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Case Report

Leiomyosarcoma of the Dermis - A Rare Presentation with a History of Recurrence

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ABSTRACT

Cutaneous Leiomyosarcoma is a rare tumor with an incidence of 2-3% of all the superficial smooth muscle tumors. Cutaneous leiomyosarcoma is divided based on the prognosis into leiomyosarcoma arising from the dermis and subcutaneous plane. Metastasis, recurrence and death are more common in subcutaneous leiomyosarcoma when compared with dermal LMS. Dermal leiomyosarcoma is a tumor of an adult with a male preponderance with a peak age incidence of 60 years. Commonest site is the extremities. These tumors are usually smaller in size. Histopathologically, two growth patterns are recognized, diffuse and nodular. Atypia and mitosis more than one per 10 high power fields are the diagnostic criteria that help in distinguishing cutaneous leiomyosarcoma from leiomyoma. Markers like smooth muscle actin and caldesmon are helpful in arriving at the diagnosis. Treatment is wide local excision with 2-5cm margin. The depth, size, location and grade alter the prognosis.

Key words: Subcutis, Extremities, Smooth muscle actin.

INTRODUCTION

Leiomyosarcoma is a tumor of the smooth muscles. Leiomyoma arising in the dermis and in the subcutaneous plane as such is a rare entity. The incidence is 2-3% among the soft tissue tumors. [1] The histological criteria to diagnose leiomyosarcoma arising from the cutaneous location are very much different from the leiomyosarcoma arising from deep soft tissue such as the uterus. And the prognosis also varies between the two. This presentation is done to bring to the reader's notice that when considering the differential among the spindle cell neoplasm arising from the dermis, leiomyosarcoma should be considered. Immunohistochemistry is useful in ruling out the differentials.

CASE PRESENTATION

A sixty three year old male attended the department of surgery with a painless mass in his left thigh. The mass was a nodular lesion on his left thigh measuring 8x5x4cm. It was a fixed mass. He gave a history of surgery for a similar mass at the same site two years back. He was diagnosed as Leiomyosarcoma then and was treated surgically. He did not have a regular follow up. MRI of the lesion was taken and a biopsy was sent to our department from the recurrent lesion. A provisional diagnosis of malignant spindle cell neoplasm was made and excision was advised. Later wide local excision was done, the specimen was fixed in formalin and sent to our department for histopathological analysis.

Investigations

MRI - Showed an exophytic lesion with lobulated margin measuring 8x4.9x4.7cm in the skin and in the subcutaneous plane. The report was given as sarcoma.

Gross- We received a wide local excision specimen from the thigh mass. The skin and the mass measured 14x10x6cm. The tumor was a nodular, exophytic lesion covered with skin and measured 8x5x4cm. On cut surface, it was gray- white, firm, [Fig 1] and was apparently circumscribed. There was an all-around skin and subcutaneous margin ranging from 2 to 6 cm.

Fig1: Shows a nodular, exophytic gray- white, firm and apparently circumscribed lesion [arrow] covered with skin measuring 8x5x4cm.

Histopathology- Sections studied from the lesion show squamous epithelium and malignant spindle cells arranged in interlacing fascicles in the dermis [Fig 2]. There was a clear zone between them. The individual cells have eosinophilic cytoplasm, cigar shaped nucleus with perinuclear vacuoles and showed a moderate amount of pleomorphism and atypia, [Fig 3&4]. Mitosis was 2/10 HPF, [Fig5]. Inflammatory cells composed of lymphocytes and plasma cells were seen. There was no necrosis in the multiple sections studied. Margins were free of tumor invasion.

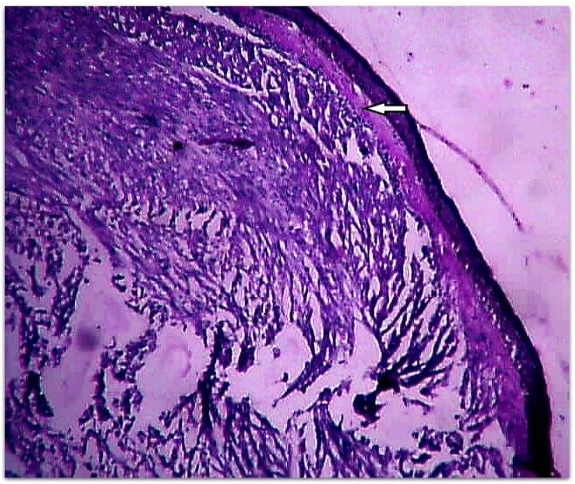

Fig 2: Show squamous epithelium and a malignant spindle cells arranged in interlacing fascicles in the dermis with a grenz zone between them[arrow], [H&E, 4x]

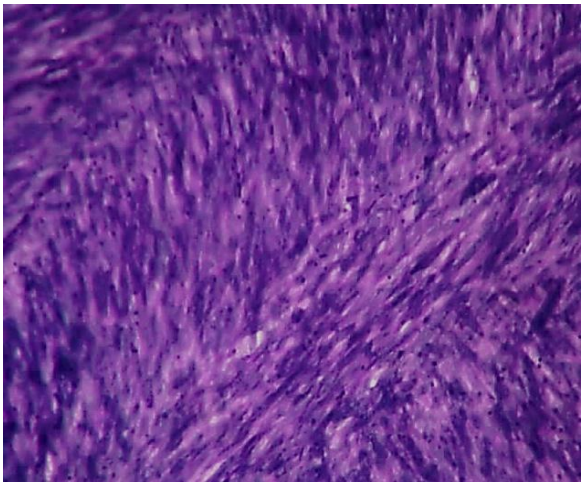

Fig3: Shows tumor cells arranged in interlacing fascicles, [H&E, 10x].

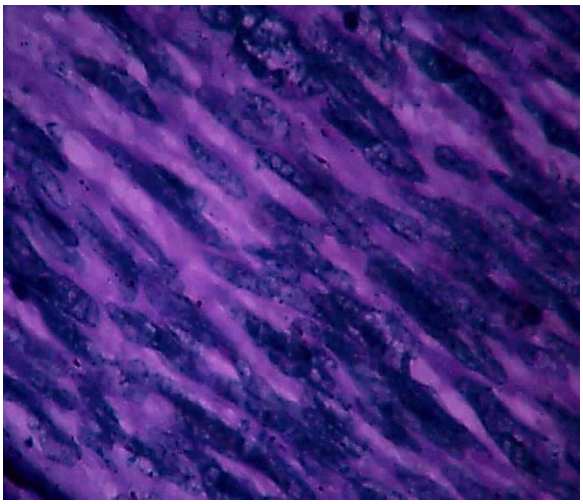

Fig 4: The individual cells have eosinophilic cytoplasm, cigar shaped nucleus with perinuclear vacuoles and showed a moderate amount of pleomorphism and atypia,[H&E,40x]

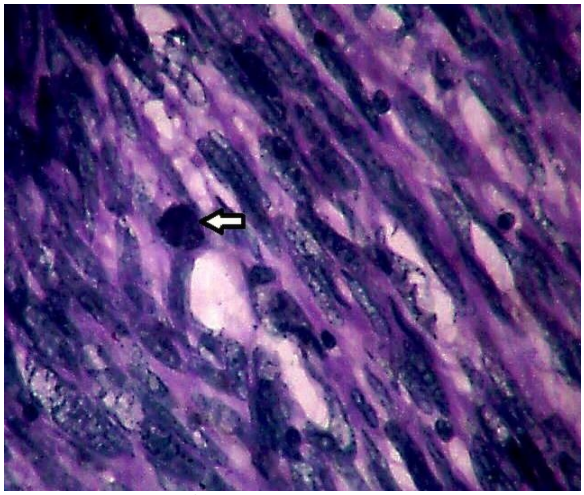

Fig5: Shows mitotic figures [arrow],[H&E,40x]

The following differential diagnosis was considered: [2]

- Dermatofibrosarcoma
- Fibrosarcoma
- Malignant Peripheral Nerve sheath tumor
- Monopahsic synovial sarcoma
- Atypical fibroxanthoma
- Angiosarcoma
- Spindle cell squamous cell carcinoma

Dermatofibrosarcoma protuberans [DFSP] also present as an apparently circumcised nodule in the dermis. Histopathologically too, this lesion is in the dermis with a clear zone in between. The spindle cells of this lesion are arranged in a storiform pattern and infiltrate the appendages and the subcutaneous tissue. This was in contrast to our case in which pleomorphic spindle cells were arranged in interlacing fascicles. Immunohistochemistry aid in the diagnosis. DFSP is positive for CD34 and leiomyosarcoma [LMS] is positive for Smooth muscle actin[SMA], Muscle specific actin [MSA], Caldesmon and Calponin. [3]

Fibrosarcoma arising from the dermis is rare. Fibrosarcoma of the cutaneous origin, mostly arise from preexisting DFSP. Our case did not show benign storiform component of DFSP nor did it show long sweeping fascicles that are characteristic for fibrosarcoma. Fibrosarcoma arising from DFSP show dim CD 34 positivity.

Malignant peripheral nerve sheath tumor [MPNST] usually arise from preexisting neurofibroma. Classical picture of MPNST like marbled effect, asymmetrically tapered spindle cells, buckled nucleus, frequent necrosis and S100 positivity were missing in our case.

Monophasic synovial sarcoma is frequently accompanied by hyalinised vascular channels and calcification along with monotonous spindle cells. These features were not seen in our case.

Atypical fibroxanthoma is a tumor of a sun damaged skin of the elderly. Usually a smaller lesion [2cm], this tumor also exhibit grenz zone. This tumor is composed of bizarre tumor cells with marked pleomorphism, mitosis and necrosis. Dilated vascular channels are seen in and around the tumor and vascular invasion is also noted. Bizarre cells, abundant mitotic figures, necrosis and vascular invasion were lacking in our case. Atypical fibroxanthoma gives variable positivity for CD34 and SMA. In cutaneous leiomyosarcoma, SMA is 100% positive as in our case. [3]

High grade angiosarcoma even though composed of spindle cells also reveals well differentiated areas with dissecting and anastomosing vascular channels. These features were absent in our case. Furthermore, angiosarcoma is positive for CD34.

Spindle cell squamous cell carcinoma arises from a well or moderately differentiated squamous cell carcinoma. This of course was not seen in our case. Cytokeratin is the marker for spindle cell squamous cell carcinoma. [3]

The location of the tumor in the dermis, fascicular arrangement, eosinophilic cytoplasm and cigar shaped nucleus, the presence of moderate atypia, the presence of more than 2 mitosis/ high power field, absence of involvement of the

subcutis and absence of involvement of the margins was considered in arriving at the diagnosis. It was reported as Dermal Leiomyosarcoma - Nodular pattern [Grade1] [FNCLCC grading system]
IHC was done to confirm the diagnosis. Smooth Muscle Actin [SMA] was positive which confirmed the diagnosis of Leiomyosarcoma, [Fig6].

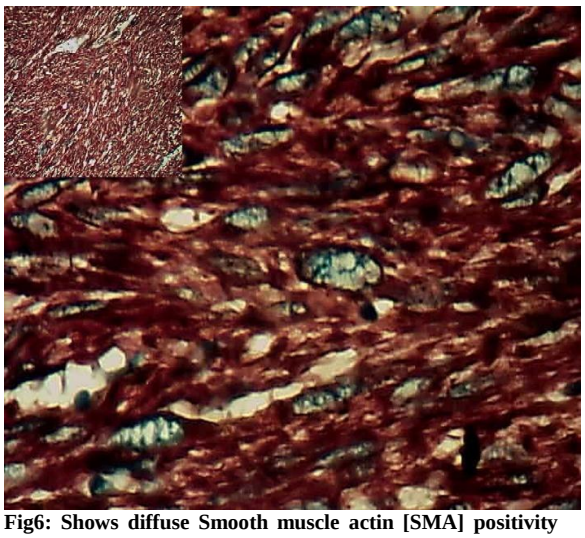

Fig6: Shows diffuse Smooth muscle actin [SMA] positivity [40x]

DISCUSSION

Cutaneous Leiomyosarcoma is a rare tumor with an incidence of 2-3% of all the superficial smooth muscle tumors. [1] Leiomyosarcoma [LMS] can be categorized based on the site of origin as leiomyosarcoma of the soft tissue and cutaneous leiomyosarcoma. Cutaneous leiomyosarcoma is further subcategorized into those LMS arising from the dermis and from the subcutaneous plane. This subdivision is important as the prognosis of these variants differs. [2,3] Metastasis, recurrence and death are more common in subcutaneous LMS when compared with dermal LMS. [2]

Origin of dermal leiomyosarcoma is from the pilar muscle, dartos muscle in the scrotum and areolar smooth muscle in case of nipple skin. [3] Scars and irradiation are proposed as the predisposing factors in some dermal LMS. [1,2] The cell of origin in case of subcutaneous leiomyosarcoma is from the vessel walls. Hence, subcutaneous LMS is categorized under smooth muscle tumors. A familial form of cutaneous LMS associated with renal cell carcinoma linked to fumarate dehydrogenase mutation is also reported. [1]

Dermal LMS is a tumor of an adult, has a male preponderance [2:1] and occurs around 50 -70 years. [2] Peak age incidence is around 60 years. The commonest site is the extremities. [1,4] Hair bearing areas like scalp and trunk is also involved. [3] The tumor is usually nodular and solitary. Multinodular cutaneous LMS is a pointer for metastasis from a soft tissue lesion. They are usually smaller and the size ranges from 0.5-3cm.

Histopathologically, two growth patterns are recognized, diffuse and nodular. Nodular pattern is more cellular and exhibit more atypia and mitosis. Diffuse pattern is composed of well differentiated smooth muscle, which is less cellular and have inconspicuous mitotic figures. [5] The cells in both these patterns have eosinophilic cytoplasm, blunt end nucleus with perinuclear vacuoles arranged in interlacing fascicles. Atypia and mitosis more than one per 10 high power fields are the diagnostic criteria to distinguish cutaneous leiomyoma from cutaneous leiomyosarcoma. [6] Atypia and mitosis may not be diffuse. Histological hotspots are described where numerous mitosis and atypia are seen focally. P53 marker studies were done to differentiate cutaneous leiomyoma from cutaneous leiomyosarcoma. [7]

Histopathological differentials include Dermatofibrosarcoma, Fibrosarcoma, Malignant Peripheral Nerve sheath tumor, Monophasic synovial sarcoma, Atypical fibroxanthoma, Angiosarcoma and Spindle cell squamous cell carcinoma. [2,6] Immunohistochemistry is a rescuer in case of dilemmas. Leiomyosarcoma is positive for Smooth muscle actin [SMA], Muscle specific

actin, Desmin, Caldesmon and Calponin. Although caldesmon is more specific it is less sensitive than SMA.

Dermal LMS tends to recur. [8] Metastasis is rare. Tumors that recur tend to metastasize more because they are larger and involve deeper tissues. Lung, and nodes are the two favored sites for metastasis. The advanced age, depth, size, location and grade alter the prognosis. [9] Subcutaneous involvement, the size more than 5cm, acral location and high grade tumor are associated with bad prognosis. Treatment is wide local excision with 2-5cm margin. Mohs' surgery is also done on this tumor.

CONCLUSION

- Cutaneous Leiomyosarcoma can be of either dermal or subcutaneous origin
- Categorizing it is important as dermal LMS is relatively benign when compared to subcutaneous LMS
- Though dermal LMS recur, it rarely metastasizes or cause death.
- Prognosis of dermal LMS depends upon size, site, depth and grade.
- It should be considered as a differential in a superficial spindle cell lesions.

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