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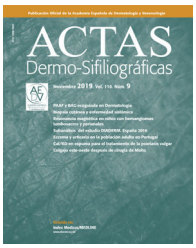

CASE AND RESEARCH LETTERS

Clinical and Ultrasound Features of Dermatofibromas in Pediatric Patients

Caracterización clínico-ecográfica de dermatomiofibromas en edad pediátrica

To the Editor:

Dermatofibroma (DMF) is a benign tumor of myofibroblastic origin that is rare in pediatric patients. The entity was first described in the literature by Hügel in 1991 as *plaque-like dermal fibromatosis*.[1] The term *dermatofibroma* was later coined by Kamino.[2]

These tumors usually present clinically as a solitary plaque or nodule, and may be multiple, poorly defined, with mild erythema or hyperpigmentation, and are usually asymptomatic, although they may cause mild pruritus. In the pediatric form, the lesions occur predominantly in the cervical region (56%) and tend to remit spontaneously.[3] The adult form, however, tends to be located on the shoulders or on the proximal surface of the limbs. It is more common in women (96%) and does not tend to remit spontaneously.[4] The clinical differential diagnosis is very broad[4] (Fig. 1).

Diagnosis of DMF is histologic. It consists of a proliferation of fusiform cells (fibroblasts and myofibroblasts) arranged with the long axis parallel to the surface of the epidermis and sparing adnexal structures. Immune staining may help with the diagnosis. Expression of vimentin is constant, that of muscle-specific actin and smooth-muscle actin is variable, and focal expression of CD34+ may be seen in some cases.[5,6]

High-frequency ultrasound, which is increasingly common in dermatologic practice, can help with the clinical diagnosis and may be helpful in the follow-up of these benign tumors.

We took all the patients under 16 years of age with a diagnosis of DMF from the anatomical pathology database of Hospital Universitario de Fuenlabrada, Spain, and we selected those cases in which an ultrasound study had been performed.

The clinical data are shown in Table 1. The study group contained 3 girls and 5 boys. Age at diagnosis ranged between 20 months and 15 years. The lesions were located on the neck (6 patients), on the left shoulder (1 patient), and on the occipital region (1 patient). The lesions had appeared between a month and a year before consultation. Clinically, the lesions were nodular or plaque-like, poorly defined in 6 of the patients (well demarcated in 2 cases), and slightly erythematous or hyperpigmented. One of the patients presented 2 lesions. Three of the lesions were painful and 2 were pruritic; the rest were asymptomatic.

An ultrasound study with a high-resolution probe (Esaote MyLab® Class c, 18 MHz linear probe) showed rounded, hyperechoic, nonencapsulated, poorly defined, dermal-epidermal lesions without Doppler flow, except in 2 cases, which presented Doppler flow on the periphery.

Clinical diagnosis of DMF may be difficult and high-resolution ultrasound may help with the differential diagnosis and with guiding treatment, essentially in large or pruritic lesions.

The ultrasound differential diagnosis includes lesions such as dermatofibroma, which usually presents as dermal lesions that may extend to the subcutaneous cellular tissue, with poorly defined, spiculated borders, without vascularization, and with changes in the echogenicity of the adjacent tissue[7]; plaque-like neurofibroma, which also tends to present as hypoechoic lesions located in the dermis, with well-defined borders and posterior reinforcement.[8] Other lesions, such as hypopigmented blue nevus, piloleiomyoma, smooth-muscle hamartoma, and connective tissue nevus, tend to be better defined. Dermatofibrosarcoma protuberans has a jellyfish-like appearance, with an oval shape and focally poorly defined borders; it invades the subcutaneous tissue in the form of hypoechoic tentacle-like projections with focal hyperechoic areas and posterior reinforcement and abundant vascularization.[9] Desmoid fibromatosis and scars tend to present with a more elongated morphology, although cases of DMF with a linear configuration have been reported.[10]

A watch-and-wait approach is a valid treatment option, as this is a benign tumor with little tendency toward recurrence. Excision is sometimes required due to the symptoms.

In our experience, given the clinical suspicion and a high-resolution ultrasound study compatible with the diagnosis, an appropriate approach would be clinical and ultrasound follow-up, with biopsy reserved for lesions with atypical characteristics, such as high Doppler flow, and excision of symptomatic lesions only. This conservative approach is valid for all ages, as it avoids unnecessary excisions and the risk of cosmetic defects, especially in pediatric patients, in whom most cases of surgery require general anesthesia or sedation.

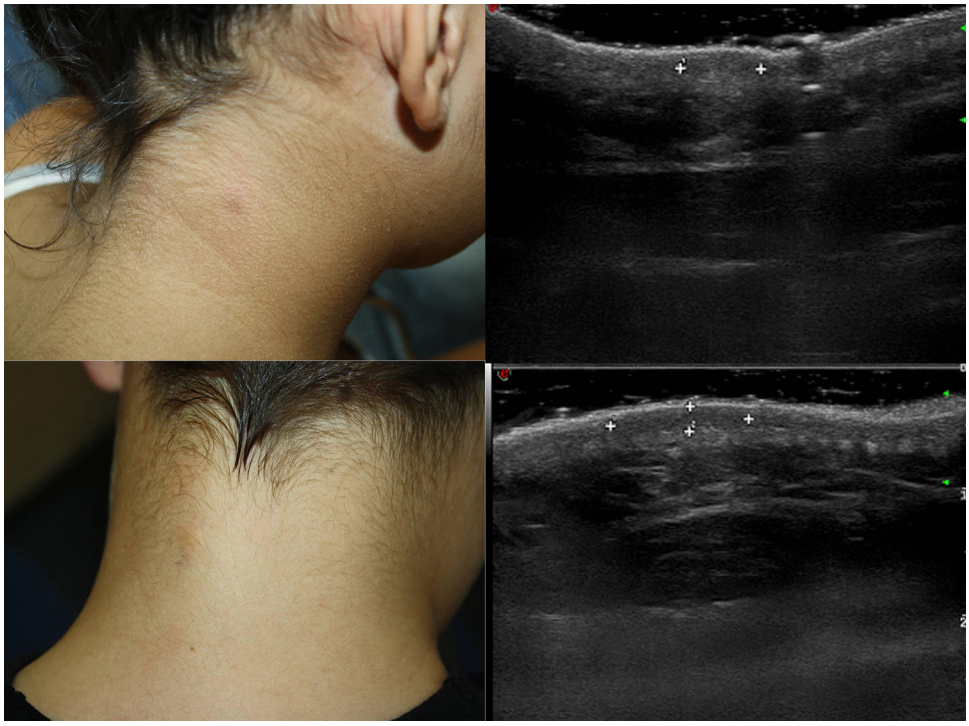

Figure 1 Clinical and ultrasound characteristics.

Table 1 Clinical and ultrasound characteristics of the patients.

	Sex	Age	Location	Time	Clinical features	Physical examination	Ultrasound features
1	Female	15 y	Posterior cervical	1 mo	Pain, stable	Poorly demarcated, hyperpigmented nodule	Poorly demarcated hypoechoic dermal lesion without Doppler flow
2	Male	11 y	Posterior cervical	Months	Asymptomatic, stable	Poorly demarcated nodule	Poorly demarcated hypoechoic dermal lesion without Doppler flow
3	Male	11 y	Posterior cervical	1 y	Asymptomatic, growth	2 well-demarcated nodules	Poorly demarcated hypoechoic dermal lesion without Doppler flow
4	Female	11 y	Left shoulder	Months	Pain	Poorly demarcated nodule	Hypoechoic dermal lesion, increased Doppler flow in points on periphery
5	Male	10 y	Posterior cervical	Months	Asymptomatic	Poorly demarcated erythematous nodule	Poorly demarcated hypoechoic dermal lesion with increased Doppler flow
6	Male	10 y	Posterior cervical	Months	Asymptomatic, slight growth	Well-demarcated, slightly hyperpigmented nodule.	Poorly demarcated hypoechoic dermal lesion without Doppler flow
7	Female	9 y	Occipital	1 y	Occasional pruritus, progressive growth	Poorly demarcated erythematous nodule	Poorly demarcated hypoechoic dermal lesion without Doppler flow
8	Male	20 mo	Posterior cervical	8 mo	Pruritic, fluctuating size	Greyish-erythematous plaque	Poorly demarcated hypoechoic dermal lesion without Doppler flow

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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Cocaine/Levamisole-Associated Autoimmune Syndrome: A Case Report☆

Síndrome autoinmune cocaína-levamisol. Presentación de un caso

To the Editor:

A 55-year-old man visited the emergency department with clinical signs and symptoms that had appeared some days earlier, consisting of progressive dyspnea accompanied by fever of up to 39 °c, productive cough, and flat, red-violaceous reticulated lesions with a purplish appearance on the left outer ear (Fig. 1), left malar region, right nostril, and both arms. The patient was an ex-intravenous drug abuser and had a history of stage-2 infection with the human immunodeficiency virus (HIV), hepatitis B virus, hepatitis C virus, which was cured spontaneously, and presented chronic neutropenia. Laboratory tests showed leukopenia and thrombocytopenia, creatinine levels of 6 mg/dL, procalcitonin of 3.74, C-reactive protein (CRP) of 313, and a glomerular filtration rate of 9 mL/min. Antineutrophil cytoplasmic antibodies (ANCAs) and lupus anticoagulant were positive, whereas cryoglobulins were negative. A chest x-ray showed bilateral disperse alveolar-interstitial infiltrates and the patient was therefore diagnosed with lobar pneumonia and acute kidney failure.

A punch biopsy of one of the skin lesions was performed and revealed massive occupation of the capillaries by mixed thrombi, with some trapped polymorphonuclear cells, without the characteristic abnormalities of leukocytoclastic vasculitis (Figs. 2 and 3). The differential diagnosis

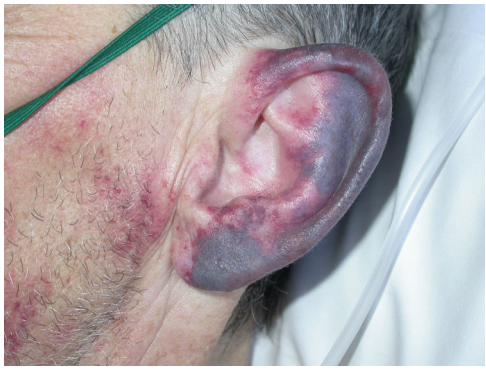

Figure 1 Purplish lesion on the outer ear.

included thrombotic thrombocytopenic purpura, disseminated intravascular coagulation, cryoglobulinemia, and retiform purpura associated with cocaine use. The first 2 entities were ruled out clinically and the third was ruled out due to the absence of cryoglobulins. The patient subsequently admitted to using cocaine. The skin lesions remitted

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