

Dermatomyofibroma: A Case Report And Review Of The Literature

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Abstract

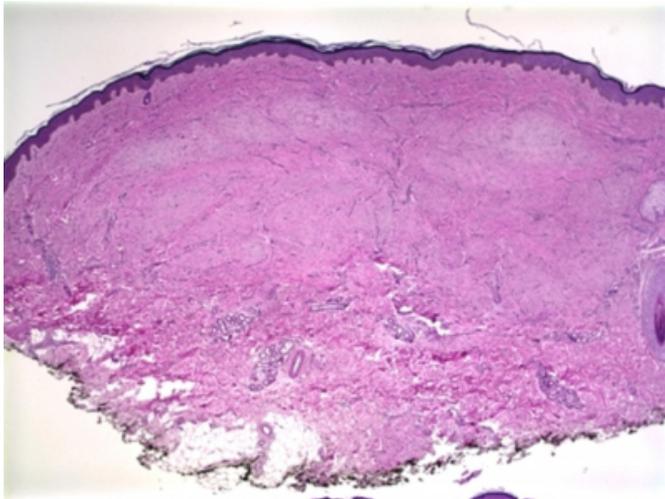
Dermatomyofibroma is a rare, benign cutaneous tumor derived from myofibroblasts. It is predominantly found in young women, and only a few cases have been reported in males. This entity is not generally well known and often misdiagnosed. We report such a case of dermatofibroma occurring in a 23-year-old male with a brief review of the English literature.

CASE REPORT

A 23-year-old male patient presented with a 0.5-cm raised skin nodule on the upper right arm. The biopsied lesion showed a plaque-like, ill-demarcated dermal fibrous proliferation extending into the subcutaneous adipose tissue (Figure 1).

Figure 1

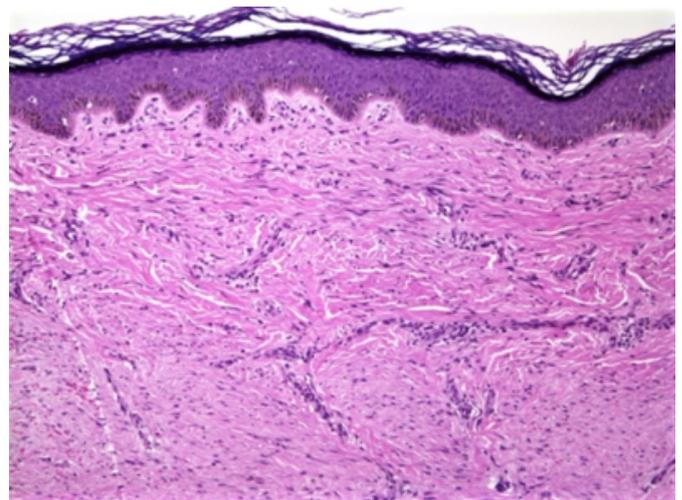
Figure 1: Low magnification.



The overlying epidermis was normal. The tumor was composed of uniform, slender spindle cells, arranged into well-defined elongated and intersecting fascicles with a predominantly parallel arrangement to the skin surface (Figure 2).

Figure 2

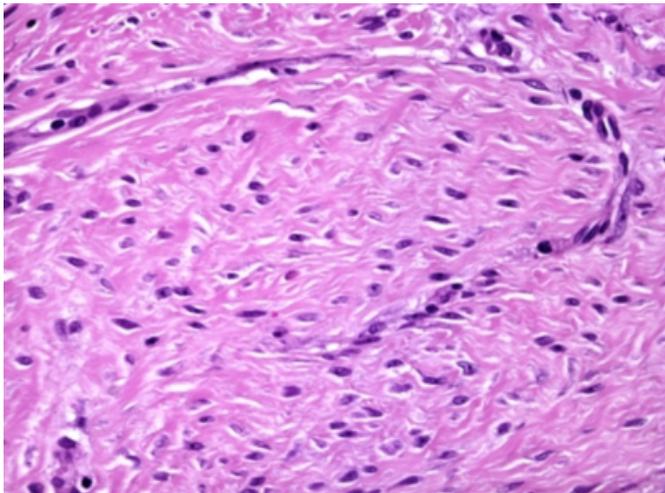
Figure 2: Higher magnification.



The tumor cells had faintly eosinophilic cytoplasm and elongated, vesicular, and uniform nuclei (Figure 3).

Figure 3

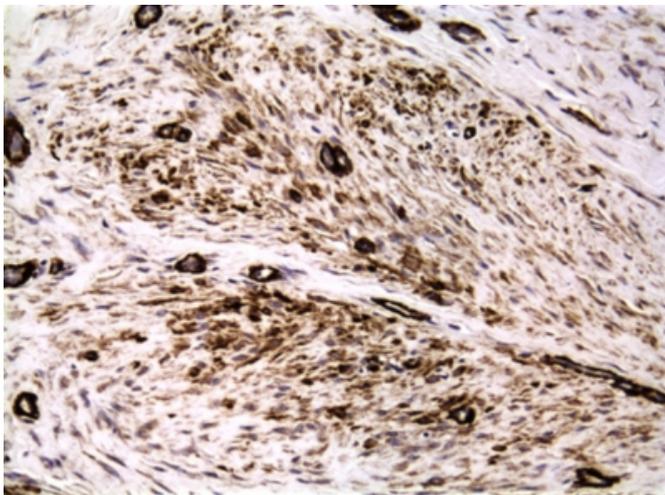
Figure 3: Higher magnification.



There was no significant cytological atypia or mitotic figures. Immunohistochemical studies showed that the tumor cells were strongly positive for smooth muscle actin (SMA) (Figure 4) and vimentin, and there is no expression of S100 protein or CD34. A diagnosis of dermatomyofibroma was made based on the clinical, histopathological, and immunohistochemical findings.

Figure 4

Figure 4: The tumor cells are positive for smooth muscle actin.



DISCUSSION

Dermatofibromyoma was first described as a distinct entity by Kamino et al in 1992 [1]. Since then several additional cases have been reported [2,3,4,5,6,7,8,9]. Most cases occur in the adolescent and young females with a mean age 30 years. The lesion is typically located in the shoulder, axilla or forearm. Clinically, the patient usually presents with a firm

red-brown plaque or nodule measuring from 1 to 2 cm in greatest diameter. One giant lesion measuring 15 cm in diameter has been reported [8].

Microscopically, dermatomyofibroma typically involves the reticular dermis in a plaque-like fashion with extension to the upper part of the subcutaneous septa, and the epidermis is often spared without significant changes. The tumor is composed of bland spindle cells that are arranged in intersecting bundles and fascicles paralleling to the skin surface. The cytoplasm of the neoplastic cells are pale eosinophilic and the nuclei are elongated or oval. There is no cytological atypia or mitotic figures in the tumor. By immunohistochemistry stains, the tumor cells are usually positive for vimentin with variable expressions of muscle actin and alpha-smooth muscle actin (SMA), but negative for desmin, CD34, S100, and epithelial markers [1,5,10]. Electron microscopic studies have revealed a mixture of fibroblasts, myofibroblasts, and undifferentiated mesenchymal cells in the tumor [11].

The major differential diagnoses of dermatomyofibroma include dermatofibroma, neurofibroma, cutaneous leiomyoma, and hypertrophic scar. Dermatofibroma usually shows marked acanthosis and pseudoepitheliomatous hyperplasia of the overlying epidermis, and the tumor cells are negative for SMA. Neurofibroma contains spindle-shaped cells with fusiform or wavy comma-shaped nuclei and pale pink-blue cytoplasm, and the tumor cells are positive for S100 but negative for SMA. Cutaneous leiomyoma commonly presents as a well-circumscribed round subcutaneous nodule, and the tumor cells have pink cytoplasm and elongated nuclei with blunt ends and are positive for SMA. An unusual subtype of dermatomyofibroma, the hemorrhagic dermatomyofibroma

[12] may have abundant extravasated erythrocytes, scattered inflammatory cells, numerous capillaries, and sieve-and slit-like spaces, which can mimic a Kaposi's sarcoma, but the spindled tumor cells are negative for CD34 and HHV8.

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