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Original Research Article

Experience of Treating Spindle Cell Tumours: Case Series in Tertiary Care Centre in Punjab

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Abstract:

Sarcomas can present differently in different parts of the body and showcase varied histopathological features and tend to recur locally and metastasize to distant sites. We are reporting our series on patients with spindle cell tumors. The mass was evaluated through computed tomography and the histology was confirmed by biopsy. Wide surgical resection of the mass was done and the patients were referred to radiotherapy for further treatment. These cases showcase the scenario of many patients in developing countries where the patients are negligent, lost and present later with grave consequences.

Introduction

Not all soft tissue tumors of the abdominal wall are desmoids, and sarcomas must be considered in the diagnosis and management of neoplasms of this site. The increased risk of relapse observed with local recurrence is likely related to the risk of subsequent local failure since presentation status did not significantly influence distant failure and tumor-related mortality rates.

Sarcomas are a rare group of malignant tumors arising from the mesenchymal tissue, which make <1% of all adult malignancies [1]. Sarcomas usually present as a painless mass and rarely present with distant metastasis especially in the lungs [2]. Spindle cell carcinoma (SpCC), also referred to as sarcomatoid carcinoma, is a biphasic tumor composed of conventional SCC and malignant spindle cells. The characteristic spindle cell phenotype of the neoplastic cells in SpCC is the result of epithelial-mesenchymal transition.

Similar to conventional SCC, SpCC has been etiologically related to cigarette smoking and alcohol consumption. It has been suggested that SpCC may develop after radiation exposure; however, some authors believe that this is not a major etiologic factor. Macroscopically, SpCC can present either as an exophytic, polypoid lesion or as a flat, ulcerated tumor. Microscopically, the SCC component may be well-, moderately- or poorly differentiated, keratinizing or nonkeratinizing, and transition between the two components may be abrupt or gradual. The spindle cell component usually forms the bulk of the tumor. Spindle cells are often pleomorphic, with large hyperchromatic nuclei, prominent nucleoli, and numerous mitoses. Sometimes, only spindle cells are present. If such tumors are less cellular, they may mimic benign reactive lesions. Foci of osteosarcomatous, chondrosarcomatous or rhabdosarcomatous differentiation may be present, particularly in patients who had previously been treated by radiotherapy Spindle cell sarcoma is one of the rare varieties of undifferentiated soft-tissue sarcomas. Due to its rarity, only a few cases have been described in medical literature [3]. Here, we report a series of cases involving the intermuscular planes of spindle cell carcinomas.

Materials and Method

Case 1:

A 26 year female presented with chief complaint of mass over the left flank for 3 years. Swelling was associated with localized pain: deep seated, dull aching, relieved with medication. On examination, 7 x 3 cm well defined lump present in the left lumbar region which was hard in consistency, fixed to underlying muscle and ribs with no overlying skin changes.

Ultrasound whole abdomen showed well defined hypoechoic vascular solid lesion in muscular plane of left lateral abdominal wall in lumbar region;? Desmoid tumor of left lateral abdominal wall.

FNAC showed collection of spindle cells along with collagenous stromal fragments against hemorrhagic background- features suggestive of Spindle Cell Tumor. All the blood works up was done and was found to be in normal limits.

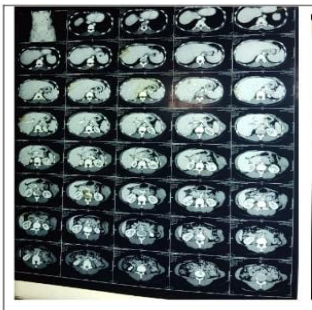
Patient underwent Wide Excision under GA with intraoperative findings of 7 x 3 cm mass in the left flank in the intramuscular plane indenting left 11th and 12th ribs.(Fig 2) Primary closure was achieved post excision. Alternate sutures removed on postoperative day 12 and all removed on postoperative day 14

CECT whole abdomen: Well defined homogenous isodense mass lesion measuring 8.5 cm x

4.5 cm x 4 cm noted in left subcostal/lumbar region intermuscular plane of left external and internal oblique muscles.

Lesion shows homogenous enhancement post contrast administration. No evidence of internal calcifications. A linear tract seen extending to skin- ? Neoplastic etiology.

Histopathological Findings: Benign fibroblastic and myofibroblastic tumor and dense collagenous deposits within well-defined capsule and tumor free margins- features s/o Spindle Cell Tumor.



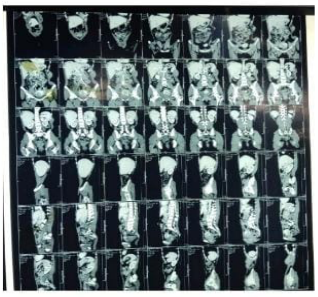


Fig 1 : CECT whole abdomen s/o Left Anterior Abdominal Wall intermuscular soft tissue dense enhancing mass lesion

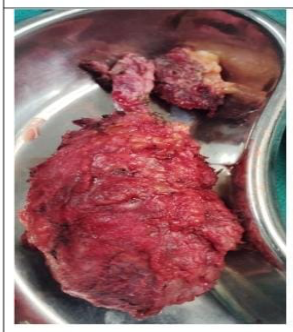


Figure 2: Excised specimen of abdominal lump

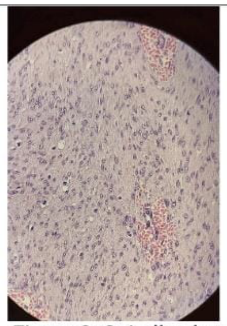


Figure 3: Spindle shaped plump cells; Well defined capsule, blood vessels and collageneousstroma visualised(hematoxylin-eosin)

Figure 1-3:

Case 2:

62 year old female presented with recurrent swelling of about 8*6 cm in epigastric region for the past 3 years. FNAC suggestive of mesenchymal neoplasm and PET CT suggestive of soft tissue lesion in superficial plane in supraumbilical region with fat planes of lesion focally indistinct with abdominal muscle and no distant metastasis.

Wide local excision including bilateral rectus muscle segment and wound coverage by STSG was done. Patient kept on regular follow up and no recurrence signs till date. Histopathology report was suggestive of dermatofibrosarcoma protuberans.

Case 3:

A 52 year old male presented with an ulcerative lesion over the lower abdominal wall. MRI was suggestive of soft tissue lesion in anterior abdominal wall with ill-defined fat planes and right rectus muscle. Wide local excision including bilateral rectus muscle segment followed by prolene mesh and pedicled right ALT flap coverage was done.(Fig 4,5,6) Patient was on regular follow up upto 1 year post surgery and showed no signs of recurrence.

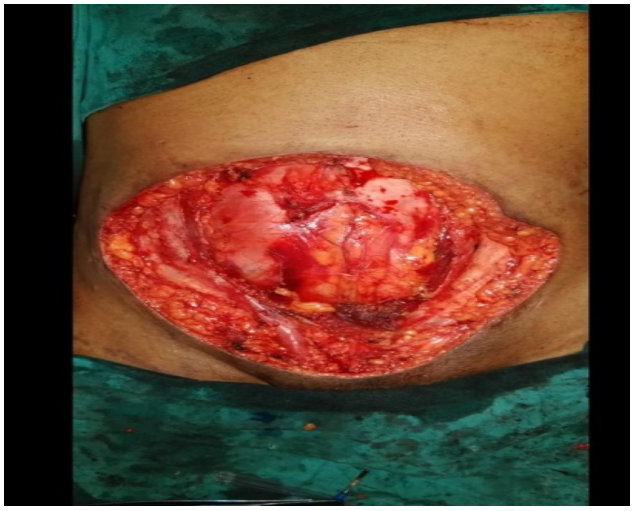


Figure 4: Post wide local excision of mass over lower abdominal wall

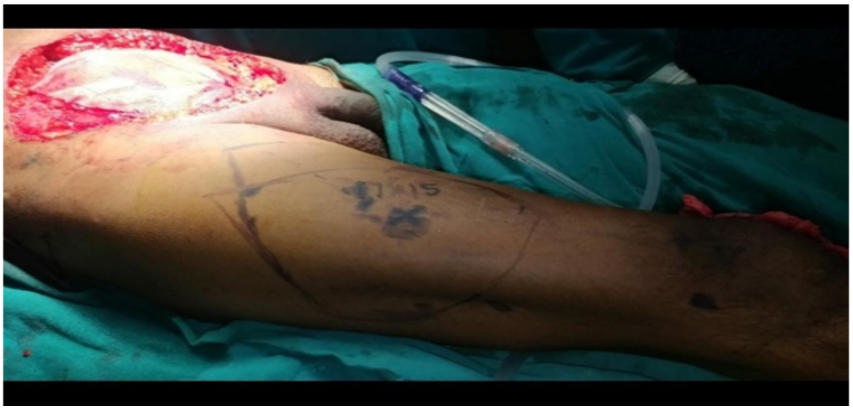


Figure 5: Marking for pedicled ALT flap for wound coverage

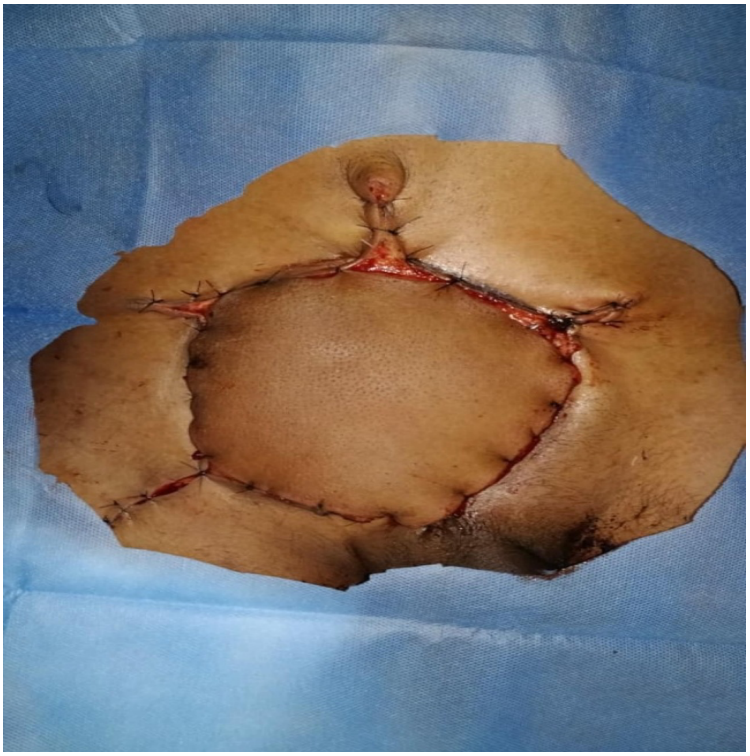


Figure 6: Wound coverage over lower abdomen by pedicled ALT flap

Discussion

Soft tissue tumors of the abdominal wall though clinically similar have many distinct histologic subtypes. Some tumor variants are known for their aggressive biologic behavior. Included among these are soft tissue sarcomas, desmoid tumor and dermatofibrosarcoma. Soft tissue sarcomas are mesenchymal neoplasms comprising 1% of adult malignant growths, having high incidence of local recurrence and propensity for distant metastasis [4]. Desmoid tumors and Dermatofibrosarcoma are rare, slow growing neoplasms recognized for their progressive, locally infiltrative nature.

Spindle cell sarcomas affect people of almost any age and sex [5]. Two separate studies by Feng et al. and Smith et al. showed the median age at presentation of 57 years [6]. In contrary to these studies, the age at presentation in our case is quite early (37 years). In the same study by Smith et al., the median tumor size found was 9.87 cm. This sort of presentation is quite rare as patients generally tend to present earlier with the mass with a median duration of 20 weeks [6], whereas our patient completely ignored the mass until it grew to a massive size and started hindering his day-to-day activities. This highlights that in poverty and destitution, people ignore medical attention until the very last stage [7].Due to their rarity, delay or misdiagnosis is common for sarcomas especially in settings with limited facilities. [8]

The evaluation of a patient with a suspected soft-tissue sarcoma includes history, imaging and biopsy [9]. MRI is the preferred modality for the evaluation of soft-tissue masses of the extremities, trunk, head and neck [10]. All patients diagnosed with sarcoma should also have a computed tomography scan at the time of diagnosis and in follow-up [9]. In our case, the mass was initially identified as liposarcoma. Later, repeat biopsies revealed spindle cell sarcoma. The rarity of the case and the little experience in diagnosing rare cases might have led to this error in histological diagnosis [11].

A definitive evaluation on the relationship between site (abdominal wall vs extremity) and subsequent outcome adjusted for clinical and pathologic factors cannot be achieved with adequate statistical power because of the relatively small number of patients with abdominal wall tumors. However, a comparison of 5-year disease-specific and recurrence-free survival rates for these sites controlling for biological factors suggests that sarcomas of the abdominal wall and extremities have similar outcome predictors and natural histories. Thus, it seems reasonable to include abdominal wall sarcomas with extremity sarcomas in the design of clinical trials [12,13].

The treatment of choice for most abdominal wall soft tissue tumors is surgical resection with an adequate margin. Depending upon the extent of tumor involvement abdominal wall resection is done which may require reconstruction, usually performed simultaneously. Various reconstruction options ranging from split thickness skin graft to local/ regional flaps and free tissue transfer have been described and require involvement of plastic surgeons. Local flaps are mainly based on random vascularisation in the base of the flap, while local perforator flaps have a pedicle. Distal flap can be used when a donor site is far from defects. Depending upon resection of sheath, reconstruction can be performed with or without mesh placement.

In our case, the lack of proper communication between involved hospitals and doctors might be one of the reasons for loss to follow-up of one patient. Besides his financial constraints and ignorance, inadequate counseling about the diagnosis may have led to the discontinuity of care, which is very common in developing countries [14,15]. Due to limited resources in hospitals, it is common in developing countries to refer patients to other centers for further care [16]. Even tertiary care centers like ours cannot provide comprehensive care to the patient. This creates a disadvantage in continuity-of-care for many patients. Hence, one of the main focuses regarding this patient would be to ensure adequate follow-up. To avoid such circumstances in the future, it is better if the health care team takes charge and arranges continuity-of-care for the patient so that any future patients do not leave without completing their course of treatment. Use of telemedicine is proven to be highly effective to ensure follow-up in developing countries [17]. It is, of course, necessary to address the main underlying issues like the financial burden for the patient and aim for sustainable treatment strategies.

Conclusion

In the study of case series, the clinical behavior and management of soft tissue tumors of the abdominal wall were evaluated. Not all abdominal wall neoplasms are desmoids, and sarcomas must be considered in the differential diagnosis. Abdominal wall desmoids, STS, and DFSP demonstrate a broad spectrum of biologic behavior. Initial biopsy is usually mandatory to facilitate treatment planning. Desmoid tumors remain exclusively a local problem. Dermatofibrosarcoma protuberans of the abdominal wall is virtually always a local issue, with rare distant metastases associated with its fibrosarcomatous variant. Prognostic factors associated with adverse survival outcomes are defined for abdominal wall sarcomas, and they are identical to factors predicting outcomes for other sites. The treatment approach for all 3 diseases remains to be aggressive, complete surgical resection to achieve local control requiring abdominal wall reconstruction most of the time necessitating the need of a reconstructive surgeon. Repair of abdominal wall defects, with excellent functional results, can be

sufficiently achieved with prosthetic mesh reconstruction. Stratification on the basis of prognostic factors for distant disease failure and tumor-related mortality will facilitate selection of patients with STS for adjuvant systemic therapies.

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