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Case Report

Sclerosing epithelioid fibrosarcoma- A rare entity with a challenging presentation

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ABSTRACT

Fibrosarcoma is a malignant tumour arising from fibroblasts. Several variants have been described and these prove to be a diagnostic challenge owing to its many mimics, thus often requiring ancillary testing such as immunohistochemistry. The sclerosing epithelioid variant is a poorly recognised entity that poses a therapeutic dilemma. This is a case of Fibrosarcoma-sclerosing epithelioid variant reported in a 62-year-old male who presented with an exophytic lesion on the sole of the foot and was operated for the same. This article stresses on the systematic approach to such a case which will enable the pathologist to arrive at an accurate diagnosis.

1. Introduction

Adult type fibrosarcoma has largely become a diagnosis of exclusion. Fibrosarcomas represent lesser than 1% of all adult soft tissue sarcoma.[1] It is a rare sarcoma composed of monomorphic epithelioid cells with prominent hyalinised sclerotic collagenous stroma and a herringbone architecture.[2] Certain variants are particularly rare including sclerosing epithelioid fibrosarcoma which shares morphologic, immunohistochemical and cytogenetic features with low grade fibromyxosarcoma. It is a diagnostic challenge due to a significant number of histologic mimics.

2. Case Report

We report a case of a 62-year-old male who presented to the surgical OPD with complaints of a swelling over the left sole for 3 years following a history of trauma with recent discharge of blood mixed pus. It showed a progressive increase in size over this time period and was associated with pain. Incision and drainage was done for the abscess following which regular dressing and a complete course of antibiotics was taken. One month later, he presented with an exophytic growth over the same site for 8 days. On examination, an ulcero nodular growth was present on the plantar aspect measuring 10*9*8cm. FNAC done from the lesion showed cellular smears comprising of atypical tumour cells in singles, associated with myxoid stromal elements. These tumour cells were small with hyperchromatic nucleus, coarse chromatin with irregular nuclear border, indistinct nucleoli and scant vacuolated cytoplasm and was reported as Malignant round cell tumour (Figure 1). Edge Biopsy done from the same showed closely arranged spindle to oval cells in fascicles. Mitosis of 5-10/hpf seen.

Features were suggestive of Fibrosarcoma. Following this, MRI of the left foot was done which showed a large aggressive, vascular, diffusely enhancing soft tissue mass involving midfoot, encasing flexor and extensor tendons with lytic destruction of bones- soft tissue sarcoma. Subsequently CECT Chest and abdomen revealed well-defined nodules in both lower lobes of lungs which were suspicious of metastasis. Five days

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later, the patient underwent below knee amputation and the specimen was sent for histopathological examination. Gross examination of the specimen showed a single ulceronodular growth measuring 10*8.5*8cm, involving the plantar arches, tendons, muscle and metatarsal bone extending up to dorsum showing areas of necrosis and haemorrhage (Figure 2). Microscopy showed small round to oval cells arranged in nests and cords with stippled nuclei and scant clear cytoplasm, seen amidst sclerosed collagen. The tumour showed an infiltrative pattern within the sclerosed collagen. Very focal areas showed spindle cells in fascicles. Mitotic figures were 4/10 hpf (Figure 3). A diagnosis of Merkel cell carcinoma was given and immunohistochemistry was performed. Tumour was positive only for vimentin (Figure 3).

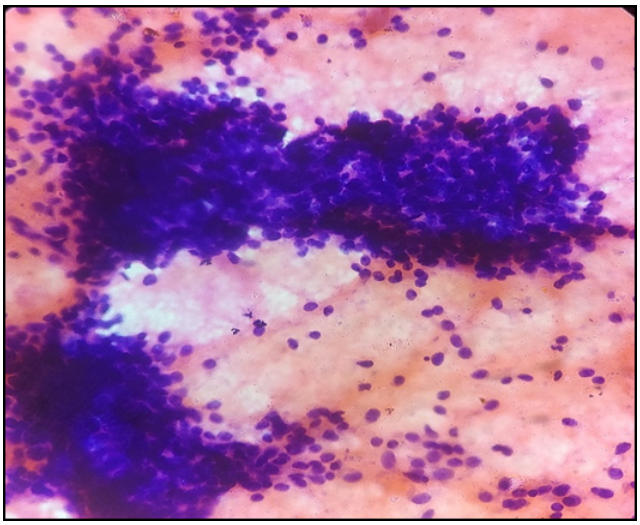

Fig. 1: Cellular smears showing small round to oval monomorphic cells in clusters. (H&E, x400)

Fig. 2: Gross examination showed an ulcero nodular growth involving the plantar arches, tendons, muscle and metatarsal bone extending up to dorsum showing areas of necrosis and haemorrhage

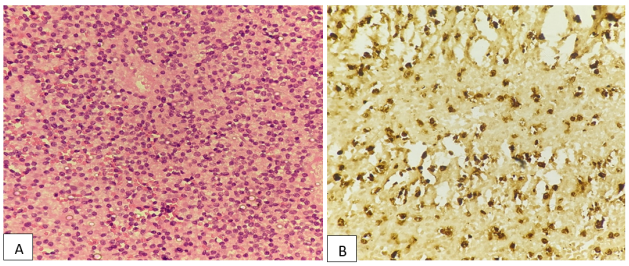

Fig. 3: A: Histopathologic examination shows small monomorphic round to oval cells in nests and cords having stippled chromatin and scant cytoplasm. (H&E, x200); **B:** Immunohistochemistry showing strong and diffuse positivity for Vimentin (x400)

The other markers CK20, EMA, SMA, CD99, NSE and TLE were all negative. However, MUC4 was not done. A diagnosis of sclerosing epithelioid Fibrosarcoma was given. The patient was then referred to the department of Oncology for further management and was started on adjuvant chemotherapy. The chemotherapeutic agents used were a combination of Adriamycin and Ifosfamide. After undergoing 2 cycles of chemotherapy, the patient decided to discontinue treatment. About 4 months later, he expired following hematemesis and breathing difficulty.

3. Discussion

Fibrosarcoma typically occurs as a deep-seated mass.[1] About 30 to 60% are found in the lower extremities but only 2.5% are confined to foot and ankle.[3] It is a diagnosis of exclusion with exception of recognisable variants like infantile and inflammatory.[4]

Sclerosing Epitheloid Fibrosarcoma, a variant of Fibrosarcoma was described by M.K Jm in 1995.[4] Majority arise in deep-seated structures in adults. Due to its rare occurrence and confusing imaging characteristics, preoperative imaging diagnosis is challenging.[5] The immunohistochemical profile was positive for Vimentin. Few cases have reported stain for S100 protein and EMA. Ultrastructural features will be those of fibroblasts. In our case, since the tumour cells were small oval to round mimicking small round cell tumour, Merkel Cell Carcinoma had to be ruled out. Hence, CK20, NSE and CD99 were done which were negative. SMA and TLE were also negative which ruled out leiomyosarcoma and synovial sarcoma respectively. MUC4 a transmembrane glycoprotein is a sensitive marker demonstrating cytoplasmic expression in Sclerosing Epitheloid Fibrosarcoma in 78% of the cases and 100 percent of cases in Low Grade Firbromyxosarcoma.[6]

4. Conclusion

Although initially diagnosed as a Merkel Cell Carcinoma considering the presentation and cell morphology, immunohistochemistry proved to be essential in this case to rule out all the possible histopathological mimics and to eventually arrive at the right diagnosis.

5. Source of Funding

None.

6. Conflict of Interest

The authors declare no conflict of interest.

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