CASE REPORT

RARE PRESENTATION OF SUPERFICIAL LEIOMYOSARCOMA OF SCALP

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ABSTRACT

Leiomyosarcomas are uncommon malignant smooth muscle tumours, mainly derived from vessels or viscera. Superficial leiomyosarcomas are rare, represent only 7 - 10 % of leiomyosarcoma cases. Superficial leiomyosarcomas can be primary or secondary. Primary superficial leiomyosarcomas arise from the dermis (cutaneous leiomyosarcoma) or subcutaneous tissue (subcutaneous leiomyosarcoma) in the skin. They have distinctly different histologic and prognostic features from each other. The most affected parts being the extremities, especially lower extremities, followed by head and neck region. In head and neck region, primary tumors are present in only 3 - 10% of cases. We present the case of a 48 year old female, with no family history of malignancy, diagnosed with primary superficial leiomyosarcoma of scalp with frontal bone lysis and intracranial extension. The peculiarity of this case is the rarity of this tumor, uncommon location and bone involvement which has been seen only in approximately 10% of the cases.

Keywords: Superficial leiomyosarcoma; scalp; bone lysis; intracranial extension

INTRODUCTION

Leiomyosarcomas account for 7% of soft tissue sarcomas, predominantly of visceral location such as the uterus, the gastrointestinal tract or retroperitoneum[1]. Superficial leiomyosarcomas are rare, malignant smooth muscle neoplasms that arise in dermis or subcutaneous tissue. They account for only 3% of all soft tissue sarcomas[2]. We report a case of superficial leiomyosarcoma of left fronto-temporo-parietal region of scalp with bone involvement.

CASE REPORT

A 48 year old housewife presented with a painless swelling on the left fronto-temporo-parietal region of scalp for the past 6 months and bleeding from the swelling following any trivial injury for 3 months. She had history of fall and acquiring an abrasion on the left frontal region.

On physical examination, there was a scalp swelling in the left fronto-temporo-parietal region, measuring approximately 13x10 cm with multiple ulcerations of the overlying skin. Surface was irregular and the swelling was fixed to underlying bone (figure 1). Basic laboratory investigations, ECG and Chest X-ray were normal. The CT Scan examination confirmed the presence of a well defined lobulated heterogeneously enhancing soft tissue lesion in left fronto- temporo-parietal region with erosion of underlying left frontal bone with an associated extradural component (3.5x2.7x3.7cm), suggestive of soft tissue tumour (figure 2).

Intraoperatively, the left frontal bone was found eroded with an intracranial component of size 4x3.5 cm without involvement of dura. Wide local excision of the extra cranial part of the tumour with nibbling of the involved bone with complete removal of intracranial part of the tumour was done. Dura was left intact and the defect was closed with rotational scalp flap. Postoperative period was uneventful.

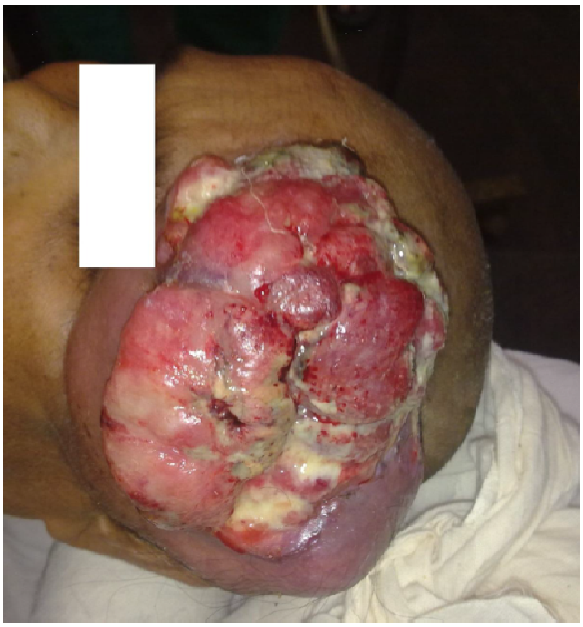

Fig 1: Scalp swelling in the left fronto-temporo-parietal region, measuring approximately 13x10 cm with multiple ulcerations of the overlying skin

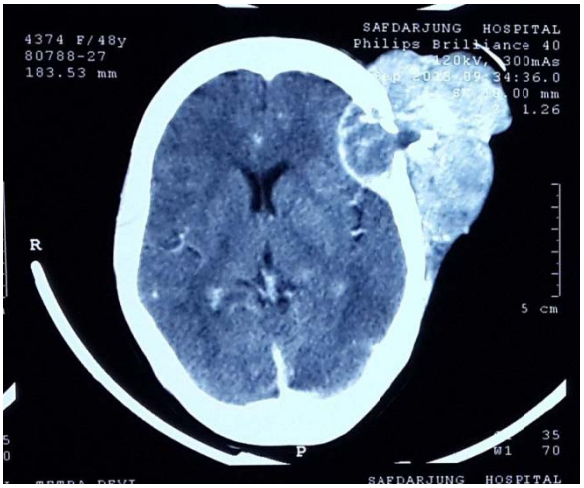

Fig 2: The CT Scan examination

Histo-pathological Examination revealed, a partially skin covered grey white tumour measuring 13x10x9.5 cm with protrusion. Microscopically, the tumour consisted of spindle cells showing severe degree of pleomorphism and high mitosis (3-5 / histopathological field), arranged in herringbone and palisading pattern. It was positive for Smooth muscle actin and Desmin and negative for Cytokeratin and S 100 protein.

DISCUSSION

Superficial leiomyosarcomas can be primary or secondary[3]. In the head and neck region, primary tumours are present in only 3 – 10 % of cases. Primary leiomyosarcomas can be cutaneous, derived from erector pili or dartous muscle or subcutaneous type from the smooth muscle wall of blood vessels[4].

Patients usually present with pain. Other common symptoms are pruritis, bleeding and burning sensation[1]. There is no age group preponderance. There is also no certain preponderance for males or females[5]. The cause is unknown; however there is some correlation with radiation, chemical exposure, trauma and chromosomal defects[5]. In our case though we couldn't find the exact cause, trauma was thought to be a possible one.

Cutaneous leiomyosarcomas grow slowly and have better prognosis than subcutaneous leiomyosarcomas, the metastatic potential being 5 – 10% and 30 – 60% respectively[6]. Most common site of metastasis is lung[4]. The factors affecting the prognosis are high mitotic index (≥ 5 mitosis per 10 histo-pathological field), high histological grade, extensive necrosis, nodular growth pattern, deep tumour and large size (≥5cm)[4].

Histologically, the tumour is composed of interlacing fasciles of elongated spindle shaped cells with eosinophilic cytoplasm and blunt tipped nuclei, which have hyperchromatic pleomorphic bizarre cells and mitoses including atypical forms in the vast majority of cases[1].

Superficial leiomyosarcoma expresses vimentin and smooth muscle actin in 100% of cases; however, desmin expression is seen in only 60% of cases. Cytokeratin (CK) and S100 stains may rarely be positive. Smooth muscle actin and desmin may assist in differentiating leiomyosarcoma from atypical fibroxanthoma and dermatofibrosarcoma protuberance which stain negatively with them. Additionally, CK and S100 stains may be helpful in differentiating leiomyosarcoma from other spindle cell neoplasms such as squamous cell carcinoma and malignant melanoma[7].

When leiomyosarcoma was documented, the whole body investigations confirmed that there was not any other site for this sarcoma, so it was documented that, this is primary leiomyosarcoma of scalp.

CT is useful in determining tumour extent, planning surgical therapy and assessing the presence of metastasis, especially in the lungs. MRI can provide additional information regarding neurovascular details, but is more useful in deeply invasive lesion[3]. Ultrasound can detect presence of tumour, presence of necrosis, sometimes bone involvement. Small tumour (<2cm) tend to be homogeneous, hypoechogeneous, with vascularisation present at Doppler ultrasound examination[4].

Treatment of superficial leiomyosarcoma consists of wide local excision (2 – 5 cm margin). Chemotherapy may only be consider for preoperative tumour volume reduction, but is not therapeutic. Radiotherapy is contraindicated for itself being a causative factor[8].

In conclusion, if a tumour of similar aspect occurs from the head and neck, we will consider the possibility of leiomyosarcoma and a careful approach is needed in early. A delayed diagnosis is correlated with large size, invasiveness into adjacent structures, with significant influence on treatment and prognosis.

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