

Case Report

A unique case of classic pleomorphic sarcoma restricted to the toes

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ABSTRACT

Over 50% of soft tissue sarcomas occurring in older adults are histologically pleomorphic and high grade. Most have traditionally been classified as malignant fibrous histiocytoma (MFH). MFH was originally defined as a malignant pleomorphic spindle cell neoplasm showing fibroblastic and histiocytic differentiation. More recently, pathologists have accepted that this morphology may be shared by a wide range of malignant neoplasms. Many sarcomas that were previously classified as pleomorphic MFH, on careful immunohistochemical and histopathologic analyses, revealed a specific line of differentiation and could be reclassified as myxofibrosarcoma (30%), myogenic sarcoma (30%), liposarcoma (4%), malignant peripheral nerve sheath tumor (2%), or soft tissue osteosarcoma (3%), whereas about 30% had no specific line of differentiation or were myofibroblastic. The term undifferentiated pleomorphic sarcoma (UPS) is now reserved for pleomorphic sarcomas that show no definable line of differentiation by current technology. The majority of extremity sarcomas occur in the lower extremity (74 vs. 26% in the upper limb). According to one of the studies conducted on 315 patients, non-metastatic soft tissue sarcoma of the lower extremity who were treated at one institution over a ten-year period. Sixty-six percent of the lesions were above the knee, and 60% were high grade. This case had a 3x3 cm ulcer at the 3rd toe in a 30-year-old male patient who subsequently underwent midfoot amputation.

Keywords: Undifferentiated pleomorphic sarcoma, Malignant fibrous histiocytoma, Myofibroblastic

INTRODUCTION

Soft tissue sarcomas account for less than 1% of all cancers. UPS, formerly called malignant fibrous histiocytoma and declassified by the world health organization in 2002, is a rare and malignant subtype.¹ These tumors are the fourth most common soft tissue sarcoma and have an incidence of about 0.08-1 per 100,000.² Sarcomas typically present in the sixth and seventh decades of life and tend to occur in the extremities, retroperitoneum, viscera, and head and neck. Here we present a case had a 3x3 cm ulcer at the 3rd toe in a 30-year-old male patient who subsequently underwent midfoot amputation.

A 30-year-old male presented to OPD with a non-healing ulcer in the right third toe associated with pain since past 6 months and was on conservative management. On physical examination general condition of the patient was stable. Ulcer of size 3x3 cm present over the planter aspect of 2nd and 3rd toe and was tender and hard.

Peripheral pulses were palpable with no loss of sensation. There was evidence of lymphadenopathy on USG showing enlarged popliteal lymph nodes as well. The other toes were normal. The patient underwent debridement with split thickness skin grafting.

Microscopic examination of the sections showed presence of benign spindle cells in small clusters and

lying singly. The morphological diagnosis at this stage was consistent with benign spindle cell lesion.

Patient came again to OPD with similar ulcer involving the 4th toe and loss of graft for which amputation of 3rd and 4th toe was done Figure 1. Gross examination of the specimen revealed irregular soft tissue tumor measuring 3x1.5x1.5cm which is attached to the 3rd metatarsal encircling the bone. Cut section of the tumor was greyish white and homogenous Figure 2.

HISTOPATHOLOGY

Microscopic examination of the sections showed infiltrating spindle cell. The tumor cells were mostly arranged in storiform pattern and short fascicles. The cells are pleomorphic showing high N:C ratio, hyperchromatic nuclei and scanty fibrillary cytoplasm. Good number of giant cells seen. Mitotic count is 28/10 HPF. The tumor is infiltrating and extending up to epidermis. Epidermis was free of tumor. Initial diagnosis of spindle cell sarcoma was made. The morphological differential diagnosis at this stage was metaplastic carcinoma, leiomyoma, malignant peripheral nerve tumor Figure 3.



Figure 1: Pre-operative photograph showing recurrent lesion (pleomorphic sarcoma).



Figure 2: Post-operative photograph following midfoot amputation.

Immunohistochemistry

On immunohistochemistry, only vimentin was positive. Cytokeratin (CK) and epithelial membrane antigen (EMA) negativity ruled out metaplastic carcinoma and phyllodes Figure 4. Smooth muscle actin (SMA), desmin, CD64, and CD34 excluded leiomyoma, stromal sarcoma, inflammatory myofibroblastic tumor, and phyllode tumor. The possibility of malignant peripheral nerve sheath tumor and liposarcoma was not considered as S-100 protein and synaptophysin were negative. Based on histological features and immunohistochemical study, diagnosis of UPS sarcoma was made

Following collaboration with department of oncosurgery midfoot amputation was done.

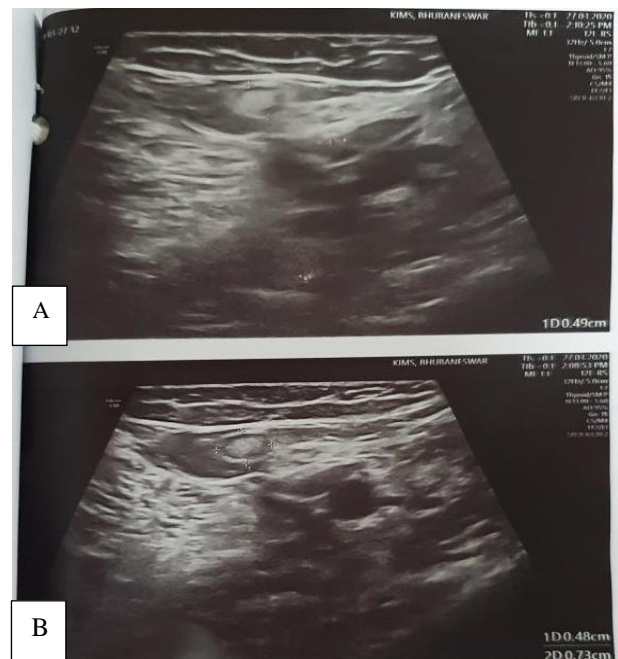


Figure 3 (A and B): Small lymphnode of 4.8 mm in popliteal fossa.

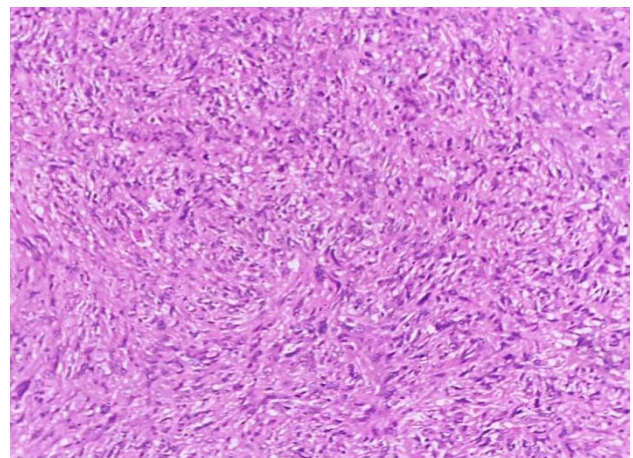


Figure 3: Histologic appearance of pleomorphic sarcoma.

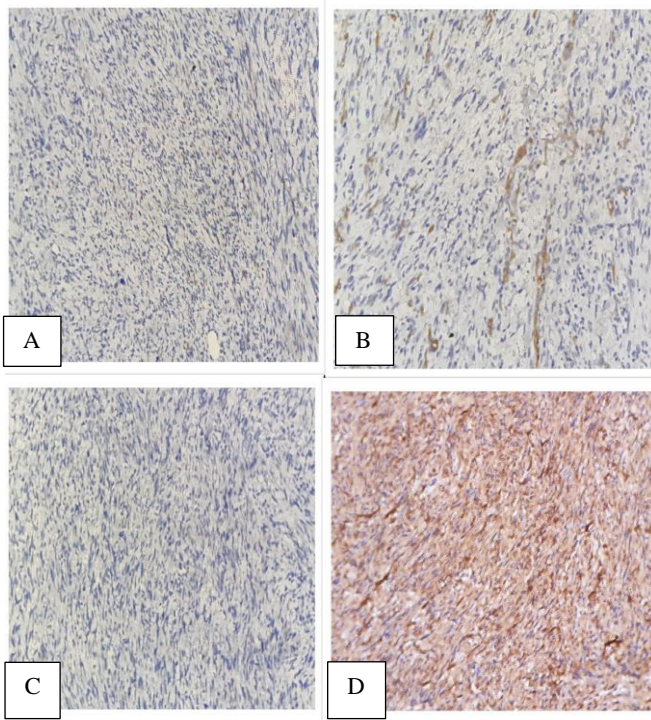


Figure 4 (A, B, and D): Undifferentiated pleomorphic sarcoma of the toes showing negative for cytokeratin, smooth muscle actin and 100. Positive for vimentin.

DISCUSSION

Tumors arising in the soft tissue form a diverse and complex group, as they may display varying degrees of mesenchymal differentiation. Most soft tissue tumors are benign and are usually cured with a simple surgical excision. Soft tissue sarcomas account for <1% of the overall human burden of malignant tumors but remain life-threatening, and approximately 40% of patients with newly diagnosed soft tissue sarcoma die of the disease, corresponding to approximately 4,000 deaths each year in the United States.³ Soft tissue sarcoma, diagnosed at an early stage, is eminently curable. When diagnosed at the time of extensive local or metastatic disease, it is rarely curable.⁴ The relatively small number of cases and the great diversity in histopathologic features, anatomic sites, and biological behaviors have made comprehensive understanding of these disease entities difficult. A better understanding is urgently needed to accelerate the development of new treatments.

Epidemiology

Benign mesenchymal tumors are 100-fold more common than soft tissue sarcomas. The annual international incidence of soft tissue sarcoma is estimated to be between 1.4 and 5.0 cases per

100,000.⁵ True incidence remains difficult to determine because of variable reporting practices and inaccurate diagnosis.

Etiology

Most soft tissue sarcomas are believed to be sporadic and have no clearly defined cause. In a small proportion of cases, researchers have identified predisposing or associated factors, including genetic factors, lymphedema, prior radiation therapy, and carcinogens.

Anatomic and age distribution

Soft tissue sarcomas can occur in any site throughout the body. A total of 45% are located in the extremities, with 30% of all lesions occurring in the lower limb (most commonly in the thigh); 38% are intra-abdominal, divided between visceral (21%) and retroperitoneal (17%); 10% are truncal; and 5% are head and neck (Figure 5).⁶ Soft tissue sarcomas become more common with increased age, and the median age at diagnosis is 65 years.⁷ However, the median age varies significantly by histologic type and subtype. In general, the median age of onset tends to be 20 to 50 years in the translocation-associated sarcomas and 50 to 70 years in the complex sarcoma types.⁸

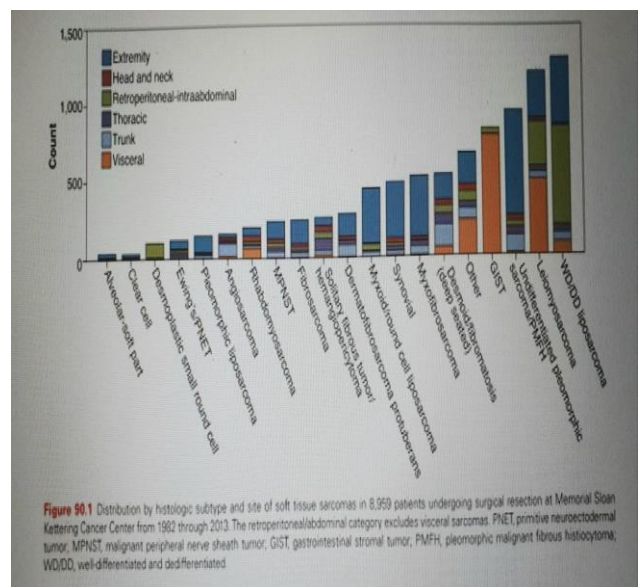


Figure 5: Distribution by histological subtype and site of soft tissue sarcomas.

CONCLUSION

Although the incidence of pleomorphic liposarcoma in the toes is very low, it is essential to perform

thorough histological analysis of all soft tissue masses, regardless of their benign appearance, because only prompt radical surgery can result in a good prognosis for the patient.

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Ethical approval: Not required

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