Case for diagnosis*

Felipe Maurício Soeiro Sampaio[1]
Fernando Gustavo Mósca de Cerqueira[2]
Curt Mafra Treu[4]
Gustavo Vieira Gualberto[1]
Ana Maria Mósca de Cerqueira[3]
Thiago Jeunon de Sousa Vargas[1]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20142507

CASE REPORT

An 11-year-old brown male presented with a single, reddish-brown nodule, 2.5 cm in diameter, with raised edges and an exudative, ulcerated center, on the distal third of the anterior surface of the right forearm (Figure 1). The lesion was asymptomatic and had been present for 2 years, with no history of local trauma. The patient was in good overall condition, with no palpable lymph nodes.

Histological examination of an excisional biopsy specimen stained with hematoxylin and eosin showed epidermal hyperplasia and a nodular, symmetrical proliferation of fusiform (spindled) and histiocytic cells in the dermis, with rounded, poorly defined borders, as well as young collagen fibers entrapping pre-existing mature collagen bundles and sparing the fat lobules (Figure 2). In the central portion, there was proliferation of spindled and histiocytic cells arranged in fascicles, with relatively large, vesicular nuclei, but no pleomorphism or evidence of mitosis. Immunohistochemical staining was positive for factor XIIIa and negative for CD34 (Figure 3).

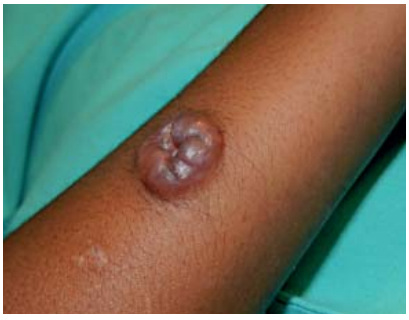

FIGURE 1: Mass lesion on right forearm

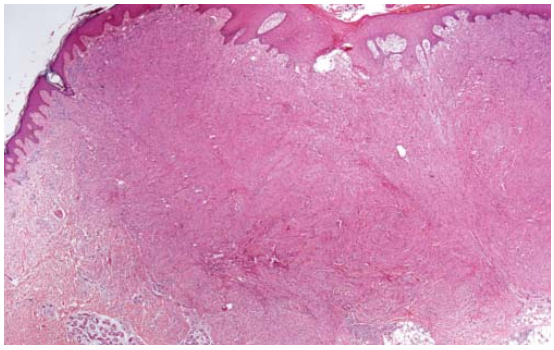

FIGURE 2: Overlying epidermal hyperplasia and dimpling of the central portion of the lesion. Nodular, symmetrical proliferation of spindled and histiocytic cells in the dermis, with rounded, poorly defined borders, sparing the fat lobules (H&E, original magnification 20x)

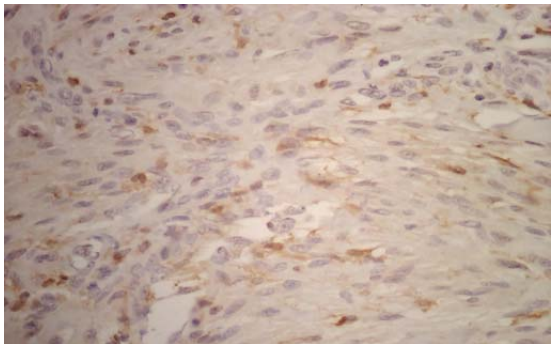

FIGURE 3: Interstitial reactivity on immunohistochemical staining for factor XIIIa

Received on 03.02.2013.
Approved by the Advisory Board and accepted for publication on 01.07.2013.
* Work performed at Hospital Federal de Bonsucesso (HFB) – Rio de Janeiro (RJ), Brazil.
Conflict of interest: None
Financial funding: None

[1] Hospital Federal de Bonsucesso (HFB) – Rio de Janeiro (RJ), Brazil.
[2] Universidade Gama Filho (UGF) – Rio de Janeiro (RJ), Brazil.
[3] Hospital Municipal Jesus – Rio de Janeiro (RJ), Brazil.
[4] Policlínica Geral do Rio de Janeiro (PGRJ) – Rio de Janeiro (RJ), Brazil.

DISCUSSION

Dermatofibromas (DFs), or benign fibrous histiocytomas, constitute a group of predominantly benign neoplasms, of unclear etiology and cell lineage, which exhibit fibroblastic and histiocytic features on histopathology.[1,2] Although widespread, the etiological hypothesis of reactive cell proliferation in response to local microtrauma remains unproven.

Clinically, DFs present as single or multiple well-circumscribed, reddish-brown papules, usually asymptomatic and slow-growing, 0.5-1.0 cm in diameter, and occurring on the lower limbs and trunk in young adults.[1,3,4,5] DFs arising in childhood may grow larger and affect unusual locations, such as the head, neck, and upper extremities. Extracutaneous involvement is rare, but has been reported in the long bones.[6]

On histopathological examination, DFs are characterized by proliferation of fibroblast-like cells, arranged in a storiform pattern in the papillary and reticular dermis, interwoven with mono- or multinuclear histiocytes with foamy cytoplasm. Perivascular lymphocytic infiltration may be present, as may increased vascularity and areas of hemorrhage, which explains the origin of the hemosiderin that is partially responsible for the tan or black aspect of the lesion in some cases. The overlying epidermis may be normal, atrophic, or acanthotic.[1]

Several histological subtypes of DF have been reported; the most common variants are the cellular, aneurysmal, atypical, and fibrocollagenous forms.[1,2,4]

The classical immunohistochemical reaction pattern of DF is positive staining for factor XIIIa, vimentin, and actin, and negative staining for CD34.[1,2,7] The importance of immunohistochemical examination lies in its ability to distinguish DF from dermatofibrosarcoma protuberans (DFSP), which is CD34-positive and factor XIIIa-negative. Other immunomarkers such as ST-3, CD10, and insulin-like growth factor-binding protein 7 (IGFBP7) can provide additional clarification in questionable cases.[7]

Depending on the clinical presentation, the differential diagnosis should also include leiomyoma, leiomyosarcoma, malignant fibrous histiocytoma, melanoma, hypertrophic scar, keloid, and Kaposi's sarcoma.[1,2,8]

No treatment are required. Complete resection is reserved for atypical cases, giant or locally symptomatic lesions, and at the cellular, aneurysmal, and atypical subtypes.[3,9]

Local recurrence has been described with lesions larger than 1.5-2.0 cm, with greater cell concentration, a high mitotic index, subcutaneous invasion and the presence of pleomorphism, hypervascularity or necrosis.[1,2] Lymph node metastases are rare, and whether they occur at all is controversial.[1,9] ❑

Abstract: We report the case of an 11-year-old male patient with a histopathological and immunohistochemical diagnosis of dermatofibroma with an atypical clinical presentation on the right forearm. Although dermatofibroma is considered a benign skin tumor, some of its differential diagnoses, such as dermatofibrosarcoma protuberans and malignant fibrous histiocytoma, are truly aggressive. Lesions with atypical clinical aspects and topology associated with specific histopathological variants are some of the criteria for complete tumor excision.
Keywords: Dermatofibrosarcoma; Histiocytoma, benign fibrous; Keloid

REFERENCES

1. Szumera-Ciećkiewicz A, Ptaszyński K. Benign fibrous histiocytoma of the skin metastasizing to the inguinal lymph node. Pol J Pathol. 2011;62:183-6.
2. Volpicelli ER, Fletcher CD. Desmin and CD34 positivity in cellular fibrous histiocytoma: an immunohistochemical analysis of 100 cases. J Cutan Pathol. 2012;39:747-52.
3. Pusztaszeri M, Jaquet PY, Williamson C. Giant hemosiderotic dermatofibroma: a case report and review of the literature. Case Rep Dermatol. 2011;3:32-6.
4. Han TY, Chang HS, Lee JH, Lee WM, Son SJ. A clinical and histopathological study of 122 cases of dermatofibroma (benign fibrous histiocytoma). Ann Dermatol. 2011;23:185-92.
5. Castellanos-González M, Rodríguez-Peralto JL, Zarco C, Ortiz-Romero P. Multiple clustered dermatofibromas: an atypical presentation of a common disease. Actas Dermosifiliogr. 2011;102:467-8.
6. Ceroni D, Dayer R, De Coulon G, Kaelin A. Benign fibrous histiocytoma of bone in a paediatric population: a report of 6 cases. Musculoskelet Surg. 2011;95:107-14.
7. Li J, Yu Y, Yang Y, Wang L, Cao J, Liang X, et al. IGFBP7, a novel immunohistochemical marker in differentiating dermatofibroma from dermatofibrosarcoma protuberans. J Eur Acad Dermatol Venereol. 2012;26:382-5.
8. Chung J, Namkoong S, Sim JH, Lee JS, Hong SP, Kim MH, et al. Deep penetrating benign fibrous histiocytoma of the foot associated with throbbing pain Ann Dermatol. 2011;23:S239-42.
9. Jung KD, Lee DY, Lee JH, Yang JM, Lee ES. Subcutaneous dermatofibroma. Ann Dermatol. 2011;23:254-7.

MAILING ADDRESS:
Felipe Maurício Soeiro Sampaio
Rua Marino da Costa, 217 – Apt. 201
Ilha do Governador
21940-210 - Rio de Janeiro - RJ
Brazil
E-mail: felipemauricio@uol.com.br

How to cite this article: Sampaio FMS, Gualberto GV, De Cerqueira FGM, De Cerqueira AMM, Treu CM, Vargas TJS. Case for diagnosis. Dermatofibroma in childhood. An Bras Dermatol. 2014;89(3):519-20.