Case Report

Myxoid pleomorphic sarcoma in a non-healing ulcer post-burn of the left foot: A rare case report

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ABSTRACT

Myxoid pleomorphic sarcoma is categorized as a soft-tissue sarcoma that most commonly appears in the lower extremities during adulthood, but rarely in the feet. We present a rare case of a primary myxoid pleomorphic sarcoma in the foot of a 45-year-old female which was rare in females. On examination, a ulceroproliferative growth of around 10 cm × 12 cm size was seen over the posterior aspect of foot involving the whole ankle region, with a thickened scar over the plantar and medial aspect of the right foot. Magnetic resonance imaging of the mass revealed a high signal intensity on T1-weighted images and heterogeneously high signal intensity on T2-weighted images without fat suppression. Histology showed variable cellularity, myxoid granular to the filamentous background, and round to spindled tumor cells. The immunohistochemistry showed positive for vimentin, acid mucins, CD34, and negative for S100. Hence, these findings were consistent with myxoid pleomorphic sarcoma. The patient was treated with below-knee amputation.

Key words: Myxoid pleomorphic, Non-healing, Sarcoma, Soft tissue, Ulcer

Liposarcoma is categorized as the second most common soft-tissue sarcoma with a peak incidence between the fourth and sixth decade of life with a slight preponderance toward the male gender. They commonly arise in the deep soft tissues of the extremities and retroperitoneum; however, the occurrence of liposarcoma in the foot or ankle is exceedingly rare. They are rare tumors of the foot with <1% occurrence [1]. Malignant fibrous histiocytoma (MFH) manifests a broad range of histologic appearances with four subtypes described: Storiform-pleomorphic, myxoid, giant cell, and inflammatory [2]. Surgery is the cornerstone of treatment for all soft-tissue sarcomas. The goal of management is surgery to eradicate all diseases in the affected area. Radiation has been clearly shown to improve the incidence of local recurrence.

CASE REPORT

A 45-year-old female came with complaints of non-healing wounds over the left-foot for 2 years. The wound was painless, progressively increase in size, associated with whitish, thick discharge, and associated deformed right foot at the ankle and first metatarsophalangeal region with a scar of the previous burn. There was a history of multiple burns over the face (10 years back), chest wall (7 years back), and foot (4 years back), while accidental fall of the patient in burning stove while cooking after an episode of seizure. A history of epilepsy for 10 years was present, and at present, she is on regular antiepileptic medication with no seizure attack after the treatment. There was no other significant medical, surgical, obstetrics, and menstrual history.

On examination, blood pressure was 128/74 mmHg, pulse rate was 74/min, and the temperature was afebrile. Local examination revealed that an ulceroproliferative growth of around 10 cm × 12 cm size was present over the posterior aspect of foot involving whole ankle region, with a thickened scar over the plantar and medial aspect of the right foot. The margins of the growth were everted (Fig. 1a). Deformed first metatarsophalangeal joint, with normal peripheral pulsations, was present.

Routine blood investigations were done and found within normal limits. No bone involvement was seen in X-ray foot. The chest X-ray was normal. Contrast-enhanced computed tomography chest was normal. Magnetic resonance imaging (MRI) of the

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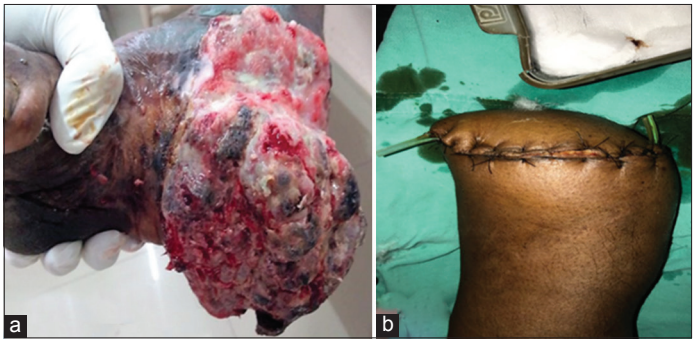

Figure 1: (a) Growth over the foot; (b) Post-operative below-knee amputation

mass revealed a high signal intensity on T1-weighted images and heterogeneously high signal intensity on T2-weighted images without fat suppression. CT chest was normal. Pre-operative tissue biopsy report showed chronic inflammatory ulcer over the foot.

The patient was offered excision with a reconstruction of the foot later but with poor cosmetic and functional results as well amputation of the limb with limb prosthesis. As per the willingness of the patient's attendant, below-knee amputation was done (Fig. 1b) and the limb was sent for histopathological examination.

On histopathological examination, the macroscopic tumor size was 9 cm × 8 cm and margins were free from tumor. Microscopic examination revealed variable cellularity, myxoid granular to the filamentous background in all cases. Tumor cells were round to spindled with variable shapes and sizes (Fig. 2). Immunohistochemistry showed positive for vimentin, acid mucins, CD34, and negative for S100. Hence, these findings were consistent with myxoid pleomorphic sarcoma.

The stump was closed with nylon 2-0, and corrugated rubber drain (CRD) placed. The CRD was removed on the 3rd post-operative day with a clean wound and no discharge. The patient was discharged on the 12th post-operative day with the removal of sutures. A follow-up course until 1 month was uneventful. The patient was suggested radiotherapy and oncology opinion for recurrence.

DISCUSSION

Soft-tissue sarcomas are benign mesenchymal tumors with more than 100 histological subtypes known so far. They are rare tumors with <1% occurrence. Detail immunohistological and ultrastructural evaluation leads to the classification of these tumors into various subtypes [3]. They can arise from any part of the bodies mainly the lower extremities, upper extremities, and retroperitoneum are some rare sites.

Considering the current classification of soft-tissue tumors, two tumors are most likely to affect the skin: Undifferentiated pleomorphic sarcoma (formerly, storiform-pleomorphic MFH) and myxofibrosarcoma (formerly myxoid MFH) [3]. Undifferentiated pleomorphic sarcomas, also known as MFH, are malignant tumors of uncertain origin whose histopathology is not very clearly defined [4]. This tumor was first described by Kauffman and Stout [5]. They described MFH as a tumor

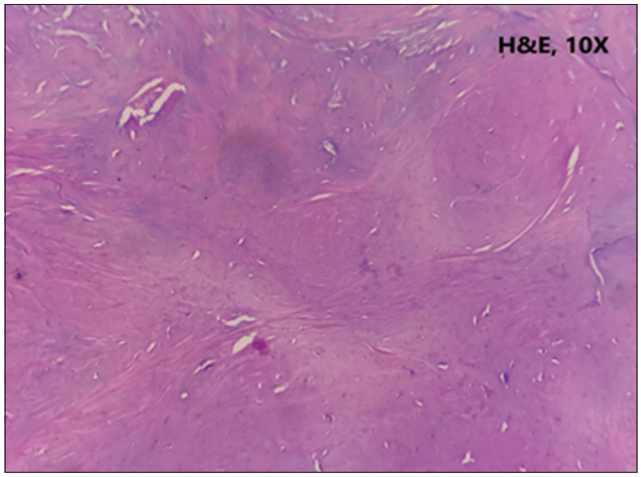

Figure 2: Histopathological image showing variable cellularity, myxoid granular to filamentous background, and round to spindled tumor cells

rich in histiocytes with a storiform growth pattern. Before 1977, it was considered as the most common soft-tissue tumor. In 2002, the World Health Organization declassified MFH as a formal diagnostic entity and renamed it as an undifferentiated pleomorphic sarcoma not otherwise specified.

MFH manifests a broad range of histologic appearances with four subtypes described: Storiform-pleomorphic, myxoid, giant cell, and inflammatory [2]. The typical age for presentation is 50–70 years, although it can appear at any age with slight male preponderance [6].

Usually, an X-ray is taken as the first diagnostic modality followed by MRI. While suspecting sarcoma, it is important to determine if a tumor is isolated (localized) or has spread (metastatic) as they may be associated with vascular, bony involvement, or lung metastasis. Thus, CT chest or X-ray chest is always done to rule out lung metastasis.

Surgery is the cornerstone of the treatment for all soft-tissue sarcomas. The goal of surgery is to eradicate all diseases in the affected area. Radiation has been clearly shown to improve the incidence of local recurrence and has become an integral part of the treatment for MFH. In spite of effective treatment, 25% develop distant metastasis [7]. The role of chemotherapy in the treatment of MFH is still not clear. Doxorubicin therapy is still under trial [8].

CONCLUSION

It is a rare presentation of a soft-tissue lesion over the foot. It is uncommon in female patients who have a history of burn due to epilepsy. Hence, Marjolin and other post-burn ulcers should be kept in mind as a differential diagnosis. Early diagnosis, metastatic workup, and early treatment with radiotherapy can benefit to patient survival.

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