Scientific Research Publishing

Open Journal of Stomatology, 2023, 13, 396-402
https://www.scirp.org/journal/ojst
ISSN Online: 2160-8717
ISSN Print: 2160-8709

Primary Cutaneous Leiomyosarcoma of the Face: A Rare Tumor that Is Difficult to Diagnose

Bakary Ouattara[1], Konan Marc Koffi[1*], Ory Opokou Alexandre Demisère[2], Koffi Laurent Boka[1], Rokiatou Kone[1], Aké Lucien Jonathan Yapo[1], Assoumou Lucien Asseke[1], Mouan Beatrice Harding[1], Pierre Breton[3]

[1]Department of Stomatology, Maxillofacial and Plastic Surgery of the Face, The University Hospital of Abidjan, Abidjan, Ivory Coast
[2]Department of Stomatology, Maxillofacial and Plastic Surgery of the Face, The University Hospital of Bouaké, Bouaké, Ivory Coast
[3]Department of Maxillofacial Surgery, Plastic Surgery of the Face and Stomatology, The University Hospital of Lyon-Sud, Lyon, France
Email: *marckoffi@ymail.com

How to cite this paper: Ouattara, B., Koffi, K.M., Demisère, O.O.A., Boka, K.L., Kone, R., Yapo, A.L.J., Asseke, A.L., Harding, M.B. and Breton, P. (2023) Primary Cutaneous Leiomyosarcoma of the Face: A Rare Tumor that Is Difficult to Diagnose. *Open Journal of Stomatology*, **13**, 396-402.
https://doi.org/10.4236/ojst.2023.1311032

Received: October 1, 2023
Accepted: November 3, 2023
Published: November 6, 2023

Abstract

Leiomyosarcoma is a rare malignant tumour of the lower limbs. Its differential histological diagnosis is difficult and is made in the presence of young scar tissue, leimyoma, dermatofibroma, melanoma, rabdomyosarcoma, sarcomatoid carcinoma, fibroxantoma, Darrier Ferrand dermatofibrosarcoma and myofibroblastic tumours. Treatment is essentially surgical, with margins of 3 to 5 centimetres. We report two observations of tumours localised to the face, including one case of a known leiomyosarcoma and another case initially diagnosed as a leiomyosarcoma which turned out to be a cellular myofibroma with no sign of malignancy after several readings. The aim of this work is to review the literature on this pathology while highlighting the diagnostic and therapeutic difficulties. Conclusion: A rare smooth muscle tumour with a high risk of local recurrence in the event of incomplete treatment, leiomyosarcoma in its dermal component is preferentially located in the head and neck. Its treatment is exclusively surgical and highly mutilating.

Keywords

Malignant Tumour, Smooth Muscle, Face, Surgery

1. Introduction

Leiomyosarcoma is a rare malignant tumor that can arise from the dermis or hypodermis. The dermal form may arise from the pilo-arctor muscle, the smooth muscle of the sweat glands, or the dartros muscle, while the hypodermal form

arises from the smooth muscle of the wall of cutaneous arterioles and venules [1]. It occurs predominantly in the lower extremities and rarely in the cephalic region. Its differential diagnosis with several tumors (myofibroblastic, leiomyoma, young scar tissue…) on standard anatomopathological examination is very difficult with the help of immunohistochemistry.

We report two cases of facial tumors, one of which was a proven leiomyosarcoma and the other which was initially diagnosed as a leiomyosarcoma but turned out to be a cellular myofibroma with no obvious histologic criteria for malignancy.

The aim of this paper is to review the literature on this condition, highlighting the diagnostic and therapeutic difficulties involved.

2. Case Report

Patient 1: Mr X, 80 years old, polypathological (diabetes, arterial hypertension, hypercholesterolemia, alcohol and tobacco abuse), referred from the dermatology department for management of a leiomyosarcoma of the cheek. Symptoms began 2 months earlier with the appearance of swelling of the left cheek requiring biopsy. Histological examination of the biopsy specimen revealed a proliferation of atypical spindle-shaped cells arranged in intersecting bundles, irregular elongated hyperchromatic nuclei with the presence of mitosis, elongated cytoplasm with a high number of eosinophils and immunohistochemistry was positive for smooth muscle actin (SMA). A diagnosis of leiomyosarcoma was made and subsequent excision was recommended. The patient was referred to us with the following findings: Clinically, a 2.5 cm/1.5cm oval swelling of the left jugal bone, indurated and mobile in relation to the deep plane; there were no palpable adenopathies, but numerous dyskeratoses of the face (**Figure 1**). Paraclinical examination: TAP scan: unremarkable, MRI: tumour had invaded the peaucier muscle without bony reaction (**Figure 2**).

An indication for surgery was given after PCR, and we excised the lesion under GA with a lateral margin of 15 - 20 mm, followed by simultaneous reconstruction of the different anatomical subunits using a frontal flap plus a

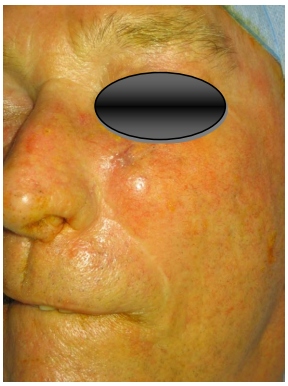

Figure 1. Indurated nodular oval jugular tumefaction.

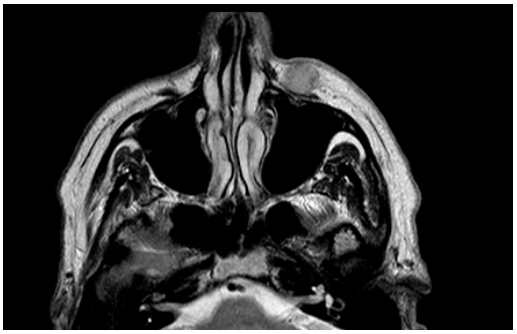

Figure 2. MRI appearance of an invasion of the eyelid muscle whith no bony reaction in front of it.

heteropalpebral flap combined with a Mustard-type flap (**Figure 3** and **Figure 4**). Microscopic examination of the surgical specimen revealed spindle-shaped cells with rounded nuclei, highly eosinophilic cytoplasm and numerous mitoses. Immunohistochemistry showed clear positivity of the spindle cells for smooth muscle actin, desmin and h-caldesmone (**Figure 5**). The diagnosis of grade 1 leiomyosarcoma was accepted.

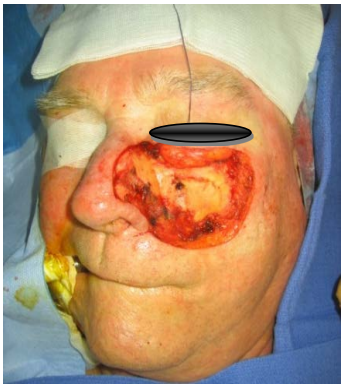

Figure 3. Carcinological exeresis with 20 mm margin laterally.

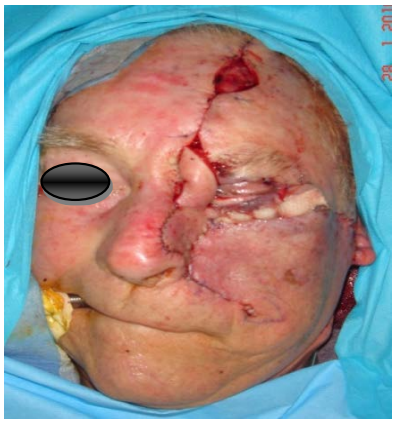

Figure 4. Reconstruction of the anatomical units by Frontal, hetero-palpebral and Mustard-types flaps.

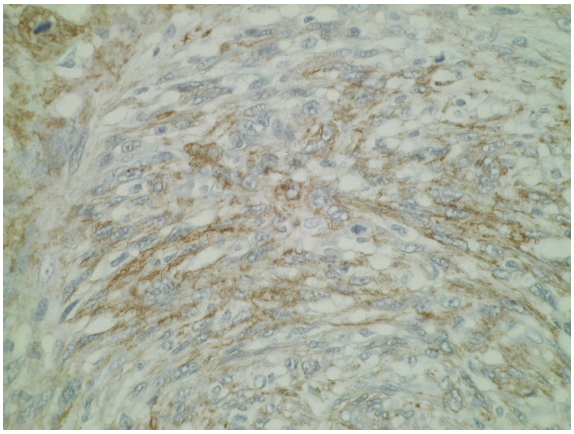

Figure 5. Spindle-shaped cells with rounded nuclei and eosinophilic, cytoplasm with many mitoses Immunohistochemistry spindle cell positivity to smooth muscle actin (AML+).

Aesthetically and functionally, the patient developed an ectropion with lacrimation, which was corrected by a Kuntz-Zymanovski flap. The carcinological evolution was unremarkable with a follow-up of 7 months.

Patient 2: Mrs Y, aged 52, with no medical or surgical history, was referred from the dermatology department for the management of a well-differentiated grade 1 leiomyosarcoma of the left nasolabial fold. Initial examination revealed an indurated scar in the nasolabial fold. Further examination revealed no metastases. Surgical excision was indicated and performed with a 10 mm lateral margin. Given the difficulty of interpreting deep sections extemporaneously, it was decided to extend the excision with multiple recuts and secondary reconstructions after definitive histology. Histology revealed a difficult differential diagnosis between scars and persistent lesions of a myofibroblastic tumour. The specimen was then sent to the Groupe Français des Sarcomes, which read it and concluded that it was a cellular myofibroma with no obvious histological criteria for malignancy. Following this diagnosis, treatment consisted of facial reconstruction using a kite-shaped nasolabial flap. The patient was followed for 5 years with no local signs of recurrence.

3. Discussion

Cutaneous leiomyosarcoma is an extremely rare tumor of smooth muscle. A distinction is made between primary cutaneous leiomyosarcomas (derived directly from the structures that make up the skin) and metastatic leiomyosarcomas (derived from distant metastases of visceral forms [2]. These leiomyosarcomas represent 3% of all soft tissue cancers, 7% of all soft tissue sarcomas, and primary forms represent 2% - 3% of all superficial soft tissue sarcomas [3]. The incidence of these tumors in the Rhône-Alpes region is 8.17% of all sarcomas [4]. They affect patients between 50 and 70 years of age, with a mean age of 60.7 years, and occur, in decreasing order of frequency, on the limbs, trunk, head and neck, most frequently in areas rich in pilosebaceous follicles. Facial localization

is uncommon 1.5. In a review of the literature, ANNEST *et al.* found that the distribution of dermal leiomyosarcoma was as follows: head and neck 48%, limbs 30%, and trunk 21% [2]. The gender distribution is controversial, but there is no gender predominance [1] [3] [5]. There are no formally identified etiologic factors, but certain elements have been implicated in the occurrence of these tumors, including trauma, ionizing radiation, burns, and sun exposure. More recently, the role of the Epstein-Barr virus in the development of leiomyosarcoma in immunocompromised individuals has been suggested [1] [6]. Other authors suggest that leiomyosarcoma originates from multipotent embryonic cells and may be radiation induced [7].

Clinically, primary cutaneous leiomyosarcoma of the face usually presents as a single, firm, sometimes painful nodule covered by skin that may be normal, erythematous, umbilicated, crusted, or ulcerated. The evolution of the initial lesion depends on the histologic type: the dermal form is characterized by a small, slow-growing nodule rarely exceeding 2 cm in size, whereas the hypodermal form grows rapidly and extends into the surrounding tissue. The differential diagnosis in this area includes basal cell carcinoma, Darrier-Ferrand sarcoma, epidermoid cyst, dermatofibroma, lipoma, neurofibroma, nevus, and benign papilloma [5] [6]. The case reported by us presents the epidemio-clinical features described in the literature, but the facial localization seems to be unusual.

The diagnostic certainty of leiomyosarcoma is based on standard anatomopathologic examination coupled with immunohistochemistry. Histology shows a well-differentiated dermal or dermo-hypodermal tumor composed of a proliferation of spindle-shaped cells with eosinophilic cytoplasm and a cigar-shaped nucleus. These cells are fasciculated or sometimes palisaded with a variable number of mitoses [2] [5] [8] [9]. Several benign or malignant pathologies can be difficult to differentiate from primary cutaneous leiomyosarcoma, including young scar tissue, leiomyoma, dermatofibroma, rhabdomyosarcoma, melanoma, sarcomatoid carcinoma, fibroxantoma, Darier-Ferrand dermatofifrosarcoma, and myofibroblastic tumor [4] [10] [11]. This histologic diagnostic difficulty occurred in our study where a myofibroma was initially diagnosed as cutaneous leiomyosarcoma. This is where immunohistochemistry comes into play. Classic immunohistochemistry of cutaneous leiomyosarcoma is positive for vimentin, desmin, smooth muscle actin, caldesmon and calponin, confirming the muscular nature of the tumor. It distinguishes leiomyosarcoma and leiomyoma (absence of mitosis) from myofibroblastic tumors, which are positive for smooth muscle actin and caldesmone and negative for desmin and calponin [1] [12] [13].

Prognostic factors have been identified: tumor size, 95% survival at 3 years if the tumor is smaller than 2 cm, 36% survival at 3 years if the tumor is smaller than 5 cm, and depth of tumor invasion; primary cutaneous leiomyosarcoma is characterized by local recurrence (30% - 50% for the dermal form and 70% for the subcutaneous form) and distant metastases, mainly in the subcutaneous form (lung, liver, bone, brain). Histopronostic factors have been identified, particularly the number of mitoses, the presence of necrosis, and tumor differentia-

tion, which allow classification into grades ranging from I to III [3] [12].

The primary treatment for leiomyosarcoma is surgical excision with a 3 - 5 cm margin. Incomplete excision or excision with inadequate margins results in local recurrence, increased risk of metastasis, and progression to a fatal outcome. This wide margin of safety makes the management of this face localized tumor a challenge. According to Jing-Yi *et al.*, Field and Helwig found local recurrence in 32% of dermal leiomyosarcomas and 47% of subcutaneous leiomyosarcomas [5]. These recurrences usually occur 1 - 5 years after the initial surgery. Metastases have rarely been described in the dermal form, in contrast to the subcutaneous form, where the incidence of metastases has been estimated to be 33%. Mohs micrographic surgery allows tissue preservation with surgical control of excision margins [3]. This surgery appears to give good results with an estimated recurrence rate of 13% compared to 30% - 40% with traditional surgery. Recurrence is a negative factor in the management of this tumor [6]. Radiotherapy is poorly defined and can never be used as monotherapy. However, in cases where surgical resection is inadequate or limited and cannot be repeated, radiotherapy is justified [8]. The role of chemotherapy in the management of these tumors is currently under debate. According to Zelek, grade II and III leiomyosarcomas deserve to be included in trials [14].

4. Conclusions

Primary cutaneous leiomyosarcoma is a rare smooth muscle tumor with a significant risk of local recurrence if incompletely treated. Long thought to be rare in the cephalic extremities, a review of the literature suggests that while the subcutaneous form is more common in the extremities, the dermal component is more common in the head and neck. Diagnosis is based on histology, always combined with immunohistochemistry. Treatment is essentially surgical, with the risk of local recurrence and metastasis if treatment is incomplete or inadequate. Given the difficulty of making a definitive diagnosis and the size of the excision margins (2 - 5 cm), which can cause problems for reconstruction, would it not be advisable to submit the biopsy specimen to the French Sarcoma Group prior to any therapeutic procedure in cases of suspected facial leiomyosarcoma?

To carry out this study, we obtained the verbal agreement of the participants.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Baccouche, D., El Euch, D., Haouet, S., Mokni, M., Cherif, F., Ouslati, F. and Ben Osman Dhahri, A. (2006) Léiomyosarcome de la lèvre. *Annales de Dermatologie et de Vénéréologie*, **133**, 988-990. https://doi.org/10.1016/S0151-9638(06)71084-5

[2] Annest, N.M., Grekin, S.J., Stone, M.S. and Messingham, M.J. (2007) A Tumor of the Head and the Neck. *Dermatologic Surgery*, **33**, 628-633. https://doi.org/10.1111/j.1524-4725.2007.33124.x

[3] Holst, V.A., Junkins-Hopkins, J.M. and Elenitas, R. (2002) Cutaneous Smooth Muscle Neoplasms: Clinical Features, Histologic Finding, and Treatment Options. *Journal of the American Academy of Dermatology*, **46**, 477-494. https://doi.org/10.1067/mjd.2002.121358

[4] Lurkin, A., Ducimetiere, F. and Ranchere-Vince, D. (2007) Evaluation des pratiques médicales et de leurs conséquences concernant la prise en charge des sarcomes en région Rhône-Alpes. Étape 1: incidence des sarcomes. *Revue de Chirurgie Orthopédique et Réparatrice de l'Appareil Moteur*, **93**, 756 p. https://doi.org/10.1016/S0035-1040(07)73269-5

[5] Lin, J.-Y. and Tsai, R.-Y. (1999) Subcutaneous Leiomyosarcome on the Face. *Dermatologic Surgery*, **25**, 489-491. https://doi.org/10.1046/j.1524-4725.1999.08290.x

[6] Clarrissa, T.F., Andrea, C., Walley, T., John, P.A. and Lisa, M.D. (2010) Superficial Leiomyosarcome: A Clinicopathologic Review and Update. *Journal of Cutaneous Pathology*, **37**, 267-276. https://doi.org/10.1111/j.1600-0560.2009.01405.x

[7] Konsem, T., Barro-Traore, F., Ouedraogo, D., Traore, A., Heid, E., and Grosshans, E. (2012) Léiomyosarcome cutané à cellules embryonnaires: une tumeur maligne cutanée rare de l'enfant. Revue Internationale du Collège d'Odonto-Stomatologie et de Chirurgie Maxillo-faciale Africain, Abidjan.

[8] Akallal, N., Afifi, Y., Bennani, B., Idrissi, M., Ait Ourhrouil, M., Senouci, K., Grosshans, E. and Hassam, B. (2004) Nodules cutanés noirs du pied. *Annales de Dermatologie et de Vénéréologie*, **13**, 207-209. https://doi.org/10.1016/S0151-9638(04)93575-2

[9] Vergier, B., Begueret, H. and Orlandini, V. (2009) Tumeurs conjonctives de la peau. Cas n˚5. Leiomyosarcome desmoplastique. *Annales de Pathologie*, **29**, 403-406. https://doi.org/10.1016/j.annpat.2009.10.017

[10] Collin, F., Gelly-Marty, M., Bui Nguyen Binh, M. and Coindre, J. (2006) Sarcomes des tissus mous: données anatomopathologiques actuelles. *Cancer/Radiothérapie* **10**, 7-14. https://doi.org/10.1016/j.canrad.2005.09.025

[11] Saiji, E. and Guillou, L. (2009) Tumeurs fibroblastiques et myofibroblastiques de la tête et du cou. *Anales de Pathologie*, **29**, 335-346. https://doi.org/10.1016/j.annpat.2009.09.003

[12] Laila, C., Kaoutar, Z., Tawfick, H., Hinde, E.F., Cherkaoui, A., Sanae, B. and Afat, A. (2007) Leiomyosarcome du larynx: à propos d'un cas. Revue Francophone de Laboratoires, **2007**, 67-69. https://doi.org/10.1016/S1773-035X(07)80185-7

[13] Perrez-Montiel, D.M., Plaza, J.A., Dominguez-Malagon, H. and Suster, H. (2006) Differential Expression of Smooth Muscle Myosin, Smooth Muscle Actin, H-Caldesmon, and Calponin in the Diagnosis of Myofibroblastic and Smooth Muscle Lesions of Skin and Soft Tissue. *The American Journal of Dermatopathology*, **28**, 105-111. https://doi.org/10.1097/01.dad.0000200009.02939.cc

[14] Zeleck, L. (2006) Chimiothérapie des sarcomes des tissus mous. *Cancer/Rdiothérapie*, **10**, 68-71. https://doi.org/10.1016/j.canrad.2005.11.003