Case for diagnosis*

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CASE REPORT

A fifty-seven year-old white female patient reported, since childhood, the presence of a lesion on the fourth left finger, which appeared after a local trauma. Ten years ago, there was a progressive increase in this lesion and 5 years ago she underwent surgical excision with histopathology indicating a benign tumor composed of smooth muscle cells. In five months, there was a recurrence of the lesion that became painful.

Physical examination revealed an irregular and indurated nodular lesion, measuring 5 centimeters, with an erythematous surface with telangiectasias, located in the dorsal aspect of the proximal phalanx of the fourth left finger extending to the metacarpophalangeal joint (Figure 1).

A new biopsy was performed and histopathology demonstrated proliferation of spindle cells with storiform pattern and areas with "cartwheel" aspect (Figure 2A). Immunohistochemistry showed positivity for CD34 (Figure 2B). Radiography revealed increase in soft tissue volume without bone involvement and ultrasonography showed a hypoechoic solid image. Upon diagnosis, the patient underwent surgical treatment, as described below in the discussion section (Figure 3).

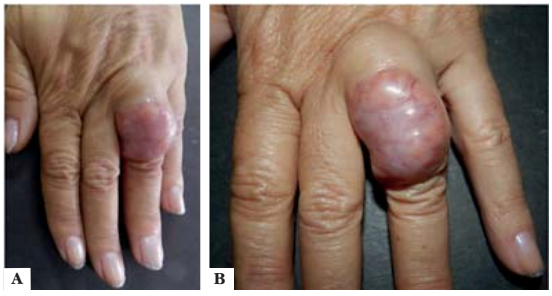

FIGURE 1: Irregular, nodular, indurate lesion with erythematous surface with teleangiectasias. **A)** Overview **B)** Lesion detail

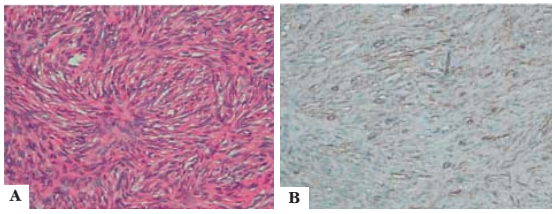

FIGURE 2: A) proliferation of spindle cells, with storiform pattern and "cartwheel" aspect in the dermis. HE200x
B) Immunohistochemistry with positivity for CD34

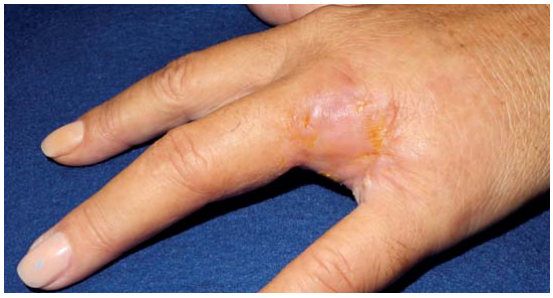

FIGURE 3: Post-operatory image showing amputation of the fourth left finger

DISCUSSION

Given the clinical appearance of the tumor, diagnostic hypotheses of dermatofibroma, schwannoma, leiomyoma, fibrosarcoma and amelanotic melanoma were formulated. The characteristic histopathological aspect, added to the immunohistochemical panel with CD34 positivity, indicated that it was a dermatofibrosarcoma protuberans (DFSP). Physical examination, and laboratory and imaging studies revealed no regional or visceral metastases. The patient was transferred to a referral hospital for cancer treatment, where she underwent surgical excision with wide safety margins, and amputation of the fourth left finger. She was asked to attend to quarter-

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ly follow-ups during the first 2 years, every 6 months for the following 3 years and once a year for 5 years.

DFSP is a fibrous histiocytic tumor of intermediate malignancy, that accounts for most of the cutaneous sarcomas.[1] It is most common among young adults between the third and fourth decades of life.[1-5] In 10-20% of cases, a local trauma is described as a triggering factor.[1,2] It has a high rate of recurrence and aggressive local growth, although its metastatic potential is low.[1-6]

During the initial phase, it manifests as an asymptomatic, hardened, violaceous, reddish-brown or slightly hyperchromic plaque, of slow growth.[1] It evolves with infiltration of adjacent tissues and development of protuberant nodules, which grow rapidly, resulting in pain, ulceration and bleeding.[2,3,6] The trunk is the most affected region.[1,3,5] Acral and genital locations are uncommon.[1]

Diagnosis is confirmed by histopathology that shows spindle cell proliferation with storiform pattern, which in some areas have the "cartwheel-like or whirlwind-like" aspect. Immunohistochemistry is useful in the differential diagnosis with positive staining for CD34, a human hematopoietic cell antigen.[1]

The treatment of DFSP is essentially surgical. Recurrence rates after conventional surgery are approximately 60%, decreasing to 20% when margins larger than 4cm are used. Mohs micrographic surgery provides significantly higher cure rates (1.6% recurrence).[1] Recent studies have described a good response to imatinib, which is indicated for metastatic and locally advanced disease.[1]

Often the indolent behavior of DFSP causes a delay in diagnosis[1] and inadequate treatment, increasing the morbidity of the disease as occurred with our patient, in whom the unusual location made it even more difficult to diagnose the tumor. ❑

Abstract: Dermatofibrosarcoma protuberans is a fibrohistiocytic tumor of intermediate malignancy with aggressive localized growth, high recurrence rate, but low metastatic potential. It appears as a hardened plaque, with slow growth, upon which the development of nodules occurs. It predominates in the trunk and is unusual in acral locations. Histopathology reveals spindle cells with storiform pattern and cartwheel-like or whirlwind-like aspect. Immunohistochemistry shows positivity for CD34. The treatment is surgical. We report a case of long evolution, with an unusual location, that relapsed after surgery, to emphasize the importance of early diagnosis and proper treatment, avoiding aggressive resections with increased morbidity.
Keywords: Dermatofibrosarcoma; Extremities; Skin neoplasms

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