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A GIANT ANTERIOR ABDOMINAL WALL LEIOMYOSARCOMA: CASE REPORT AND REVIEW OF LITERATURE

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INTRODUCTION

Soft tissue sarcomas are one of the rare condition in medical world with occurrence of approximately 1% of adult solid malignant cancer.[1] Almost 50 different histological subtype of sarcoma occur which may arise either from stem cell like population (primitive neuroectodermal tumors) or from morphological recognizable connective tissue.[2] Leiomyosarcoma (LMS) arises from smooth muscles which make up involuntary muscle present in wall of blood vessel, internal organs and skin, are rare aggressive mesenchymal neoplasms accounting 3-7% of soft tissue sarcomas.[3] LMS are divided according to their site into deep soft tissue sites as the retro peritoneum and in peripheral soft tissue sites also known as superficial LMSs. Superficial LMSs are further divided on the basis of their histogenesis and different clinical and prognostic factor into cutaneous and subcutaneous LMSs.[4] Subcutaneous LMSs account for 1-2% of all soft tissue sarcomas[5] and arises from small to medium sized blood vessels present in subcutaneous tissue.[6] Superficial LMSs can occur anywhere in body but lower extremities are most frequent site.[7] The thigh considered to be favorite site[8] and only 10-15% in trunk.[9] and leiomyosarcoma of anterior abdominal wall is very rare.[7] Here we report a case of giant subcutaneous Leiomyosarcoma arising from the anterior abdominal wall.

CASE PRESENTATION

A 35-year-old female presented with an asymmetric, voluminous tumor in the middle of her abdomen, which had gradually grown during the past one year. The patient had two children and had no family history of neoplasia. She was not addicted to tobacco and alcohol. She had a history of surgical excision of a same tumor twice 7 year back and 1 year back but previous documents and biopsy report are not available. There was no history of any radiotherapy or chemotherapy. Patient also has complaint of bleeding from the surface of tumor. Physical examination on admission revealed a tumor of size around 30 cm × 20 cm in the middle of abdomen. The tumor was fixed to the overlying skin as shown in figure 1. The skin appeared reddish with vascular engorgement with small ulceration which are bleeding as shown in figure 2.

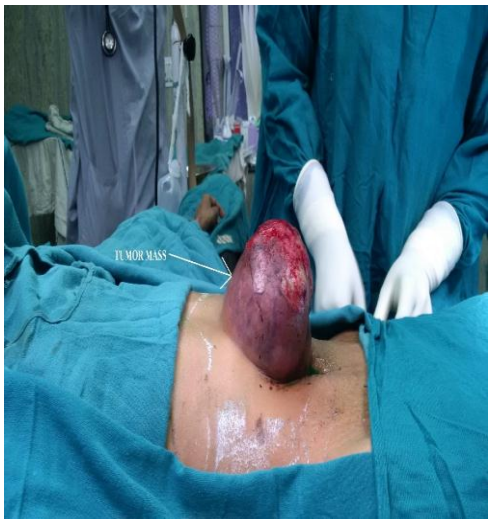

Figure 1 showing pre operative pictures of tumor mass.

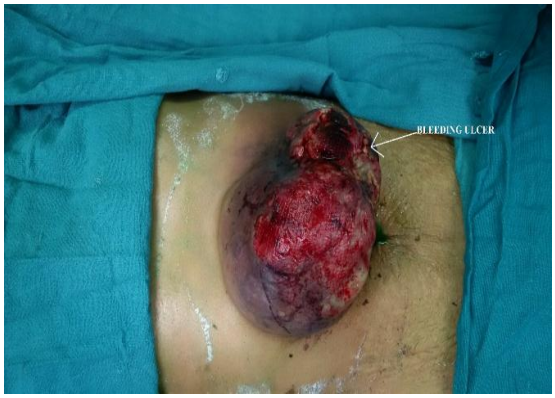

Figure 2 showing bleeding ulcer on tumor mass.

Computed tomography (CT scan) depicted a large lobulated heterogeneously contrast enhancing soft tissue mass in middle of anterior abdominal wall not involving underlying muscle and rectus sheath. There was no evidence of metastases to distant organs on imaging. Pre-operative biopsy of the lesion showed plump spindle cells in clusters, atypical and typical mitosis seen, and suggesting Leiomyosarcoma. She was subjected to surgery which consisted of a wide resection of the tumor along with overlying skin. The resection exceeded the macroscopic limits of the tumor by 2 cm in order to accomplish a tumor-free margin. The abdominal cavity was opened to make sure no internal extension of tumor as shown in figure 6. Defect was closed primarily. Post operatively stich line got infected, middle two stiches were opened to drain pus and regular dressing was done. The resected tumor as shown in figure 7, measuring 30 × 20 × 8 cm, was reddish brown in color without central necrosis and hemorrhage on its cut surface. Routine histological examination with hematoxylin and eosin revealed that the tumor is comprised of spindle shaped cells arranged in intersecting sheets and fasciculated bundles as shown in figure 3. Cell have scant to moderate amount of eosinophilic cytoplasm elongated, vesicular nuclei with blunt end and tiny conspicuous nucleolus.

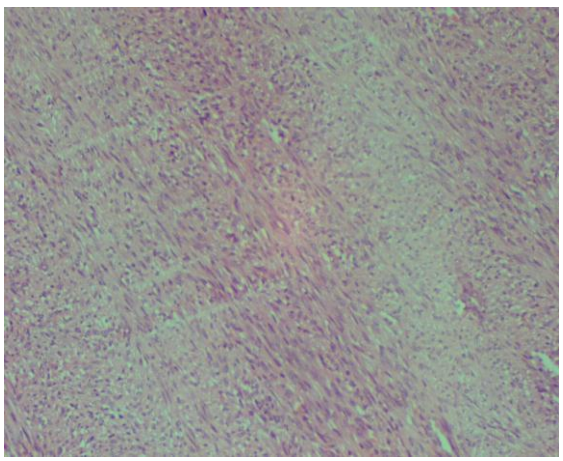

Figure 3 HPE slide showing spindle shaped cells arranged in intersecting sheets and fasciculated bundles

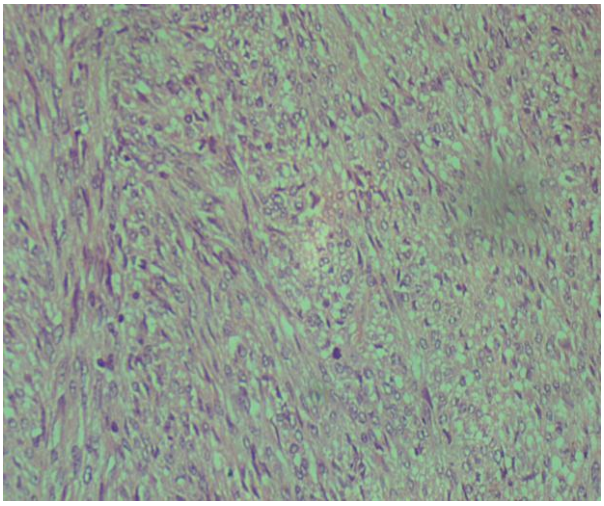

Figure 4 HPE slide showing typical and atypical mitotic figures.

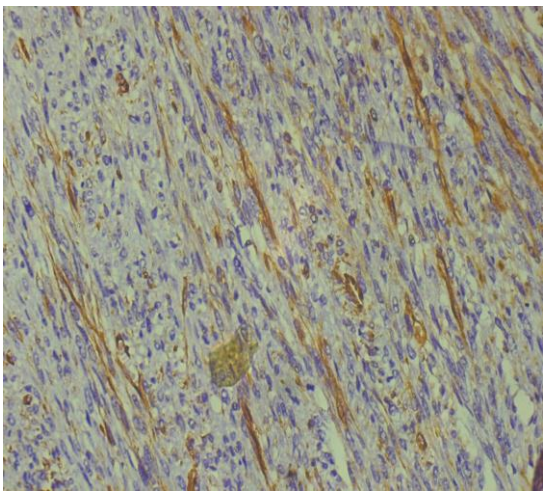

Figure 5 Immunohistochemistry (IHC) tumor cells were positive for smooth muscle actin (SMA)

Typical and atypical mitotic figures are seen as shown in figure 4. All resected margins were free from tumor. In immunohistochemistry (IHC), the tumor cells were positive for smooth muscle actin (SMA) as shown in figure 5 but negative for desmin, CK, EMA, CD117 and S-100. With these findings, the diagnosis of subcutaneous leiomyosarcoma was confirmed. Taking into consideration the large tumor size, high mitotic rate, and recurrent setting, postoperative radiotherapy was planned.

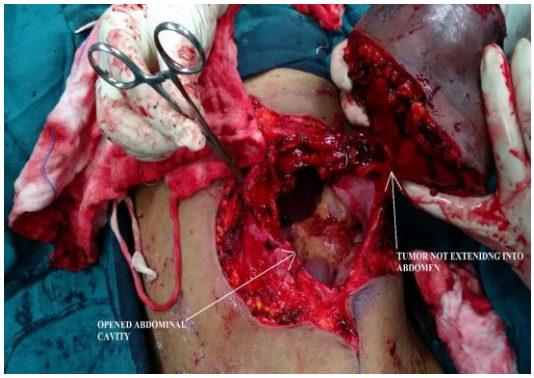

Figure 6 showing tumor free from underlying abdominal cavity

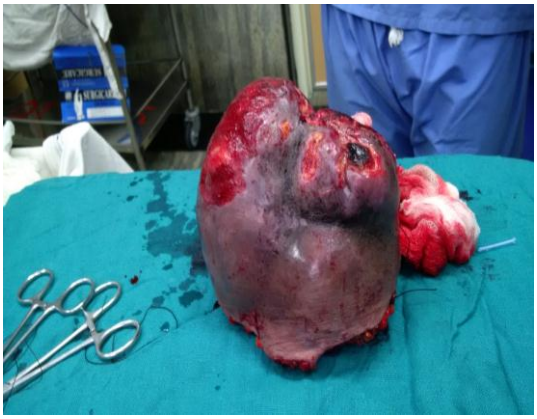

Figure 7 showing Resected tumor mass

DISCUSSION

Soft tissue sarcomas are rare tumors comprising only 1% of all adult solid tumors [10] with 50 different histological subtypes most common are liposarcoma, malignant fibrous histocytoma and leiomyosarcoma. [11] Leiomyosarcoma arising from smooth muscle cells of wall of blood vessels, internal organs and skin depending upon the site grouped into deep and peripheral of superficial which further grouped into cutaneous and subcutaneous. [4] Although soft tissue sarcoma can occur at any anatomic site,45% occur in extremities, most commonly in thigh, followed lower in frequency by visceral(20%), retroperitoneal(15%), truncal or thoracic(10%), and other location (10%). Most cases of soft tissue sarcomas are thought to be sporadic and there causes is unknown. Genetic factors, environment factors, prior radiation therapy, viral infection and immunodeficiency have been associated with the development of sarcomas. Also sarcomas have been reported to arise in scar tissue, fracture site, or anatomic region associated with prior soft tissue trauma. Genetic syndromes such as neurofibromatosis, familial adenomatous polyposis, and the li-ffraumeni syndrome have all been shown to be associated with the development of soft tissue sarcoma.[10] Soft tissue sarcoma is one of the most common types radiation associated tumor in general population, although dose response relationship is not well established.[12] Several studies have reported an increased incidence of soft tissue sarcoma after relatively high level occupation exposure to phenoxyacetic herbicides, chlrophenols, and dioxins.[13,14] The genetic alterations in soft tissue sarcoma segregate sarcoma into two major groups. The first group consists of sarcoma types with near diploid, simple karyotypes that bear few chromosomal rearrangements such as translocation in myxoid round cell liposarcoma and synovial sarcomas, APC/β-catenin mutation in desmoids tumors.[15,16,17] The second group consist of sarcoma with complex karyotypes, including dedifferentiated and pleomorphic liposarcoma, leiomyosarcoma, pleomorphic malignant histocytoma, and myxofibrosarcomas. 80-90% of patients present with the pain.[18] which can be spontaneous or induced by pressure. [19] Clinically tumor may be presents as a small to big mass with well-defined or irregular border or pedunculated mass, umblication and skin discoloration.[18] Subcutaneous leiomyosarcoma often present as a well circumscribed nodular lesion[20] with normal overlying skin.[21] and 0.6 to 5cm hemispherical skin elevation.[19] Dermal leiomyosarcoma presents as a small firm fibrous nodule of size less than 2cm in size whereas subcutaneous are larger in size with average size of 4cm.[22.23,24] leiomyosarcoma present late with median duration of 12 months[19] and may be presented as multiple nodules in rare cases. In this cases metastasis to other organs or from other sites like retroperitoneal should be excluded.[20,21] As reported by Dahl et al 4-7 patients has the history of operation for retroperitoneal leiomyosarcoma now presented with multiple superficial leiomyosarcoma.[20] However in some cases patients presentation is nonspecific[25] leads to misdiagnosed on clinical grounds.[26] One of the misleading sign is not involvement of surrounding structure and easily excised or shelled out as it is a benign. Therefore knowledge of this misleading feature is necessary to avoid delay in treatment and diagnosis. [19, 27] Delay in diagnosis is common, with the most common differential diagnosis for extremity and trunk lesion being a hematoma or a pulled muscle. Therefor physical examination should include assessment of the size of the mass, its depth in relation to superficial fascia, and its relationship to neurovascular and bony structure. Fine needle aspiration cytology has a limited role in diagnosing but may be of value in the documentation of recurrence. For most soft tissue mass, an open incisional biopsy or core needle biopsy is usually preferred. If tissue is inadequate, repeat core biopsy under image guidance is preferred. For evaluation of extent of disease MRI examination is the usually preferred procedure which provide differentiation between tumor and adjacent structure and provide excellent definition of fascial planes. Newer techniques, such as flurodeoxyglucose positron emission tomography (FDG-PET), are being used to evaluate distance metastasis. Superficial leiomyosarcoma has nonspecific features on imaging.[10, 28] Large tumors appears heterogeneous because of necrosis, cystic changes and hemorrhage, and microcalcification in 10-15% on radiographs or computed tomography scan.[22] On ultrasonography, tumor is hypoechoic solid mass with well-defined or ill-defined borders. MRI should be used

for staging purpose.[22] Surgical excision with wide margins in early stage is the treatment of choice.[18, 19, 20,22,29,30] however the safe margins width have not been clearly defined.[31] Margins of 2cm is suggested by most of the authors.[31,32] McKee et al,[33] suggested margins >1cm independently result in longer local recurrence free survival. The recurrence rate of 0-14% has been reported in Mohs micrographic surgery in management of superficial leiomyosarcomas.[34,35,36] Starling et al,[35] reported no tumor recurrence in largest series using Mohs surgery with median follow up 4.47 years with average tumor size of 4.69 cm^2 and average size of surgical defect was 14.95 cm^2. However larger number of patients with longer follow up is needed treated with Mohs surgery before coming to any conclusion.[21] Early surgical intervention that is wide local excision with safe surgical margins is the most important prognostic factor in patients with superficial leiomyosarcomas.[20] High local recurrence rate has been noted in patients with inadequate surgical margins and put the patients of subcutaneous leiomyosarcoma to potentially incurable and fatal metastatic disease.[18] Histologically, the tumor shows perpendicularly arranged fascicles of spindle cell with abundant eosinophilic cytoplasm and vacuolated and elongated blunt ended nuclei.[37] Leiomyosarcomas can be differentiated from the other spindle cell tumors with the help of immunochistochemical analysis especially anaplastic and poorly differentiated tumors.[18,21,32] The SMA smooth muscle actin has been reported to be positive in 100% of patients with leiomyosarcomas and is the most sensitive marker indicating smooth muscle differentiation.[25,30,37] High grade tumor has been reported histopathological in most cases.[24,30,38]

The depth of subcutaneous extension in histopathology report is essentially important in patients with dermal leiomyosarcomas because even with a minimal invasion can leads in late local recurrence and distant metastasis.[22] Fibrosarcoma, dermatofibroma, malignant schwannoma, malignant fibrous histocytoma, rhabdomyosarcoma, nodular fascilitis and neurofibroma are the differential histological diagnosis.[18] Patients with superficial leiomyosarcomas has been reported with 65% of cumulative five year survival.[23,32] In 225 patients of Scandinavian sarcoma grouped in 5 and 10 year overall survival rate was 69% and 49% without metastases at presentation however five year survival dropped to 16% in patients with metastases.[24]

In the patients with subcutaneous leiomyosarcomas 50-60% local recurrence has been reported.[27] 50% of tumor recur within 4-18 months in primary tumor measuring 0.5cm after initial diagnosis.[19]Recurrences are uncommon in patients with cutaneous tumors.[18,38] In 30-60% of patients with subcutaneous leiomyosarcoma metastasis have been reported and only 5% in patients with cutaneous tumors.[19,27,32] Lung is the most common site for the metastases and hematogenous spread is the most common mode.[19,20,27,38] 80% of the cases reposted metastases with in first two years of diagnosis however late metastases after many years have also reported.[21,32] Only 25% of case metastases occur in lymph node making it an uncommon site.[27] Presence of metastases is the strongest risk factor related to death.[24] Tumor size, presence of necrosis, high mitotic rate, deep seated tumor with fascia involvement and intramural vascular lesions are the adverse prognostic factors associated with subcutaneous leiomyosarcoma.[23,24,29,38,40] 95% and 30% are the survival reported for tumors smaller than 2cm and larger than 5cm respectively.[32] Advanced age, DNA aneuploidy and vascular invasion can be used to identify prognosis of patients as suggested by Gustafson et al.[23] 16 of 30 patients died of the disease in presence of two or three risk factors whereas only one out of 15 died with one or more risk factors.[23] It seems that the tumor is resistant to adjuvant chemotherapy and radiation therapy and therefore there role in treatment is controversial but there are the rare cases which has been treated successfully with theses modalities.(18,19) Haffner et al,[41] reported use of factor-alpha and melphan as isolated limb perfusion with high-dose tumor necrosis result in complete remission of an advanced cutaneous leiomyosarcoma. The response rate was 15-30% in the patients with wide spread metastatic disease when managed with chemotherapy and with the median survival was one year.[32] Tumor with larger than 5 cm and high grade adjuvant radiotherapy may be reserved in order to decreases the chances of recurrence and along with surgery in recurrent tumors.[32] The long term prognosis remains poor despite adequate local control with or without adjuvant radiation therapy in the patients with subcutaneous leiomyosarcomas.[24] In immunosuppressed patients tumor may take aggressive mode therefore long-term follow up of the patients is essential.[34]

CONCLUSION

In the end we conclude that soft tissue sarcomas are rare adult solid tumors and anterior abdominal wall leiomyosarcomas are even rarer. Misleading features of tumors and lack of knowledge about unusual site of these tumors leads to delay and diagnosis and management result in progress of disease to advanced stages. Early surgical intervention along with chemotherapy and radiotherapy outcome can be changed as these tumor as high tendency to recur locally and metastasize and required long follow-up. So this case report increases awareness about the unusual presentation of subcutaneous leiomyosarcomas and helping in early diagnosis and management preventing advancement of diseases.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest regarding the publication of this manuscript.

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