Print ISSN: 2321-6379
Online ISSN: 2321-595X

Case Series

Clinical Presentations of Soft-Tissue Sarcoma – A Case Series

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Abstract

This article will be dealing about multiple clinical presentations of various musculoskeletal tumors. They are a rare and heterogeneous group of malignancies, which are best managed by multidisciplinary teams in specialist sarcoma referral centers. Historically, the standard for local control of these tumors has always been amputation. Evolution in multimodality treatment has seen a shift toward preservation of the limb. Advances in limb sparing surgery have seen the quality of life in sarcoma patients to improve drastically; however, unplanned surgical excision of sarcomas remains a major treatment dilemma in the control of local disease. A 30-year-old female presented with swelling over lower back and on investigations revealed to have a large fibromyxoid sarcoma. Neoadjuvant was not indicated and the patient was planned for surgical excision first. A 30-year-old male presented with swelling over nape of neck associated with weakness in the right upper limb and on investigation was found to have neurofibroma. The patient underwent surgical excision first and then sent for radiotherapy. A 62-year-old male presented with the complaint of swelling over the left side neck and upper chest wall, and on investigations, the diagnosis was under dilemma between lymphangiosarcoma and liposarcoma. The patient was planned for excision. A 35-year-old male presented with swelling over the left upper limb. On investigation, malignant fibrous histiocytoma was noted and the patient was planned for excision. Although sarcomas are diverse in nature, the treatment approach is same. It is a multidisciplinary approach with the surgical excision being the mainstay of treatment in non-metastatic sarcoma and chemotherapy in metastatic carcinoma. Limb salvation is always tried. Advances in radiotherapy have helped decrease local adverse effects and sustain good local control of the disease. Hence, the importance of team management cannot be overlooked and if possible, all three modalities can be tried for better prognosis of the patient.

Key words: Sarcoma, Soft Tissue, Tumours

INTRODUCTION

Sarcomas make up <1% of all cancers. Sarcomas occur in 2–4 people/100,000 population and its >60 subtypes are divided into two broad categories: STS and bone sarcomas.[1]

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www.ijss-sn.com

Month of Submission : 09-2022
Month of Peer Review : 10-2022
Month of Acceptance : 11-2022
Month of Publishing : 11-2022

Rhabdomyosarcoma is the most common STS (soft-tissue sarcoma) in children, whereas undifferentiated pleomorphic sarcoma is the most common in adults.[2]

The analysis of large SEER database shows that the age-adjusted incidence of sarcomas arising in soft tissue is 3.1/100,000 irrespective of gender. These rates are little higher in men (3.8/100,000) than woman (2.6/100,000).[3]

Soft-tissue sarcoma (STS) is a diverse group of more than 60 neoplasms that can arise from virtually any anatomic site and can affect the very young as well as the elderly. Sarcomas are of mesenchymal origin. The tissue types of

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STS origin include skeletal muscle, adipose cells, blood and lymphatic vessels, and connective tissue or those cells with a common mesoderm origin. Taxonomy of soft-tissue sarcoma mentioned in Table 1.

Large published series demonstrate that extremity and trunk STS is more common than intraperitoneal and retroperitoneal STS.[5]

The adverse prognostic factors for soft-tissue sarcomas' are: Size, increasing age, high grade, metastasis at diagnosis, local recurrence at diagnosis (following unplanned excision), positive surgical margin, deep to muscular fascia, and high levels of tumor necrosis.[6-8]

MATERIALS AND METHODS

- Place: Department of General Surgery, M. Y. Hospital, Indore
- Duration: July 2021–July 2022
- We identified four patients (three men and one woman, mean age at presentation 41 years, range 15–79 years).

Patients were eligible for inclusion if they had histologically confirmed soft-tissue sarcoma that came in MYH Hospital, Indore, in General Surgery Department. We extracted the following information from each patient's medical record: Age at presentation, sex, disease status at presentation (primary, locally recurrent, or metastatic), prior treatment, medical history, histological diagnosis, lesion characteristics (tumor size, location, depth, grade, and involvement of neurovascular and vital structures), details of treatment (pre- or post-operative chemotherapy or radiation therapy, resected structures, surgical margins, reconstructive technique, and complications), and condition at follow-up. Staging was performed using the system proposed by the American Joint Committee on Cancer (AJCC).

Study Protocol

- After proper consent from the patient, complete history taking was done.
- Complete joint and limb examination was done with orthopedic and cancer opinion.
- All blood investigations were sent along with chest X-ray, respective limb X-rays, and complete cardiac workup of the patient.
- High-end radiological investigation was done to rule out any metastasis and to evaluate the locoregional spread and margins of the tumor for proper planning of the treatment plan.
- Biopsy of the tumor and histopathology report taken into consideration.
- Treatment plan formulated in multidisciplinary fashion.

CASE SERIES

Case Report 1

A 30-year-old female presented with swelling over lower back, apparently noticed 2 years back. The patient also had intermittent episodes of back ache. The patient underwent local excision of the mass which reoccurred and came to MYH Hospital for definitive treatment.

On examination, no lower limb sensory-motor neuropathy noticed. Size of mass was ~25 × 20 cm over lower back which was soft and cystic in consistency and not fixed to the underlying structures [Figures 1 and 2].

Figure 1: Lateral View of mass

Figure 2: Posterior Aspect

Figure 3: Post-operative image

MRI LS spine was suggestive of neurofibroma. The patient underwent excision of mass and intraoperatively "GOOEY" like content ~2 L aspirated from the mass and sent for histopathology, which was suggestive of fibromyxoid sarcoma [Figure 3].

The patient was later referred for radiotherapy.

Case Report 2

A 30-year-old male belonged low socioeconomic status and presented with the complains of swelling over nape of the neck which he apparently noticed 2 years back which gradually increased in size. The patient came to the surgery department when he developed pain in the back of neck and upper back which radiates to right upper limb with slight weakness.

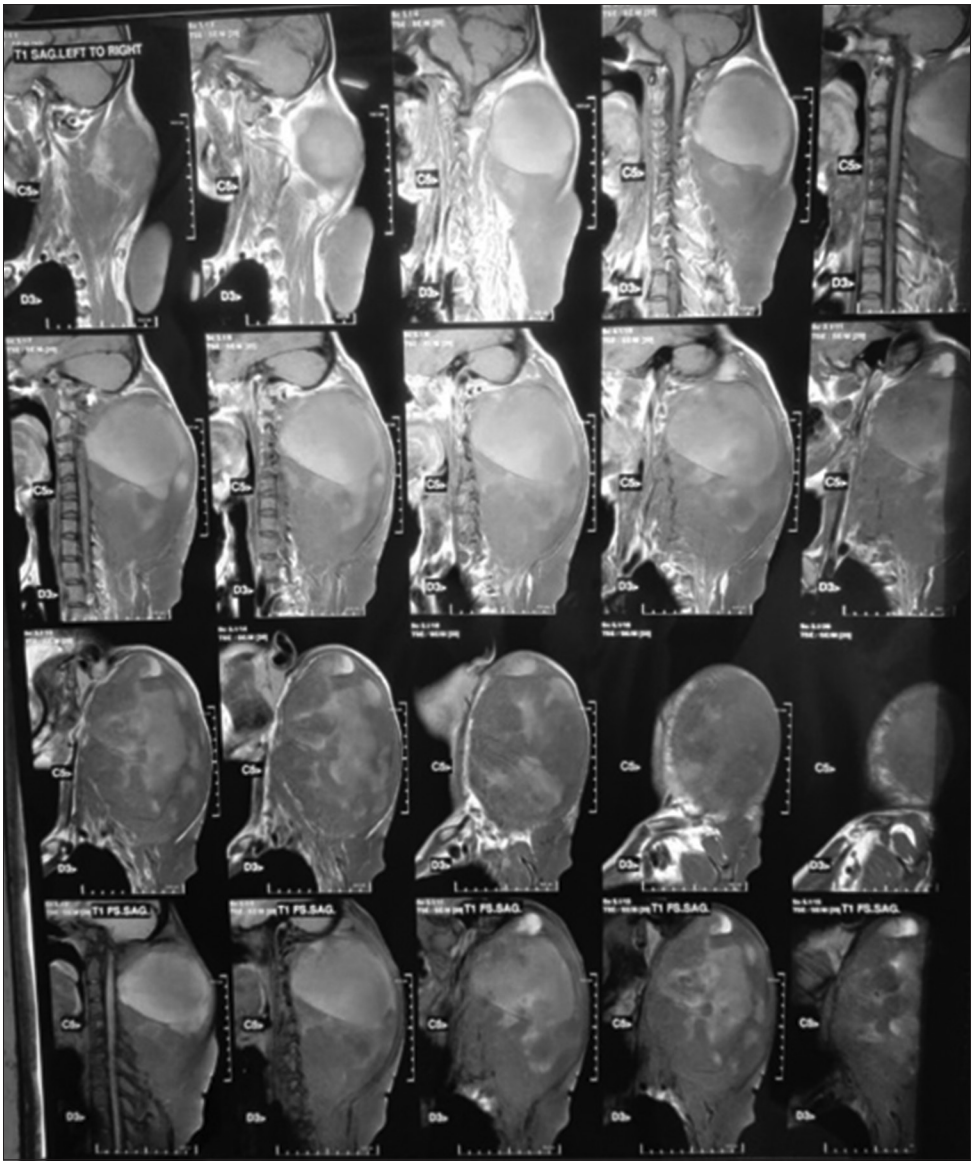

Figure 4: MRI- Showing extent of tumor

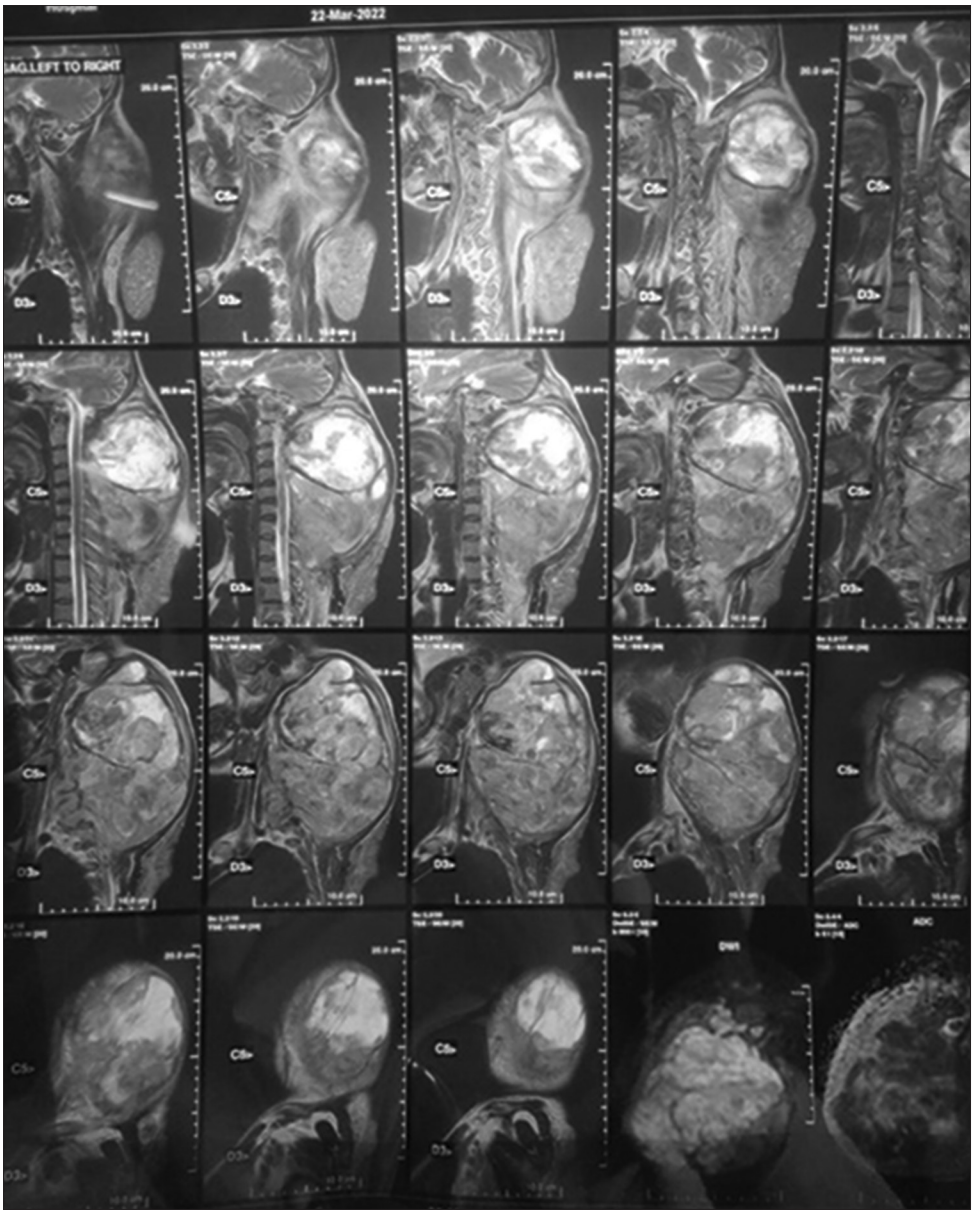

Figure 5: MRI- Showing extent of tumor

Figure 6: Lateral view

Figure 7: Posterior View

Figure 8: Pre-operative image

Figure 9: Post-operative image

On examination, a large ~20 × 20 cm soft to firm in consistency solid mass located at right side nape of neck, which was fixed to the underlying structures associated with restriction of neck movements. Similar small hard nodular multiple lesions were present over torso.

MRI was suggestive of soft-tissue sarcoma which was extended from occipital region to D3 with cord abutment at C5-6 level. True-cut biopsy suggestive of neurofibroma [Figures 4 and 5].

Excision was planned and specimen sent for histopathology. Histopathology suggestive of spindle cell tumor with neural differentiation (malignant peripheral nerve sheath tumor) [Figures 6 and 7].

The patient was sent for radiotherapy.

Case Report 3

A 62-year-old male presented with the complaint of swelling over the left side neck and upper chest wall, apparently noticed 1.5 years back which gradually increased in size with no complaint of pain, respiratory distress, facial swelling, and any neurological deficit.

On examination, large ~20 × 20 cm soft, solid mass was present over the left side of neck, non-tender, and mobile. Investigations were not clearly depictive of histological type [Figures 8 and 9].

USG suggestive of lymphatic malformation whereas CECT neck was suggestive of liposarcoma which was extending in lateral aspect of left thorax as well with no pleural extension.

Table 1: Taxonomy of soft-tissue sarcoma[4]

Tissue of origin (histogenesis)	Type of sarcoma
Adipocytic	Myxoid/round cell liposarcoma
	Pleomorphic liposarcoma
	De-differentiated liposarcoma
	Well-differentiated liposarcoma
Chondro-osseous	Mesenchymal chondrosarcoma
	Extraskeletal osteosarcoma
Fibro-/myofibroblastic	Desmoid-type fibromatosis
	Superficial fibromatosis
	Myxoinflammatory fibroblastic sarcoma
	Solitary fibrous tumor
	Dermatofibrosarcoma Protuberans
	Adult fibrosarcoma
	Low-grade fibromyxoid sarcoma
	Myxofibrosarcoma
	Sclerosing epithelioid fibrosarcoma
	Congenital fibrosarcoma
	Myofibroblastic sarcoma
Fibrohistiocytic	Giant cell tumor of soft tissue
	Plexiform fibrohistiocytic tumor
	MFH
Uncertain differentiation	Mixed tumor/myoepithelioma
	Chordoma
	Ossifying fibromyxoid tumor
	Epithelioid sarcoma
	Malignant mesenchymoma
	Alveolar soft part sarcoma
	Clear cell sarcoma
Nerve sheath	Malignant nerve sheath tumor
Pericystic	Malignant myopericytoma
	Malignant glomus tumor (and variants)
Skeletal muscle	Pleomorphic rhabdomyosarcoma
	Sclerosing rhabdomyosarcoma
	Spindle cell rhabdomyosarcoma
	Embryonal rhabdomyosarcoma
	Alveolar rhabdomyosarcoma
Smooth muscle	GIST
	Deep-seated/visceral leiomyosarcoma
	Cutaneous leiomyosarcoma
Vascular	Epithelioid hemangioendothelioma
	Angiosarcoma
	Kaposi's sarcoma
	Retiform hemangioendothelioma
	Composite hemangioendothelioma
	Kaposiform hemangioendothelioma

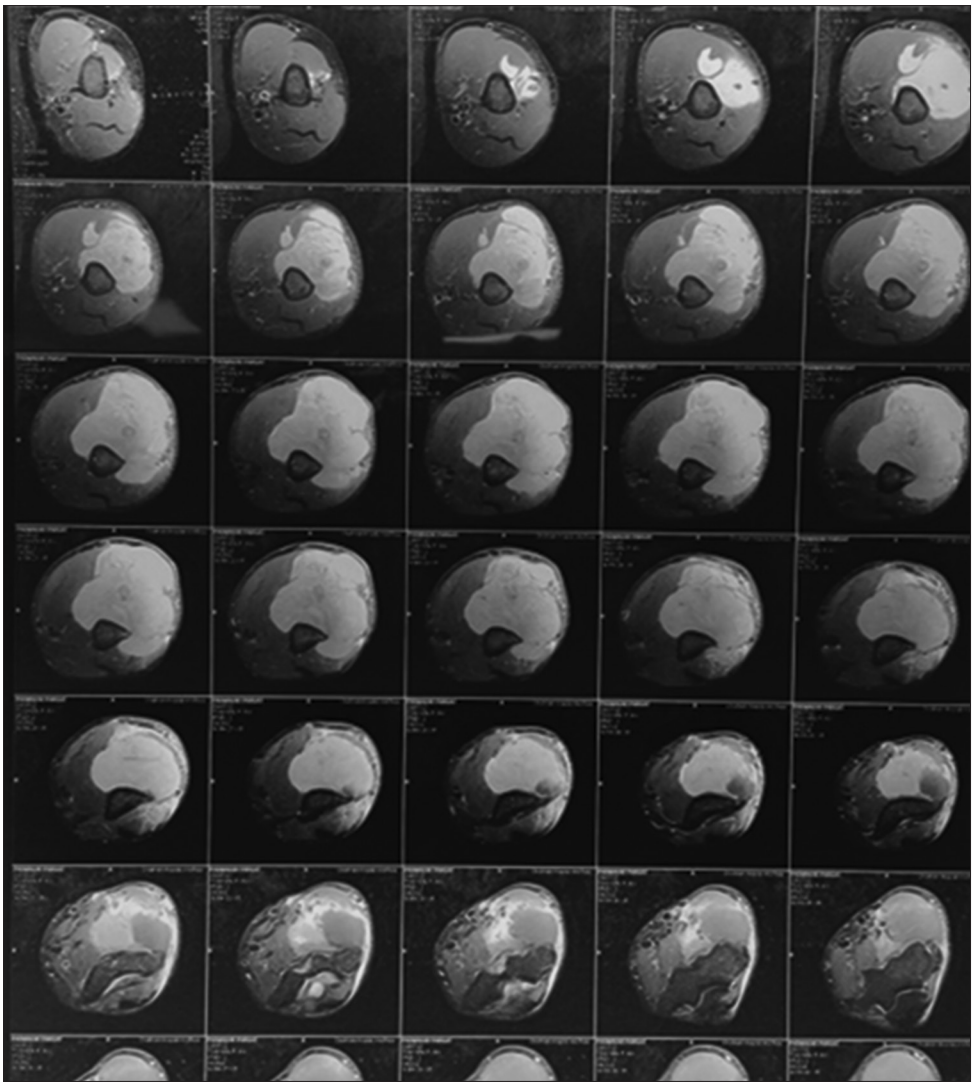

Figure 10: MRI- Showing extent of tumor

Surgical excision was planned. Intraoperatively, mass was found extending into the thorax through infraclavicular route [Figure 9]. Due to lack in proper planning, wide local excision of mass was done and specimen sent for histopathology. The patient has been asked to follow up with report to plan for thoracotomy and excision of the residual mass.

Case Report 4

A 35-year-old male presented with swelling over the left upper limb (forearm) for 4 years which gradually increased in size over the time, leading to difficulty in weight lifting and physical day-to-day activity. The patient had no complaints of sensory loss.

On examination, 10*10 cm large mass presents over lateral aspect of forearm just below cubital fossa [Figures 10 to 14.

The patient on MRI did not show any bony invasion and mass had a clear delineation from the underlying bone. Histopathology was suggestive of malignant fibrous histiocytoma.

Excision of mass was done and no functional disability in hand functioning was noted in the post-operative period.

DISCUSSION

As STS constitutes a heterogeneous group of rare tumors, management by experienced multidisciplinary team of specialists should be the standard of care from the time of diagnosis. Recent studies found that patients with STS who underwent treatment with multidisciplinary approach at high-volume hospitals had far better survival rates than those at low-volume setups without specialist care[9] and even when managed adequately have a 5-year survival rate of 62–84%.[10] For successful treatment of STS, early diagnosis is the key.

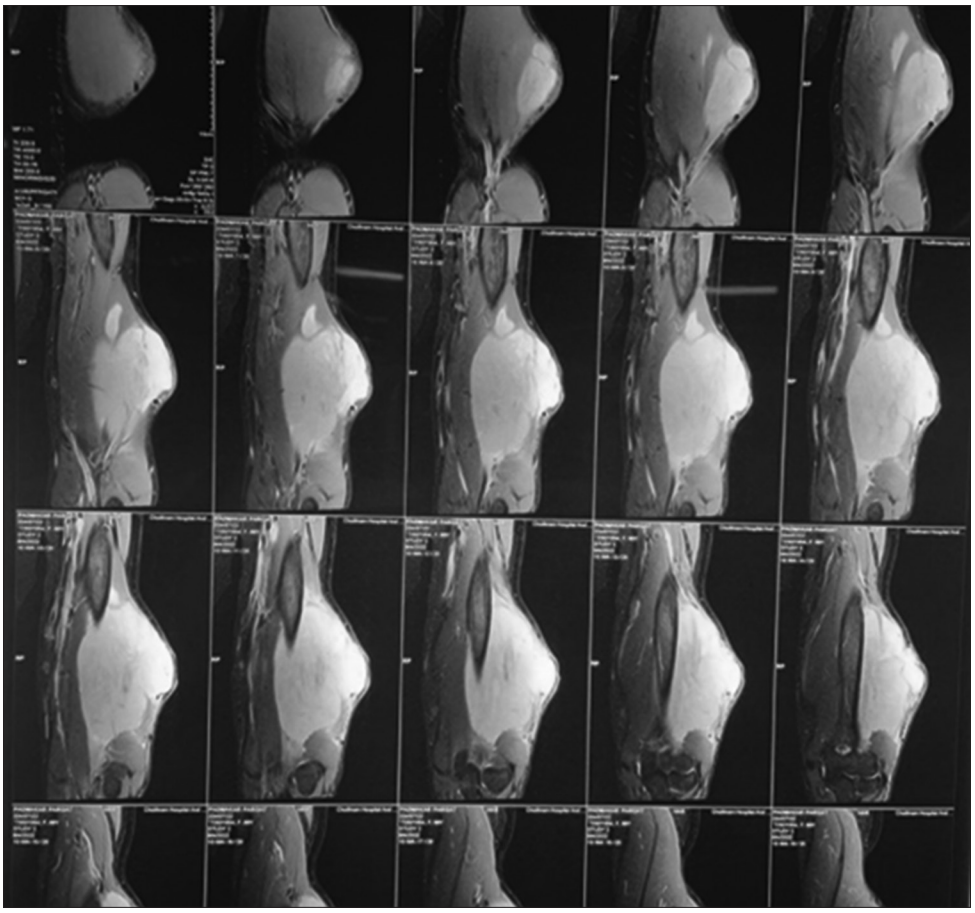

Figure 11: MRI- Showing extent oftumor

Figure 12: Intra-operative image

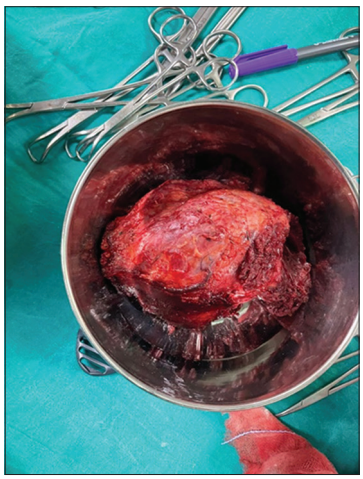

Figure 13: Excised tumor

A study by Blay *et al.* suggested lower extremity to be the most common site for sarcomas whereas, leiomyosarcoma and undifferentiated pleomorphic sarcoma as the common histologic types in adults.[11] According to the eighth edition of the *AJCC Cancer staging Manual*, site-specific staging system of STS has been developed which has used sarcoma stage, tumor size, lymph node involvement, metastasis, and histologic grade as prognostic factors suggesting the major drawback of not using histological types into consideration.[12]

Ramu *et al.* study suggests that soft-tissue sarcoma is best managed by surgical resection with or without radiotherapy. Chemotherapy is mainly reserved for the patients who present with metastasis with or without deep tissue invasion.[13]

Figure 14: Post-operative image

In this series, three of the patients were treated surgically and sent for further radiotherapy and one is waiting for further surgery in view of residual disease. Smith *et al.* have suggested in their study that limb salvage has been applied for major soft-tissue sarcomas of lower extremities and amputations have been as low as 4.1%.[14]

CONCLUSION

Clinical presentation of soft-tissue sarcoma is variable and anatomic site varies as well with commonly arising from the extremities. It presents aggressively and usually leads to functional impairment of the involved region of the patient. These sarcomas need proper clinical assessment, complete metastatic workup with reconstructive surgical plan (if the defect is large) followed by further treatment in oncology and physiotherapy department. Surgery is the main stay of treatment. Early diagnosis improves the prognosis of the patient and chances of limb salvage rate.

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How to cite this article: Ankit A, Simran S, Dawar R, Kapadia J, Verma S, Moses S, Sharma SS. Clinical Presentations of Soft-Tissue Sarcoma – A Case Series. Int J Sci Stud 2022;10(8):4-11.

Source of Support: Nil, **Conflicts of Interest:** None declared.