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Myofibroblastic dermatofibroma: an unusual variant

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Abstract

Myofibroblastic dermatofibroma (MFD) is an unusual neoplasm characterized by a predominance of myofibroblastic differentiation. It is extremely rare and it is not well described. Expressions of smooth muscle actin, CD34, S-100, desmin, CD31, and Factor XIIIa were evaluated along with hematoxylin-eosin and trichrome staining of fixed tissue specimens from a fibrohistiocytic neoplasm. The neoplasm demonstrated a storiform pattern characteristic of fibrohistiocytic origin. It was strongly and diffusely positive for smooth muscle actin and vimentin, and negative for all other stains performed. A trichrome stain showed the entire tumor to be blue, demonstrating the collagenous and fibrous tissue to a marked degree. MFD is a distinct variant of dermatofibroma characterized by a predominance of myofibroblastic differentiation.

Myofibroblastic Dermatofibroma (MFD) is thought to be an unusual variant of dermatofibroma (1). It shares features of intranodal myofibroblastoma. All reported lesions show immunohistochemical staining for smooth-muscle actin and vimentin, supporting myofibroblastic differentiation. We described a 31-year-old male with MFD with review of the literature.

Case report

Clinical features

A 31-year-old man initially presented to the Dermatology Department of the University of Medicine and Dentistry of New Jersey with a six-month history of a pink pruritic nodule on the nose, which started as a "pimple". There was a history of occasional painless bleeding. He had a one-year history of diffuse scaling and erythema on the dorsum of the hands and lateral fingers. The family history was negative for similar conditions or skin disorders. The patient had no other significant medical history and no known drug allergies. Physical examination revealed a firm, pearly pink nodule measuring 0.5 x 0.5cm in diameter. The clinical impression was of a possible basal cell carcinoma or dermatofibroma. A shave biopsy was performed.

Histopathology

The epidermis was without significant pathology and no psuedoepitheliomatous hyperplasia was evident. There was a Grenz zone in the superficial dermis, between the epidermis and a nodular dermal neoplasm. This nodule showed a network of short, ovoid cells intimately intertwined with longer cells with tapered nuclei, suggestive of myofibroblastic differentiation (Figure 1). Areas of the tumor show a storiform pattern characteristic of a fibrohistiocytic lesion (Figures 2 and 3). Immunohistochemical stains, including smooth muscle actin, CD34, S-100, desmin, CD31, and Factor XIIIa were performed. The lesion was strongly and diffusely positive for smooth muscle actin and vimentin and negative for all other stains performed (Figure 4). A trichome stain was also performed and stained the entire lesion blue, demonstrating the collagenous/

fibrous nature of the tumor. The diagnosis of myofibroblastic dermatofibroma was rendered.

Discussion

Fibroblasts are activated fibrocytes that are similar in morphology to macrophages, Langerhans cells and other epithelioid cells and are immunohistochemically positive for vimentin (2). In contrast to fibroblasts, myofibroblasts display a number of heterogeneous cytoskeletal immunophenotypes and actin isoforms; some combinations include vimentin with alpha-smooth muscle actin, desmin, or both or neither, and myosin (3,4).

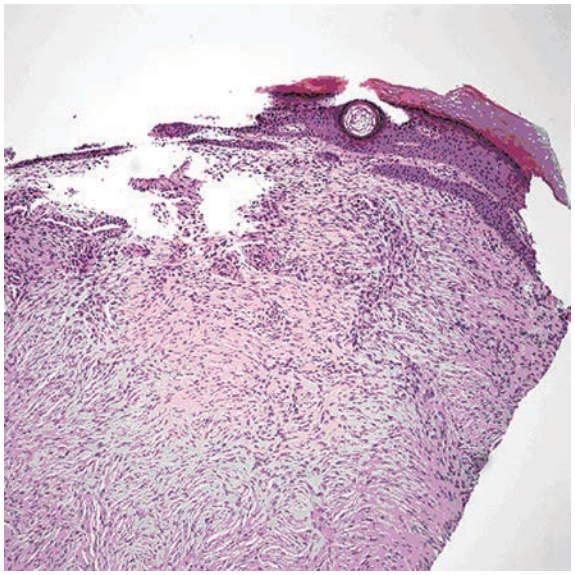

Figure 1. Low power view of the spindle cell dermal lesion (hematoxylin and eosin, x10).

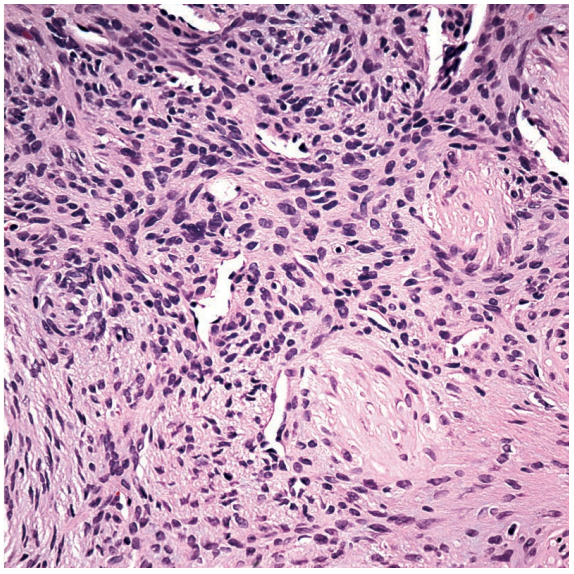

Figure 2. High power view of the tumor with smooth muscle appearing areas and a fascicular pattern (hematoxylin and eosin, x40).

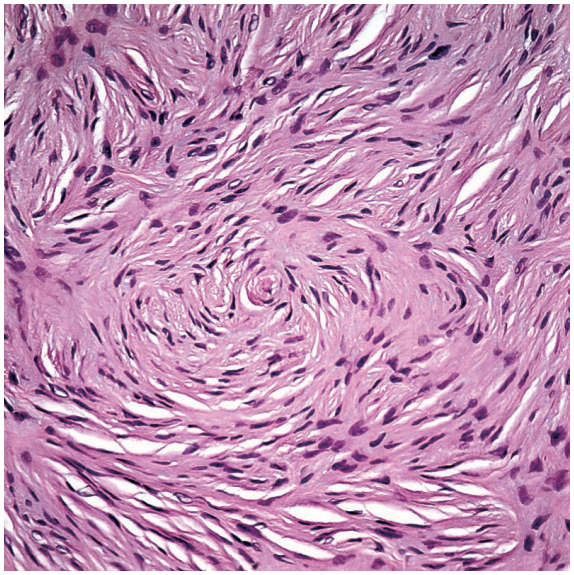

Figure 3. High power view of spindled areas of the lesion with storiform architecture (hematoxylin and eosin, x40).

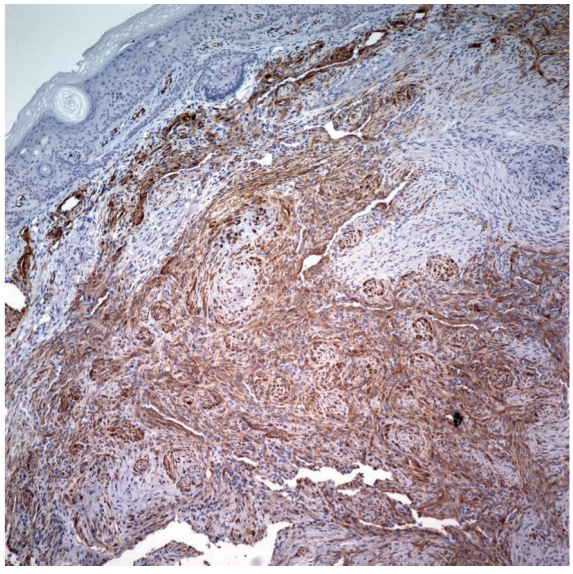

Figure 4. Tumor staining positive for smooth muscle actin, low power (smooth muscle actin, x10).

Fibroblastic tumors include histiocytic and fibrohistiocytic tumors; adipocytic and lipogenic tumors; and myofibroblastic tumors (5). Myofibroblastic tumors consist of modified fibroblasts that form the primary structure of multiple reactive and benign soft tissue lesions; the cells may be stellate or bipolar with nuclei elongated and tapered as seen in fibroblasts, or short, oval, and pale staining cells with distinct, punctuate nucleoli (4,6,7).

Dermatofibroma, or benign fibrous histiocytoma, is a fibrosing dermatosis with a significant number of dermal fibrocytes with variable number of histiocytes, lymphocytes, and multinucleated giant cells; and an overlying psuedoepitheliomatous hyperplasia is often present (2,8,9).

The epidermal hyperplasia may be so intense as to mimic basal cell carcinoma. It is unclear whether dermatofibroma is a neoplastic or reactive process (2,8,10).

MFD is a distinct variant of dermatofibroma characterized by a predominance of myofibroblastic differentiation (2,3). It is extremely rare and is not well described in the medical literature. Zelger and Zelger (1,11) described three cases in the neck and shoulder region of male adults. They described cells that were densely packed, slender and spindle shaped with strong immunoreactivity for vimentin, smooth muscle actin and CD57. Our case showed strong immunoreactivity for vimentin and smooth muscle actin, without staining for CD57, but showed all other characteristic histopathological features. The storiform architecture and tapered nuclei in some areas of the tumor argue against the diagnosis of leiomyoma.

MFD may be related to intranodal myofibroblastoma (1,12). Intranodal myofibroblastoma is a rare primary spindle cell tumor of lymph nodes, which is vimentin- and smooth muscle actin- positive, with proliferative spindle cells giving a striking histopathological resemblance to myofibroblastic dermatofibroma (12,13). MFD may also be mistaken for other spindle cell neoplasms such as dermatofibrosarcoma protuberans and can lead to unnecessary wide excisions and investigations (14).

In our patient, the lesion was on the left dorsum of the nose, a location not previously reported. Myofibroblasts occur most commonly in adults, in the periodontal ligament and around testicular seminiferous tubules; in addition, they are a major constituent of inflammatory and granulation tissue (4,13). They may not show predilection for a particular part of the body and more cases will need to be examined in order to develop definitive epidemiological data.

MFD is currently viewed as a benign tumor with limited clinical behavior (2). Appropriate investigations will prevent clinical mismanagement of this rather benign process.

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Miofibroblastni dermatofibrom: neuobičajena varijacija

Sažetak

Uvod: Miofibroblastni dermatofibrom (MFD) neobičan je tumor karakterističan po tome što kod njega preovladava miofibroblastna diferencijacija. Veoma je redak i nedovoljno opisan u literaturi.

Metode: Ispitivanje je obuhvatilo ekspresiju aktina glatkih mišića, CD34, S-100, dezmin, CD31 i faktor XIIIa, kao i histopatološke analize hematoksilin-eozin i trihromnim bojenjem fiksiranih uzoraka tkiva fibrohistiocitnih tumora.

Rezultati: Histopatološki, tumor se sastojao od vretenastih ćelija, karakterističnih za fibrohistiocitno poreklo. Imunohemijskom analizom dobijena je pozitivna reakcija na aktin i vimentin glatkih mišića, dok je na ostala bojenja reakcija bila negativna. Trihromnim bojenjem dobijena je potpuna obojenost tumora plavom bojom, što je do određene granice demonstriralo i kolagenozno i fibrozno tkivo.

Zaključak: MFD je posebna varijanta dermatofibroma sa karakterističnim prisustvom miofibroblastne diferencijacije.

