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RESEARCH ARTICLE

DARIER-FERRAND DERMATOFIBROSARCOMA OF THE FACE: TWO CASES REPORT

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Abstract

Darier-Ferrand dermatofibrosarcoma or dermatofibrosarcoma protuberans (DFSP) is a cutaneous mesenchymal tumor of intermediate malignancy. It is a rare but not exceptional tumor, accounting for 0.1% of malignant skin tumors. Histological examination is essential for diagnosis. Wide surgical excision is the standard treatment. We report two cases of Darier-Ferrand dermatofibrosarcoma of the face. A wide local excision is the gold-standard treatment, with negative margins of 3–5 cm from the tumor edge including the skin, the subcutaneous tissue, and the underlying fascia. However, this is a mutilating surgery, especially for cervicofacial lesions which is the case for our two patients For cervico-facial localizations,some authors advise the Mohs' micrographic surgery for its low recurrence rates described. Target adjuvant therapy with imatinib has been shown to reduce tumor size and improve surgical resectability and can be proposed as an option for our patients. However, its unavailability and high-cost limit its use in our context. A combination of conservative excision and adjuvant radiotherapy was reported to reduce the rate of local recurrence and should be also considered by practicians. To improve prognosis, early, codified, multidisciplinary management is essential for this pathology.

Introduction:-

Darier-Ferrand dermatofibrosarcoma or dermatofibrosarcoma protuberans (DFSP) is arare but not exceptional slowly growing cutaneous mesenchymal tumor of intermediate malignancy. It accounts for 0.1% of malignant skin tumors. It is markedby its local aggressiveness and is prone to a high riskof local recurrences. Metastases are rare and generally preceded by multiple local recurrences.

The most common locations in DFS are usually the trunk or proximal extremity but can still occur anywhere in the dermis.Complete surgical resection is considered the gold-standard therapy. The potential for recurrence of DFS is directly related to the margin of resection.

However, some cases may be inaccessible to surgery due to their location, size, foreseeable functional and/or aesthetic consequences post-surgery and a medical treatment might be considered, a treatment currently based on the molecular and cytogenetic characteristics of DFSP. It has recentlybeen shown that inhibiting PDGFR with imatinib can induce high response rates in case of unresectable or metastatic disease.

Herein, we report two cases of dermatofibrosarcoma on the face.

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Cases Report

The first case is about a 37-year-old woman, with no medical past history, who presented a mass that has been present for a year on the left cheek and increasing in volume.

The tumor was painless, and mobile in relation to the deep plane. No associated adenopathy was found.

An MRI of the face was asked but due to lack of funds, the patient went ahead with a facial scan that showed a 2.3cm long, circumscribed, well-limited, and homogeneous tissue mass.

The patient benefited from an excisional biopsy, which revealed at the anatomopathological examination, a Darier-Ferrand dermatofibrosarcoma withinsufficient resection margins. The immunohistochemical stain for CD34 was positive.However, the patient refused to proceed with a second surgery for wide excision in order to prevent recurrence.

The second case is a 53-year-old man, asmoker, who presented a mass that has been present for 8 months, located on the nasolabial fold and increasing in volume. The mass itself was 3 cm long but there was a wide induration around the mass that had no exact limit. A biopsy conducted revealed a Darier-Ferrand dermatofibrosarcoma and the immunohistochemical stain for CD34 was positive. The facial scan also showed a homogeneous tissue mass.

Discussion:-

DFSP is a rare mesenchymal tumor, representing 1.8% of all soft tissue sarcomas and only 0.1% of all cancers.

The first person todescribe this tumor was Taylor in 1890 and then Darier and Ferrand in 1924.

It is characterized by slow infiltrative growth, a local aggressiveness, a high potential for local recurrence if not properly treated, and extremely rare distant metastases.

Dermatofibrosarcoma can occur at any age, but is most common inadults, between the ages of 20 and 50, with a slight male predominance.

DFSP can occur in any part of the body, with a predominance of the trunk and extremities. Cervical localization is rare.

A wide local excision is the gold-standard treatment, with negative margins of 3–5 cm from the tumor edge including the skin, the subcutaneous tissue, and the underlying fascia. However, this is a mutilating surgery, especially for cervicofacial lesions which is the case for our two patients; a wide excision can leave them with serious aesthetic consequences. Reconstructive surgery may be required in almost every instance to restore tissue defects using a local skin flap, skin graft, or myocutaneous flap.

To make this surgery less disfiguring, while still being carcinological, some authors have achieved 3 cm and fewer margins of exeresis with the Mohs' micrographic surgery, where proof of complete excision of the tumor is confirmed. This technique is suitable for cervico-facial localizations, but obtaining such a technical platform and experienced personnel is far from being within everyone's reach.

Some authors like R. Behbahaniand Al described good results with this technique following the literaturewith low recurrences although the study of Smola et al showed a recurrence of 1.2% at 21 (6 – 36) months for lateral margins of 1 – 3 cm.

Similar recurrence rates were found by McPeak in 82 patients, and a series of 119 patients treated by wide excision. The head and neck are more common sites for recurrence, and it is unclear whether this reflects the difficulties in excising DFSP over this area.

As for adjuvant treatment, imatinib mesylate can be used for DFSP.

Dermatofibrosarcoma protuberans is characterized by chromosomal rearrangements resulting in the production of platelet-derived growth factor B, eventually leading to autocrine growth stimulation of dermatofibrosarcoma

protuberans. Imatinib functions as an inhibitor of platelet-derived growth factor receptors, thus blocking this autocrine stimulation. Therefore, imatinib might perhaps eventually be used as an adjuvant therapy in cases in which obtaining sufficient surgical margins is impossible.

Han and Al reported a neoadjuvant treatment with imatinib that significantly reduced the preoperative lesion size for four patients and the smallest pretreatment lesion in that case series was a 3-cm, freely movable, exophytic nodule on the medial aspect of the ankle, resulting in a less extensive surgical defect, and that allowed placement of a full-thickness skin graft over a wound that extended to the periosteum. So Imatinib appears to be a safe oral medication with the ability to significantly shrink the lesions of DFSP.

Imatinib has been reported to be efficacious only in tumors exhibiting chromosomal translocation (17;22), which occurs in 90% of all DFSP lesions.

Target adjuvant therapy with imatinib has been shown to reduce tumor size and improve surgical resectability and can be proposed as an option for our patients. However, its unavailability and high-cost limit its use in our context.

In cases of positive or inadequate margins, recurrence, or unacceptable functional or cosmetic results after wide excision, a combination of conservative excision and adjuvant radiotherapy was reported to reduce the rate of local recurrence by 5%.

Dermatofibrosarcoma protuberans is a radioresponsive disease with excellent local control after conservative surgery in combination with radiation therapy. However, radiation therapy should be considered for patients with large (>5 cm) or recurrent tumors. Radiation therapy should also be considered for patients when complete resection with negative margins is not possible or when repeated attempts at wide surgical margins would result in significant morbidity. Radiation therapy can be delivered either preoperatively or postoperatively. However, there is a high risk of malignant transformation that can occur with radiotherapy.

Conclusion:-

Dermatofibrosarcoma protuberans is a rare tumor soft tissue tumor, that can appear at any age; in many locations especially the trunk and rarely the face.

Wide surgical excisions are the best treatment with 5cm margins however the cervicofacial location should be considered before making that decision as it can lead to a mutilating result. The Mohs' micrographic surgery proved to be efficient by authors but an adjuvant treatment by target therapy as Imatinib must be also considered regarding its results.

A combination of conservative excision and adjuvant radiotherapy was reported to reduce the local recurrence rate and should also be considered by practicians.

To improve prognosis, early, codified, multidisciplinary management is essential for this pathology.

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