CASE REPORT

PSEUDOMYOGENIC HEMANGOENDOTHELIOMA: A CASE REPORT

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Pseudomyogenic hemangioendothelioma (PMH) is a rare, intermediate-grade vascular neoplasm with distinctive clinicopathological features. Also termed epithelioid sarcoma-like hemangioendothelioma due to its histological resemblance to epithelioid sarcoma, PMH typically presents as multiple discontinuous cutaneous or subcutaneous nodules, intramuscular masses, and occasionally lytic bone lesions. The lower limbs are most frequently affected, followed by upper limbs, trunk, and head and neck. Despite its infiltrative nature, PMH demonstrates low metastatic potential, and only a limited number of cases are reported annually in the literature. We report the case of a 17-year-old male with a slow-growing chest wall mass present for eight years. MRI revealed a solitary, ill-defined subcutaneous lesion in the right pectoralis region measuring 5.5×4.5×3.5 cm, hypointense on T1 and hyperintense on T2-weighted images. Gross examination showed a lobulated lesion with a homogeneous white cut surface. Histology demonstrated sheets and fascicles of plump spindle cells with eosinophilic cytoplasm, vesicular nuclei, and small nucleoli infiltrating skeletal muscle, accompanied by neutrophilic infiltrates, reactive bone formation, osteoclast-like giant cells, and low mitotic activity. Differential diagnoses included rhabdomyoma, inflammatory myofibroblastic sarcoma, and epithelioid sarcoma. Immunohistochemistry revealed focal CD31, diffuse FLI1, and cytokeratin positivity, with retained INI-1 and negative CD34, supporting PMH diagnosis. Although demonstration of FOSB gene rearrangement is recommended by WHO for definitive confirmation, this was not available in our setting. Molecular studies or FOSB immunohistochemistry were advised for further validation. This case highlights the diagnostic challenges of PMH and emphasizes the importance of integrating morphology, immune-profile, and molecular studies for accurate diagnosis.

Keywords: Pseudomyogenic hemangioendothelioma; Clinicopathological features; Vascular neoplasm

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INTRODUCTION

Pseudomyogenic hemagoendothelioma (PMH) is a very rare, intermediate-grade, rarely metastasizing vascular neoplasm.[1] It is also known as epitheloid sarcoma-like hemangoendothelioma due to its striking histological resemblance to epitheloid sarcoma.[2,3] PMH presents as painful or painless multiple discontinuous cutaneous and subcutaneous nodules (75%), intramuscular nodules (50%), and lytic bone lesions (20%). Common sites of involvement include lower limbs (55%), upper limbs (20%), trunk (20%) and head and neck (5%).[4] Incidence of PMH is reported to be significantly low, with only a handful of cases documented in medical literature each year.

CASE PRESENTATION

We present a case of 17 years old male with a slow-growing mass in the right chest wall for 8 years. On MRI, “There was an ill-defined, solitary subcutaneous lesion in right pectoralis region, hypointense on T1 and hyperintense on T2-weighted images, measuring 5.5×4.5×3.5 cm”. Grossly, the lesion was lobulated with ill-defined margins and homogenous white cut surface. Histological examination revealed sheets and loose fascicles of plump spindled cells with vesicular chromatin, small nucleoli and abundant eosinophilic cytoplasm mimicking rhabdomyoblasts infiltrating into skeletal muscle. There were admixed inflammatory cells predominantly neutrophils, foci of reactive bone formation, few osteoclast-like giant cells and 2-3 mitosis/10 HPF were noted FIG1. Rhabdomyoma, inflammatory myofibroblastic sarcoma and epitheloid sarcoma were considered in differentials. Tumour cells showed focal CD31, diffuse FLI1 and diffuse cytokeratin expression FIG2. Retained INI-1 and negative CD34 excluded the possibility of epitheloid sarcoma. All other markers were negative, negating other differentials. Based on bland myoid-appearing spindle cell morphology, co-expression of vascular and epithelial markers and retained INI1 expression, PMH was favoured. According to WHO 5th edition of soft tissue and bone tumours, demonstration of FOSB gene rearrangement is a desirable criteria for confirmation of diagnosis. FOSB IHC was not available in our set up. Therefore, we suggested molecular studies/IHC for FOSB rearrangement for further confirmation.

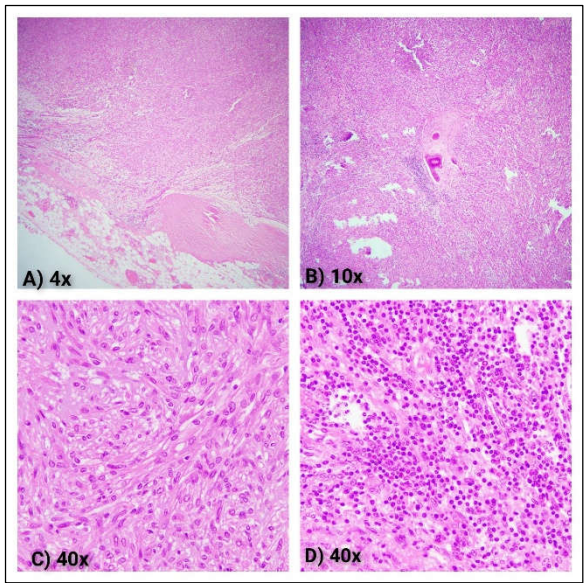

Figure-1: A. Ill-defined margins with fat infiltration. B. Focus of reactive bone formation. C. Bland spindle cells with vesicular nuclei and abundant eosinophilic cytoplasm. D. Admixed inflammatory cells comprising neutrophils, eosinophils, lymphocytes, and plasma cells.

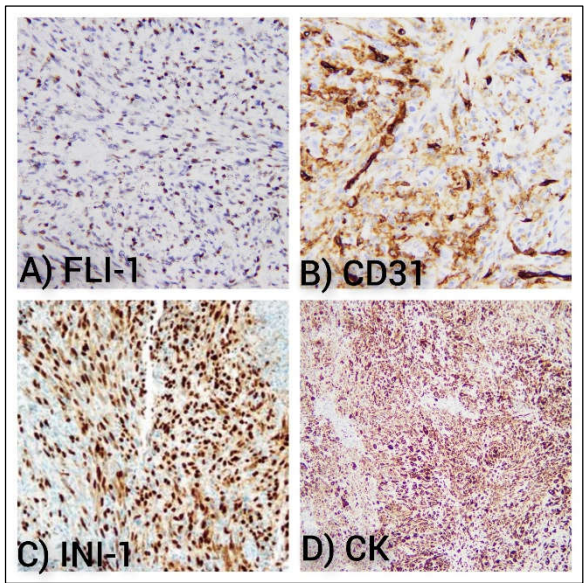

Figure-2: A. FLI-1 nuclear positive in tumour cells. B. CD31 membranous positive in tumour cells. C. INI-1 retained expression in tumour cells. D. Cytokeratin diffuse positive in tumour cells

DISCUSSION

In 1992, Mira *et al* described this tumour in five patients for the first time and named it as "Fibroma-like variant of epitheloid sarcoma" because of lack of classical-granuloma like pattern of epitheloid sarcoma in all five cases.[2]

In 2003, Billing *et al*[1] described this tumor in seven patients and labelled it as "Epitheloid sarcoma-like hemangioendothelioma". Billing *et al* observed that although none of the 7 cases displayed architectural evidence of vascular differentiation like vascular channel formation y*et al*[1] of them demonstrated immunohistochemical evidence of endothelial differentiation.

In 2011, Hornick and Fletcher[3] described this tumour in 50 patients and proposed the name "Pseudomyogenic hemangioendothelioma". They observed that despite similarities with epitheloid sarcoma, all 50 cases exhibited positive expression for cytokeratin, CD31, FLI1, negative CD34 and retained INI1 expression favouring endothelial differentiation. Based on myoid-appearing morphology and endothelial differentiation they designated it PMH.

Although clinically vast majority of cases present as multifocal nodules in different planes, in our case, the patient presented with solitary growth. Unifocal disease has also been reported by Mc Clinity *et al*, Ye *et al* and Pradan *et al*.[4,5]

PMH is caused by balanced translocation t (7;9) (q22; q13)[6], resulting in fusion of SERPINE1 to FOSB. Lately, alternative gene fusion ACTB-FOSB was reported in certain cases. The clinicopathological features and behaviour do not differ between PMH with SERPINE1-FOSB and ACTB-FOSB. Tumours with ACTB variant present as solitary lesions.[5,6]

Grossly, our case had ill-defined margins with fat infiltration similar to cases reported by Righi *et al*, Pradan *et al*, inyan *et al*.[5,6]

Microscopically, PMH shows plump spindle to epithelioid cells with vaguely nodular or plexiform architecture and prominent neutrophilic infiltrate.[1,2] Necrosis and marked nuclear pleomorphism are rare.[6]

PMH co-express keratins and vascular markers. However, as per Yin Pi hung *et al*, diffuse nuclear immunoreactivity for FOSB (>50% of cells) was observed in 48 of 50 (96%) cases of PMH. Nuclear staining of FOSB is a consistent finding in the majority of cases, making it as the most useful diagnostic marker of PMH.

PMH is treated by surgical excision. 60% of patients develop local recurrence. Telatinib is an Effective Targeted Therapy for PMH.[6]

CONCLUSION

In conclusion, the clinical case we reported is an extremely rare, commonly misdiagnosed entity. PMH can be easily confused with its histological mimicker epitheloid sarcoma. Despite the ominous presentation, PMH has an indolent clinical course with a small risk of metastasis. It is necessary to carefully examine the tumour in question and render the diagnosis with caution to avoid misdiagnosis and aggressive treatment.

Lack of classic necrobiotic nodular pattern of epitheloid sarcoma, deceptively bland myoid-appearing spindle or epitheloid morphology, fascicular or sheet-like arrangement, rare mitosis, presence of abundant stromal neutrophils and retained expression of INI-1 immunostain are the morphological clues to the diagnosis of PMH. As this is a very rare entity, sound knowledge of clinical presentation, morphology, radiology and prognostic significance is very important to avoid serious consequences in terms of patient management.

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