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Case Report

Recurrent cutaneous leiomyosarcoma of the anterior abdominal wall: a rare tumor at an unusual site

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ABSTRACT

Cutaneous leiomyosarcomas are uncommon neoplasms, accounting for 1-3% of all the soft tissue sarcomas. They encompass a variety of clinicopathological features that makes their diagnosis difficult. They are divided into various categories, as this division has an important prognostic implication. We describe a rare case of recurrent cutaneous leiomyosarcoma of the anterior abdominal wall in a 50 years old female.

Keywords: Leiomyosarcoma, Recurrence, Cutaneous, Abdominal wall

INTRODUCTION

Soft tissue sarcomas comprise approximately 0.7% of all malignant neoplasms, and leiomyosarcomas (LMS) have been reported to account for 3-7% of soft tissue sarcomas.[1,2] LMS are divided into those involving deep soft tissues sites as the retroperitoneum and those involving peripheral soft tissue sites (superficial LMS). Based on their clinical and biological differences, superficial LMS are further sub-divided into site related two groups, cutaneous and subcutaneous LMS.[3,4] Cutaneous LMS are thought to arise from the arrector pili muscles in the dermis and may have extension into the underlying tissues.[5] Subcutaneous LMS originate from small to medium sized vessels in the subcutaneous tissue and have more aggressive behaviour. These sarcomas occur in adult life with a peak in 50-70 years of age group with male preponderance.[3,6] They may arise anywhere on the body with predilection for the lower extremities. The trunk is rarely involved by LMS.[7] Preoperative misdiagnosis is frequent for LMS because of their rarity and non-specific presentation. We herein report the clinical, cytological, histopathological and immune-histochemical findings in a 50 years old female with recurrent cutaneous leiomyosarcoma arising in the anterior abdominal wall.

CASE HISTORY

A 50 years old female patient presented to surgical out-patient department of our hospital with a lump in the upper abdomen which had been progressively increasing in size since last 2 months. On inspection, there was a lump in the right hypochondriac region and scar marks in the hypogastric, umbilical and epigastric region of the abdomen. On palpation, an irregular, firm, tender swelling, measuring 3 cm x 3 cm, with overlying skin showing congestion was noted in the right hypochondriac region as shown in Figure 1. Inguinal lymph nodes were not palpable. Her past history was significant as she had similar swelling in the epigastric region, seven months back for which she had surgery in our hospital. She also revealed that she had similar swelling in umbilical region, 5 years back for which she had surgery, but records pertaining to it were unavailable with the patient. Her systemic examination and investigations, including complete blood count, fasting blood sugar level, liver

function test and renal function test, were within normal limits. Hepatitis B virus surface antigen and human immunodeficiency virus enzyme-linked immunosorbent assay were non-reactive. A clinical diagnosis of malignant tumor was considered and fine needle aspiration cytology (FNAC) was advised. On FNAC, smears were highly cellular and showed numerous spindle cells with cigar shaped blunt-ended nuclei. Occasional cells showed prominent nucleoli. The cells exhibited moderate nuclear atypia and few mitotic figures could be identified as shown in Figure 2. On immunocytochemistry, the tumor cells were positive for vimentin, smooth muscle actin (SMA), desmin and were negative for pancytokeratin (CK), Epithelial Membrane Antigen (EMA), myogenin, CD34 and S100. Based on these features, a diagnosis of malignant mesenchymal tumor with a possibility of leiomyosarcoma was made. The swelling was excised with a 5 cm resection margin and submitted for histopathological examination. The defect was reconstructed with omentum and prolene mesh. The skin defect was covered with rotational skin flaps and split skin grafting.

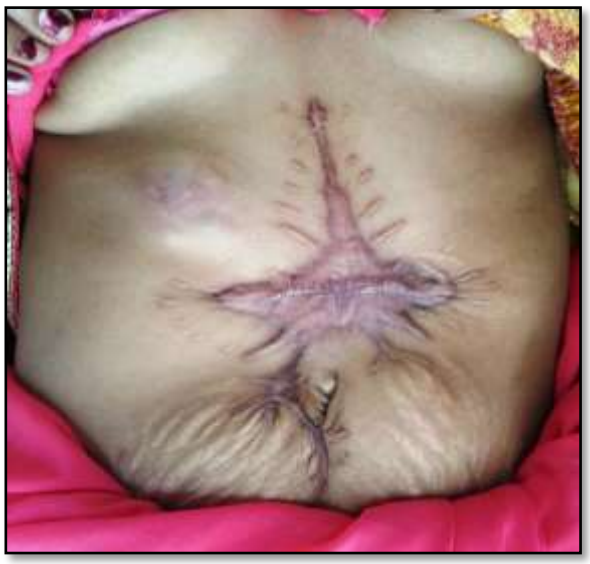

Figure 1: Mass in right hypochondriac region adjacent to the scar marks of previous surgery on clinical inspection.

Gross inspection of the specimen received revealed a grey brown, irregular, nodular soft tissue mass, measuring 4 cm x 4 cm x 2 cm and partly covered with skin. The cut surface of the mass was greyish white, solid, homogenous and fleshy.

Microscopic sections showed a poorly delineated dermal neoplasm extending into the underlying subcutis. The neoplasm was composed of bundles of elongated spindle cells arranged in interlacing fascicles, with intensely pink, fibrillary cytoplasm and pleomorphic nuclei with coarse irregularly dispersed chromatin. Mitotic figures, including atypical (15 mitotic figures per 10 high power fields) were evident with an occasional tumor giant cell. Focal necrosis was noted. The overlying epidermis was thinned out with areas of ulceration as shown in Figure 3a & 3b. The surgical margins were free of the tumor. A provisional diagnosis of malignant spindle cell neoplasm of the skin, which typically includes a host of diagnostic possibility, namely fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumor, monophasic synovial sarcoma, malignant fibrous histiocytoma and spindle cell variant of squamous cell carcinoma, was considered and immunohistochemistry was advised for confirmation. On immunohistochemical analysis, the tumor cells were positive for SMA, desmin and vimentin as shown in Figure 3c and negative for CK, EMA, S100, CD-117, CD34 and myogenin. Based on these features, the diagnosis of a cutaneous leiomyosarcoma was established.

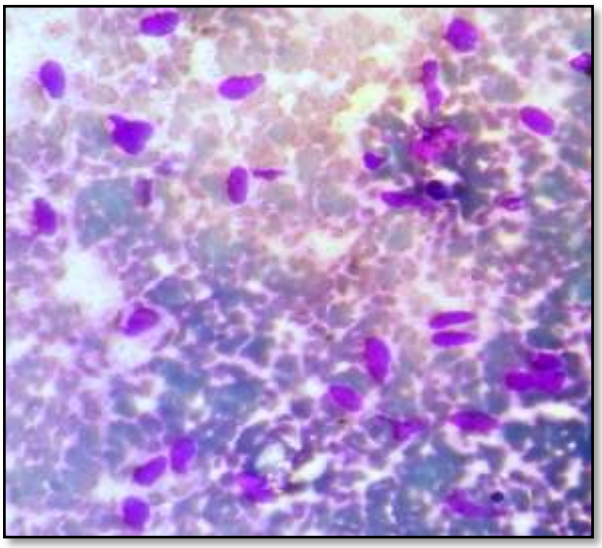

Figure 2: Photomicrograph of cytological smears showing spindle shaped cells (MGG, x 40).

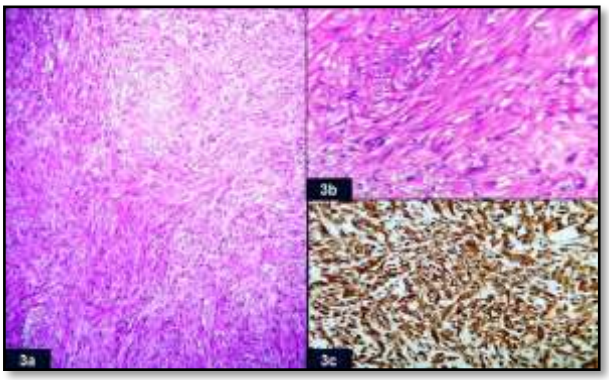

Figure 3: (a) Tumor composed of interlacing fascicles of elongated spindle cells (H & E, x10); (b) Tumor cells exhibiting intensely pink, fibrillary cytoplasm and pleomorphic hyperchromatic nuclei (H & E, x40); (c) SMA positivity of the tumor cells (IHC, x20).

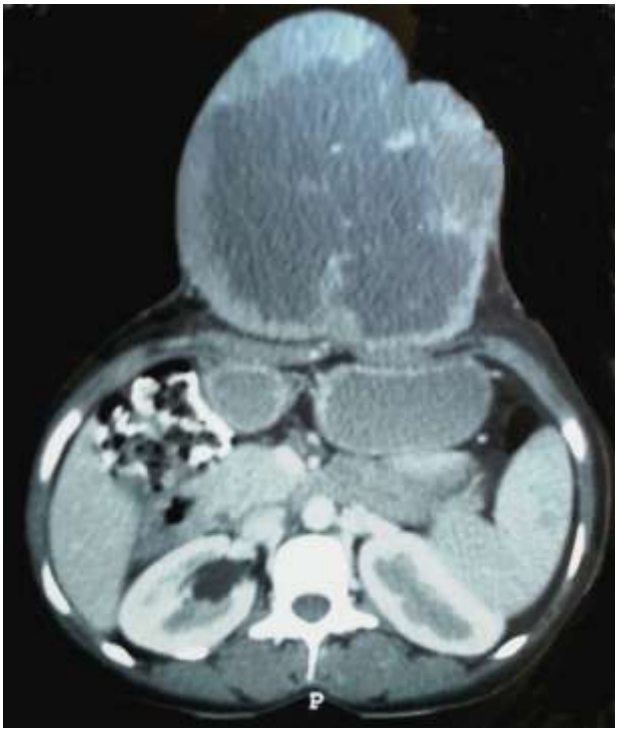

Figure 4: CT abdomen demonstrating a large heterogeneously enhancing abdominal mass.

Simultaneously, based on her past history, her previous records available in the surgery department were investigated, which revealed that she had presented seven months back with a large abdominal mass. On physical examination, the mass was 12 cm x 10 cm x 10 cm in size and was located at the epigastric region. It was firm in consistency, non-mobile, non-tender. Overlying skin showed multiple dilated blood vessels. Radiological workup was also done at that time. Ultrasonography, revealed a large solid lobulated exophytic mass in the epigastric and supraumblical region. On computer tomography (CT) scan abdomen, a large heterogeneous enhancing mass with central hypodense necrotic area in the anterior abdominal wall (supraumblical) was noted which infiltrated the left rectus muscle, however, no intraperitoneal infiltration was observed as shown in Figure 4. Tru-cut biopsy of the mass was done. Histopathological examination supported by immunohistochemistry confirmed the diagnosis of leiomyosarcoma. On the basis of which, surgical excision with wide margins of at least 2 cm was performed. The histopathological findings were identical to the current findings of the patient. So, based on the past history as well the current clinicopathological findings in this patient, a final diagnosis of recurrent cutaneous leiomyosarcoma of the anterior abdominal wall was made. Although a complete resection with wide surgical margins had been performed, adjuvant postoperative radiation therapy was administered. The patient is being followed up and is symptom free since last 6 months after the resection of the recurrent tumor.

DISCUSSION

LMS, being exceedingly rare entity, was initially described by Montgomery and Winkelman in 1959.[8] Among them, cutaneous LMS are rare, constituting 1-3% of all soft tissue sarcomas. They occur as solitary, slow growing lesion majority conceding with hair bearing areas of body. Very few cases have been described in literature involving the abdominal wall.[6] Etiology of these tumors is yet uncertain though predisposing factors like traumatic scar, previous radiation exposure and leiomyomas have been reported in the literature.[9] Cutaneous LMS have variety of clinical presentations but the usual symptom is pain, which can either be spontaneous or induced by pressure like in our case. Its surface can be smooth, ulcerated or, rough, verrucous and sometimes indurated and hemorrhagic. On histopathology cutaneous LMS are like other soft tissue sarcomas which are fairly cellular, comprising of spindle shaped cells arranged in fascicles. Individual cell is elongated shaped with abundant eosinophilic cytoplasm, the nucleus is centrally located and blunt ended or "cigar shaped". Criteria for malignancy include the presence of mitoses of at least one per 10 high power fields, high cellularity, significant nuclear atypia and tumor giant cells. Histological variants of cutaneous LMS can lead to diagnosis difficulties as in the case for epithelioid, granular cell, desmoplastic, inflammatory and myxoid LMS.[10] Morphological differential diagnosis includes a host of other malignant spindle cell neoplasms, thereby necessitating the use of a panel of immunohistochemical markers to arrive at a definitive diagnosis. Spindle cell lesions, very often confused with superficial LMS include dermatofibrosarcoma protuberans, nodular fasciitis, fibrosarcoma, neurosarcoma, dermatofibroma, neurofibroma, rhabdomyosarcoma, atypical fibro-xanthoma, malignant fibroxanthoma and synovial sarcoma.[11] Immunohistochemistry plays a key role in differential diagnosis and in pleomorphic or poorly differentiated cases.[7] Majority of the cases are positive for smooth muscle actin (SMA), desmin, heavy caldesmon and smooth muscle myosin. Amongst them SMA is the most sensitive marker and has been reported to be positive in 100% of the cases of LMS.[3,12] In this patient, the characteristic histological features and positivity for SMA clinched the final diagnosis.

These tumors rarely metastasize but relapse may be seen in up to 50% cases, therefore the best way of management is wide local excision with 3-5 cm wide margins and a depth that includes subcutaneous tissue and fascia in order to avoid recurrences.[13-16] The response to radiotherapy and adjuvant therapies is still not well defined in these tumors.[17]

Several factors are correlated to prognosis. These include tumor size, high mitotic rate, presence or absence of necrosis, intratumoral vascular invasion.[18,19] Never-theless, the prognosis often remains poor. The survival

rate for tumors smaller than 2 cm was of 95%, while in tumors that exceeded 5 cm, survival drops to 30%.[20]

CONCLUSION

Cutaneous leiomyosarcoma of the anterior abdominal wall is a rare clinical entity which can have multiple recurrences, so long term follow-up is mandatory even after wide surgical excision of the primary tumor. This neoplasm should always be kept in mind while encountering a large abdominal mass or a malignant spindle cell neoplasm of the skin. Both cytology and histopathology along with immune markers play a pivotal role in its definitive diagnosis.

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