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REVIEW

Dermatofibrosarcoma Protuberans☆

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Abstract Dermatofibrosarcoma protuberans is the most common skin sarcoma, although its incidence is very low compared with other skin tumors. It presents as a slow-growing indurated plaque on which nodules develop over time. The lesion arises in the dermis but can invade subcutaneous tissue, fascia, muscle and even bone. *COL1A1-PDGFB* translocation is specific to dermatofibrosarcoma protuberans, and the presence of this fusion contributes to diagnosis in certain cases. A review of the literature provides evidence that recurrence is much lower after Mohs micorgraphic surgery than after conventional wide local excision. In the case of metastatic disease or when surgery would be mutilating, another recently approved treatment is the tyrosine kinase inhibitor imatinib.
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PALABRAS CLAVE
Dermatofibrosarcoma protuberans;
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Dermatofibrosarcoma protuberans

Resumen El dermatofibrosarcoma protuberans es el sarcoma de piel más frecuente aunque su incidencia es muy baja comparada con otros tumores cutáneos. Se presenta clínicamente en forma de placa indurada de crecimiento lento sobre la que aparecen nódulos a medida que el tumor progresa. Se localiza inicialmente en la dermis desde donde infiltra el tejido celular subcutáneo, la fascia, el músculo e incluso el hueso. La translocación COL1A1-PDGFB es específica del dermatofibrosarcoma protuberans y sirve de ayuda en el diagnóstico de determinados casos. Según la revisión de las series publicadas en la literatura, el porcentaje de recidivas con cirugía micrográfica de Mohs es mucho menor que el encontrado cuando se emplea cirugía convencional con márgenes amplios. Para casos metastásicos o en aquellos donde la cirugía pueda ser mutilante se dispone recientemente del imatinib, fármaco de la familia de los inhibitores de la tirosina quinasa.
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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a cutaneous tumor representative of the advances in diagnosis and treatment in oncology gained through an understanding of molecular biology. Certain cases can be diagnosed through the presence of a specific translocation and there is a promising protein tyrosine kinase inhibitor that has opened up the possibility of treatment of advanced disease.

DFSP was first described in 1890 by Taylor[1] as a sarcoma resembling a keloid. Darier and Ferrand[2] were the first to recognize DFSP as a unique entity in 1924. One year later, Hoffman[3] coined the terms *Darier-Ferrand tumor* or dermatofibrosarcoma protuberans in reference to the particular tendency of this tumor to form protruding nodules on the skin.

DFSP is currently defined as a slow-growing infiltrative skin tumor with a high rate of local recurrence but low metastatic capacity. According to the World Health Organization,[4] DFSP is classified as a fibrous, fibrohistiocytic, or histiocytic tumor. Weiss and Goldblum,[5] in the book *Enzinger and Weiss's Soft Tissue Tumors*, considered it as a fibrohistiocytic tumor of intermediate malignancy.

Although DFPS has traditionally been classified as a fibrohistiocytic tumor, the histogenesis of DFSP remains uncertain. According to different studies, the origin of DFSP may be fibroblastic,[6,7] histiocytic,[6,8] or neural.[9,10] $CD34^+$ dermal dendrocytes have been proposed as another possible origin.[11,12] However, many of these studies have contradictory results and none clearly demonstrate which type of cell DFSP is derived from.

The expression in DFSP of nestin, an intermediate filament expressed on neuroectodermal stem cells, suggests that DFSP originates from pluripotent neuromesenchymal stem cells.[13–15] This hypothesis, which considers the origin of certain tumors to be a mutated pluripotent stem cell, is currently the most widely accepted for DFSP. This type of nestin-positive mesenchymal stem cell is found in the hair follicle.[14]

As for most sarcomas, DFSP does not have a well-established risk factor and its etiology is unknown. In 1951, Pack and Tabah[16] suggested that local trauma in the region of the tumor was an etiologic factor on the basis that such an event was reported by 13% of the patients in their series. Subsequently, Taylor and Helwig[17] found that local trauma had occurred in 19 out of a series of 115 cases (16.5%). Since then, there have been numerous reports of DFSP in a region affected by trauma. A history of trauma, which is present in 10% to 20% of cases, could trigger the appearance of the tumor or just be a coincidence.

Several cases of DFSP have been reported in women in whom tumor growth starts or accelerates during pregnancy.[18] In fact, extensive expression of progesterone was found in 3 cases of DFSP in pregnant women,[19] and attempts have been made to link this finding with a possible hormonal etiology of DFSP, although results so far have been inconclusive. What does seem clear is that tumor onset is not related to sun exposure as no studies report such an association.

Epidemiology

DFSP is an uncommon tumor, with an estimated incidence of between 0.8 and 5 cases per million inhabitants per year.[20–22] The annual incidence seems to be greater in blacks than in other races,[22,23] and it appears to affect men and women equally.[17,24,25] Although extensive series of cases show a higher incidence in men than women, in the review of 2885 cases performed by Criscione and Weinstock,[22] there was a slightly higher incidence in women and in the series of 143 patients studied by Martín et al.,[26] 63% of the patients were women.

For all races and both sexes, incidence peaks between 30 and 50 years,[22] although congenital cases and cases in elderly individuals have been reported.

DFSP is most frequently located on the trunk, as reflected by all the large series in which 40% to 60% of the cases appear at this site,[16,17,22,26,27] particularly on the shoulder girdle and back. The second most frequent site is the limbs, which account for 20% to 30% of cases. The head and neck are involved in 10% to 15% of cases; tumors on this site typically present on the scalp and the supraclavicular region.[16,17,22,26,27]

Clinical Characteristics

DFSP usually presents as a small plaque with a flesh, brownish, pinkish, or even violaceous coloration. In this initial stage, it might go unnoticed by the patient and often be confused with a benign tumor, given that it is an asymptomatic, nonspecific lesion. The tumor grows slowly in this initial plaque stage. Three different appearances are possible.[26] The first is morphea-like, in which the lesion appears as an indurated plaque with a flesh, whitish, or grayish color. In the second, the atrophoderma-type tumor presents as a soft, depressed flesh-colored plaque with an atrophic appearance. The third, an angioma-type tumor, is less common and resembles vascular lesions such as flat angioma. As the tumor grows, it infiltrates more deeply and spreads, and nodules start to develop on the surface (Fig. 1A–D). The time taken for the transition from the plaque phase or nonprotruding phase to the nodular phase is highly variable, with a range of less than 1 month to up to 50 years.[26,27]

The size of the tumor depends essentially on the duration of growth. Normally, when the tumor is seen in the clinic, it usually has a size of 1 to 5 cm,[27] but sizes of greater than 20 cm have been reported.[16]

The tumor is usually located in the dermis and infiltrates the subcutaneous cellular tissue, and so it is usually mobile with no fixation to deeper structures, although long-standing tumors can invade the fascia, muscle, periosteum, and bone.[16,23,27,28]

Histopathologic Characteristics

Macroscopically, DFSP appears as a single, fairly well-delimited mass in the dermis. It has a firm consistency and a yellowish or gray color. In the macroscopic examination, infiltration of subcutaneous cell tissue is usually evident (Fig. 2A). Microscopically, DFSP appears as a

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