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Cutaneous leiomyosarcomamimicking a routine lipoma on the facein a young male- A case report.

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ABSTRACT
Cutaneous leiomyosarcoma (LMS) is a malignant smooth muscle sarcoma (SMS) accounting only for less than 3% of all cutaneous soft tissue neoplasms. The need for immunohistochemical markers to arrive at a definitive diagnosis is very imperative in these cases. The subcutaneous LMS may recur after surgery but rarely metastasize to other organs. This current case report highlights a rare case of leiomyosarcoma that was diagnosed as lipoma based on history and clinical examination findings. However, the incisional biopsy findings had prompted towards a sarcoma which needed a secondary surgeryafter whicha final diagnosed of cutaneous leiomyosarcoma was established .
Keywords: leiomyosarcoma,lipoma, soft tissue sarcoma, sarcoma

I. INTRODUCTION

Cutaneous leiomyosarcoma (LMS) is rare malignant smooth muscle sarcoma (SMS) accounting only for less than 3% of all cutaneous soft tissue neoplasms. This origin of the tumour is described to be from the hair erector smooth muscle in the dermis.[1]Classically, LMS originates from the precursor mesenchymal stem cells that would eventually differentiate into smooth muscle cells. A history of radiotherapy is the most significant risk factors while hereditary retinoblastoma (RB1 gene deletion) and Li-Fraumeni syndrome (mutation in the TP53 gene) can lead to development of LMS amongst other STS. [2,3] The occurrence of a leiomyosarcoma on the face is extremely rare approximately accounting to 1%–5% of all cases. [4] The role ofimmuno-histochemical markers in marking a definitive diagnosis is unprecedented for these tumours.[5]The alarming features of this sarcomas is the rate of local recurrenceswhich are reported to range from 14% to 42% after surgery[6]Cutaneous LMSmay rarely metastasize to other organs and thus must be excised surgically and studied for its borders/ invasion under histo-pathology.[1]

This current case report highlights a rare case of leiomyosarcoma that was diagnosed as lipoma based on history and clinical findings. However, the incisional biopsy findings had prompted for a secondary surgery which was finally diagnosed as cutaneous leiomyosarcoma.

II. CASE REPORT

A 26-year-old male patient came to surgical outpatient clinic with complaints of a painless swelling on his left cheek since 2 years. The swelling had graduallyprogressed to attain the current size and is reported with no relevant history of trauma or radiation exposure. Secondary changes such as recent increase in size/ change in pigmentation/ discharge were not reported. The family history, drug history and medical/ surgical histories were non-contributory. Physical examination showed a 2 X 3 cm sized swelling in the left cheek region just inferior-lateral to the lateral canthus of eye. The swelling had a smooth surface with no secondary changes. On palpation it was non-tender, was soft in consistency associated with no palpable cervical lymph nodes. Based on the presenting symptoms and examination findings, a provisional diagnosis of lipoma was given.

The swelling was considered for an excision biopsy. The histopathology showed features a deep dermal lesion with interlacing fascicles of spindle-shaped cells with varying degrees of intermingled collagen. The spindle cells were showing an elongated blunt-ended nuclei with mitotic figures and pleomorphic hyperchromatic nuclei suggestive of soft tissue sarcoma. Patient and his attender were explained about the diagnosis

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and consent was obtained for the need of further surgery. A wide local excision was done with safety margin of 2.0 cm was carried out removing the previous scar with limberg flap. The post-surgical specimen (Figure 3A) was sent for a histopathologic examination. The histology showed XXXX suggestive of Soft tissues sarcoma (SST).

Figure 1: Initial presnaton of swelling on the face.

Figure 2: Immediate post operative view after ressection of the tumour .

The immunohistochemical staining showed positive findings of smooth muscle actin, vimentin, and CD34 and negative findings of cytokeratin, S-100, factor 8, and Melan-A.(See Figure 3B) There were mitotic figures but no invasion into surrounding tissues or horizontal or deep margins. A final diagnosis of cutaneous leiomyosarcoma was given. Post-operative period was uneventful. The patient is currently on follow up and had shown no recurrences till date.

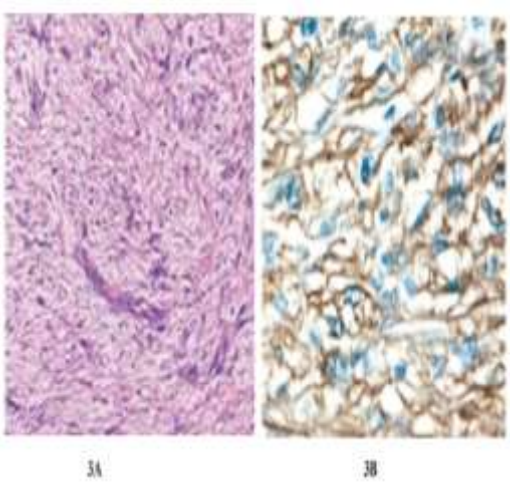

Figure 3: A) Histopathology of lesion showing featues of typical smooth muscle sarcoma (H & E stained at 40X) ; B) IHC positive reaction for smooth muscle actin (100X)

III. DISCUSSION

Cutaneous leiomyosarcoma (LMS) is a rare soft tissue neoplasm which occurs in the extremities commonly than face. The source of smooth muscles in the face may be around the sweat glands, the arrectorpili muscle (hair follicles) and the vascular endotheliala. [6] The pre-existing leiomyomas, trauma, radiation, and chemical or ultraviolet exposures have all been risk factors for this malignancy. [8] These risk factors were absent in the current case making the diagnosis a difficult task. LMS is a very rare soft tissue tumour (prevalence of 1%–5%)reported to occur in those aged between 50- 70-years.[6]This was again leading to dilemma as the current case had a young patient in his twenties. Absence of symptoms leads us towards lipoma which is common tumour of the skin. LMS were reported to have suddent onset growth, ulceration or secondary changes as per previous reports. [5]

LMS should be distinguished from other possible differentials such as the dermatofibrosarcomaprotuberans, atypical fibroxanthoma, angiosarcoma, and spindle cell squamous cell carcinoma which is only possible via histological examinations.[9] Histologically, LMS is characterised by a sharply marginated fascicles of spindle cells with elongated hyperchromatic nuclei, abundant eosinophilic cytoplasm and wide range of pleomorphism. [7]The cutaneous LMS will be positive findings for vimentin, desmin, and smooth muscle actin staining while CK, S-100, and CD34 are rarely positive. [7]In the current case, vimentin,

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smooth muscle actin, and CD34 were found positive and the CK and S-100 were negative leading to definitive diagnosis ofleiomyosarcoma.

The most effective treatment for a LMS of the face or skin is the wide local resection with 3–5 cm of clearance to prevent recurrence. The resection must be done to the depth of subcutaneous tissue and fascia. [4,7] which was accordingly adopted for the current case. The margin clearance for this tumour is reported to be 1.5cm in some papers with no recurrence after 15 months while other reported a need for 2-5 cm. [10] Considering the possibility of organ metastasis, the tumour was resected with 2 cm clear margins and currently under regular follow-up. He showed no immediate recurrence or late complications after surgery.

IV. CONCLUSION:

In conclusion, if a tumour mimicking a routine lipoma clinically may turn out to be soft tissue sarcoma or cutaneous leiomyosarcomas as in the current case. Thus, a careful diagnostic approach, detailed histopathological study and promt surgical treatment is needed for these cases.

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Conflict of interest: Nil declared

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