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PHOTOLETTER TO THE EDITOR

Atrophic dermatofibrosarcoma protuberans with minimal clinical manifestation

Hugo Marques Barreiros[1], *Pedro Nuno Serrano*[1], *José Carlos Parreira*[2], *Elvira Bártolo*[1]

1. Department of Dermatology, Hospital Garcia de Orta, Almada, Portugal;
2. Department of Plastic Surgery, Hospital Garcia de Orta, Almada, Portugal.

Corresponding author:
Hugo Barreiros, Department of Dermatology, Hospital Garcia de Orta, Avenida Torrado da Silva, Pragal, 2801-951 Almada, Portugal. E-mail: *hbarreiros@gmail.com*

Abstract

Dermatofibrosarcoma protuberans is an uncommon soft tissue neoplasm. In the vast majority of cases it presents as a nodule or a firm tumor that can reach massive dimensions producing the protuberant nodules for which it is named.

We report a case of a 34-year-old woman presented at our department with an 8-year history of a small and discretely erythematous supraclavicular atrophic plaque. Skin biopsy lead to the diagnosis of dermatofibrosarcoma protuberans and a wide local excision of the tumor was performed in collaboration with the Plastic Surgery department.

In this clinical case we describe an uncommon variant of the disease with minimal clinical manifestation that can cause serious diagnostic difficulties. The small and discrete atrophic plaque of our patient could have been easily ignored with serious clinical and prognostic implications for the patient. (*J Dermatol Case Rep*. 2013; 7(1): 27-28)

Key words:

dermatofibrosarcoma protuberans, sarcoma, carcinoma

Dermatofibrosarcoma protuberans (DFSP) is a relatively uncommon soft tissue neoplasm with an incidence of 0.8—4.5 cases in a million per year.[1] In 1924, Darier and Ferrand[2] recognized DFSP as a clinical entity, but its characteristic microscopic pattern — fibroblastic proliferation of tumor cells arranged in a storiform or cartwheel pattern — was first described by Taylor and Helwig in 1962. DFSP is locally aggressive through peripheral tumor extensions, slow infiltrative and tends to recur after conservative initial surgical intervention. Metastization is rare (below 5%), and when it occurs generally follows several attempts of reexcision of tumor recurrences.

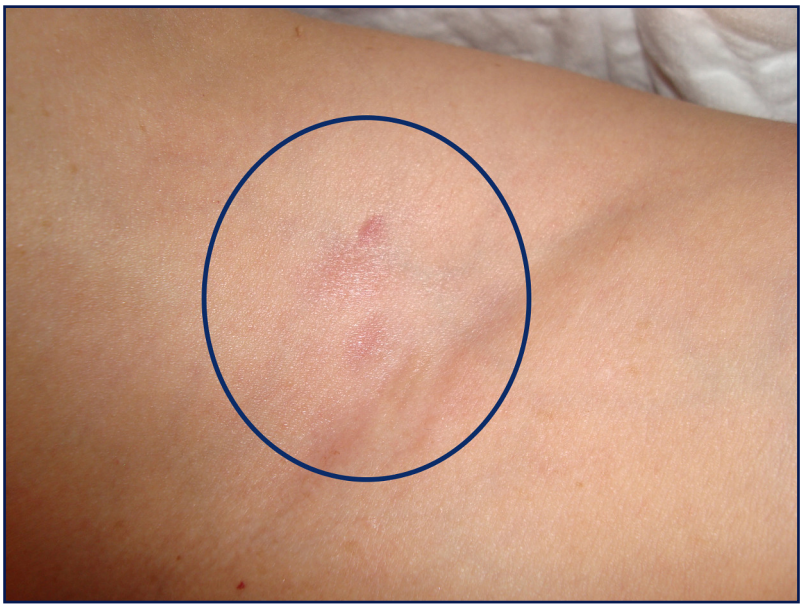

Figure 1
Small erythematous left supraclavicular atrophic plaque.

A 34-year-old woman presented at our department with an 8-year history of a small and discretely erythematous supraclavicular atrophic plaque (Fig. 1). Diagnostic impressions lead to clinical diagnosis of morphea or granuloma annulare. Skin biopsy showed a tumor composed by monomorphous spindle cells occupying all the dermis and invading the hypodermis. Immunohistochemical staining was positive to vimentin and CD34, negative to CD68, XIIIa factor and S100 (Fig. 2). Diagnose was established: dermatofibrosarcoma protuberans. Laboratory tests that assess disease extension were performed. Computed tomographies of the head, chest and abdomen were normal. Wide local excision (margins of 5 cm) was performed in collaboration with the Plastic Surgery department. The 12 month follow-up showed no recurrence.

Martin *et al*[3] distinguished three clinical forms of non-protruding DFSP: morphea-like (childhood), atrophoderma-like (congenital) and angioma-like (uncommon). As suggested by Bakry *et al*[4] such atrophic lesions can be clinically hard to diagnose as they resemble benign lesions such as morphea, idiopathic atrophoderma, atrophic scar, anetoderma or lipoatrophy.

Dermatofibrosarcoma protuberans in the vast majority of cases presents as a large plaque with multiple nodules on its surface. It can reach massive dimensions producing the protuberant nodules for which it is named. In this clinical case we describe a variant of the disease that can cause serious diagnostic difficulties. The small and discrete atrophic plaque of our patient (Fig. 1) could have been easily ignored. It was the long standing history of the lesion that was the key for performing the biopsy that lead to the diagnosis.

Trauma is often considered the eliciting factor for the growth of the tumor. Chromosomes 17 and 22 have been incriminated in the pathogenesis of DFSP with translocation t (17:22) involving COL1A1 (collagen type 1α 1 gene) and platelet-derived growth factor β (PDGF β) genes.[1] Wide surgical excision with a safety margin of 3-5 cm including the underlying fascia remains the mainstay of treatment. Advantages of Mohs micrographic surgery include the effectiveness and tissue conservation. Mohs micrographic surgery is increasingly being accepted as the treatment of choice for DFSP. Radiation therapy and imatinib mesylate are alternative treatments for unresectable tumors or additional adjuncts.[5]

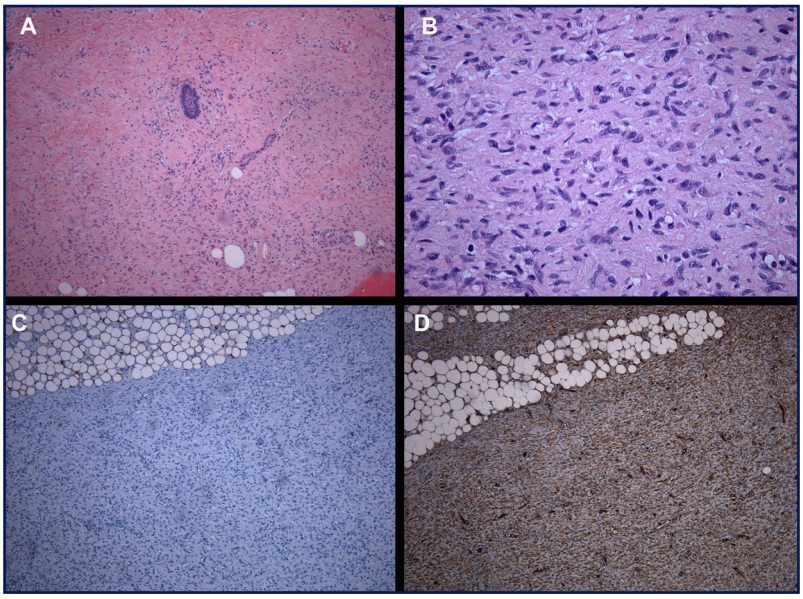

Figure 2

A photomicrograph showing (A) (H&E x100), tumor occupying all dermis and invading the hypodermis (B) (H&E x400), spindle cell tumor (C) (S100 x100), S100 is negative (D) (CD34 x100) tumor cells show positivity for CD34.

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