Dermatomyofibroma: a case report of a rare disease

Dermatomiofibroma: relato de caso de doença rara

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Abstract: Dermato myofibroma is included in the group of benign cutaneous mesenchymal neoplastic lesions of fibroblastic and myofibroblastic lineage. It's a rare disease and there are approximately only one hundred cases described worldwide in the medical literature up to now. The present study reports the case of a young woman with typical clinical cutaneous lesion and histopathological diagnosis of dermato myofibroma. Special stains were carried out which showed preserved collagen fibers and immunohistochemistry was positive for vimentin and negative for actin and S100. As it is a rare disease, the histopathological findings are of great importance but clinical suspicion is possible in typical cases such as this one.
Keywords: Fibroblasts; Myofibroma; Skin neoplasms

Resumo: O Dermatomiofibroma está incluído no grupo de lesões neoplásicas mesenquimais benignas de linhagem fibroblástica e miofibroblástica da pele. É uma doença rara, havendo aproximadamente 100 casos descritos na literatura mundial até o momento. Este artigo relata o caso de uma mulher jovem com apresentação clínica típica e diagnóstico histopatológico de dermatomiofibroma. Foram realizadas colorações especiais que mostraram preservação das fibras colágenas e a imunohistoquímica revelou positividade para vimentina e negatividade para actina e S100. Por se tratar de doença rara, os achados histopatológicos são de grande importância, mas a supeição clínica é possível em casos típicos como este.
Palavras-chave: Fibroblastos; Miofibroma; Neoplasias cutâneas

INTRODUCTION

Dermatomyofibroma was described by Kamino[1] in 1992, in a series of 9 cases as a clinical pathological entity. However, there were previous reports of 25 similar cases observed by Hügel[2] in 1991, in the German medical literature and denominated by him, in a non specific way as 'die plaqueformige dermale fibromatose.[2,3] It represents a rare and distinct benign mesenchymal cutaneous neoplasia of fibroblastic and myofibroblastic differentiation.[4] Its neoplastic etiology is not yet completely established but recent clinical studies with electronic microscopy [5,6] and immunohistochemistry strongly suggest this possibility. Clinically it is predominant in young women, with small plaque or nodule that can vary from erythematous bronwish to normochromic, lonely, asymptomatic more palpable than visible, of fibro- elastic consistency and normally located on the shoulder,[2,3,7] resembling a keloid. Clinical suspicion is difficult and diagnosis can only be made by histopathologic analysis. Immunohistochemistry reveals the nature of lesions showing positivity for vimentin and actin in most cases and negativity for S100 and desmin, among others. Treatment is surgical. The. excision of the tumor promotes complete cure and there are no reports in

Approved by the Editorial Board and accepted for publication on 25.11.2010.
* Work carried out at the Pedro Ernesto University Hospital (HUPE – UERJ)- Rio de Janeiro(RJ), Brazil
Conflict of interest: None / *Conflito de interesse: Nenhum*
Financial funding: None / *Suporte financeiro: Nenhum*

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the medical literature of recurrence or metastases[1-4,7-10]. There are approximately 100 cases published up to now.

CASE REPORT

Eighteen-year old female patient, Caucasian, student, single and from Rio de Janeiro. The patient reported story of asymptomatic lesion, on the left shoulder for 2 years without improvement with the use of corticosteroids. Dermatological examination showed plaque slightly erythematous, with 1,5 cm at its largest diameter and located on the anterior area of the left shoulder (Picture 1- clinical lesion). Palpation allowed to notice nodule of approximately 2 cm, of elastic consistency, painless and movable. Physical examination and laboratory parameters were within normal limits. The diagnostic hypotheses were granuloma annulare and tuberculoid leprosy as it was noticed hypoesthesia in a small area at the edge of the lesion. It was carried out incisional biopsy that showed, in the reticular dermis, monomorphic fusiform cells parallel to the skin surface that was preserved as well as the papillary dermis and the annexed structures (Pictures 2 and 3). Masson's trichrome staining was positive for collagen fibers (Pictures 4 and 5). The diagnosis made was dermatomyofibroma. Immunohistochemistry was positive for vimentin and negative for S100 and actin. The proposed treatment was surgical excision. There has not been recurrence of the lesion up to now and the patient is in attendance.

DISCUSSION

In 1991 Hügel[1] described for the first time a skin alteration that he called " plaque type dermal fibromatosis" in a series of 25 patients. One year later Kamino et al.[2] reported 9 similar cases and determined a pathological entity with distinct and peculiar characteristics naming it dermatomyofibroma. In Kamino's initial series the average age was 29,8 and 8 out of 9 cases were located on the shoulders[1], as it was in our patient. Other locations that were also described were posterior cervical region, armpits, lower limbs, chest and abdomen. It affects mainly young women[4], as the case reported here but there are reports of the disease in men and children.[3,4,8,10] Clinically, dermatomyofibromas are nodules or asymptomatic plaques, well circumscribed, of oval or anular shape, erythematous bronwish to normochromic, lonely, with 1-2 cm of diameter and fibro-

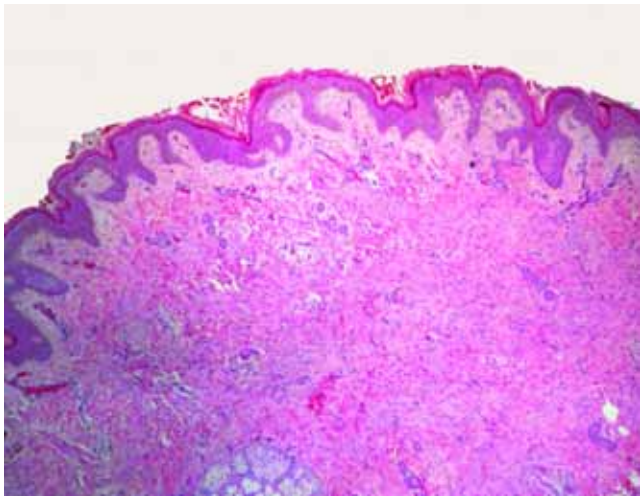

Figura 2: Staining by HE&;shows in the reticular dermis bundles of monomorphic fusiform cells parallel to the skin surface that is preserved as well as the papillary dermis and the adnexal structures

Figure 1: Erythematous plaque with 1,5 cm at its largest diameter located on the anterior area of the right shoulder

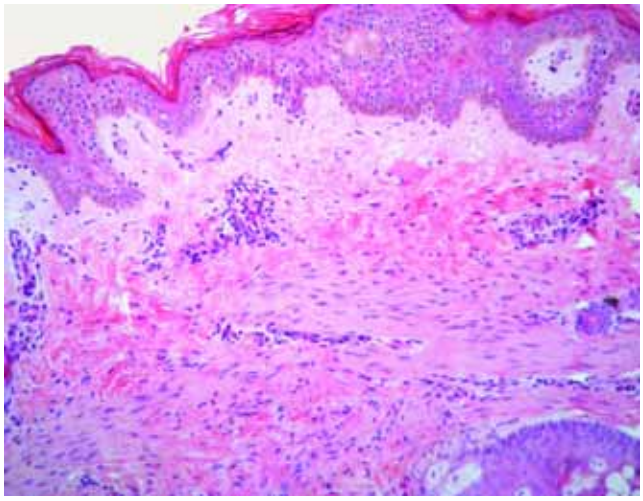

Figure 3: Staining by HE showing, at its biggest enlargement, the bundle of fusiform cells of the tumor parallel to the skin .surface

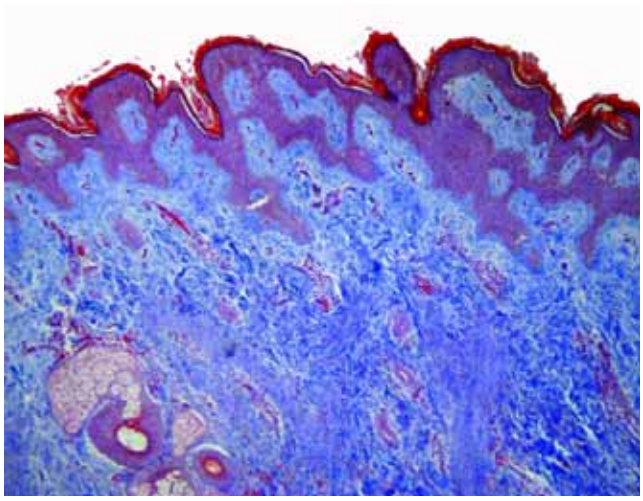

Figure 4: Staining by Masson's trichrome showing preservation of bundles of collagen and of das adnexal structures

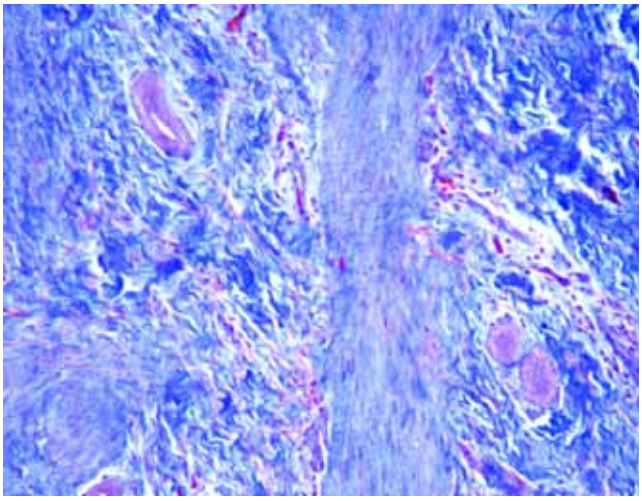

Figure 5: Staining by Masson's trichrome, biggest enlargement

elastic consistency. Multiple[10] or extensive [11,12] lesions have already been reported. Histologically[1], the lesion is located in the reticular dermis, and for this reason is more palpable than visible. It adopts a plaque morphology with extension superficial to the cutaneous tissue. It is constituted by proliferation of monomorphic fusiform cells, forming elongated and interlocking fascicles which adopt a disposition parallel to the skin surface. It is observed a preserved dermal segment above the lesion and the cells are separated by tick collagen fibers. The fascicles described are found around the annexes, without causing invasion or obstruction. The elastic fibers are also preserved. The fusiform cells present enlarged nuclei, with one or two nucleoli, showing scarse atypia and little or almost none mitotic activity. Masson's trichrome staining is useful as it stains the collagen fibers. Van Gieson's staining for elastic fibers reveals preservation of them among the fascicles of the lesion. Immunohistochemistry confirms its nature, showing positivity for vimentin and actin in most cases and negativity or slightly positive for smooth muscle actin-specific. They are negative for desmin, factor XIIIa, CD4, ALK1, S-100. As for the origin of this entity the majority of the authors agrees with its neoplastic nature instead of reactive, based primarily in the absence of story of trauma or surgery, histological alterations of recent or old bleeding, necrosis or other degenerative diseases as well as for electron microscopic findings.[5,6] Differential diagnosis should be made with other cutaneous myofibroblastic lesions such as dermatofibroma, leiomyoma pillar, neurofibroma, cutaneous myofibromas, infancy fibrous hamartoma, extra-abdominal fibromatosis and due to its special prognostic implications with the dermatofibrosarcoma protuberans[9] in plaque. Treatment is simple excision of the lesion which promotes full cure. There are neither reports of cases that became malignant nor reports of metastases or local recurrence.[1-4,7-10] Our case adds to the medical literature one more case of a rare benign neoplasia and highlights the fisibility of clinical suspicion due to the typical characteristics of our patient such as sex, affected area, lesion consistency. Palpation in the dermatologic examination is extremely important for the diagnosis of this condition. ❑

ACKNOWLEDGEMENTS

The authors acknowledge the invaluable help from the Instituto Nacional do Câncer (INCA) by the gentle partnership in carrying out the immunohistochemistry technique.

ERRATUM

The last name of the author was published wrong in one issue of 2011. The correct name of the author is Carolina Cotta Zimmermann and the abbreviation is Zimmermann CC.

The article that should be corrected is:

Macedo PM, Mann D, Zimmerman CC, Alves Mde F, Daxbacher EL. Dermatomyofibroma: a case report of a rare disease. An Bras Dermatol. 2011;86(1):120-3.

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How to cite this article/*Como citar este artigo*: Marques PM, Mann D, Zimmermann CC, Alves MFGS, Daxbacher ELR. Dermatomyofibroma: a case report of a rare disease. An Bras Dermatol. 2011;86(1):120-4.