Case Report

Giant subcutaneous leiomyosarcoma at knee joint: malignant tumour at rare site

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ABSTRACT

Primary subcutaneous leiomyosarcoma (LMSs) are uncommon of all soft tissue malignancies. Its frequency is about 1% to 2%, with a predilection for middle age, and found more frequently in the lower extremity. They can be of two types according to its primary site of origin: deep subcutaneous and superficial cutaneous. The physical appearance of this tumour resembles more with benign condition and can easily be mistaken for non-malignant condition. Hence, it is important to diagnose it correctly and it is important to differentiate it from other spindle cell neoplasm.

A case of giant subcutaneous leiomyosarcoma is presented here which was found around knee in lower extremity. The diagnosis was suspected by histopathology and confirmed by immunohistochemistry.

***Key words**: Leiomyosarcoma, malignant, lower extremity, subcutaneous, knee joint*

Introduction

Cutaneous leiomyosarcoma is a rare smooth muscle tumour which arises from the dermis or subcutaneous tissue of the skin. [1] Leiomyosarcoma (LMSs) are classified into two groups-first involving deep soft tissues sites as the retroperitoneum and other involving peripheral soft tissue sites. Hair bearing area of thigh is more common site. [1, 2] The peripheral soft tissue LMSs are divided into cutaneous and subcutaneous subtypes on the basis of their histogenesis and differential clinical and prognostic features. [2, 3] Subcutaneous LMS is thought to arise from small to medium sized blood vessels in the subcutaneous tissue. [1] Usually patient does not complain of any pain but on examination it is usually a tender entity. This case report is of a 65 year old male patient with giant sized subcutaneous LMSs which was located at the left knee joint. The findings are correlated with histopathological and immune- histochemical findings.

Case report

A 65 year old male, presented with painless progressively increasing swelling present at the left knee joint since 3 months. History revealed that, it started with a small nodule one year back after trauma, which progressively increased in size and was excised outside 6 months back. After about 3 months of removal of the first tumor another swelling appeared

at the same site. Clinical examination revealed a 12 cm X 8 cm partially ulcerated skin covered mass present over left knee joint fixed with the skin and underlying structure with extension up to the lateral aspect of leg. Inguinal lymph nodes were palpable. A clinical diagnosis of soft tissue sarcoma was made. The mass was excised and keeping in view of recurrence of short duration, it was sent for histopathological examination.

Pathological findings

Gross examination: Excised specimen covered with skin and measuring 12 cm X 8 cm X 5.2 cm was received. The tumour was located at the subcutaneous region, reaching up to underlying muscle. The cut surface revealed predominantly solid nodular greyish white, firm, homogenous mass with whorled appearance at places. Secondary changes in the form of cystic degeneration, haemorrhage are also seen. (Fig. 1) Inguinal lymph nodes were also received.

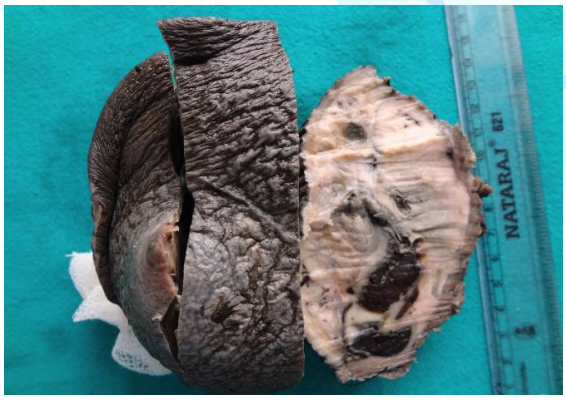

Fig.1 Skin covered excised mass with an area of ulceration

Microscopic examination: The tumour comprised of spindle cells, predominantly arranged in fascicles with focal areas of haemorrhage. The spindle cells had eosinophilic fibrillary cytoplasm with hyperchromatic but vacuolated and elongated blunt end (cigar shaped nuclei). The tumour cells reveal marked nuclear pleomorphism with the presence of occasional tumour giant cells. Mitotic figures were quite frequent, around 15-25 per 10 high power field. (Fig. 2) All the margins were free including the attached muscle at the resected base. Lymph nodes were free of tumour.

Tumour seems to arise from the thin to medium sized blood vessel wall. Immunohistochemically, tumour cells showed positivity for Vimentin and Smooth Muscle Actin (SMA), in the tumour proper as well as around the blood vessel wall. (Inset 1 & 2 of Fig. 2) With these findings, the diagnosis of subcutaneous leiomyosarcoma was confirmed.

Postoperative period was uneventful. Taking into the consideration the large tumour size, high mitotic rate, and recurrent setting, postoperative radiotherapy was given. On consequent follow up of the patient, no clinical sign of local recurrence was found.

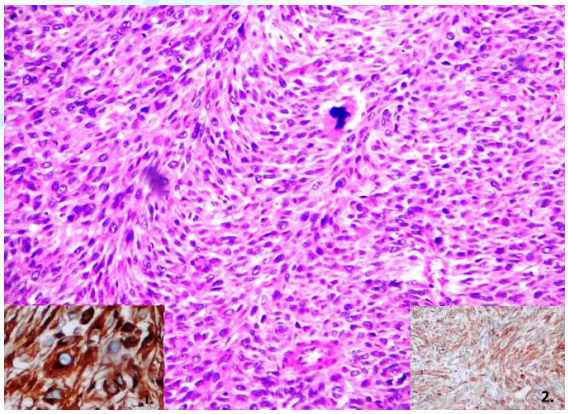

Fig. 2 Tumour comprised of spindle cells, predominantly arranged in fascicles, eosinophilic fibrillary cytoplasm with hyperchromatic, vacuolated and elongated blunt end (cigar shaped nuclei)
Inset 1- show strong positivity with Smooth Muscle Actin (SMA)
Inset 2- show moderate positivity with vimentin (H & E stain; 100X)

Discussion

Leiomyosarcoma is a rare neoplastic

disease that was first described by Montgomery and Winkelman in 1959. [2] According to their study the tumour develops most commonly in the lower limb. LMSs are divided into those involving deep soft tissues sites as the retroperitoneum and peripheral soft tissue sites. Peripheral soft tissue LMSs, are subdivided into cutaneous and subcutaneous LMSs on the basis of their histogenesis and different clinical and prognostic implications. [2, 3] Origin of tumour from these two areas cutaneous and subcutaneous region seems to be different. When cutaneous, the tumour arises from erector Pilli muscle. When it is subcutaneous in origin it is thought to arise from small to medium sized blood vessel wall, [1-5] as observed in the present case. Although most tumour present as a subcutaneous nodule in the extremities, usually measuring 30 mm or less in diameter in the reported cases. In this case it presented as giant form which is also a rare finding.

In a large series reported by Fields and Helwig, 95% of the patients with primary cutaneous leiomyosarcoma presented with a solitary nodule with a median size of 1.8 cm at the time of presentation with pain in 24% cases. [6] Clinical diagnosis of cutaneous leiomyosarcoma is difficult and diagnosed only after histopathological examination. Immunohistochemical analysis plays a key role in differentiating leiomyosarcoma from other spindle cell tumours. Histologically, the tumour is characterized by the presence of perpendicularly arranged fascicles of spindled cells, with abundant eosinophilic dense cytoplasm with vacuolated and elongated blunt ended nuclei. Differential diagnosis can be considered are, desmoplastic malignant melanoma, spindle cell sarcoma, spindle cell angiosarcoma, fibrosarcoma, malignant fibrous histiocytoma and malignant peripheral nerve sheath tumour. [3] Immunohistochemical examination plays a key role to reach a definite diagnosis: the tumour expresses vimentin and smooth muscle actin. Vimentin was moderately positive; smooth muscle actin was intensely positive. The smooth muscle actin (SMA) is the most sensitive marker and has been reported to be 100% positivity in leiomyosarcoma. Wide local excision with 3 to 5 cm surgical margin, has been recommended depending on the size of the tumour and recurrent presentation of tumour, is the primary treatment for the cutaneous leiomyosarcoma. [3-7]

Factors correlated with the adverse prognosis with subcutaneous leiomyosarcoma include tumor size (>5cm), high mitotic rate, presence of necrosis, deep seated tumours with fascial involvement, and intratumoral vascular invasion. [1] Adverse prognostic factors present in our case were: giant size, high mitotic rate and deep seated tumour. In spite of giant tumour size and history of recurrence, our case lack necrosis and vascular invasion.

Considering the giant size and history of recurrence, treatment done in this case was wide excision with free flap reconstruction. It was combined with adjuvant chemotherapy and radiation therapy. Inadequate excision or local excision without adequate margin leads to recurrence and increase risk of metastasis with high mortality rate. Previous surgery landed into recurrence in this case. In 30-60% of patients with cutaneous leiomyosarcoma, the disease complicated with metastases, usually by haematogenous route. Lung is considered as most frequently affected organ. The metastases were noted in 1 to 3 years after the diagnosis of the primary tumour. [1-3]

Conclusion

Subcutaneous leiomyosarcoma is a rare entity, which must be taken into consideration when encountered with a malignant spindle cell neoplasm of the skin. A thorough clinical and laboratory examination is essential in order to achieve fast diagnosis and curative excision of this aggressive tumour. Despite the claims of radical surgical treatment, there are chances of recurrence and the prognosis remains poor. Hence, long term follow up of patients is necessary.

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References

1. Jena S, Bhattacharya S, Roy S. Giant subcutaneous leiomyosarcoma of anterior abdominal wall. Case Rep Surg 2014;18:1-4.
2. Ciurea ME, Georgescu CV, Radu CC, Georgescu CC, Stoica LE. Cutaneous leiomyosarcoma-Case report. J med Life 2014;15(7):270-273.
3. Bali A, Kangle R, Maitrayee R, Hungund B. Primary cutaneous leiomyosarcoma: A rare malignant neoplasm. Indian Dermatol Online J 2013;4:188-190.
4. Agale SV, Grover S, Zode R, Hande S. Primary cutaneous leiomyosarcoma. Indian J Dermatol 2011;56:728-730.
5. Skoulakis C, Chimona TS, Tsirevelou P, Papadakis CE. Subcutaneous leiomyosarcoma of the neck: a case report. Cases J 2010;52(3):1-4.
6. Fields JP, Helwig EB. Leiomyosarcoma of the skin and subcutaneous tissue. Cancer 1981;47 (1):156-69.
7. Wascher R, Lee M. Recurrent cutaneous leiomyosarcoma. Cancer 1992;70:490-492.