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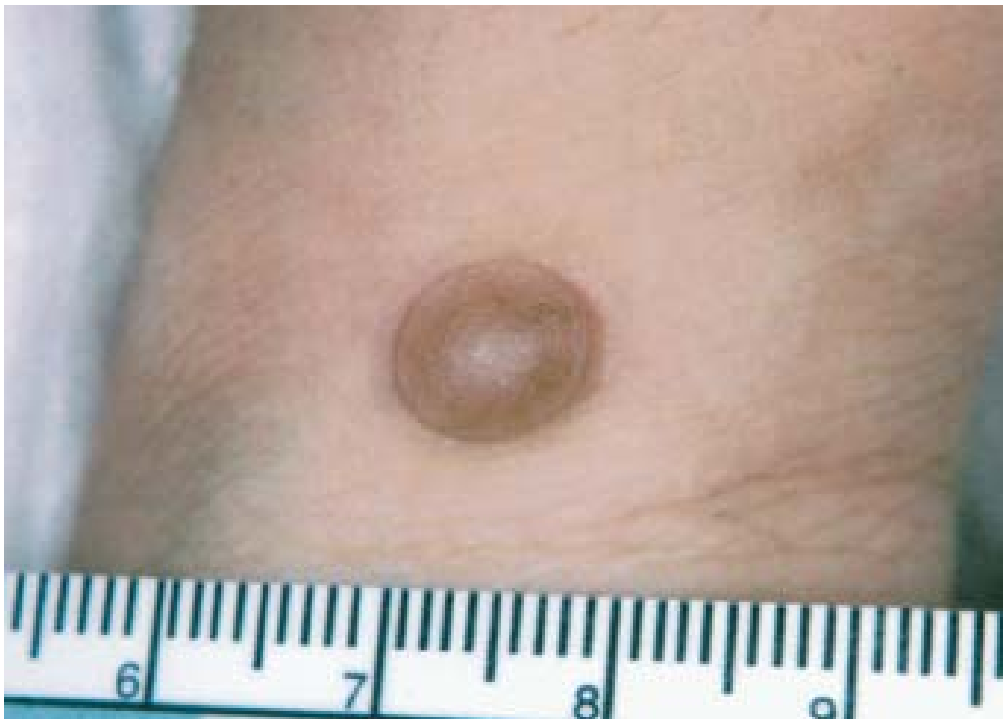

A 16-year-old male without significant past medical history presents with a firm, non-tender, pedunculated lesion on his distal left forearm. The lesion began as a small brown papule and has grown in size over 1½ years without associated symptoms.

What is your diagnosis?

PLEASE TURN TO PAGE 157 FOR DISCUSSION.

The Diagnosis

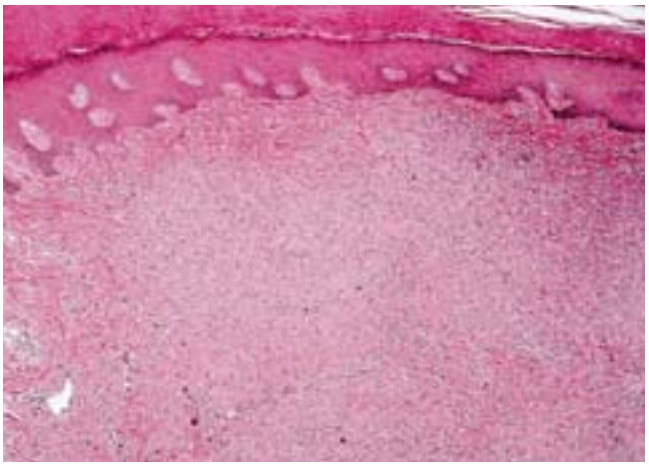

FIGURE 1A. The hyperplastic epidermis is separated by a Grenz zone from a dermal collection of cellular spindle cell tumor in the dermis (H&E; original magnification, x 4).

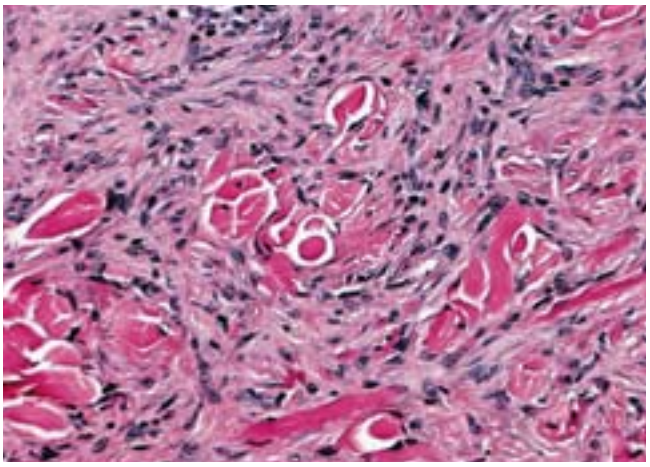

FIGURE 1B. Proliferations of plump dendritic cells are intermingled with thick eosinophilic collagen bundles (H&E; original magnification, x 20).

Discussion

The differential diagnosis of the lesion includes Spitz nevus, juvenile xanthogranuloma, dermatofibroma, dermatofibrosarcoma protuberans, verruca, neurofibroma, traumatic neuroma, solitary mastocytoma, and eccrine poroma. The lesion was excised and the histology was consistent with dermatofibroma.

Dermatofibroma (DF), also known as benign fibrous histiocytoma, is a common cutaneous lesion, usually presenting on the extremities as a solitary, slow-growing, firm dermal nodule with color ranging from red and red-brown to blue-black. DF may arise anywhere on the body but is rarely found on the palms or soles.[1] These lesions typically present in adults, but have been reported in children as young as 1 year of age.[2] On examination, the presence of a central "dimple" or "pucker" sign is a clinically useful diagnostic feature. Variants with unusual clinical morphologies such as polypoid,[3] atrophic,[4,5] and giant DF[6-8] have been reported. The latter may grow as large as 8 cm in diameter.[9]

DF is most commonly observed as solitary lesions in healthy individuals. Multiple dermatofibromas are rare and usually observed in patients with immune suppression. They have been reported in renal graft recipients[10]; in patients with HIV infection[11]; in patients with autoimmune disorders such as myasthenia gravis, systemic lupus erythematosus, Sjogren's syndrome, pemphigus vulgaris, and ulcerative colitis[12,13]; and in patients with atopic dermatitis.[14] The mechanism by which immunosuppression leads to the development of multiple dermatofibromas is not known.

Treatment of DF is usually not necessary unless recurrent trauma to the lesion interferes with function. Spontaneous regression has rarely been reported.[15] Excision is generally curative. Cryotherapy has also been reported to be effective.[16]

Histologically, DF are typically composed of a dermal collection of dendritic cells, lymphocytes and macrophages, separated from a hyperplastic epidermis by a Grenz zone (Figure 1A). At higher magnification, proliferations of plump dendritic cells are seen intermingled with thick eosinophilic collagen bundles, usually at the periphery of the lesion (Figure 1B). Immunostaining for factor XIIIa is positive on the dendritic cells in 90% of cases.[17] Histologic variants of DF include cellular, epithelioid, aneurysmal, atypical,[18] myxoid,[19] granular cell,[20] keloidal,[21] clear cell,[22] lichenoid, erosive, and ulcerated DF.[23]

Clinical and histologic DF variants can contribute to diagnostic confusion. Thus, recognizing the range of presentation of DF may help prevent misdiagnosis. The case presented here has an unusual morphology and is most likely a polypoid DF.

The pathogenesis of DF remains poorly understood. Some authors argue that DF is a reactive process since many develop after a history of trauma (*eg*, insect bite, shaving trauma, vaccination[15,24]); others have recent evidence that it is a neoplastic growth.[25] At present, it is not known whether trauma directly or indirectly affects the proliferations of fibroblast-like spindle cells in the dermis. Previous studies have demonstrated high expression of platelet-derived growth factor beta-receptor mRNA and protein,[26] as well as cutaneous tissue growth factor mRNA[27] in DF. The role of these molecules in the pathogenesis of DF is not known. Further investigation at the molecular level is needed to clearly delineate the factors that trigger the development of DF.

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