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CASES FOR DIAGNOSIS

Congenital Tumor With a Vascular Appearance

I. Cervigón, L. M. Torres, and Á. Palomo
Servicio de Dermatología, Hospital Nuestra Señora del Prado, Talavera de la Reina, Toledo, Spain

Medical History

A newborn girl presented in the dermatology department with a tumor in the right antecubital fossa; the tumor prevented full extension of the arm.

Physical Examination

A fixed fibrous, nonulcerated, erythematous nodule 2 cm in diameter was observed (Figure 1). The patient presented no other similar lesions and no enlarged organs were found on palpation.

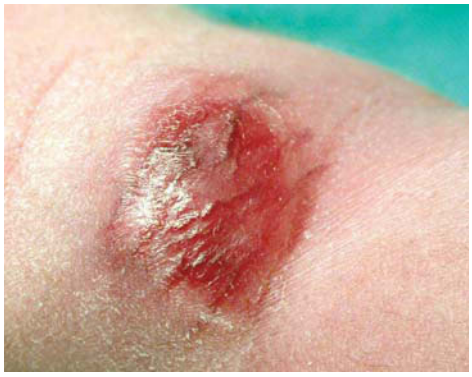

Figure 1.

Additional Tests

The patient was referred to the pediatrics department for general evaluation and to rule out internal lesions. Diagnostic imaging, including x-ray, ultrasound, and computed tomography, revealed no signs of bone or organ involvement.

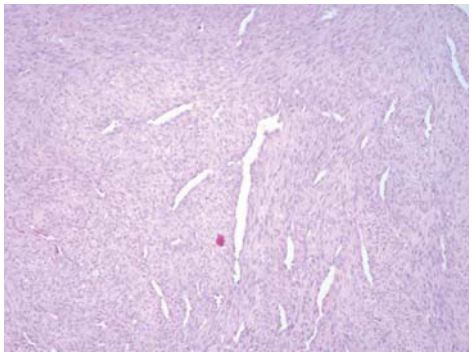

Figure 2. Hematoxylin–eosin. ×100.

Histopathology

The histologic study revealed a proliferation of fusiform cells in the superficial and deep dermis; the cells showed no nuclear abnormalities and were arranged in bands and fascicles, surrounding thin-walled vascular structures (Figure 2). Immunohistochemistry showed that the cells were positive for vimentin and smooth muscle α-actin and negative for S100, myoglobin, cytokeratins, and desmin.

What is your diagnosis?

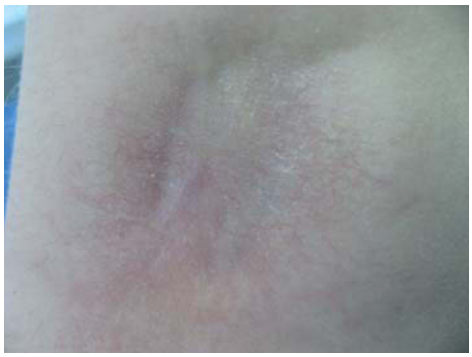

Figure 3.

Correspondence:
Iván Cervigón González
Servicio de Dermatología
Hospital Nuestra Señora del Prado
Carretera de Madrid, km 114
45600 Talavera de la Reina, Toledo, Spain
icervigon@sescam.jccm.es

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Diagnosis

Solitary infantile myofibromatosis

Treatment and Course

It was decided to adopt a wait-and-see attitude, with periodic follow-up of the patient. The tumor gradually regressed and resolved completely after 4 years, leaving no sequelae and allowing free movement of the arm (Figure 3).

Comment

Infantile myofibromatosis is a benign tumor that typically occurs during infancy.[1] There are 4 variants: solitary infantile myofibromatosis, multicentric myofibromatosis with and without involvement of the viscera, and solitary adult myofibromatosis.[2] Lesions are located in the dermis and hypodermis, though they may occasionally reach the muscle and bone; more than 30% of the multicentric forms affect the viscera.[2]

Although myofibromatosis is the most common form of fibrous tumor in infancy, the incidence is low.[2] Eighty percent of cases are either congenital or commence in the first 2 years of life; the disease is rare in late infancy and adult life.[2]

The etiology of this disease is unknown. Most cases are sporadic, though patterns of autosomal dominant and recessive inheritance have been reported in some patients.[3]

Clinically, the lesions present as solitary or multiple plaques, nodules, or masses ranging in diameter from 0.5 to 5 cm; they are painless and of a firm consistency, rarely ulcerate or bleed, and may have a keloid or vascular appearance.[2] Tumors involving soft tissue may remit spontaneously in 1 or 2 years, probably due to massive apoptosis and necrosis.[2] The adult forms, however, do not remit spontaneously.[4] Histologically, the disease presents as a well-defined dermal nodule with a biphasic appearance[2] and abundant peripheral fusiform cells arranged in short fascicles. These cells show no nuclear abnormalities, though there may be occasional mitotic figures; they express smooth muscle α-actin and are negative for S100, cytokeratins, and desmin.[2] Electron microscopy shows them to be contractile myofibroblasts. The central area contains vascular structures with irregular lumens and a hemangiopericytic pattern.

The differential diagnosis should include tumors such as leiomyoma, leiomyosarcoma, neurofibroma, neuroblastoma metastasis, desmoplastic fibroma, fibrosarcoma, and hemangiopericytoma.[2]

Treatment should be conservative and a wait-and-see attitude should be adopted with solitary and multicentric forms that do not affect the internal organs.[2] Close monitoring of patients is, however, of vital importance, even in solitary forms, in order to rule out the subsequent appearance of new skin, bone, or visceral lesions. Surgery is the treatment of choice in aggressive solitary forms, when lesions are in high-risk areas, and in myofibromatosis affecting the internal organs.[2] Treatment with interferon or chemotherapy may be attempted in unresectable lesions or lesions that recur after surgery.[2]

Prognosis for solitary and multiple lesions that do not affect internal organs is excellent as the tumors remit spontaneously, though some do recur.[2] Myofibromatosis involving internal organs, however, has a mortality of over 70%.[2]

Conflicts of Interest
The authors declare no conflicts of interest.

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