Superficial leiomyosarcoma of the face in a human immunodeficiency virus infected albino

Case Report

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ABSTRACT

Introduction : Superficial leiomyosarcoma is a skin neoplasm of smooth muscle origin. It is commonly seen on the extremities, and it rarely affects the face. This is a presentation of a clinical case of superficial leiomyosarcoma in a human immunodeficiency virus (HIV) infected patient with albinism, and the treatment carried out with its evolution.

Observation : A 47 years-old albino woman with an HIV infection presented with a recurrent skin tumour of the face. The symptomatology was non-specific. The treatment consisted on a healthy margin surgical resection, and a reconstruction with a local bilobed pedicled flap. The histopathological and immunohistochemical analysis were in favour of a superficial leiomyosarcoma, and permit to confirm the surgical margins. The follow-up of the patient was favourable with no recurrence after 22 months.

Conclusion : Superficial leiomyosarcoma is a differential diagnosis to take into consideration in front of a clinically suspected malignant skin tumour of the face. With albinism and HIV infection, two major risk factors of skin neoplasms, oral and maxillofacial surgeons should be aware of this. Its treatment is essentially surgical, with good results over time.

Key Words: face, leiomyosarcoma, neoplasm, resection, spindle cell

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INTRODUCTION

Superficial leiomyosarcomas are rare malignant tumour from smooth muscle origin. They represent approximately 2% to 3% of soft tissue sarcomas, and about 0.04%of all neoplasms. They are commonly seen in men, with a pic of frequency between the fifth and eighth decades of life [1,2].

Superficial leiomyosarcoma can be classified into three subtypes with different clinical features and prognosis : cutaneous or dermal, subcutaneous or hypodermal, and secondary metastatic forms. The primary cutaneous leiomyosarcoma seems to derive from the erector pili muscle. It behaves as a non-aggressive neoplasm with a low risk of distant metastasis but with a tendency to local recurrence. The subcutaneous type arises from the smooth muscle of the vessels and is characterized by high rates of locoregional recurrence and distant metastasis [3- 5].

Leiomyosarcoma can occur in all parts of the body, but the most affected sites are the lower extremities, followed by the arms and the trunk. The face is rarely affected, and it takes up approximately 1% - 5% of all cases [3,6]. Generally, it occurs as a solitary and slowly enlarging lesion. There are numerous differential diagnoses, including all malignant spindle cell neoplasms. Thus, there is a need of a panel of immunohistochemical markers to confirm the

diagnosis [7- 9].

We report a clinical case of a superficial leiomyosarcoma of the face in an HIV infected patient with albinism, successfully treated with a free margin resection.

OBSERVATION

A 47 years-old albino woman presented with a progressive swelling of the left face during the last 5 months. She had HIV infection for the past 7 years with a good compliance to the antiretroviral treatment. She had a past history of a surgical resection of a same lesion on the same location 2 years ago. The patient was a healthy looking individual with normal vital signs. Extraoral examination revealed a 5 cm ulcerated swelling of the left parotid region, with necrosis and suppuration. On palpation, it was firm in consistency, tender to palpation, and not adherent to the underlying deep tissues. There was neither bleeding, nor evidence of cervicofacial lymphadenopathy. The treatment consisted

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on a surgical resection with superficial surgical margins of 1.5 cm, and deeper until the superficial parotidomasseteric fascia (figures 1 and 2), and an immediate reconstruction with a local bilobed flap (figure 3). The histopathological findings of the excised specimen revealed a dermal lesion consisting on a dense fibrous stroma and a highly cellular matrix consisting of pleomorphic round or spindle-shaped cells with hyperchromatic nuclei, and interspersed histiocyte-like cells and multinucleated giant cells. The mitotic count was 6 to 8 per 10 high power fields. The periphery consisted mostly on spindle shaped-cells arranged in a storiform pattern. Based on these findings, the first diagnosis was undifferentiated pleomorphic sarcoma. Immunohistochemistry was then performed. The tumour showed a high positivity to α-SMA, a moderate positivity to Bcl2, ad a negativity to CD34, CD99, and S100 markers. Thus, a final diagnosis of superficial leiomyosarcoma (pleomorphic type) was rendered. All of the resection margins were free of tumour. The follow-up shows good results (figure 4). At 30 months of follow-up there was no evidence of recurrence or metastasis.

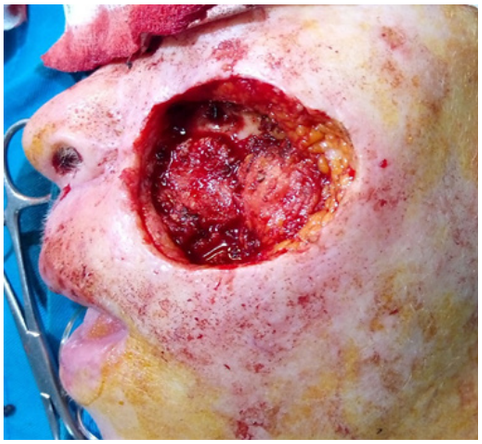

Figure 1 : Surgical wound after resection with free margins

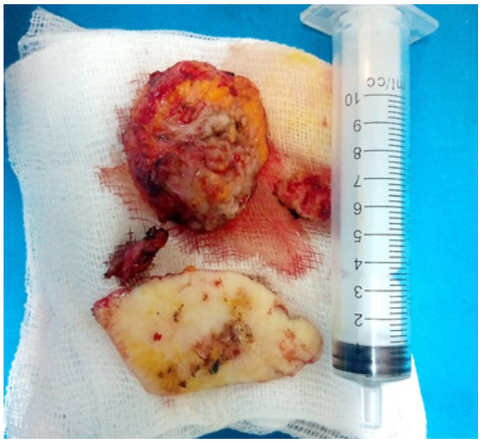

Figure 2 : Resected lesions

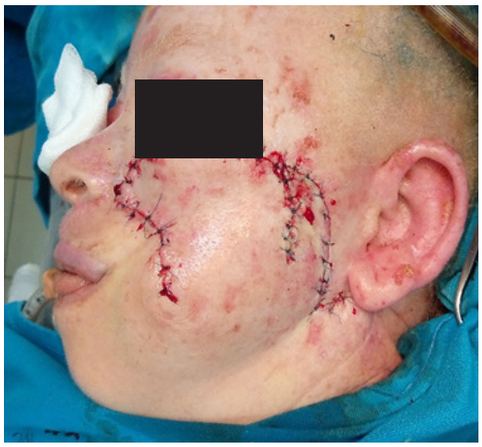

Figure 3 : Wound closure with a local bilobed flap

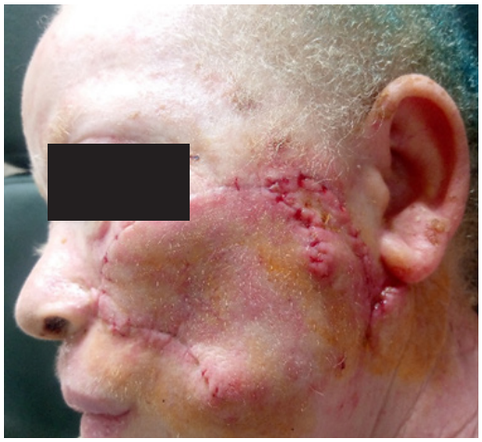

Figure 4 : Postoperative appearance after 3 weeks

DISCUSSION

Superficial leiomyosarcomas are malignant tumours that derive from the erector pili smooth muscle, or from the smooth muscle of the vessels found in the subcutaneous tissue. They can occur on any part of the body, but are mostly seen in extremities ant the trunk. The face is rarely affected with a frequency of 1% to 5%. The pic of frequency is between 50 to 70-years. Trauma, radiation, sun exposure and chemical substances are recognized to be predisposing factors [3-5,7,8]. They appear to be more common in caucasian individuals [2,9], maybe due to the lack of melanin in their skin. In our patient, the conjunction of this melanin deficiency and the sun exposure of her face could explain the occurrence of this lesion with this rare location.

Clinically, the superficial leiomyosarcoma appears as a firm slow-growing solitary nodule, with an indolent growth pattern. The size is less than 5 cm at the time of the diagnosis, and it can show ulceration due to fixation to the epidermis. Pain is usually present during palpation. Other reported symptoms include paraesthesia in the territory of the lesion, pruritis, bleeding, and burning [5,7,8].

The necrosis and suppuration seen in our patient were probably secondary to an infection of the exposed ulceration. The clinical presentation of superficial leiomyosarcoma is non-specific. Thus, the final diagnosis is based on the histological findings and the immunohistochemical examination [8]. The tumour is characterized by fascicles of elongated spindle-shaped cells with eosinophilic cytoplasm and hyperchromatic nuclei. It shows greater cellularity and mitotic activity. Marked nuclear pleomorphism and atypical giant cells can be observed [2,6,7,10]. Immunohistochemically, positive α-SMA, desmin, vimentin, and caldesmon are the best indicators for leiomyosarcomas. PS100 is sometimes positive, whereas CD34 is negative. Other immunohistochemical stains performed to differentiate leiomyosarcomas from other spindle-cell lesions include EMA, CD34, CD117, CEA, HMB45, Mart-1, Melan A and CK7; all of them are negative in LMS [3,5,9,10,11]. As histological findings in our case lead to confusion with undifferentiated pleomorphic sarcoma (another spindle cell tumour), we performed an immunohistochemistry which confirm the final diagnosis.

Complete surgical excision is the gold standard for localized tumours [1,2,4]. They should be resected to the depth of subcutaneous tissue and fascia [3,5]. There is no consensus regarding the appropriate safety margins. In the reported literature, the classic recommendations suggested margins of 3-5 cm where they can be achieved. however, similar results have been observed with margins of 1 cm, without higher local recurrence rates and with improved morbidity [1,5]. Since it can be difficult to achieve these 3-5 cm margins on the face, the histological evaluation of surgical margins is a key-point in establishing the probability of future recurrence [5]. To date, the role of radiation therapy in superficial leiomyosarcomas is still a topic of debate. Effectiveness of radiotherapy in this setting has not been determined especially that it has been considered as a causative factor itself [7-9].The metastasis rates for cutaneous and subcutaneous leiomyosarcomas are 4% (range, 3%-14%) and 43% (range, 21%-62%) respectively [9]. In our patient we could consider that the first lesion previously resected two years ago on the same location was leiomyosarcoma, even if it was not documented. The second surgery we performed allows us to have better result with no resurgence after 30 months. The final prognosis depends on the site of the tumour, its extension, and the quality of the resection [11].

CONCLUSION:

Superficial leiomyosarcoma of the face remains a rare entity. Despite his low frequency, it is a differential diagnosis to take into consideration in front of a clinically suspected malignant skin tumour of the face. With albinism and HIV infection, two major risk factors of skin neoplasms, oral and maxillofacial surgeons should be aware of this lesion. There is a need of immunohistochemistry to confirm the final diagnosis, and the aggressive surgery with safe margins is the treatment of choice.

Competing interests

The authors declare no competing interests.

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