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Surgery

Giant Darier-Ferrand Dermatofibrosarcoma of the Scalp: A Case Report

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Abstract | **Case Report**

Dermatofibrosarcoma (DFS) is a slow-growing fibrous tumor of the skin with a very high risk of local recurrence but low metastatic potential. In this work, we report a case of a 43-year-old male patient who was treated in the university hospital of Ibn Sina of Rabat for a giant DFS of the scalp. This entity represents a tumor whose prognosis and risk of progression are mainly linked to the diagnostic delay and the quality of the first excision. Late diagnosis makes excision and reconstruction surgery difficult like in our case. The chances of cure in the case of well-conducted primary surgery are significantly higher than those of salvage surgery. Improving the prognosis requires early and codified multidisciplinary care, hence the importance of raising awareness and informing the general practitioner for early diagnosis and correct referral of these patients to specialist centers.

Keywords: Giant Tumor, Dermatofibrosarcoma, Scalp Neoplasm, Resection Margin.

INTRODUCTION

Darier-Ferrand dermatofibrosarcoma (DFS) is a low- to intermediate-grade soft tissue sarcoma originating in the dermis of the skin. It is a rare fibrous cutaneous tumor representing 0.1% of malignant skin tumors and 2 to 6% of all soft tissue sarcomas. DFS preferentially affects young adults between 20 and 50 years of age; there is no clear gender predominance. It usually presents as a nodular skin mass located mainly on the trunk and extremities. Its aggressiveness is primarily local, there is no lymphatic dissemination, and the rate of general metastases is less than 5%. Therapeutic management involves surgery with wide margins of 3 to 5 cm to reduce the recurrence rate; marginal excision is accompanied by a local recurrence rate of around 40% [1, 2]. Its prognosis is linked to the quality of its surgical excision.

CASE PRESENTATION

A 43-year-old patient with no significant medical history presented with a large, painless multinodular mass on the left scalp, which had been increasing in size for the past 2 years (Fig. 1). Clinical examination revealed a well-demarcated, firm, and painful and bleeding skin tumor (16 x 13 x 7 cm) located on the temporal, parietal and frontal regions of the scalp. A skin biopsy was performed and showed proliferation of atypical fibroblasts, confirming the diagnosis of dermatofibrosarcoma protuberans of Darier-Ferrand type. Radiological examinations (CT-scan and MRI) showed localized tumor extension into the subcutaneous tissues with no signs of endocranial spread or metastases. No bone involvement was observed. After discussion with the patient, a radical surgical excision of the tumor is performed with a sufficient safety margin to minimize the risks of local recurrence followed by milling of the bone triggering the process of directed healing. (Fig. 2) The result was very satisfactory at 1 year post-surgery. (Fig. 3) A minimal loss of substance opposite the left frontal sinus was observed and is planned for reconstruction by a local flap after the ending of the inflammatory process.

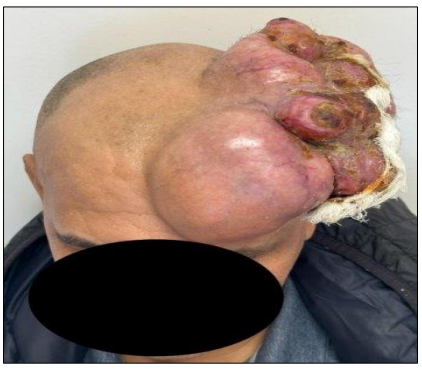

Figure 1: Image showing the DFS of the left region of the scalp

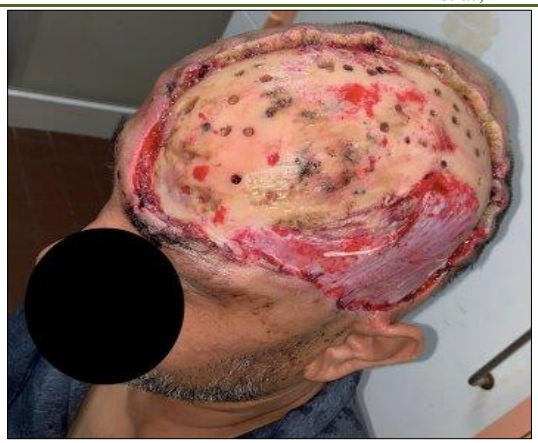

Figure 2: Three days after surgery

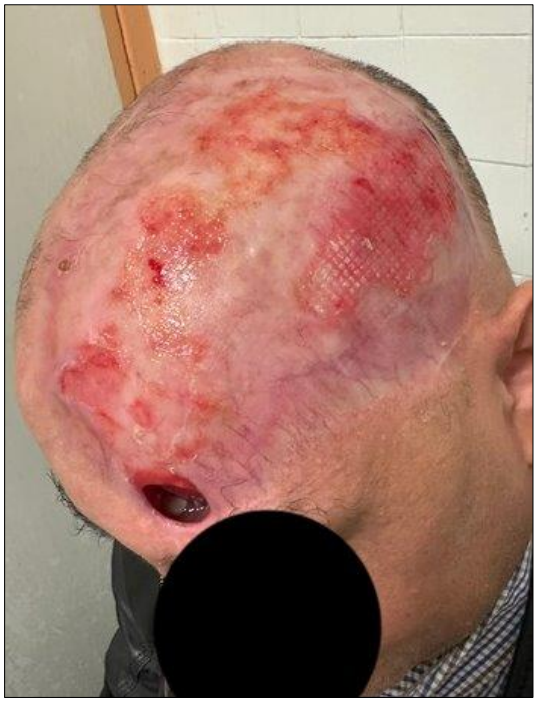

Figure 3: One year after surgery

DISCUSSION

Dermatofibrosarcoma is a slow-growing mesenchymal cutaneous tumor with high local aggressiveness and a high risk of recurrence. It generally develops in the trunk or extremities. In the majority of cases, the tumor remains asymptomatic for a long time. Cranial (scalp) and maxillofacial involvement is rare, particularly in cases of extension of the cranial vault and intracranial. It affects more young adults between 20 and 50 years of age with a male predominance, although cases in children and newborns have been described. It represents nearly 0.1‰ of all cancers, for 2-6% of soft tissue sarcomas with an incidence in the black population estimated at 6.5 cases/million inhabitants/year (twice that of Caucasians) [3]. Craniofacial location represents 1 to 10% of all possible locations; predominantly on the scalp, forehead, and cheek.

Repeated trauma (10-20%), a "t" chromosome translocation [17COL1A1 (Collagen Type 1 Alpha 1); 22 PDGFB (Platelet-Derived Growth Factor Beta)], or a previous scar (burn, vaccination, surgery, wound) appear to be the currently listed etiological factors causing this sarcoma [4]. None of these could be found in our patient. This is a specialized connective tissue sarcoma because it is histologically of fibroblastic origin, originating from the dermis of the skin, and is of low to intermediate grade. The histoprognostic grading of the French National Federation of Cancer Control Committees classifies this sarcoma as Grade 1 [Certain histological type; Mitotic index = 10 – 19 mitoses/10 fields (1 field = 0.1734 mm^2)], therefore at low metastatic risk and overall ten-year survival equal to 85% [2]. Generally, its initial macroscopic appearance is that of a fibrous plaque or a pink dermal nodule, evolving into a multinodular plaque years later. Its infiltrative capacity dictates its

wide excision. The cutaneous infiltrate can reach the subcutaneous muscular fascia.

Diagnosis is often delayed due to the slow and tolerated growth of the tumor. If left untreated, tumors become very large. They push away the surrounding tissue and adhere to the deep layer or ulcerate, becoming painful and bleeding. This same observation was observed in our case. The size of the lesion can be extremely variable, generally depending on the time to consultation. It averages 1 to 5 cm, up to 40 cm. In the reported case, the tumor size is larger than the average observed in the literature.

This could be explained by the longer consultation time, leading to a diagnostic delay observed in our setting. Typically, most lesions remain stable or progress gradually for some time before accelerated growth is observed. The average time between the onset of the lesion and the first request for treatment is generally 7 to 9 years [5]. Confirmation of the diagnosis is provided by pathological examination, possibly supplemented by immunohistochemistry.

In the reported case, the pathological examination confirmed the clinically suggested diagnosis. Immunohistochemistry with CD34+ reactivity also supported the diagnosis.

CT and MRI provide information on the deep invasion of the tumor. CT allows for a better analysis of bone structures, as the tumor appears as a soft tissue mass with tissue density without any particular specificity.

Surgery is the only treatment with proven efficacy in eradicating the tumor and preventing recurrence. This surgical approach consists of wide excision with 2 or 3 cm margins on healthy skin and extending deep to the underlying fascia in a single block. Some authors recommend [6], widening the excision margin to 5 cm on the surface, thus sacrificing a healthy anatomical barrier at depth. Countries with high-level technical facilities use the Mohs technique with frozen section examination. This involves first removing the majority of the tumor, then performing cryogenic sections on the underside of the surgical specimen, which allows for the collection of horizontal tissue slides. After reading, the invaded areas are treated again in the same manner until no tumor tissue is found on the sections. Teams using this technique have shown that lateral excision margins of 3 to 2.5 cm are sufficient. Recent data suggest that radiotherapy can significantly reduce local recurrences and improve surgical outcomes by allowing more conservative interventions [7]. However, chemotherapy, although combined with radiotherapy in certain palliative situations, has not shown significant efficacy in terms of overall survival, despite the various protocols tested.

DFS has a good prognosis when treatment is well conducted but nevertheless requires lifelong clinical monitoring, with some cases of recurrence having been described very late.

Conclusion

Protuberant dermatofibrosarcoma of the scalp remains uncommon. The fear associated with this cranial location is the exhaustive development of its tumor properties, which are recurrent with risk of sarcomatous transformation and underlying tissue infiltration. The standard therapeutic arsenal combines surgery, radiotherapy and/or molecular therapy to guarantee a complete cure. The other delicate parameter of this condition is the delay in consultation or even the primary consultation in dermatology. Therefore, a multidisciplinary consultation is essential for efficient management and adequate and broad public awareness.

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References

1. Marks LB, Suit HD, Rosenberg AE, Wood WC. Dermatofibrosarcoma protuberans treated with radiation therapy. Int J Radiat Oncol Biol Phys. 1989 Aug; 17(2):379- 84.
2. Rutgers EJ, Kroon BB, Albus-Lutter CE, Gortzak E. Dermatofibrosarcoma protuberans: treatment and prognosis. Eur J Surg Oncol. 1992 Jun; 18(3):241-8
3. COURTNEY J, GORLICK RG, GELLER DS. Dermatofibrosarcome Protubérant (Dermatofibrosarcome de Darier-Ferrand). Lid Shriv Sarcoma Init. June 2009;6(3) & 2010;7(6).
4. HANDOLIAS D, MACARTHUR G. Dermatofibrosarcome de Darier-Ferrand. Orphanet, Octobre 2008 ORPHA 31112 OMIM 607907
5. Kasse, A., Dieng, M., Deme, A., Fall, M. C., Drabo, B., Timbely, G., ... & TOURE, P. (1999).
6. Elamrani, D., Droussi, H., Boukind, S., Elatiqi, K., Dlimi, M., Benchamkha, Y., & Ettalbi, S. (2014). Le dermatofibrosarcome de Darier et Ferrand, une tumeur cutanée particulière: à propos de 32 cas et revue de la littérature. Pan African Medical Journal, 19(1), 196.
7. Mark RJ, Bailet JW, Tran LM, Poen J, Fu YS, Calcaterra TC. Dermatofibrosarcoma protuberans of the head and neck: A report of 16 cases. Arch Otolaryngol Head Neck Surg. 1993 Aug;119(8):891–6.