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Dermatofibrosarcoma protuberans rapid growth after incisional biopsy

Crecimiento rápido de un dermatofibrosarcoma protuberans después de la biopsia incisional

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare malignant tumor with dermal origin, forming a dense arrangement of spindle cells in a storiform pattern. The diagnosis is based on the clinical aspect, histopathological and immunohistochemical evaluation. A typical feature is its ability to invade surrounding tissues, making it difficult to determine the real boundary of the lesion. By this fact, local recurrences are frequent after excision with wide margin. We have reported a case of DFSP with rapid growth after incisional biopsy.

RESUMEN

Dermatofibrosarcoma protuberans (DFSP) es un tumor maligno raro con origen dérmico, que forma un arreglo denso de células fusiformes en un patrón storiforme. El diagnóstico se basa en el aspecto clínico, la evaluación histopatológica e inmunohistoquímica. Una característica típica es su capacidad de invadir los tejidos circundantes, lo que hace difícil determinar el límite real de la lesión. Por este hecho, las recurrencias locales son frecuentes después de la escisión con amplio margen. Hemos reportado un caso de DFSP con crecimiento rápido después de la biopsia incisional.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare malignant tumor which has dermal origin, forming a dense arrangement of spindle cells structured in irregular installments resulting in storiform pattern. It shows aggressive local growth, high recurrence rate, but low metastatic potential. The incidence of this tumor is estimated to vary from 0.8 to 5 per million cases each year worldwide, accounting for 1 to 6% of soft tissue sarcomas. It typically affects patients between 20 and 50 years of age. However, there are case reports in the age extremes. It is slightly predominant in males.¹

CASE PRESENTATION

Thirty-one-year-old male patient, previously healthy, reports appearance of tumor lesion, hardened and initially painless, in the left arm, nearly two and a half years ago (*Figure 1*). He denied previous surgery or trauma on the place before the onset of the injury. After evaluation by other doctors, injury drainage

was performed with removal of purulent exudate and amoxicillin and clavulanate were prescribed. Discrete improvement with decreased size of the lesion was observed at the time.

Four months ago, he noticed progressive growth of the lesion. On physical examination he showed well-defined tumor lesion on the left upper limb, measuring approximately 5 cm in diameter, containing central necrotic crust, which was erythematous and painful at the edges. There was no palpable lymph node enlargement or systemic signs. The tuberculin skin test (TST) was positive (40 mm). Ultrasonography of the lesion showed heterogeneous expansive process located in the soft parts of the left arm measuring about 4.5 x 2.5 cm with no evidence of fluid level. Computed tomography scan showed mass with heterogeneous contrast enhancement, measuring approximately 5.2 x 4 cm without invasion of muscles. Incisional biopsy was performed, with anatomic pathology test, revealing spindle cell neoplasm showing moderate pleomorphism and scattered mitosis with dilated blood

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vessels in between. The immunohistochemical study was diffusely positive for vimentin and CD34 and negative for smooth muscle actin, desmin and S100 in neoplastic cells. The morphologic setting and the immunohistochemical profile were consistent with DFSP (Figure 2).

The patient returned one month after the biopsy date, reporting lesion growth and pain in the left upper limb (Figure 3). Tomography revealed mass with hypodense center suggesting cystic degeneration, with larger diameter of 6.2 cm, without bone invasion. He underwent extensive surgical resection of the lesion with 3 cm margin. The

anatomic pathological examination of the specimen revealed margins with no lesions. The patient is being followed in oncology and undergoes local adjuvant radiotherapy.

DISCUSSION

Most cases of DFSP has its root in the translocation of chromosomes 17 and 22 resulting in inadequate expression of the platelet derived growth factor B locus (PDGFB). It is histologically composed of monomorphic fusiform cells with low mitotic index, arranged around vessels and that infiltrate the subcutaneous fat with honey-comb pattern. Early lesions may have a neoplastic-cell-free



Figure 1. Nodular lesion on left upper limb with central crust.



Figure 3. Lesion on the left upper limb 30 days after biopsy.

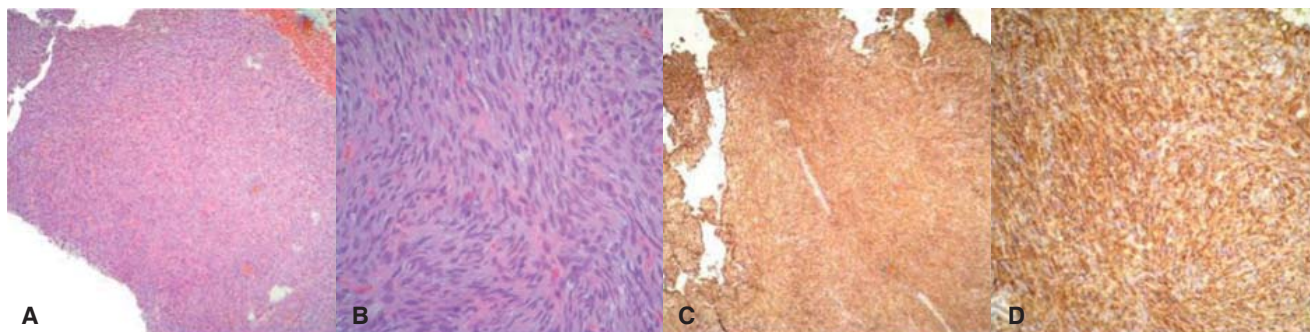


Figure 2. A) Nodular proliferation of spindle cells with storiform pattern HE 40x; B) Detail of spindle cells HE 400x; C) Immunohistochemical testing for CD34 diffusely positive in cellular proliferation; D) Detail of CD 34 immunostained cells.

zone (Grenz Zone) between the tumor and the epidermis.² A typical feature is its ability to invade surrounding tissues via subcutaneous irregular projections, which may not be detected at first sight, making it difficult to determine the real boundary of the lesion. By this fact, local recurrences are frequent after excision with an apparently wide margin.³

The immunohistochemical profile of the tumor is positive for CD34, vimentin and also CD99, and negative for factor XIIIa, CD 44, CD 117, SMA, desmin and S-100, which is useful in the differential diagnosis with dermatofibroma.⁴ The dermatofibroma is negative for CD34 and there are macrophages positive to CD68.⁵ As reported, the case patient presented positivity for vimentin and CD34 and immunonegativity for desmin and S100, confirming the diagnosis of DFSP. The difficulty to differentiate the histopathology pattern of the DFSP and the giant dermatofibroma (benign fibrous histiocytoma) hypercellular variant, is common. Among the histopathologic diagnosis, the most important are plaque-like CD34-positive dermal fibroma, dermatofibroma, spindle cell melanoma, leiomyosarcoma and perineurioma. In myxoid variants of DFSP, some important differential diagnosis must be considered, such as pleomorphic sarcoma, myxoid liposarcoma and myxofibrosarcoma.⁵

The lesions are located preferably in the trunk, lower limbs, upper limbs and head and neck, in descending order.^{6,7} The initial clinical setting consists of a hardened plate with purplish or bluish margins of slow growth for months to years. During its evolution it may present various colors and aspects, with sclerodermiform morphology or atrophic telangiectasias. It increasingly becomes a firm nodule, bonded to the local skin, but not profoundly. There may be a rapid growth phase occasionally, causing the typical bulging lesion, which can ulcerate, bleed or cause local pain.⁸

The diagnosis is based on the clinical aspect of the lesion and incisional or excisional biopsy for histopathological and immunohistochemical evaluation. A diagnosis based only on clinical data is not often possible to be done, because of the similarity of DFSP with other tumors. In our case, the differential diagnoses were scrofuloderma, amelanotic melanoma, Merkel cell carcinoma, squamous cell carcinoma and cutaneous metastasis. Computed tomography can be used to eliminate the adjacent bone involvement to extensive or recurrent lesions, such as the reported case. Further tests in search of distant metastasis are not justified in the absence of clinical symptoms or adverse histologic

prognosis since lymph-node or hematogenous metastases are rare and slow.⁹

The chosen treatment is lesion resection with negative margins and among the options are surgical approach and Mohs micrographic surgery. In 1978, seven patients with DFSP who had micrographic surgery without recurrence, were reported by Mohs. The pathology processing is the main difference between wide local excision (three dimensional en bloc removal of the tumor) and Mohs micrographic surgery (MMS) and in MMS the surgeon can know the clear surgical margin. However, the problem with MMS is the prolonged procedure time, especially if a wide reconstruction is needed, labor-intensive and the challenge is to achieve a satisfactory local control.^{10,11} It was described that the local recurrence rate related to the conventional surgery is up to 30% of cases and the rate in MMS, according to several series, is as low as 0 to 6.6%.^{12,13}

The resection margin needed to achieve local control is still undefined, although a minimum of 2 cm of surgical margin has been recommended.³ The use of Imatinib Mesylate is currently indicated for recurrent, unresectable or metastatic disease. According to studies, its benefit is related to the performance of conservative surgery, reduction of local recurrence and extended lifespan when incorporated into the treatment of localized disease. Imatinib Mesylate is a tyrosine kinase inhibitor which works against activated PDGFB and the intention of the neoadjuvant therapy with this drug is to convert the unresectable tumor into a resectable tumor or to reduce the comorbidities from the surgery.¹⁴⁻¹⁶

As evidenced in the clinical case, DFSP is a tumor with high local tissue invasiveness and in this specific case it grew significantly after the biopsy. To our knowledge this is the first case description of the DFSP growth after biopsy, suggesting an important relationship between trauma and tumor development. According to several studies, a history of antecedent trauma in the region of the tumor can be elicited in 10 a 20% of patients.¹⁷⁻¹⁹ The sites of trauma reported are surgical²⁰ or burning scars,²¹ immunization,²² postradiation,²³ tattoos¹⁵ and site of a central venous line.²⁴ The development of the tumor after the trauma ranges from 2 months to 20 years. However, shorter periods tended to predominate, similar to the case reported.²⁵

CONCLUSIONS

Considering the reported case, we emphasize the importance of knowledge about the clinical manifestations

of DFSP in order to include it in the differential diagnosis of bosselated tumors, seeking an early surgical approach and, thus, avoiding functional sequelae and esthetic damage. As reported in this case, there may be a real relationship between trauma and the growth of the lesion. We suggest future research to establish this association.

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