Dermatomyofibroma*

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Abstract: We report a case of dermatomyofibroma that, to our knowledge, is the second case reported in Brazil. About 100 cases have been reported worldwide. Dermatomyofibroma represents a rare, benign mesenchymal neoplasm of fibroblastic/myofibroblastic differentiation, with prolonged evolution and little or no symptoms. It most commonly occurs in young women and male children. Dermatomyofibroma can be easily confused with other clinical entities, which could lead to unnecessary treatments. Therefore, it is important that dermatologists and pediatricians suspect and start to consider this hypothesis in their diagnostic exercises.

Keywords: Immunohistochemistry; Myofibroblasts; Skin cancer

INTRODUCTION

Dermatomyofibroma (DMF) is a rare benign cutaneous neoplasia of fibroblasts and myofibroblasts first described by Hügel in 1991 and later by Kamino *et al.* in 1992, who proposed the current term.[1-3] The lesions typically arise on the shoulder of young women as an ill-defined asymptomatic fibroelastic plaque or single nodule, more palpable than visible, varying in color from normochromic to brown erythematous.[1,3-5] It is most often oval with 1cm-2cm in diameter. However, some cases with 13cm x 15cm and multiple DMFs have been reported.[5,6] Lesions are benign, but they are often confused with malignancies, leading to unnecessary treatments. Diagnosis is confirmed histopathologically. Immunohistochemistry may be useful to distinguish in doubtful cases: in most cases, it is positive for vimentin and smooth muscle actin, and negative for S-100, desmin, and other markers.[5] Clinicians may opt for a conservative excision or clinical follow-up.[7]

CASE REPORT

A 31-year-old white female patient sought treatment reporting an asymptomatic erythematous macula for the last six years. The lesion of a few millimeters in size was located on the posterior region of the right shoulder, with no history of local trauma. In the last year, it became more erythematous, a little harder, and increased in size until it became stable. We observed an oval brown erythematous cutaneous nodular lesion. The surface was flat with a fibroelastic consistency and 2 cm in diameter (Figure 1).

Diagnostic hypotheses included: dermatofibrosarcoma protuberans (DFSP) in the plaque stage; scar; keloid; dermatofibroma; tumor of the skin appendages; and granuloma annulare.

Histopathology showed no epidermal changes. Dermis and hypodermis revealed spindle cell proliferation without atypia, exhibiting wavy nuclei, pink cytoplasm, non-defined cell limits, and parallel orientation to the epidermis into the dermis and perpendicular to the hypodermis. The elastic fibers showed no changes in silver impregnation (Figures 2 and 3).

Immunohistochemistry was positive for vimentin, smooth muscle actin, factor XIIIa, and Ki-67 (less than 5% of the cells), and negative for desmin, CD34, and S-100 protein (Table 1).

Based on histological findings and corroborated by immunohistochemical analysis, we diagnosed DMF.

The patient was reevaluated fifteen months after the last visit and a full term pregnancy without complications. Clinically, the lesions had become more pigmented with apparent reduction in consistency, suggesting a regression of DMF. However, a new incisional biopsy demonstrated the persistence of DMF.

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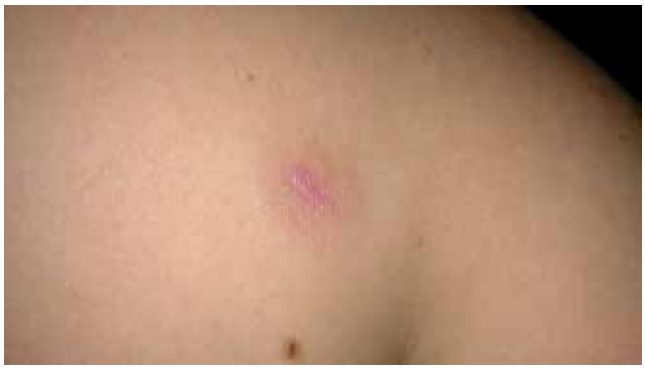

Figure 1: Brown erythematous macule, about 3 cm in size, asymptomatic, and biopsy scar on the posterior region of the right shoulder

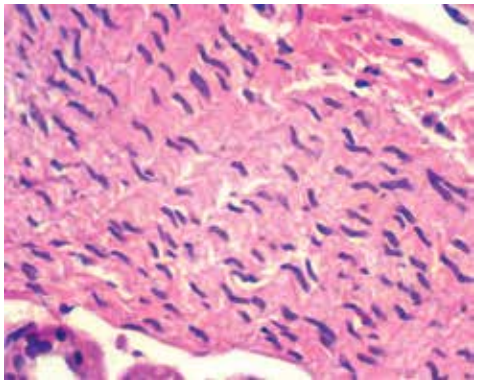

Figure 2: Fusiform cells without atypia, with wavy nuclei; pink cytoplasm, non-defined cell limits boundaries (HE, high magnification)

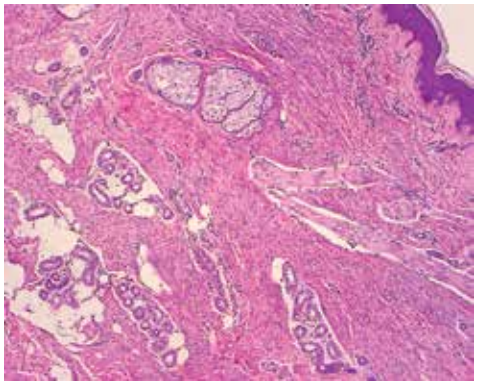

Figure 3: Fusocellular proliferation through the skin appendages, without destroying them or changing their morphology (HE, low magnification)

Table 1: Results of immunohistochemical markers

Positive reaction	Negative reaction
Vimentin	Desmin
Smooth muscle actin	CD34
Factor XIIIa	S-100 protein
Ki-67 (less than 5% of the cells)	

DISCUSSION

In 1991, Hugel reported the disease that he called plaque-like dermal fibromatosis in 25 patients.[2] In 1992, Kamino *et al.* reported 9 cases of DMF with a higher prevalence in young women, located mostly in the shoulder and axilla regions, but with wide anatomical distribution.[3] Other locations mentioned were neck, chest, inguinocrural region, thigh, and forearm.[1,3,8]

The average age of affected women was 31.4 years, and armpit and shoulder involvement was more common. In males, the average age was 12.3 years, and the most affected site was the neck.[7,9]

DMF is rare in pediatric patients and, when it occurs, affects mostly boys.[7] The reasons for the female predilection in young adults, and for the male predilection in children, have not yet been fully established. Studies demonstrate spontaneous regression of DMF in boys after infancy, while in girls it continues to evolve to stabilize after puberty, possibly because of female hormones.[9] If androgenic hormones indeed influenced the regression of DMF in boys, one would expect it not to occur in women with hyperandrogenism, an absent condition in our case. In this sense, both the androgenic hormonal profile and immunohistochemistry for estrogen receptors and androgen could be useful to verify a possible correlation with clinical data. That would help clarify the fact that DMF affects predominantly male children and young women. However, Tardío *et al.* found no positivity for estrogen and progesterone receptors in their 12 pediatric cases.[7]

DMF belongs to a heterogeneous group of benign, intermediate, and malignant cutaneous mesenchymal tumors, which shows fibroblastic/myofibroblastic differentiation, mainly composed of spindle cells.

Clinically, it is mistaken for dermatofibroma, leiomyoma, pilomatrixoma, tumor of the skin appendages, nevus sebaceous, several cysts, granuloma annulare, atrophodermia, and, most importantly, DFSP, for which histology and immunohistochemistry are useful differential diagnosis.[8]

Histologically, it reveals poorly defined proliferation of fibroblasts and myofibroblasts in the reticular dermis and in the subcutaneous tissue surface[8] sparing the papillary dermis, the most superficial dermis, and associated structures.[6,9] It also shows the typical fascicles of spindle cells with elongated nuclei containing one or two nucleoli,[6] and poorly delimited eosinophilic cytoplasm. In the reticular dermis, they are arranged parallel to the epidermis; in the superficial subcutaneous tissue, sometimes at perpendicular arrangement. Mitotic figures,[6] cytologic atypia, or inflammatory infiltrate are rarely seen. Fine collagen fibers are seen between fusiform cells.[9] Thick and fragmented elastic fibers are also very characteristic, which rules out hypertrophic scar in which there is loss of elastic fibers.[9,10] Contrary to the literature, our case showed no alterations of elastic fibers. On the other hand, the second histopathological examination revealed a degree of collagen loss, which would explain the subsequent partial clinical regression of the lesion.

Immunohistochemistry helps with the non-characteristic histological pictures. Fusiform cells were positive for calponin, actin smooth muscle, and vimentin, an intermediate filament of myogenesis.[5-7,9] No reactivity for S-100 protein, desmin, and CD34 was observed, ruling out several differential diagnoses.[9,10]

Unlike two other reports, in the present case, immunohistochemistry was negative for factor XIIIa and smooth muscle actin.[5,6]

DMF treatment consists of simple excision. No malignancy, recurrence, or metastasis have been reported.[1,2] When surgical excision may result in cosmetic damage, as in cases of DMF in childhood, clinical follow-up would be acceptable, with no need to remove the lesion.[7]

The prognosis is usually good. On the other hand, the diagnosis of DMF requires adequate clinical and histological evaluation, since it may be mistaken for aggressive neoplasias. Despite the almost exclusive prevalence in young women and male children, the possibility of congenital DMF should not be ruled out.[7] ❑

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