

Sclerosing Epithelioid Fibrosarcoma: A Case Report

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Abstract: Sclerosing epithelioid fibrosarcoma is an unusual variant of fibrosarcoma composed of bland epithelioid cells deposited in a densely hyalinised collagenous matrix. A 41 year male patient admitted with complains of pain in right hip since 6 months. He was a known case of fibrosarcoma (soft tissue) operated 3 years ago. On examination, old scar on medial aspect of right thigh was seen. Tenderness was present over right greater trochanter. A biopsy was performed which was diagnosed as Sclerosing epithelioid fibrosarcoma. Sclerosing epithelioid fibrosarcoma is a rare, slow growing, sarcoma of deep soft tissue with bland epithelioid tumor cells in nests and cords set in hyalinized fibrous stroma. As it is a clinically aggressive but histologically low-grade sarcoma with unsatisfactory treatment results and also has a high recurrence rate with high metastatic potential, it is important to consider this rare entity as a possible differential diagnosis in all suspicious soft tissue sarcomas.

Keywords: Sclerosing epithelioid fibrosarcoma; Clinically aggressive; Histologically low-grade.

INTRODUCTION

Sclerosing epithelioid fibrosarcoma is an unusual variant of fibrosarcoma composed of cytologically bland epithelioid cells deposited in a densely hyalinised collagenous matrix.

Sclerosing epithelioid fibrosarcoma was originally described by Meis-Kindblom et al in 1995. They reported 25 cases of peculiar Sclerosing epithelioid variant of fibrosarcoma simulating an infiltrating carcinoma. They suggested that Sclerosing epithelioid fibrosarcoma is a relatively low-grade fibrosarcoma[1].

High rate of recurrence and metastasis have been reported for this entity [1,2].

Here we present a case of Sclerosing epithelioid fibrosarcoma which is clinicopathologically distinct variant of the adult fibrosarcoma group.

CASE HISTORY

A 41 year male patient admitted with complains of pain in right hip since 6 months. Patient was a known case of fibrosarcoma (soft tissue) operated 3 years ago. On examination, healed scar on medial aspect of right thigh was seen. Tenderness was present over right greater trochanter. Hip movement was of full range. Clinical differential diagnosis were secondaries in greater trochanter, fibrosarcoma and radiation necrosis of femur.

A biopsy was performed and gross specimen consists of multiple grey white to grey brown tissue bits, largest measuring 1.5 x 1.5 cm. Microscopy shows soft tissue fragments with a tumor composed of hypocellular and hypercellular areas. The hypocellular areas show densely hyalinised stroma and hypercellular areas show neoplastic cells arranged around blood vessels. The tumor cells are round to oval and have angulated vesicular nuclei with faintly eosinophilic cytoplasm (Fig. 1&2). Immunohistochemically, tumor cells showed a strong positivity for vimentin (Fig.3).

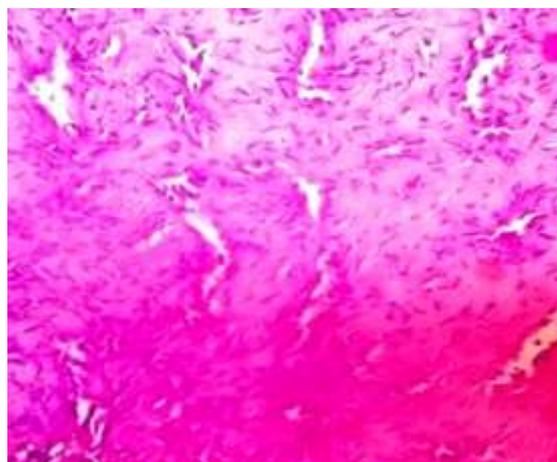


Fig. 1: Microphotograph showing hypocellular and hypercellular areas [Haematoxyline& Eosin, 10x]

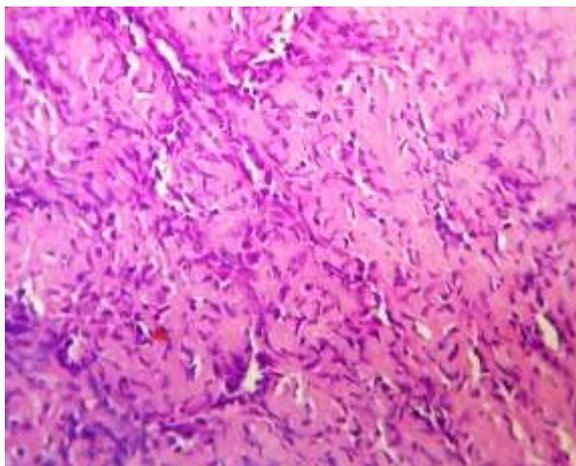


Fig. 2: Microphotograph showing bland, epithelioid tumor cells [Haematoxyline & Eosin, 40x]

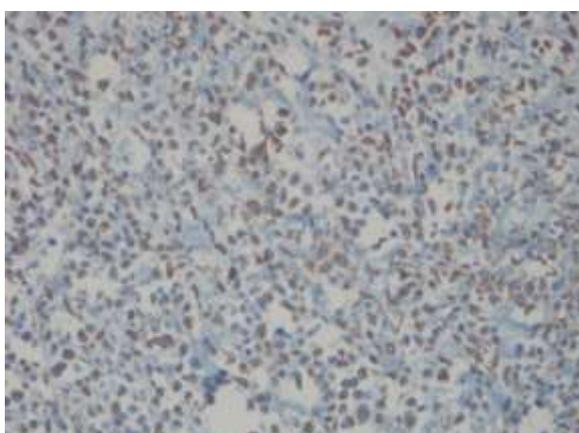


Fig. 3: Immunohistochemistry shows strong positivity for vimentin [Vimentin, 40x]

DISCUSSION

Sclerosing epithelioid fibrosarcoma is an uncommon tumor of deep soft tissue, usually affecting adults between 14 to 87 years of age (mean age – 45 years) [1]. The patient usually present with deep seated mass that is painful in one-third of cases [3]. This rare tumor usually occur in lower extremities/limb girdle but cases have been described in oral and maxillofacial region, trunk, upper extremities, sacrum, base of the penis and head and neck region[1,2,4,5].

Grossly, the tumor is grey white, usually well circumscribed and lobulated with occasional examples showing cystic or myxoid change, measuring mostly 5-10 cm. in greatest diameter [3].

Histologically, this lesion is characterised by nests and cords of uniform and relatively small round to ovoid epithelioid cells, which usually have a clear cytoplasm, set in an extensive, hyalinized stroma. This histological growth pattern in the form of infiltrating epithelioid cells with prominent sclerosis is highly suggestive of a carcinoma. Foci with a more fascicular pattern of conventional fibrosarcoma, also with myxoid and even chondro-osseous matrix are commonly

present. The chromatin pattern of the nuclei is finely stippled to vesicular with small and basophilic nucleoli. Polymorphism is minimal and mitotic figures are usually scarce whereas some tumors may have a high mitotic rate with more than 5 mitoses per 10 high power fields[1,3,6].

Immunohistochemically all tumors stain strongly with vimentin, and up to one half cases show membranous pattern of immunoreactivity with epithelial membrane antigen.

Neural markers including S-100 protein and NSE are positive in small number of cases [3]. Standard therapy is wide surgical excision with long term follow-up [3].

In spite of low grade histology, Sclerosing epithelioid fibrosarcoma has high recurrence rate and high metastatic potential. In a study of 25 cases done by Meis-Kindblom *et al.*, persistence disease or local recurrence was observed in 53% and metastasis in 43% of cases[1]. In the study of 16 cases done by Antonescu CR *et al.*, recurrence rate was 50% and distant metastasis was observed in 86 % of the patients [2]. Patients with tumour on trunk, those with large tumour, and those of male gender may have a worse prognosis [3].

CONCLUSION

Sclerosing epithelioid fibrosarcoma is a rare, slow growing, sarcoma of deep soft tissue with epithelioid tumor cells in nests and cords set in hyalinized fibrous stroma. It is a clinically aggressive but histologically low-grade sarcoma with unsatisfactory treatment results. As Sclerosing epithelioid fibrosarcoma has high rate of local recurrence and high metastatic potential this rare entity should be kept in mind in all suspicious soft tissue lesions.

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