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Darier and Ferrand Dermatofibrosarcoma: Do Not Miss the First Exeresis

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Abstract **Case Report**

Individualized as a true anatomical and clinical entity in 1924 by Darier and Ferrand, dermatofibrosarcoma is a rare cutaneous tumor of intermediate malignancy, which is characterized by its slow evolution, its local aggressiveness and a high power of local recurrence and the rarity of metastases [1]. It affects preferentially 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 [5]. Its prognosis is related to the quality of its surgical removal. We report a case of an advanced and recurring darier and ferrand dermatofibrosarcoma of the back on histological evidence, evolving for 8 years of an 53 year old man, he was operated 4 years before he showed up at our department at the same location without any documents, computed tomography was performed showing an image of a tumor process centered on the left trapezius muscle, hyper vascularized and locally infiltrating, the decision of the staff was to make a large exeresis with margins as described in the literature with immediate coverage by a long dorsal flap.

Keywords: Darier and Ferrand, Dermatofibrosarcoma, Reccurent, Long Dorsal Flap.

INTRODUCTION

Intermediate between the harmless fibro- ma and the dreaded sarcoma Darier et Ferrand dermatofibrosarcoma (DFS) is a low to intermediate grade soft tissue sarcoma arising from the dermis of the skin. It is a rare cutaneous fibrous tumor representing 0.1% of cutaneous malignancies and 2-6% of all soft tissue sarcomas, occurring at any age but especially in adulthood with a slight male predominance. It presents clinically as a firm reddish plaque or nodule [3]. Despite its distinct histological presentation, its histogenesis remains undefined [4]. Its aggressiveness is primarily local, there is no lymphatic dissemination and the rate of systemic metastases is less than 5%. Therapeutic management involves surgery with wide margins of 3 to 5 cm to reduce the recurrence rate; marginal resection is associated with a local recurrence rate of about 40% [1, 5]. We report an uncommon case of an advanced and recurring darier and ferrand dermatofibrosarcoma of the back, evolving for 8 years of an 53 year-old man.

CASE REPORT

An 53-year-old Mediterranean man, of low socio-economic level, with a history of chronic smoking at the rate of thirty packs per year not weaned at the time of admission.

Initially the patient reported a purplish nodule progressively increasing in size for which the patient consulted 4 years after its appearance thus benefiting from an exeresis without any documents, a few months after his gesture the lesion recurred locally evolving progressively and slowly in size thus motivating him to consult in our training. On admission, the patient was stable, with a normal colored conjunctiva, weight 52 kg without any notion of weight loss. Locally, he presented a large left cervical--dorsal mass measuring 19 cm, painful, fixed, hard consistency and purplish color with the presence of several telangiectasias, not bleeding. (Fig 1)

After the agreement of the anaesthetists, a large surgical exeresis with wide margins was made, the dissection reached the upper edge of the scapula thus allowing to make bone cuts followed by a cove- rage by a long dorsal flap thus allowing to create a new loss of substance for which a round block was made. (Fig 3)

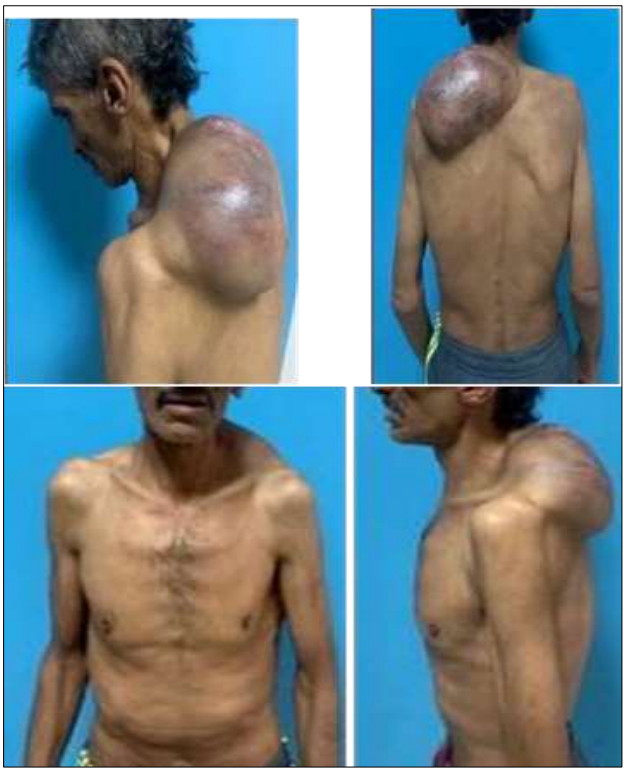

Fig. 1: Patient admission photos

An anatomopathological study was re- quested and came back in favor of a dermatofibro- sarcoma of darier et ferrand with immunohistochemical confirmation.

A CTAP angio scan was performed, showing an image of:

- Individualization of a voluminous tumoral process centered on the left trapezius muscle measuring 13x19x16.6 cm, vascularized by branches coming from the two subclavian arteries and drained by dilated veins that drain into the subclavian veins, the process comes into contact with the semispinous muscles of the neck and thorax, the rhomboid muscle, and the homolateral subspinous muscle with loss of the fatty separation line. It also comes into contact with the left subscapularis muscle with respect for the fatty separation line. It also comes into contact with the left acromioclavicular joint and the spinous processes of C7, DI, D2 and the internal edge of the scapula without detectable bone lysis. (Fig 2)

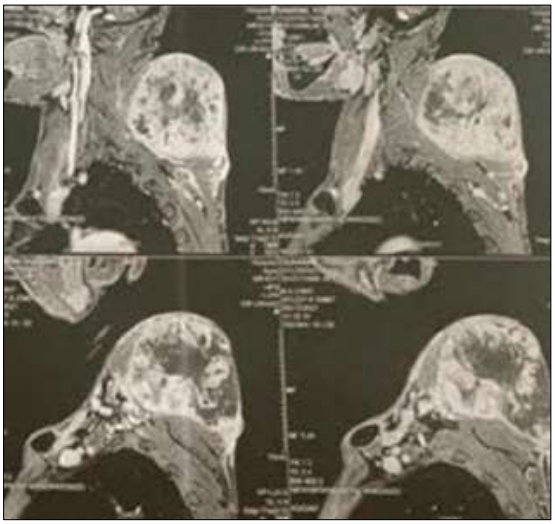

Fig. 2: A CTAP angio scan showing an image of a voluminous tumoral process centered on the left trapezius muscle measuring 13x19x16.6 cm

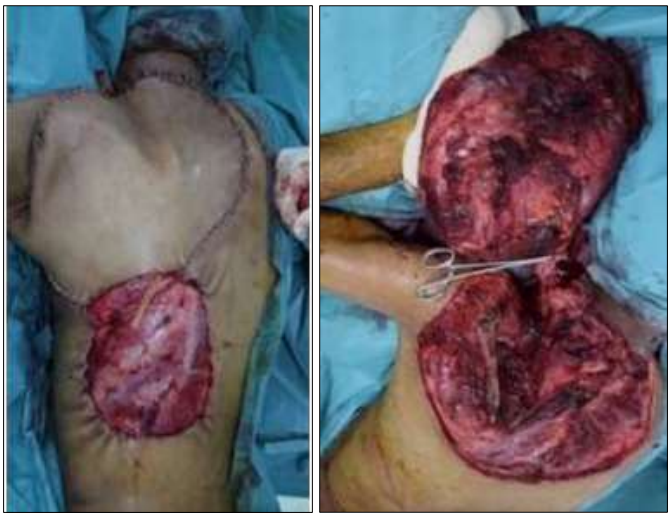

Fig. 3: Per--operative photos

The anatomopathological study came back in favor of a dermatofibrosarcoma of Darier et Ferrand with healthy limits, the closest of which is 3 cm and a non-tumoral bone tissue.

The patient was followed as an outpatient for a directed healing of the donor area, thus allowing its budding and a semi-$-thick skin graft at D 16 post-op. He completely healed and comes regularly to the consultation for control. (Fig 4)

Fig. 4: Three months post-operative

DISCUSSION

DFS is a rare mesenchymal tumor first described by Taylor in 1890 [6] and by Darier and Ferrand in 1924. It is characterized by slow infiltrative growth, local aggressiveness and high potential for local recurrence if not managed appropriately which means that we don't have to miss the first exeresis. However, distant metastasis is very rare and usually occurs as a late sequela after local recurrence [7]. DFS can occur at any age, but is most common in patients between the ages of 20 and 50 years [6]. Although it occurs primarily in adults, various case series have reported an incidence of 6--20% in children, and congenital cases have also been observed [8].

The most common anatomic site affected by DFSP is the trunk (42-72%), with the majority of cases occurring in the chest and abdomen. Between 16% and 30% of DFS cases are located in the proximal extremities (particularly the legs) and approximately 16% in the head and neck region [9, 10].

DFS usually manifests as pink, reddish-brown, or purplish spots limited to the skin. Over time, the tumors may develop into multiple «protruding» nodules that invade subcutaneous tissue, fascia, muscle, and even bone [11]. In our case, infiltration of adjacent muscle structures and features of hypervascularization, a marker of malignancy, were evident [12, 13].

The treatment of choice is wide local excision, with negative margins of 3–5 cm from the tumor edge including the skin, the subcutaneous tissue, and the underlying fascia unless other structures are affected as in our patient's case.

Mohs micrographic surgery is an alternative to wide excision that is considered by some as the preferred

treatment for DFS. It consists of a removal method that offers precise microscopic control of the entire tumor margin while maximizing the conservation of healthy tissue [14].

Our patient underwent wide excision of the tumor. Since wide excisions usually cause note worthy distortion and leave patients with serious cosmetic problems, reconstructive surgery may be required in almost every instance to restore tissue defects using a local skin flap, skin graft, or myocutaneous flap.

In cases in which primary flap reconstruction is performed simultaneously with tumor excision, there should be no doubt as to the adequacy of the excision, since the presence of a flap may prevent the detection of local recurrence. In general, the reconstructive challenge in DFS such as in the trunk, extremities, head, and neck is related to large tissue defects which need covering when vital structures are exposed. In our case, a immediately long dorsal flap was chosen due to the bone exposure.

As for adjuvant treatment, imatinibmesylate can be used to treat unresectable, recurrent, and/ or metastatic disease in patients with at [15] translocation [16].

CONCLUSION

Positioned between the harmless fibroma and the frightening sarcoma, Darier and Ferrand dermatofibrosarcoma is a rare fibrous tumor of the skin, difficult to diagnose, of very slow local onset and associated with rare metastases. It is characterized by a tendency to recur. Due to the high rate of recurrence, clinical monitoring is necessary. Its diagnostic and therapeutic problems require a reliable histological diagnosis confirmed by immunohistochemical studies. Th first surgery should be wide in lateral margins ranging from 3cm to 5cm.

Conflict of Interest: No conflict of interest

REFERENCES

1. Marks LB, Suit HD, Rosenberg AE, Wood WC. Dermatofibrosarcoma protuberans treated with radiation therapy. Int J RadiatOncolBiol Phys. 1989 Aug;17(2):379–84.
2. Morel M, Taïeb S, Penel N, Mortier L, Vanseymortier L, et al. Imaging of the most frequent superficial soft-tissue sarcomas. Skeletal Radiol. 2011;40(3):271–284.
3. Torreggiani WC, Al-Ismail K, Munk PL, Nicolaou S, et al. Dermatofibrosarcoma Protuberans MR Imaging Features. AJR. 2002;178(4):989– 993.
4. Stojadinovic A, Karpoff HM, Antonescu CR, Shah JP, et al. Dermatofibrosarcoma Protuberans of the Head and Neck. Ann Surg Oncol. 2000;7(9):696–704.
5. Rutgers EJ, Kroon BB, Albus-Lutter CE, Gortzak E. Dermatofibrosarcoma protuberans: treatment and prognosis. Eur J Surg Oncol. 1992 Jun;18(3):241–8.
6. Stamatakos M, Fyllos A, Siafogianni A, Ntzeros K, Tasiopoulou G, Rozis M, Kontzoglou K. Dermatofibrosarcoma protuberans: a rare entityandre view of the literature. J BUON. 2014;19(1):34–41.
7. Snow SN, Gordon EM, Larson PO, Bagheri MM, Bentz ML, Sable DB. Dermatofibrosarcoma protuberans: a report on 29 patients treated by Mohsmicro graphic surgery with long-term follow-up and review of the literature. Cancer. 2004;101(1):28–38.
8. Reddy C, Hayward P, Thompson P, Kan A. Dermatofibrosarcoma protuberans in children. J Plast Reconstr Aesthet Surg. 2009;62(6):819–23. 9.Taylor HB, Helwig EB. Dermatofibrosarcoma protuberans. A study of 115 cases. Cancer. 1962;15(4):717–25.
9. Serra-Guillén C, Llombart B, Sanmartín O. Dermato- fibrosarcoma protuberans. Actas Dermo-Sifiliográficas. 2012;103(9):762–77.
10. Stivala A, Lombardo GAG, Pom- pili G, Tarico MS, Fraggetta F, Perrotta RE. Dermatofibrosarcoma protuberans: our experience of 59 cases. OncolLett. 2012;4(5):1047–55.
11. Kau T, Lesnik G, Arnold G, Hausegger KA. Sono-graphy of dermatofibrosarcoma protuberans of the groin. J Clin Ultrasound. 2008;36(8):520– 2.
12. Llombart B, Serra C, Requena C, Alsina M, Morgado-Carrasco D, Través V, et al. Sarcomascutáneos: directrices para el diagnóstico y tratamiento. Dermatofibrosarcoma protuberans. Actas Dermo Sifiliográficas. 2018;109(10):868–77.
13. Ratner D, Thomas CO, Johnson TM, Sondak VK, Hamilton TA, Nelson BR, Et al. Mohs micrographic surgery for the treatment of dermatofibrosarcoma protuberans. J Am Acad Derma.