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Case Report

Leiomyosarcoma Of The Scalp.

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Abstract

We describe a case of an elderly male subject with a primary diagnosis of leiomyosarcoma of the scalp, whose peculiarity lies in both the rarity of this tumor and its unusual site.

Keywords : leiomyosarcoma, scalp, soft tissue tumors.

INTRODUCTION

Soft tissue sarcomas are a very rare disease, comprising about 1% of malignancies.

These are rare, aggressive mesenchymal neoplasms that originate from smooth muscle. The most frequent sites of occurrence are the gastrointestinal tract, retroperitoneum, and in women the uterine site is common.

Superficial leiomyosarcomas in particular account for 7-10% of total leiomyosarcoma cases, and the most frequent sites are the extremities, especially the lower extremities, followed by the head and neck region.

The etiology is unknown; some correlations with radiation and chemical exposure, trauma, and genetic mutations have been established [1,2] and they have been associated with bilateral hereditary retinoblastoma (the result of mutations or deletions on the RB 1 gene) [3].

CASE REPORTS

A lesion was received in our O.U. of Pathology Anatomy whose macroscopic feature described a cutaneous lozenge entirely occupied by a nodular formation of 3x2x1 cm of whitish color and tense-elastic consistency.

The microscopic feature appeared to be characterized by a proliferation, at the dermal site, of a rich component of spindle-shaped cellular elements, sometimes gathered in intersecting bundles (f**ig.1,2**).

Said cells showed acidophilic cytoplasm with ill-defined boundaries and frequent perinuclear vacuolization.

Zonal nuclear pleomorphism, areas of necrosis and more than 15 mitoses x10HPF were also observed (**fig.3**).

Immunohistochemical investigations showed extensive and diffuse positivity for smooth muscle actin (clone a1A4) (**fig.4**), generic muscle actin (clone HHF35) **(fig.5**) and focal positivity for Desmin (**fig. 6**), negativity for CD34, HMB45, Pancitokeratin, pS100 and CD68.

The proliferative index, assessed by Ki-67, affected more than 75% of the neoplastic cells (**fig. 7**).

The lateral and deep margins of resection were affected of the neoplasm described above (**fig.8**)

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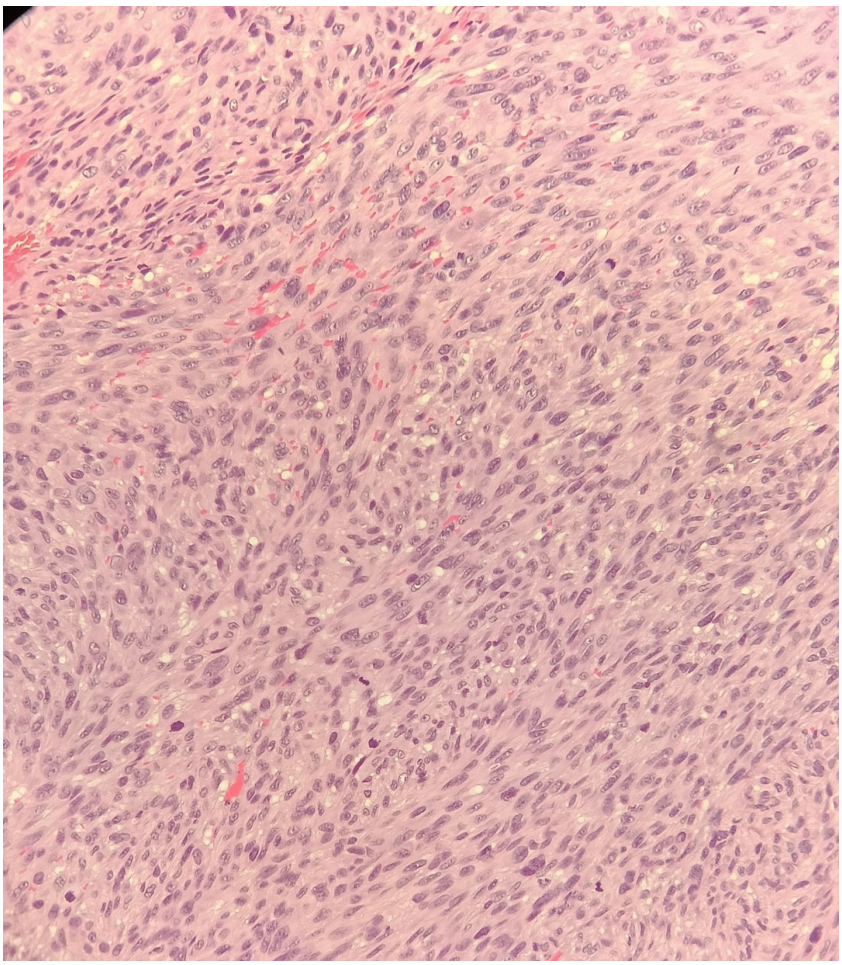
Figure 1.



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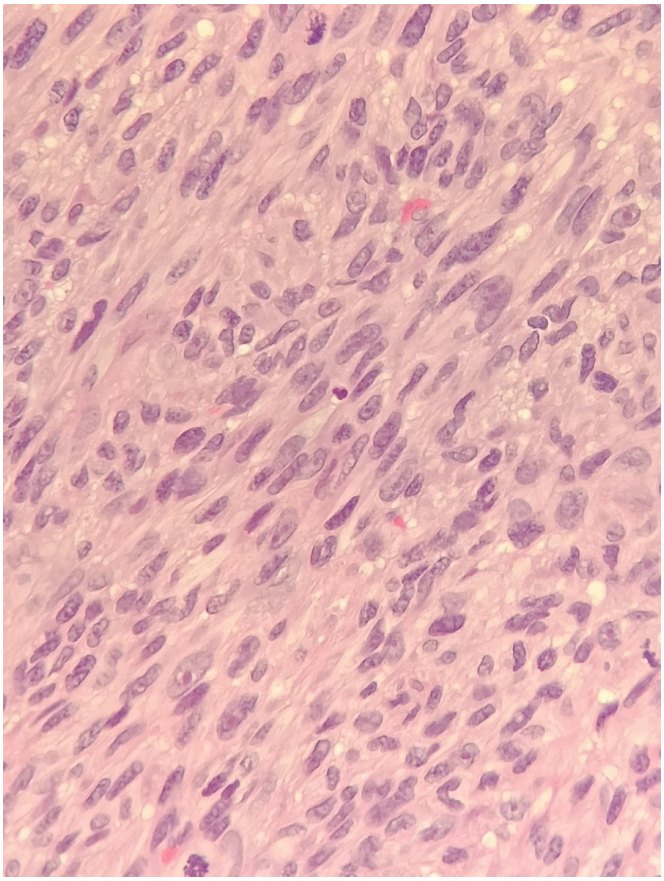

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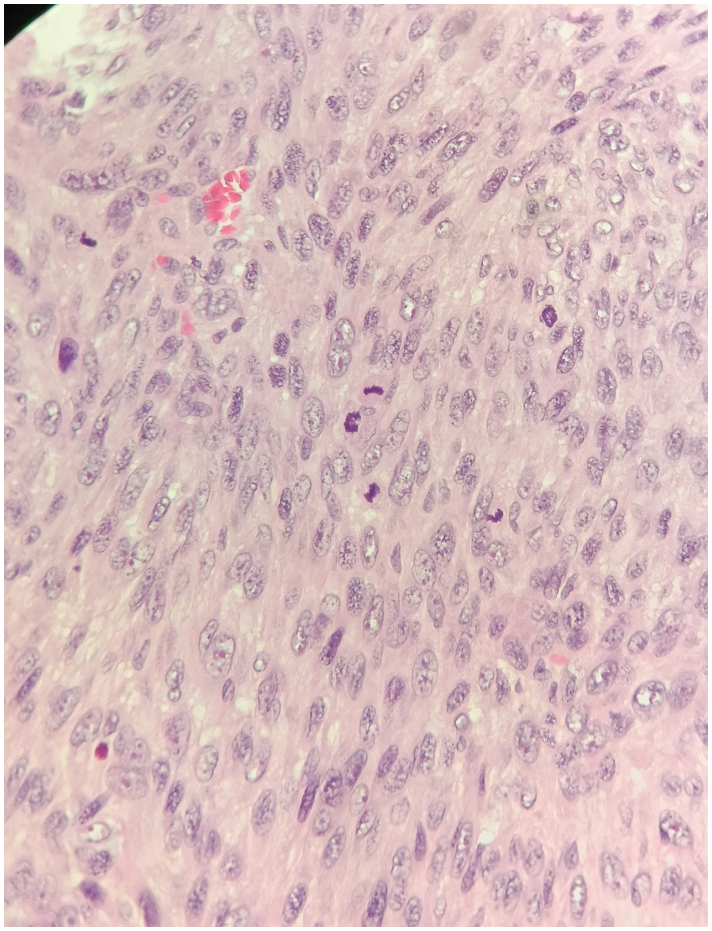

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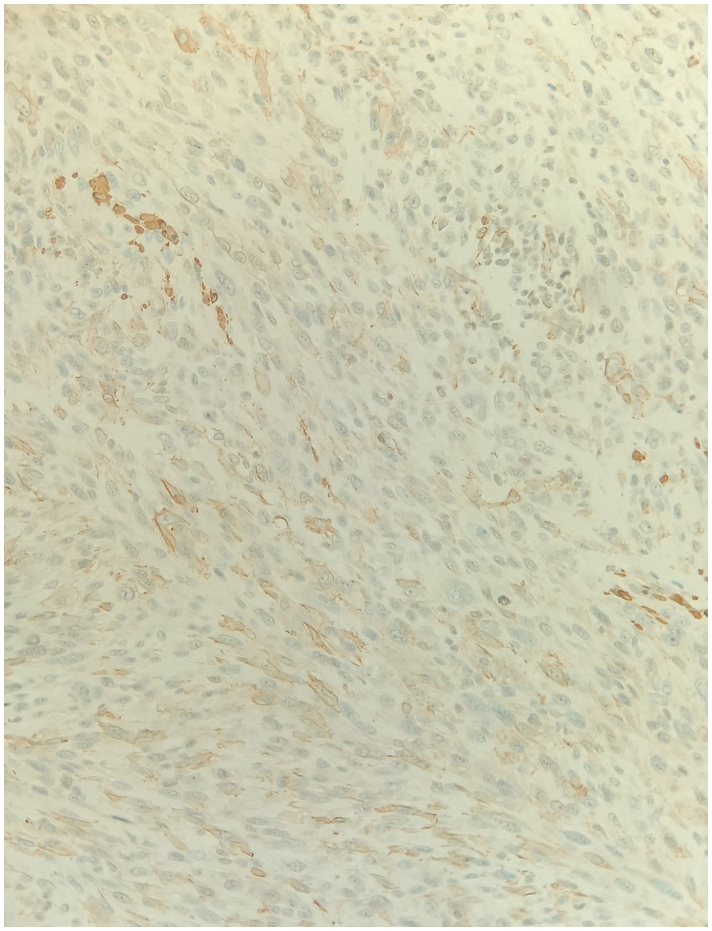

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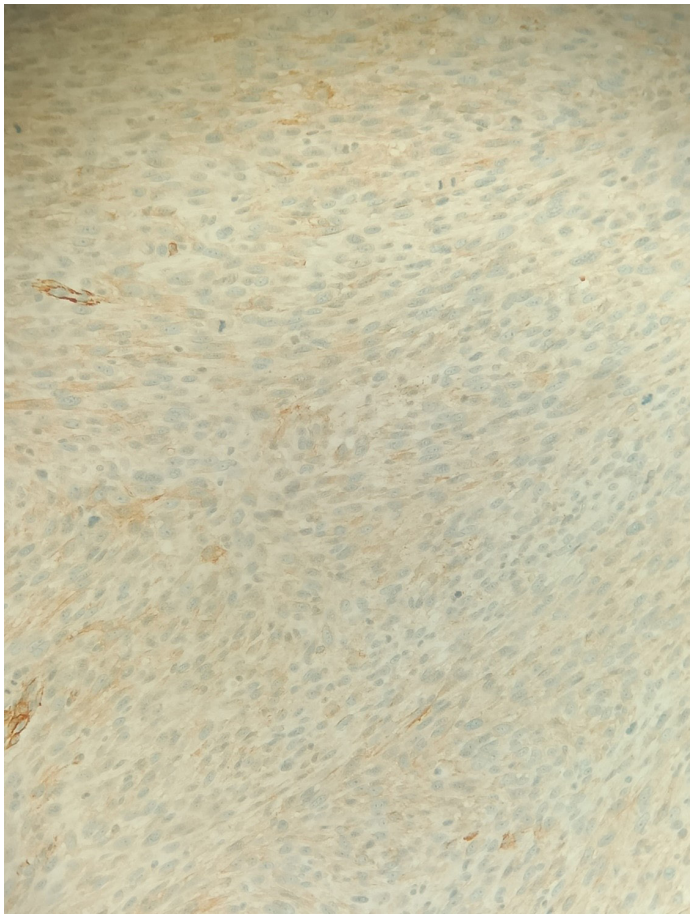

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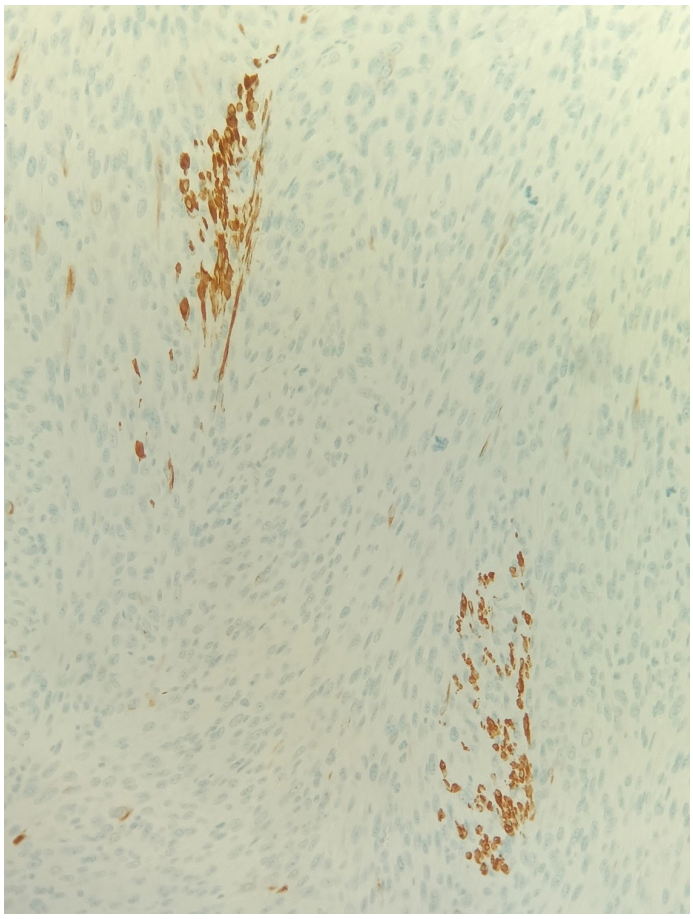

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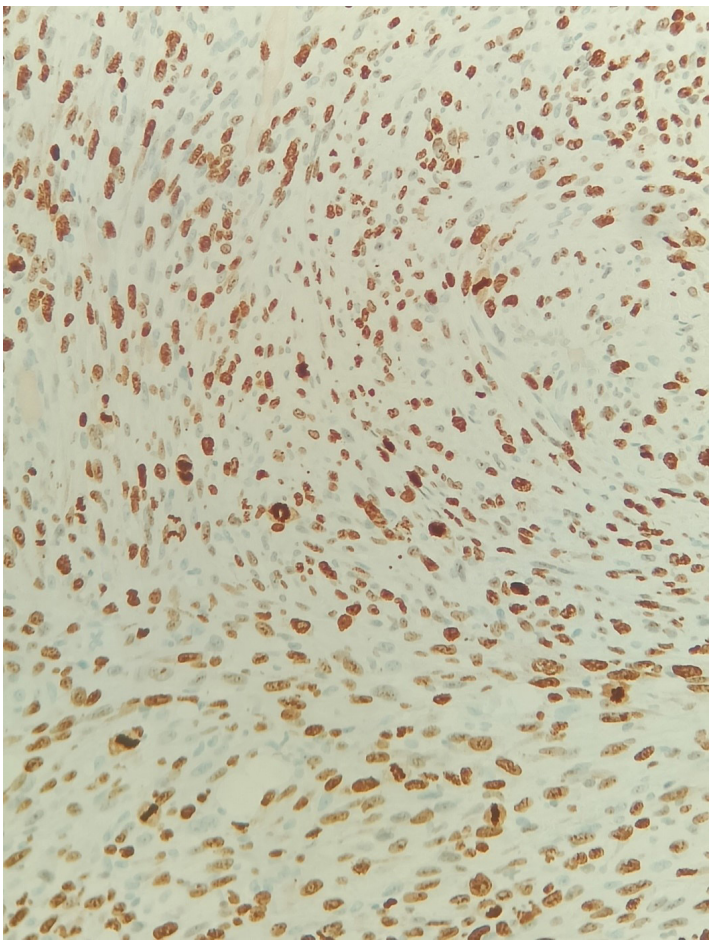
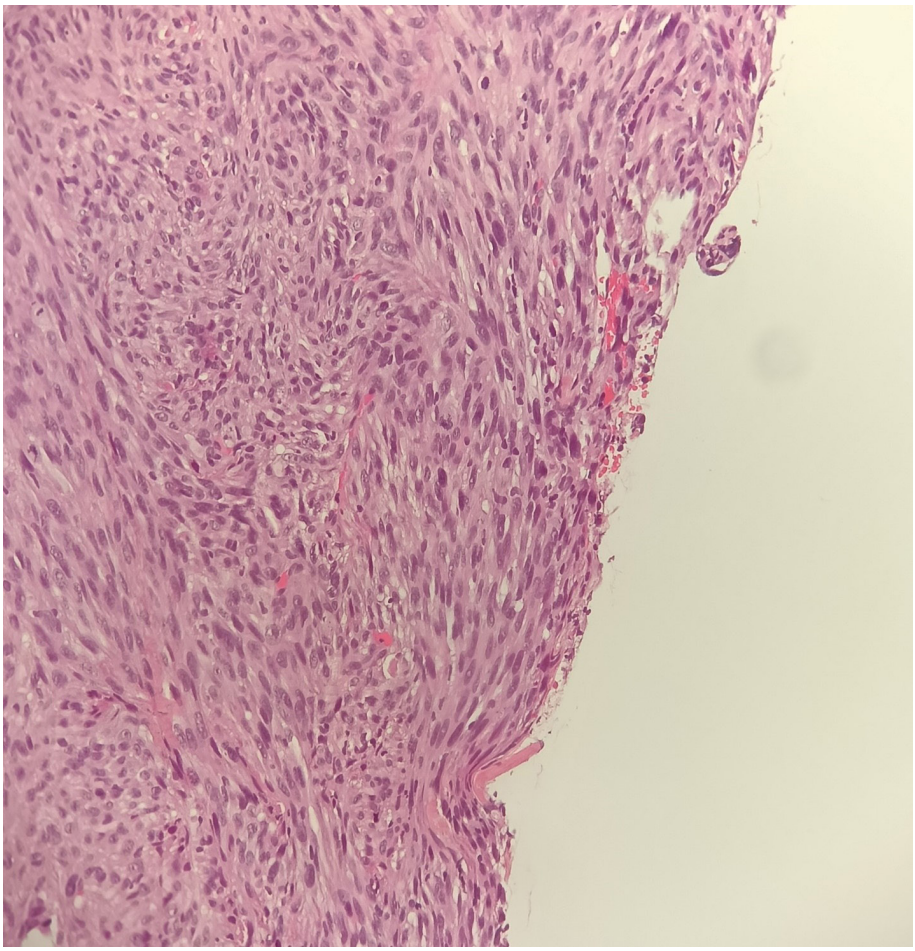


Figure 8.



DISCUSSION

Superficial leiomyosarcomas are divided into primary and secondary tumors. Primary tumors are usually solitary nodules, while multiple nodules depose more for lesions of secondary nature (mainly from neoplasms originating from the retroperitoneum and uterine site) [4,5], with a growth pattern varying from diffuse to nodular.In the head and neck region, the most common site of cutaneous leiomyosarcomas [6], primary lesions are present in only 3-10% of cases.

Primary leiomyosarcomas may be cutaneous, derived from the arrector pili, or subcutaneous, arising from the smooth muscle wall of blood vessels [7].

Any age group can be affected; cases have also been reported in pediatric age [8, 9], although mostly elderly subjects are affected [10, 11].

Subcutaneous leiomyosarcomas occur most frequently in the lower extremities, while for cutaneous leiomyosarcomas the most frequent site is the head and neck region.

Cutaneous tumors tend to be slow-growing and small (<2 cm), while subcutaneous tumors are usually larger and faster-growing.

The metastatic potential of the two superficial forms differs: a risk of 5-10% in the cutaneous type and 30-40% in the subcutaneous type.

The histologic features of leiomyosarcomas are: fascicular growth, medium-sized spindle cells, elongated “cigar-like” nuclei, eosinophilic cytoplasm, and perinuclear vacuoles.

The most common site of metastasis is the lung.

Both types, cutaneous and subcutaneous, have a high recurrence rate (about 70%), higher in subcutaneous tumors. The 5-year survival is 61-100%, and the prognosis is better in the cutaneous forms.

The differential diagnosis of leiomyosarcomas includes: lipoma, dermatofibroma, dermatofibrosarcoma, neurofibroma, spindle cell melanoma, squamous spindle cell carcinoma, and atypical fibroxanthoma.

Radical surgical excision represents the main treatment of these lesions.

CONCLUSIONS

Superficial leiomyosarcoma of the scalp is an unusual tumor both in location and frequency. Therefore, this neoplasm should also be considered in the differential diagnosis of lesions of the head and neck region.

The peculiarities of this case were the patient’s age, uncommon location, and involvement of the lateral and deep margins, with need for careful follow-up to treat local recurrences.

Availability Of Data

The data supporting the results of this study are available from the corresponding author upon request.

Conflicts Of Interest

The authors declare that there is no conflict of interest regarding the publication of this study.

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