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CUTANEOUS LEIOMYOSARCOMA OF SCALP – A RARE CASE ENTITY

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ABSTRACT

Cutaneous leiomyosarcoma is a rare malignant spindle cell tumour which commonly develops in the bone or soft tissue. Most common sites are the head, neck, arms, legs. Spindle cell lesions of the head and neck are quite diverse with great clinical and biological heterogeneity. Some are malignant while many others are benign or simply reactive in nature. Here, we present a case of a 35 year old male who presented with complaints of a swelling on the scalp for 4 months with no pain and discharge. On examination the swelling was 3 x 2.5 x 1cm on scalp on right occipital region and clinical diagnosis was lipoma. Excision biopsy was done and histopathological examination showed features of spindle cell sarcoma, which on immunohistochemistry was confirmed as cutaneous liposarcoma. This case is reported here due to its extreme rarity.

KEYWORDS: *Cutaneous leiomyosarcoma; Spindle cell sarcoma; Malignant tumors of scalp; Smooth muscle tumors.*

INTRODUCTION

Cutaneous leiomyosarcoma is a rare malignant smooth muscle tumor and it occupies approximately less than 3% of cutaneous soft tissue neoplasms with a predilection for middle-aged to elderly male. This tumor can originate either in the hair erector muscle and be located in the dermis.[1] Moreover, cutaneous leiomyosarcoma appearing in the dermis and subcutaneous tissues involves approximately 2%–3% of all incidence of soft tissue sarcomas.[2] Morphologic differential diagnosis includes a host of other malignant spindle

cell neoplasms, thereby necessitating the use of a panel of immunohistochemical markers to arrive at a definitive diagnosis.[10]

Leiomyosarcoma can occur in all parts of the body; it is predominantly found from lower extremities and is characterized by its comparative slow growing nature. Leiomyosarcoma of the scalp is extremely rare and takes up to only 1%–2% of the total leiomyosarcomas has been reported. Cutaneous leiomyosarcoma may recur but rarely metastasize to other organs. Local recurrence rates after surgical excision have ranged from 14% to 42%.[3,4]

CASE REPORT

A 35 year old male, came to the surgery out patient department with a painless swelling in the right occipital region of scalp for the past 4 months, which gradually increased in size. On examination, swelling of 3 × 3cm was noted, which was firm in consistency, no warmth, no tenderness, no discharge was noted. Resection was done and specimen was sent for histopathological examination. Gross examination findings showed an encapsulated greyish Cut section of the tumor showed greyish white area and a soft tissue mass measuring about 3×2.5×1 cm. The tumor size was measuring 3×2×1cm. (Fig 1, 2); A malignant neoplasm with pleomorphic spindle cells arranged in a fasicles, focal storiform appearance with focal myxoid areas and mitosis (1 to 2 / 10 high power field). Histologic type shows spindle cell sarcoma probably of smooth muscle differentiation and no necrosis identified (Fig 3). Pathologic Stage Classification - pT2 Tumor more than 2 cm to less than 4cm. (pTNM, AJCC 8th Edition). On further immunohistochemistry for confirmation, SMA showed Positive, percentage of cells with nuclear positivity: 90-95 % Average intensity of staining was Strong (Figure 4). Ki67 showed Positive, percentage of cells with nuclear positivity was 35-40 % (Figure 5). CD34 shows negative for tumor cells and S100 also showed negative for tumor cells. All of the above findings helped in the confirmation of the diagnosis of cutaneous leiomyosarcoma.

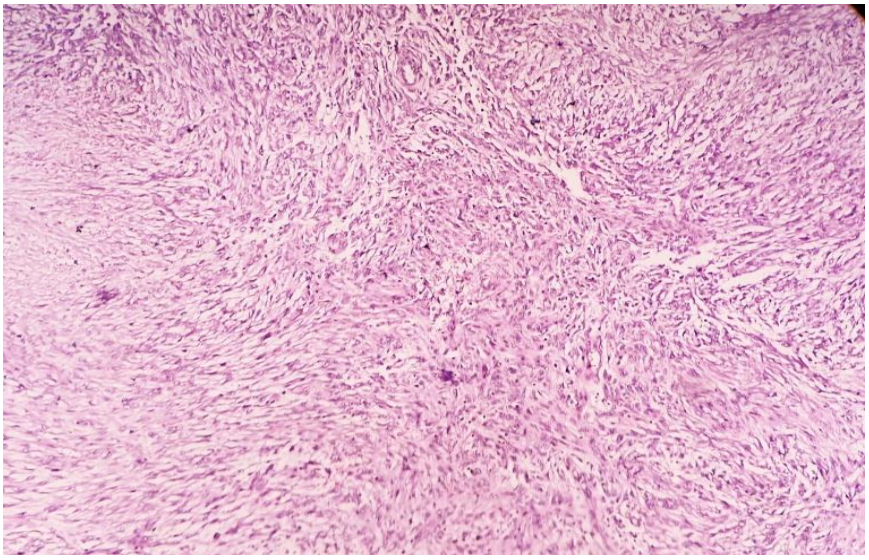

Figure 1: (H & E; 4x).

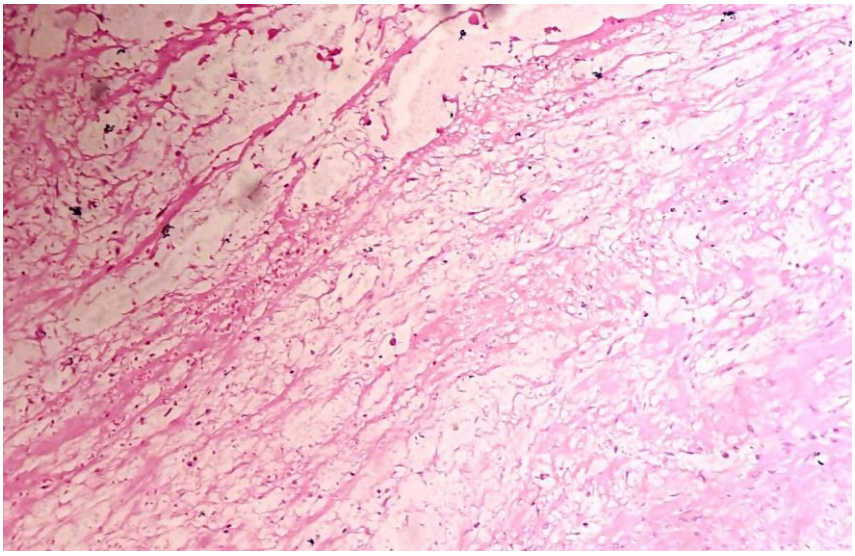

Figure 2: (H & E; 20x).

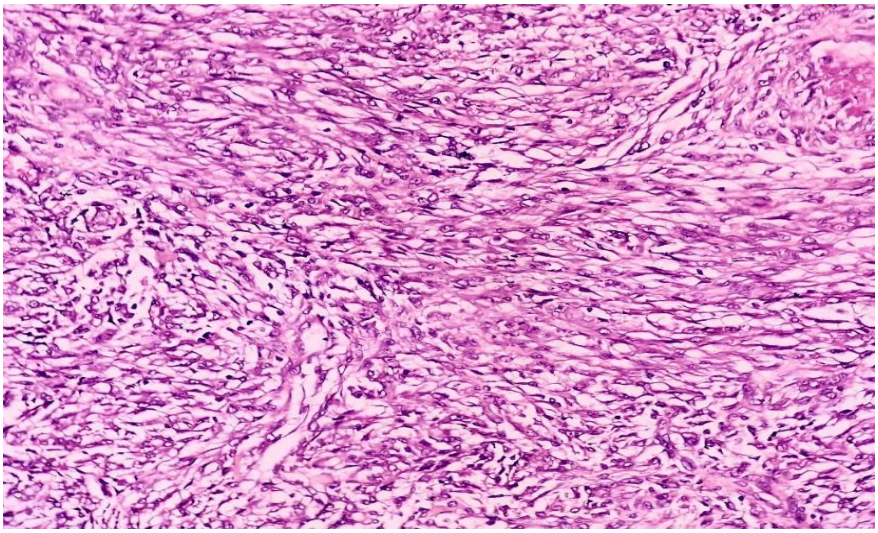

Figure 3: (H & E; 40x).

Immunohistochemistry markers

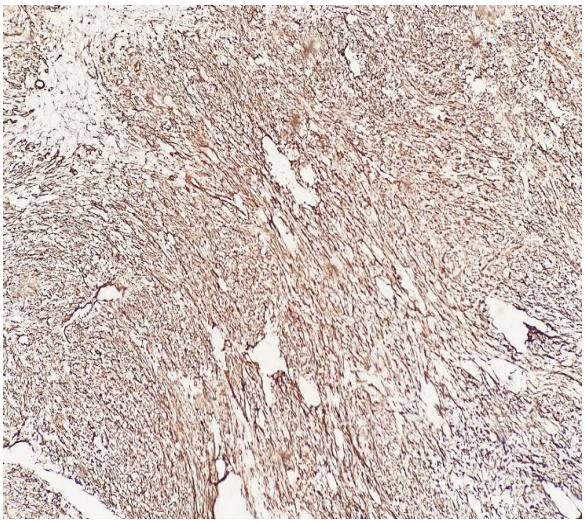

Figure 4: SMA: Positive

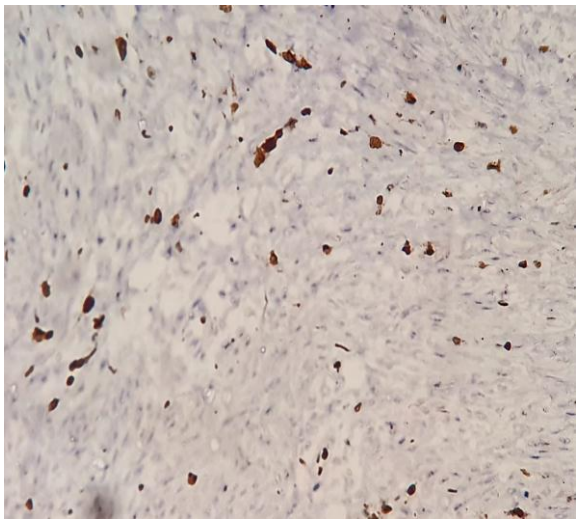

Figure 5: Ki67: Positive.

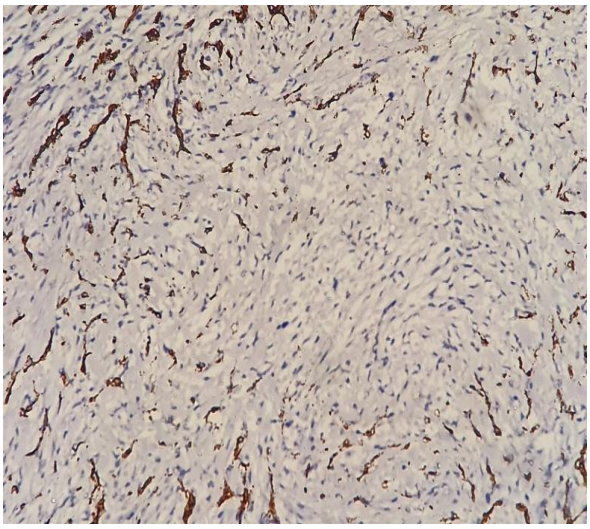

Figure 6 CD34: Negative

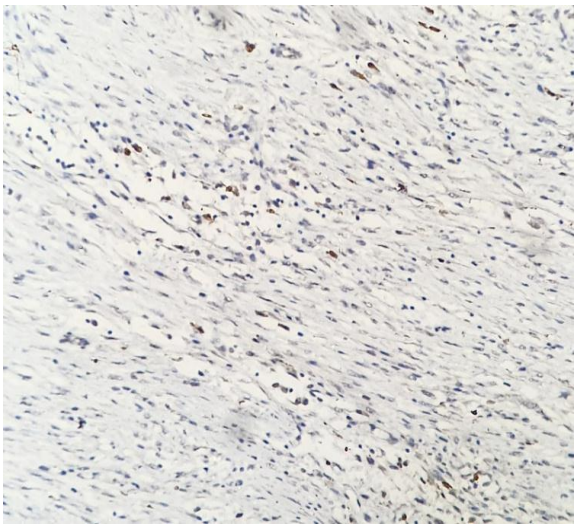

Figure 7 S100: Negative.

DISCUSSION

Cutaneous leiomyosarcoma is a rare type of tumor that occurs in the skin and subcutaneous tissues, which present less than 3% of skin and soft tissue neoplasms. The etiology of leiomyosarcoma is unknown; some relations with radiation, chemical exposure, chromosomal defects are noted.[1] Leiomyosarcoma of the skin originates from the smooth muscle around sweat glands, the arrector pili muscle of cilia and the vascular smooth muscle.[1,2]

Leiomyosarcoma of the scalp is extremely rare, which takes up to approximately 1%–3% of all soft tissue sarcomas.[5] However, in our case, the cutaneous subcutaneous leiomyosarcoma was found in the patient's scalp region.

Leiomyosarcoma should be distinguished mainly from atypical fibroxanthoma, dermatofibrosarcoma protuberans, angiosarcoma, and spindle cell squamous cell carcinoma through histological examination.[9] In cutaneous leiomyosarcoma, positive findings are shown in Ki67 and smooth muscle actin (SMA) staining. S-100 and CD34 are mostly negative. It is difficult to distinguish from some other tumors and if S-100 is found positive, then it should be discriminated from different skin cancers. In particular, S-100 immunohistochemistry is known as a useful immunoassay to differentiate it from other spindle cell neoplasms, squamous cell carcinomas, and malignant melanoma. In our case, immunohistochemical examination the smooth muscle actin and Ki 67 were found positive. CD34 and S-100 were found negative thereby the lesion was finally diagnosed as leiomyosarcoma.[10] Therefore, careful examination of cytological details in multiple sections, clinicopathological correlation and immunohistochemistry are necessary for definitive diagnosis.

The most effective treatment for cutaneous leiomyosarcoma would be a wide resection that includes approximately 2.5 to 4 cm of normal tissue to prevent recurrence, and should be resected to the depth of subcutaneous tissue and fascia.[12] Therefore, we need to monitor the patient due to possibility of a recurrence of leiomyosarcoma for a long time. The local recurrence rate of cutaneous leiomyosarcoma is known to range from 30% to 50%.[7] Hence, a long-term follow-up is necessary regarding the possibility of distant metastasis that may involve other parts of organs.

CONCLUSIONS

As cutaneous leiomyosarcoma is a very rare entity and the clinical presentation can be very nonspecific, careful histopathological and immunohistochemical examination is very necessary to give a confirmed diagnosis. Surgical excision is the unique and most effective way to treat this condition and avoid local recurrence. Recent advances in the identification of genetic alterations and immunohistochemical markers will improve the definitive management and proper treatment.

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