Dermatofibrosarcoma protuberans with fibrosarcomatous transformation*

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To the Editor,

Dermatofibrosarcoma protuberans (DFSP) is an uncommon sarcoma of the skin and soft tissue with an intermediate level of malignancy. DFSP with fibrosarcomatous differentiation (FS-DFSP) is a rare variant of DFSP, involving greater aggression, higher rates of local recurrences and a higher metastatic potential. We describe the case of a 70-year-old woman who presented with a painless tumor in her lumbar area that had first been noticed eleven months previously. The tumor had slowly increased in size but over the last two months it had become fast-growing and started bleeding. On physical examination, we observed an ulcerated, violaceous to red-brown, indurated tumor with prominent vessels (Figures 1-2). A punch-biopsy showed a dense growth of spindle cells forming a storiform pattern, with little pleomorphism and a low mitotic index, confirming the DFSP diagnosis. The CT scan discarded invasion of the underlying muscle tissue or bone, lymph node involvement and distant metastases. The patient underwent wide, local excision with macroscopic margins of 3cm. The final histopathological analysis (Figure 3) revealed fibrosarcomatous areas, containing fascicles of spindle-shaped cells arranged in a herringbone pattern, and abundant, atypical cells with an elevated mitotic index, unlike the usual histological appearance of DFSP. In addition, these transformed areas showed a weak expression of CD34 staining compared with the ordinary DFSP area. Therefore, the diagnosis of FS-DFSP was made. The surgical margins were free, and the patient has been free of disease for five months since surgery ended. DFSP is an un-

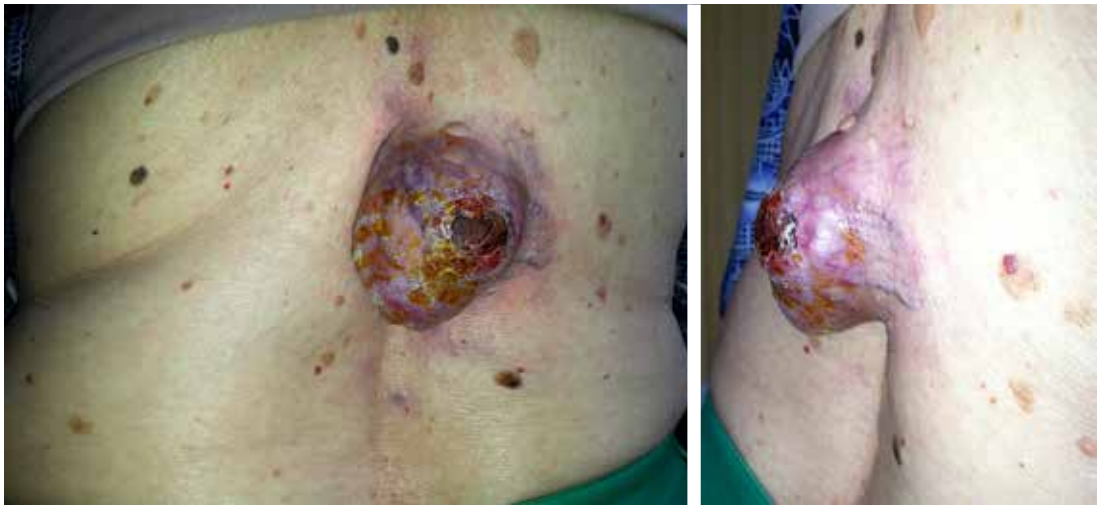

Figure 1: Ulcerated, violaceous to red-brown, indurated tumor with prominent vessels, measuring 60 x 50 x 45mm, in the lumbar area.

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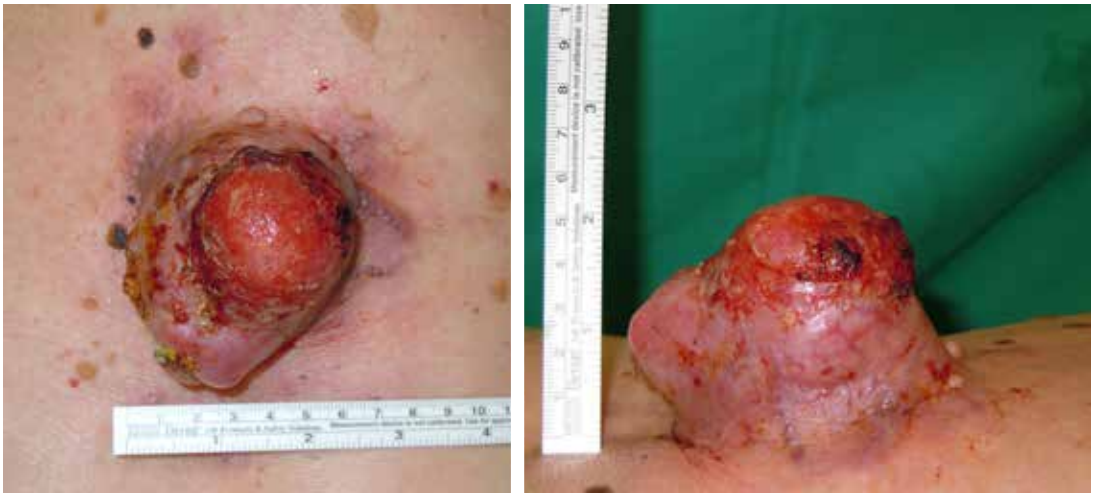

Figure 2: Clinical aspect of the tumor 3 weeks later. Note the rapid growth in the central transformed area

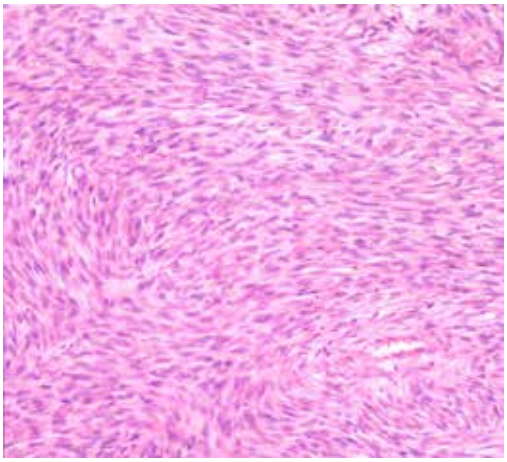

Figure 3: Hematoxylin-eosin stain: fibrosarcomatous areas with a fascicular pattern of spindle-shaped cells

common sarcoma of the skin and soft tissue with an intermediate level of malignancy, which favors young to middle-aged adults. The most commonly involved anatomic sites are the trunk and proximal extremities. FS-DFSP is a rare variant of DFSP, involving greater aggression, higher rates of local recurrences and a greater metastatic potential, affecting the lungs in particular.[1] Given their clinical similarities, differentiating these two entities relies on histopathology, although a sudden rapid growth should lead the clinician to suspect transformed DFSP. Wide excision with margins ≥2 cm represent the gold standard in the treatment of FS-DFSP, since this prevents recurrences and metastases.[2] Micrographic Mohs surgery can achieve tumor clearance with smaller margins, avoiding unnecessarily large surgical defects. Hence, some authors suggest the latter should be the treatment of choice instead of conventional surgery.[3] Radiation should be considered for cases that are not amenable to complete resection, while the PDGFR-inhibitor imatinib has proven effective in locally advanced, irresectable or disseminated FS-DFSP with translocation between chromosomes 17 and 22.[3] ❑

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