwannoma, Undifferentiated pleomorphic sarcoma and Myxoinflammatory fibroblastic sarcoma. Areas of ancient features in schwannoma mimics PHAT, but negative S100 protein and Strong CD34 expression excludes Ancient Schwannoma. Absence of necrosis , higher nuclear and cellularity clearly excludes undifferentiated pleomorphic sarcoma.

Studies conducted by Wei et al published the presence of an unblanced translocation of chromosome 1 and 3 and chromosome 1 and 10 ,with breakpoints mapped to TGBR3 and or OGA (MGEA5) seen in this tumours and other related tumours like MIFS. This feature led to hypothesis that origin of these tumours are similar with MIFS which had an aggressive course. Although the cases were less in number this finding should be considered mostly in a setting of recurrence.(2)

CONCLUSION

PHAT is a soft tissue tumour of low malignant potential which can mimics tumours ranging from benign lesions such as neurilemmoma to malignant tumours such as undifferentiated pleomorphic sarcoma. Hence awareness of this entity is necessary for management and followup. In our case we had described a case of PHAT with Early PHAT like areas mimicking HFLT. On follow up of the patient after one year shows no evidence of recurrence. Considering the rarity of the tumour ,future studies with large case series are needed to confirm the histogenesis .

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