

WHAT IS THE DIAGNOSIS

Annular plaque on the back

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CLINICAL PRESENTATION

Female patient, 35 years old, with no relevant history, consulted for presenting clinical picture of 1 year of evolution, characterized by annular plaque with hyperpigmented border and pale center, approximately 1 cm in diameter, located in the right scapular area (Fig. 1 a and b). The lesion is asymptomatic and does not refer to previous treatments. At dermoscopy, a brown border with a lighter center and marked follicles were observed.



Fig. 1a. Clinical image, annular plaque, hyperpigmented border.



Fig. 1b. Dermoscopic image: brown border with lighter center and prominent follicles.

WHAT IS THE DIAGNOSIS?

A skin biopsy was performed and sent for histopathological study which revealed a tumor consisting of a “plaque” proliferation of fascicles of spindle cells arranged parallel to the epidermis and located in the middle and deep dermis. The nuclei are elongated, vesicular, with fine chromatin. They are arranged between collagen fibers. The adnexa are entrapped but not destroyed by the tumor. S100 immunohistochemistry is negative (Figure 2 a and b).

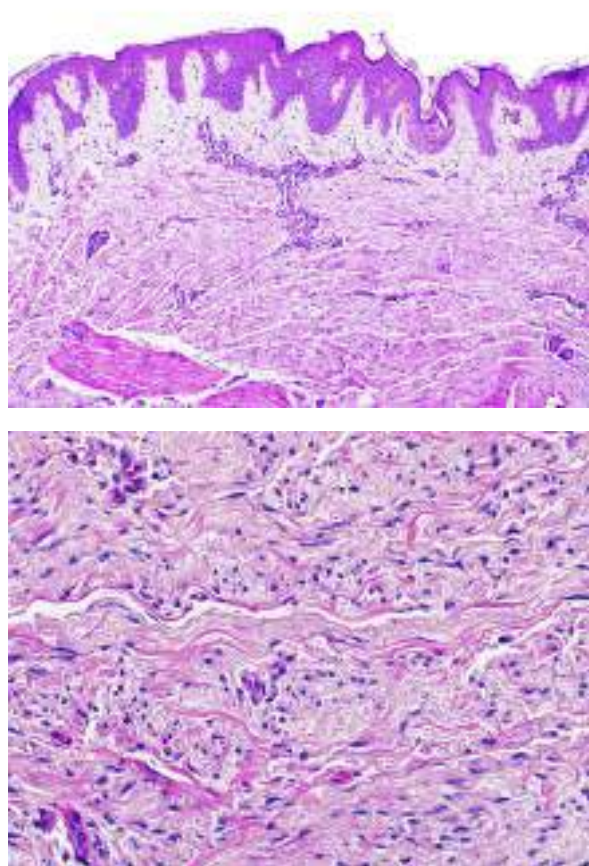


Fig 2a and 2b. Tumor consisting of a “plaque” proliferation of fascicles of spindle cells arranged parallel to the epidermis and located in the middle and deep dermis.

With the clinical findings and the pathology result we arrived at the diagnosis of **Dermatomyofibroma**.

COMMENTARY

Dermatomyofibroma (DMF) is a benign and acquired cutaneous tumor of infrequent presentation, which is usually underdiagnosed because it is little known.

It was first described in 1991 by Hügel, who named it “plaque dermal fibromatosis” when he published a series of 25 cases. However, a year later, Kamino et al. described 9 cases with their clinical and histological characteristics, proposing the name “Dermatomyofibroma.”¹ In 2009, Mentzel et al. reported a series of 56 cases with ages ranging from 3 to 51 years, and a predominance of females.²

This lesion has a mesenchymal origin, formed by a proliferation of spindle cells corresponding to fibroblasts and myofibroblasts as demonstrated by structural and immunohistochemical studies. It is frequently observed in young adults with predominance in the female sex (about 90%), although there are cases reported in children, in them, the predominance is male.² The reason why FMD is more common in females is unknown; however, it is hypothesized that this lesion continues to grow in women after childhood due to hormonal effects, while in men it would regress spontaneously.

In adults it is preferentially located in or around the shoulder, axilla, upper trunk, neck and arm,³ while in children, the most frequent location is the cervical region.² Clinically, it presents as an oval or annular nodule or plaque, well demarcated, somewhat indurated, with a smooth surface, 1 to 10 cm in diameter, brownish red in color, without ulceration or desquamation, asymptomatic, with slow growth. It is usually a single lesion, although there are cases of multiple presentations or satellite lesions around the DMF.¹

Dermoscopy shows a lesion similar to that seen in lentigo-type dermatofibroma,⁴ composed of a reticular pigimentary network giving rise to a regular mesh with the presence of fine lines and the presence of small multifocal hypopigmentations can be observed.

Histologically, there is a uniform, dense, well-demarcated proliferation formed by monomorphous spindle cells in fascicles or bands that are arranged parallel to the epidermis and can reach the deep dermis and hypodermis and correspond to fibroblasts and myofibroblasts. There is no atypia or mitosis. These cells are located around the adnexal structures and elastic fibers without destroying or altering them.⁴ Immunohistochemically, calponin is positive and is the most constant marker. Other muscle markers such as caldesmin and desmin are negative. Meanwhile, smooth muscle actin is inconsistent.⁵

Among the differential diagnoses are dermatofibroma, desmoid tumor, piloleiomyoma and dermatofibrosarcoma protuberans, in pediatric age should also be considered fibrous hamartoma of infancy, connective tissue nevus and smooth muscle hamartoma.^{2,6}

The treatment is surgical excision, which if performed with an adequate margin is not associated with recurrence.⁶

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