Recurrent Fibrosaroma of the Thigh: A Rare Clinical Presentation

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Abstract: Soft tissue sarcomas are rare mesenchymal tumours usually occur in the extremities or the retroperitoneum. Fibrosarcoma is diagnosed in about 10% of the cases of soft tissue sarcomas. These are usually managed with extensive local resection and post-operativeradiotherapy. Retroperitoneal sarcomas are prone to local recurrences while distant metastases are more common in extremity lesions. Our case presented to us with recurrent masses in the left leg and pain in the masses. The patient was operated thrice in a period of 6 years and local resection was done. The patient was given radiotherapy after the third surgery. The case was a rare third local recurrence within a short period of 6 years without any distant metastasis. Post –operative radiotherapy after the local excision is a must to prevent recurrences.

I. Introduction

Soft tissue sarcoma (STS) is a family of rare tumors that can occur anywhere in the soft tissues of the body — fat, muscle, connective tissue and nerves. Soft tissue sarcomas (STS) are malignancies of mesenchymal origin that represent approximately 1% of cancers in adults¹ and have been estimated to occur approximately 30 cases among every one million people. Fibrosarcoma is a rare variant the incidence of which is about 10% of all soft tissue sarcomas². It has two variants the infantile subtype occurring in children less than 1year of age and the adult subtype which occurs in adolescent children and young adults. The management of fibrosarcoma involves local resection with a recurrence rate of about 50% with haematogenous metastases occurring in about 25% of the cases².

II. Case Report

An 18 year old female presented to us with complaints of a mass in the left lower thigh being felt by the patient for the past 2 months and pain in the swelling for 15 days. The patient had a history of a similar swelling in the left calf 3 years back for which local resection was done. The patient underwent imaging studies including an MRI and a CT scan which revealed a well circumscribed soft tissue mass. A tumor marker study was done which was positive for vimentin, SMA and factor XIII. An immunological diagnosis of dermatofibroma was made and the patient was taken up for local resection. Post-operative histology revealed spindle cells arranged in whorls leading to a diagnosis of fibrosarcoma. The patient was symptom free for a period of 2 years when she experienced pain in the left thigh. Examination revealed a mass in the left thigh. Imaging studies (CT and $MRI_{fig.1}$) revealed a soft tissue mass extending from the popliteal fossa inferiorly to the neck of the femur superiorly. A chest X ray and abdominal USG was done which found no metastasis. The tumour was stage according to AJCC staging as T2b N0 M0 and stage III⁴. The patient was undertaken for an extensive local resection. Per operatively the tumour showed extensive soft tissue involvement. The tumour was resected with a healthy margin of 1cm and sent for a histopathological examination which revealed features suggestive of fibrosarcoma. The patient was the advised local radiotherapy post operatively and is currently undergoing physiotherapy.

III. Discussion

Soft tissue sarcomas most commonly present as an asymptomatic mass. The size at presentation usually depends on the location of the tumor. Tumors in the distal extremities are often small when discovered, whereas tumors in the proximal extremities and retroperitoneum can become quite large before they are apparent. Soft tissue sarcomas grow in a centrifugal fashion and compress surrounding normal structures, but rarely does impingement on bone or neurovascular bundles produce pain, edema and swelling³. Most soft tissue sarcomas that recur do so within two to three years of the completion of treatment. The pattern of recurrence is related to the anatomic site of the primary tumor. Distant pulmonary metastases are the typical pattern in patients with extremity sarcomas, with disease recurring in up to 20% of these patients. Patients with retroperitoneal or intra-abdominal sarcomas tend to have local recurrences. Other less common sites of metastasis in patients with soft tissue sarcomas include bone (7%), the liver (4%), and lymph nodes (less than 4%). Our case presented as a

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rare third local recurrence of the malignancy and there was no evidence of distant metastasis. The patient also complained of pain in the swelling which is a rare presenting complaint in cases of soft tissue tumors.

The tumour is staged using the American Joint Committee on Cancer (AJCC) staging criteria for soft tissue sarcomas which rely on the histologic grade, the tumour size and depth, and the presence of distant or nodal metastases (Table 1).⁴ In the present case the tumour was T2bN0M0.

The treatment modality is then decided upon depending on the stage of the tumour⁴. In the present case which was stage IB, we did wide local resection of the tumor with healthy margins and post- operative local radiotherapy was given.

IV. Conclusion

In conclusion a third recurrence within a span of 6 years without distant metastasis is a rare presentation of fibrosarcoma, which itself is a rare malignancy in India. It is imperative to go for post-operative radiotherapy in order to prevent local recurrences.

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Fig. 1



Fig. 2

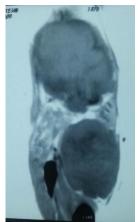


Fig. 3

Table 1.

Table 1.						
Primary tumor (T)						
TX	Primary tumor cannot be assessed					
Т0	No evidence of primary tumor					
T1	Tumor ≤ 5 cm in greatest dimension*					
T1a	Superficial tumor					
T1b	Deep tumor					
T2	Tumor> 5 cm in greatest dimension*					
T2a	Superficial tumor					
T2b	Deep tumor					
in any of the	cial tumor is located exclusively above the superficial fascia without invasion of the fascia; the deep tumor is located following: exclusively beneath the superficial fascia, superficial to the fascia, with invasion of or through the fascia, ficial to and beneath the fascia.					
Regional lyn	nph nodes (N)					
NX	Regional lymph nodes cannot be assessed					
N0	No regional lymph node metastasis					
N1	Regional lymph node metastasis [†]					
† The present	the of positive nodes (N1) in M0 tumors is considered stage III.					
Distant meta	astasis (M)					
M0	No distant metastasis					
M1	Distant metastasis					

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Table 2.

Stage IA	T1a	N0	M0	G1, GX	Local resection (margins>1cm) + Local radiotherapy
	T1b	N0	M0	G1, GX	
Stage IB	T2a	N0	M0	G1, GX	Wide local excision + Local radiotherapy
	T2b	N0	M0	G1, GX	
Stage IIA	T1a	N0	M0	G2, G3	Wide local excision + Local radiotherapy
	T1b	N0	M0	G2, G3	
Stage IIB	T2a	N0	M0	G2	Wide local excision + Local radiotherapy
	T2b	N0	M0	G2	••
Stage III	T2a, T2b	N0	M0	G3	Wide local excision + Local radiotherapy
8	Any T	N1	M0	Any G	Wide local excision + Lymphadenectomy + Local radiotherapy
Stage IV	Any T	Any N	M1	Any G	Chemotherapy +Pallaiative Surgery