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CASE REPORT

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Sclerosing epithelioid fibrosarcoma – a rare variant with myriads of differentials

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Abstract

Sclerosing Epithelioid Fibrosarcoma is a rare form of soft tissue sarcoma, with increased propensity for local recurrence and metastases mainly pulmonary and pleural in about 50% of cases. It carries morphological resemblance with many sarcomas, hence detailed and comprehensive immunohistochemical examination is mandatory for definitive diagnosis. Hereby, we present a case in a 39 year old male, who presented with a painless right thigh swelling. The histopathological examination revealed nests of epithelioid cells embedded in a densely sclerotic matrix, and immunohistochemistry showed MUC4 and Vimentin positivity, rendering the final diagnosis of Sclerosing Epithelioid Fibrosarcoma.

Keywords Rare malignant mesenchymal neoplasm, Sclerosing epithelioid fibrosarcoma, MUC4 positivity

Introduction

Sclerosing epithelioid fibrosarcoma (SEF) is a rare and distinctive deep-seated variant of fibrosarcoma typically occurring in adults. Enzinger and Weiss were the first to describe a variant of fibrosarcoma with epithelioid cells in 1988 (Enzinger et al. 1988). It has a chance of local recurrence and late metastasis, usually pulmonary and pleural in 50% of cases.

Histopathology and Immunohistochemistry plays a significant role in its diagnosis, as it carries wide range of differentials. The classic histologic features of SEF include nests and cords of uniform small epithelioid cells embedded in a densely sclerotic matrix. We, hereby report a rare case of Sclerosing Epithelioid Fibrosarcoma in a 39 year old male, demonstrating the detailed and comprehensive clinical, radiological, histology and immunohistochemical examination for a definitive diagnosis.

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Case presentation

A 39 year old gentleman presented with history of right thigh mass since one month, gradually increasing in size. On clinical examination, a firm to hard painless swelling on the medial aspect of right thigh measuring around 12.5×10 cm, mobile in relation to superficial plane and adherent to deeper plane. Femoral pulsation was palpable. No inguinal lymphadenopathy seen and with no neurovascular deficit. MRI revealed a well circumscribed heterogenous intensity lesion in the medial musculofascial lower adductor compartment of the upper thigh msg 8.2×5.9x7.8 cm with mild perilesional oedema. Findings were in favor of the neoplastic lesion. Further, biopsy was advised and was reported as poorly differentiated neoplasm of mesenchymal origin. He was undertaken for wide local excision and reconstruction of soft tissue sarcoma of right thigh was done. The patient was stable and was uneventful post-operatively.

Pathological findings

Grossly, the specimen measures around 12.5×10.5x6cm. On serial sectioning hard grey white fleshy mass is seen msg 9×7.5x6cm (Fig. 1A). Microscopically, small round to ovoid epithelioid cells arranged in nests, cords and

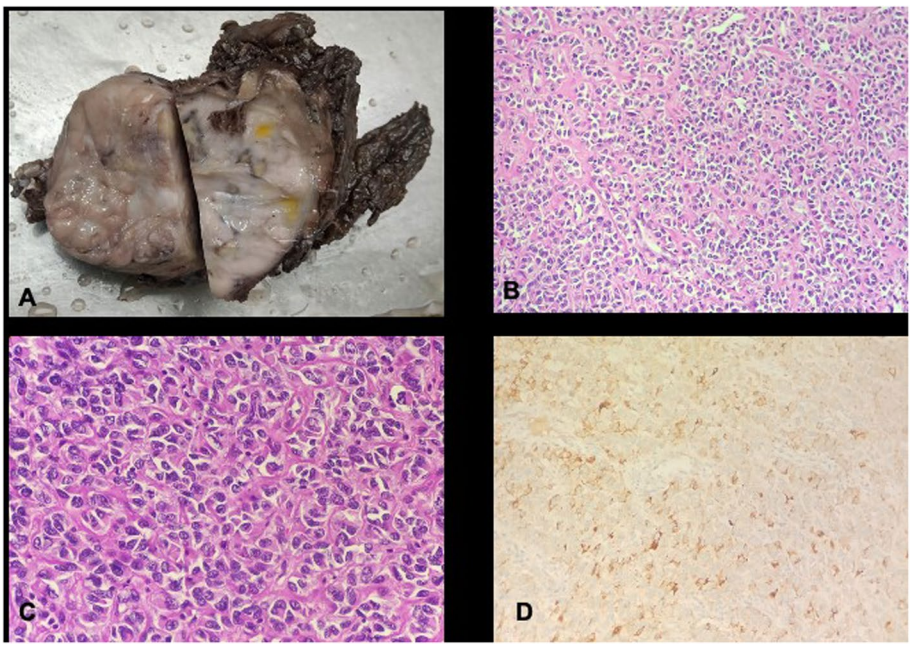

Fig. 1 **A** Gross photograph of excised Soft tissue hard and fleshy mass. **B** Nests of small to round epithelioid looking cells separated by hyalinised and sclerotic stroma. (hematoxylin and eosin stain X100). **C** High power view of round cells with sclerotic stroma and less frequent mitosis. (H and E X400). **D** IHC stain for MUC-4 showing strong immunoreactivity in neoplastic cells. [IHC- MUC-4 stain x400)

sheets with prominent hyalinized and sclerotic collagenous stroma is seen with 6–8 MF/10mm2 (3–4 mitosis/ mm2) and 30% areas of necrosis. FNCLCC Grade 2 was rendered (Fig. 1B and C). Futher it was subjected to immunohistochemistry panel, and was found that the tumour cells showed strong immunoreactivity to MUC4 (score 4+) (Fig. 1D). Based upon the aforementioned morphological and immunohistochemistry features the final diagnosis of Sclerosing epithelioid fibrosarcoma was rendered. The patient was advised for radiation therapy and is on follow-up.

Discussion

Sarcomas are malignant tumors with mesenchymal differentiation. Sclerosing Epithelioid Fibrosarcoma is a low grade sarcoma and is a rare variant, which tends to behave aggressively and carries poor prognosis (Genevay et al. 2003). The local recurrence rate is about 50% and metastases are not uncommon. The tumor is genetically heterogenous with various genomic imbalances and fusion genes. Histologically, it comprises of epithelioid cells arranged in cell clusters or cords in an abundant hyalinized collagenic stroma. (Meis-Kindblom et al. 2002). Average age of presentation is 45 years and is commonly localized in proximal limb, trunk, head and neck. (Meis-Kindblom et al. 2002) The myriads of differentials includes benign fibrous proliferations and neoplasms such as nodular fascitis, myositis ossificans, hyalinizing leiomyoma, ossifying fibromyxoid tumor and myoepithelial tumors. Other diagnostic considerations include sclerosing lymphoma, melanoma, clear cell sarcoma, alveolar rhabdomyosarcoma, extraskeletal myxoid chondrosarcoma, epithelioid malignant peripheral nerve sheath tumor, epithelioid sarcoma and epithelioid haemangio endomthelioma (Table 1). Hence, a wide panel of immunohistochemistry will allow distinction between these entities, which includes AE1/AE3, LCA, S-100, HMB- 45, SMA, desmin, CD34, MUC-4 and SATB2. In our case, morphology showed cords of epithelioid cells in a dense collagenous stroma and the tumor cells showed strong immunoreactivity with MUC-4, hence the findings were consistent with SEF. The preliminary treatment includes surgical excision of tumor as wide as possible. The efficacy of adjuvant therapy is not yet demonstrated. Chemotherapy including Adriamycin and ifosfamide was proposed in some cases. However, long term follow- up is recommended, as there are chances of local recurrence and metastasis (Antonescu et al. 2001).

Conclusion

Sclerosing Epithelioid Fibrosarcoma is a rare and aggressive malignant sarcoma, that occurs in middle age, with predilection in lower extremities. The tumor bears diagnostic challenge with overlapping features with varied histological mimics, commonly with Low grade Fibromyxoid sarcoma. Therefore, a detailed and

Table 1 Morphological differential diagnosis of Sclerosing Epithelioid Fibrosarcoma

Morphological Differential diagnosis	Immunohistochemical distinguishing markers
Sclerosing epithelioid fibrosarcoma	**MUC-4 (+)**
Ossifying fibromyxoid tumour	CD10 (+), S-100 (+)
Sclerosing Rhabdomyosarcoma	MyoD1 (Strong), Myogenin
Metastatic carcinoma	Cytokeratin expression
Low grade fibromyxoid sarcoma	Morphological clue is enough for definitive diagnosis
Osteosarcoma	SATB2 (+)
Epithelioid hamenagioendothelioma	Cytokeratins and CAMTA1 (+)

comprehensive panel of Immunohistochemistry and molecular evaluation is essential to come to a definitive diagnosis. Long term follow up of patient is required due to increased propensity local recurrence and metastases.

Authors' contributions
Dr. Prachi and Dr. Hema Malini Aiyer has contributed towards manuscript writing, editing and reviewing this manuscript.

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Availability of data and materials
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Declarations

Ethics approval and consent to participate
Ethical consent is obtained for this case report.

Competing interests
All authors declare no conflict of interest.

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