



Pleomorphic Liposarcoma of Finger—a Rare Entity

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Introduction

Liposarcoma (LPS) is a tumor of adipose tissue and is one of the common soft tissue sarcomas of adults [1]. They comprise approximately 16–18% of all malignant soft tissue tumors [2]. They can be categorized into three different subtypes by clinicopathological and molecular characteristics: well-differentiated/dedifferentiated liposarcoma, myxoid/round-cell liposarcoma, and pleomorphic liposarcoma (PLS) [1, 3]. Among these rarest is PLS which represents 5 to 15% of liposarcomas [1, 4]. PLS usually is seen in elderly patients (> 50 years) and has an equal sex distribution [1, 2]. It commonly occurs in the extremities more commonly in the lower. Patients usually present with a firm, enlarging mass. Investigations which can help for diagnosis and management include ultrasonography (USG), magnetic resonance imaging (MRI), computed tomography (CT) scan, or positron emission tomography–computed tomography (PET-CT) [1, 2, 5]. Surgical excision remains the mainstay of treatment [1, 5, 6]. Grossly, PLS usually appears as relatively well circumscribed unencapsulated tumor with yellow to gray tan appearance [1]. Microscopy shows pleomorphic spindle-shaped to small round cells admixed with a variable number of pleomorphic lipoblast which is diagnostic of PLS [4]. PLS is considered as high-grade sarcoma with local recurrence and metastatic rates of 30 to 40% and an overall 5-year survival rate of 55 to 65% [1, 4]. Here, we report a case of PLS in a 67-year-old male who presented with swelling over the left ring finger.

Case History

A 67-year-old male presented with swelling over the left ring finger on the palmar aspect of 8-month duration. Initially, swelling was small in size but gradually increased to present size of about 5 cm × 4 cm for 6 months. The patient gave a history of restricted finger movements. There was no history of pain/discharge from the swelling. Overlying skin was stretched (Fig. 1). X-ray of the left hand showed soft tissue density along the ventral aspect of the proximal portion of the left ring finger with mild periosteal reaction. USG of the left ring finger showed a large, well-demarcated, isoechoic, highly vascular, solid mass with few cystic spaces along the ventral aspect. The mass was wrapping around the proximal phalanx of the left ring finger, extending dorsally without obvious cortical breach. The possibility of a soft tissue arteriovascular malformation was considered. MRI of the left wrist showed intensely enhancing, multilobulated soft tissue lesion abutting the flexor tendon on the palmar aspect of the 4th proximal phalanx extending to the dorsal aspect (Fig. 2). Probable diagnosis of synovial sarcoma was made. USG abdomen and pelvis and contrast-enhanced computed tomography (CECT)

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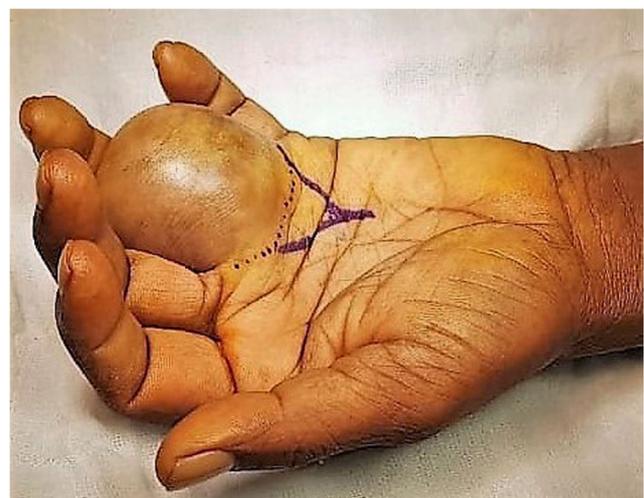


Fig. 1 Nodular mass on the palmar aspect of the left ring finger



Fig. 2 MRI lateral view, showing intensely enhancing, multilobulated soft tissue lesion without bone erosion

thorax did not show any evidence of metastasis. Fine-needle aspiration cytology (FNAC) showed pleomorphic, polygonal to spindle cells, arranged in tight clusters and singles displaying marked pleomorphism, bizarre hyperchromatic nuclei, and cytoplasmic vacuoles containing occasional brown pigment. Cytological features were suggestive of soft tissue sarcoma favoring clear cell sarcoma. Amputation of 4th left ring finger along with the swelling was done with primary closure (Fig. 3). Gross appearance of the specimen was a nodular soft tissue swelling at proximal end on the palmar aspect of ring finger measuring 5 cm × 4 cm × 4 cm. The cut surface of the lesion showed a relatively well-circumscribed tumor. The tumor was lobulated and yellowish with myxoid areas. Focal areas of necrosis and calcification were noted



Fig. 3 Postoperative status—amputation of left ring finger with primary closure



Fig. 4 Cut surface of the mass, showing well-circumscribed tumor with yellow lobulated areas and necrosis

(Fig. 4). The tumor appeared to be abutting underlying tendon sheath and extending upwards to dermis sparing the overlying skin. Histopathology showed a well-circumscribed, partly encapsulated tumor in subcutis and reticular dermis containing pleomorphic spindle-shaped to small round cells admixed with variable pleomorphic, multivacuolated lipoblast with bizarre, hyperchromatic, irregular to scalloped nuclei (Fig. 5). Adipocytes with enlarged hyperchromatic nucleus were also seen (Fig. 6). Tumor cells showed frequent mitosis (10–12/high-power field). Cytoplasm was moderate to abundant which was eosinophilic to vacuolated. At places, spindle cells were in fascicles and were arranged around blood vessels in

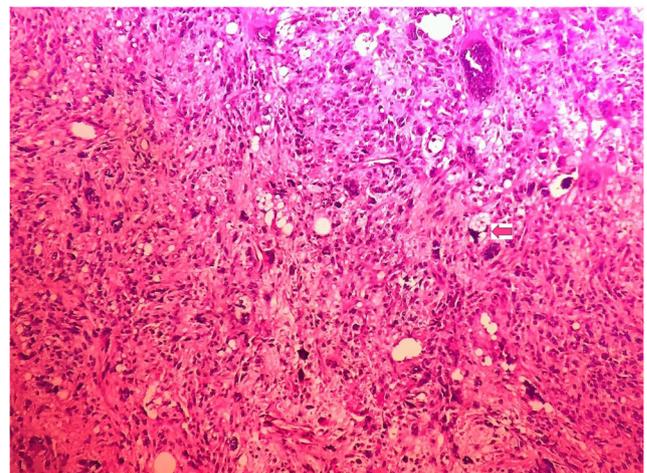


Fig. 5 Microscopy showed pleomorphic spindle-shaped to epithelioid cells, pleomorphic, multivacuolated lipoblast with hyperchromatic, irregular to scalloped nuclei, H&E, × 10

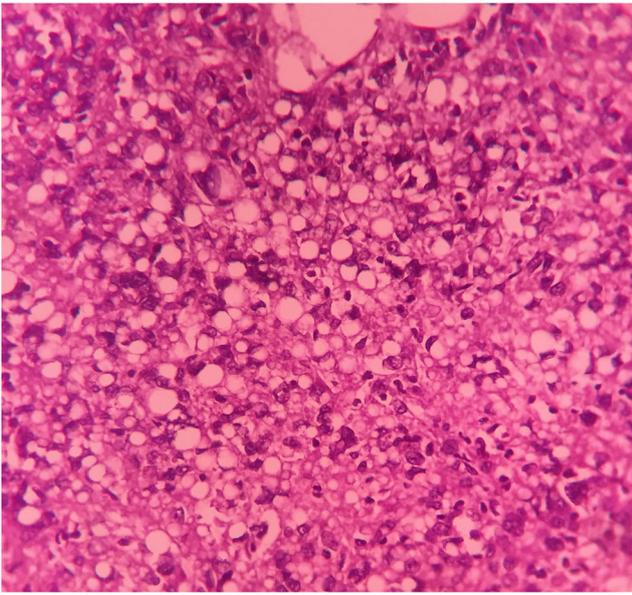


Fig. 6 Adipocytes with an enlarged hyperchromatic nucleus, H&E, $\times 10$

the hemangiopericytoma-like pattern. Multinucleated tumor giant cells were also seen (Fig. 7). Stroma showed myxoid areas and extensive necrosis (40%). There was no angiolymphatic invasion or invasion of the tumor into underlying tendon or bone. The surgical resected margin was free of tumor. Histopathological features were suggestive of pleomorphic liposarcoma—grade 3 with pathological staging pT2. Immunohistochemistry (IHC) showed tumor cells positive for vimentin (Fig. 8), negative for smooth muscle actin (SMA), desmin, and CD34. Ki-67 index was 20% (Fig. 9). IHC was consistent with high-grade pleomorphic liposarcoma. The patient received three cycles of chemotherapy on subsequent follow-up and is doing well during treatment.

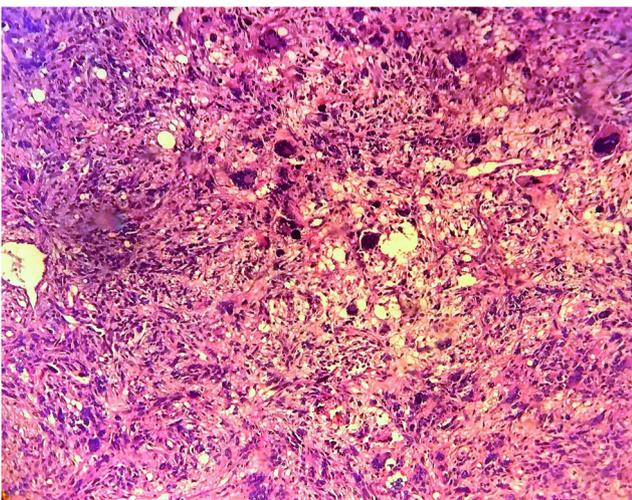


Fig. 7 Multinucleated tumor giant cells, H&E, $\times 10$

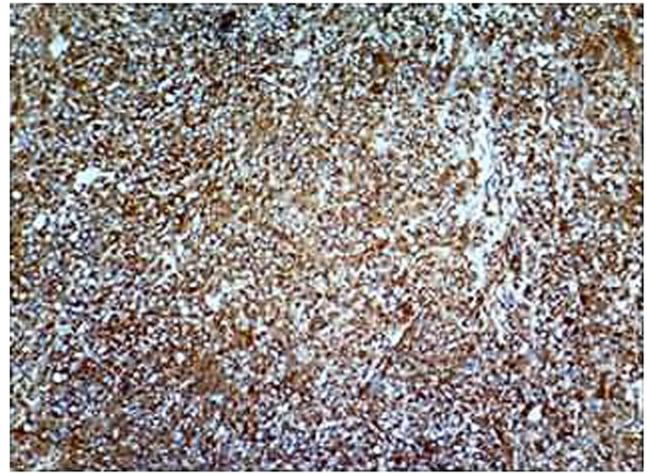


Fig. 8 Immunohistochemistry—vimentin positive

Discussion

Pleomorphic liposarcoma is the rarest of all liposarcomas, represents 5 to 15% of all liposarcoma [4]. PLS typically occurs in elderly patients (> 50 years) with an equal sex distribution [1, 2]. Most common site of occurrence is the extremities (lower > upper limbs), whereas trunk and retroperitoneum are less frequently affected. Approximately 15% of these occur in the upper extremity, and only 4% occur in hand [5]. Rare sites of involvement include mediastinum, paratesticular region, scalp, abdominal and pelvic cavities, and orbit [7]. Patients usually present with painless soft tissue swelling or mass of prolonged period. Other symptoms include rapid growth with skin ulceration and occasionally pain [1, 5]. Our patient was a 67-year-old male, who presented with a swelling on the palmar aspect of the left ring finger with restriction of movements for 8 months.

Radiological investigations which can aid in diagnosis include X-ray, USG, CT scan, and MRI. Plain radiograph detects the fat density of lipomas, soft tissue calcification, and even bone erosion [5]. Typically on CT and MR imaging, they

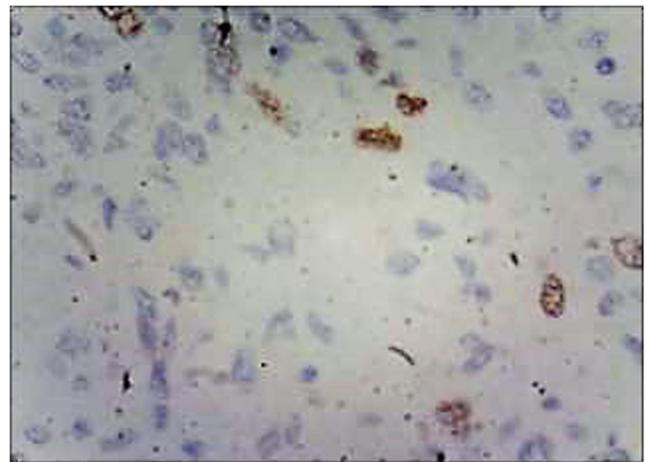


Fig. 9 Ki 67 index (20%)

show inhomogeneous appearance with areas of necrosis and irregular enhancement [2]. In our patient, X-ray of the left hand showed soft tissue density along the ventral aspect of the proximal portion of the left ring finger with mild periosteal reaction. Ultrasound of left ring finger mass showed a large, well-demarcated, isoechoic, highly vascular, solid mass with few cystic spaces. MRI of the left wrist showed intensely enhancing, multilobulated soft tissue lesion abutting the flexor tendon on the palmar aspect of the 4th proximal phalanx extending to the dorsal aspect. FNAC showed features of soft tissue sarcoma.

Pleomorphic liposarcoma is treated by surgical excision. When it occurs in the fingers, amputation is the treatment of choice [5]. The reconstructive surgeon must be well versed in microsurgical techniques, such as nerve or vessel repair and grafting, variety of local flaps, and bone fixation techniques to ensure optimal hand function [6]. In our case, amputation of 4th left ring finger along with the mass with primary closure was done.

Grossly, pleomorphic liposarcomas are usually well circumscribed with partial encapsulation. They are firm in consistency and grayish yellow in appearance with variable myxoid areas. Necrosis may be seen in tumors of large size. Histopathology shows spindle to epithelioid cells with marked pleomorphism with a variable amount of atypical lipoblasts and bizarre multinucleate giant cells in interlacing bundles with areas showing a hemangiopericytoma-like pattern. Areas of coagulative necrosis and lymphovascular invasion can be observed [1]. Grading of soft tissue sarcoma depends on summative scores on tumor differentiation, mitotic rate, and amount of tumor necrosis. According to French Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) system, pleomorphic liposarcoma is graded as grade 3 [8].

On IHC, PLS shows positivity for S100 protein and vimentin and negative for cytokeratin, epithelial membrane antigen, and desmin. The Ki67 index component varies from 10 to 40% with an average of 30% [1]. In our case, IHC showed positivity for vimentin, negative for SMA, desmin, and CD34. Ki-67 index was 20%, suggesting a diagnosis of high-grade pleomorphic liposarcoma.

Chemotherapy is advised in patients with tumor size between 5 and 10 cm [5]. Chemotherapy regimen includes

doublets of doxorubicin/ifosfamide or gemcitabine/docetaxel, cyclophosphamide, and dacarbazine. Response rates are 25 to 35% with the survival of 12 to 18 months [4, 6, 9]. The probability of local recurrence, metastasis, and mortality rate ranges between 25 and 45%, 32 and 44%, and 32 and 50% respectively. As compared to deep-seated tumors, subcutaneous PLSs have a much more favorable clinical outcome [1]. In our case, the patient has presently completed three cycles of chemotherapy containing adriamycin, ifosfamide, cyclophosphamide, human granulocyte colony-stimulating factor (G-CSF).

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