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Desmoplastic Fibroblastoma (Collagenous Fibroma)

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Case Presentation:

Patient: P.B. is a 55 year-old Caucasian female.

History of Present Illness: This patient presented for evaluation of an enlarging lesion on the 3rd digit of her right hand. The lesion first appeared approximately three years prior as a skin colored papule proximal to the nail fold. The lesion remained relatively quiescent and asymptomatic for two years. After this time, the patient self-treated the lesion with over-the-counter 17% topical salicylic acid for three weeks. The topical salicylic acid treatment was discontinued secondary to significant irritation and bleeding. Over the next 2-3 months the lesion rapidly enlarged, reaching its current size. The patient does not report any associated pain, pruritus, dysesthesias or symptoms associated with vascular compromise.

Medical History/Surgical History: Non-insulin dependent diabetes mellitus, condyloma acuminata, obesity, cesarean section (1990), dental surgery (2011)

Family History: Nonmelanoma skin cancer, diabetes mellitus

Medications: Metformin, vitamin E, B-complex

Previous Treatment: Salicylic acid 17% (topical)

Physical Examination: There is a large, firm, pink, pedunculated tumor measuring 4.5 x 4.5 cm emanating from the proximal nail fold on the 3rd digit of the right hand.

Studies: Patient refused MRI and X-ray imaging.

Biopsy: *Phoenixville Hospital Department of Pathology (S12-01093, 03/07/2012); subsequently sent for consultation to Pennsylvania Hospital. Right long finger: "On H&E sections the mass is sparsely populated by bland spindle cells with dense collagenization with focal myxoid areas. The overlying skin is unremarkable. The differential diagnosis includes collagenous fibroma and other sclerosing/desmoplastic lesions. Immunohistochemical stains are negative for CD34, S100, EMA and SMA. These immunohistochemical stains excluded perineurioma, sclerosing epithelioid fibrosarcoma, and desmoplastic melanoma. The combined findings are consistent with the diagnosis of collagenous fibroma (desmoplastic fibroblastoma)."*

Diagnosis: The combined findings are consistent with the diagnosis of **collagenous fibroma (desmoplastic fibroblastoma)**.



Figure 1a, Figure 1b — Large, firm, pink, pedunculated tumor measuring 4.5 x 4.5 cm emanating from the proximal nail fold on the 3rd digit of the right hand.

Figure 2a — Magnification 2x, **Figure 2b** — Magnification 10x, **Figure 2c** — Magnification 20x - The tumor is sparsely populated by bland spindle cells with dense collagenization with focal myxoid areas.

Figure 3 — Post-excision; central nail deformity with longitudinal ridging.

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Discussion:

Desmoplastic fibroblastoma (DF), also known as collagenous fibroma is a distinct yet uncommon benign fibrous soft tissue tumor that typically occurs in the subcutaneous tissue or skeletal muscle in adults. Since first described in 1995 by Evans, fewer than 100 cases have been reported in the English literature. Clinically, the lesion usually presents as a well circumscribed, firm, painless, slow growing mass of several months duration and can range greatly in size; up to 20 cm. It most commonly appears in males with a peak incidence in the fifth and sixth decade of life. Lesions can appear in a variety of locations with the most common sites being the forearm, shoulder, upper leg and feet.

Histopathologically, the tumor appears well defined with normal overlying epithelium. The tumor is composed of sparsely distributed stellate or spindled fibroblastic cells with bland cytologic features separated by abundant intercellular collagen fibers. Blood vessels, necrosis and mitotic figures are typically absent. Immunohistochemical stains are useful in diagnosis with the tumor cells showing a fibroblastic-myofibroblastic compatible profile. The cells are intensely positive for vimentin and show variable expression for SMA. Staining for desmin, S100, CD34 and EMA is typically negative.

The differential diagnosis of desmoplastic fibroblastoma includes entities both benign and malignant. These tumors include: nodular fasciitis, fibroma of tendon sheath, nuchal fibroma, sclerotic fibroma, neurofibroma, solitary fibrous tumor, fibromatosis and fibromyxoid sarcoma.

Interestingly, a few recent studies have found desmoplastic fibroblastomas to be associated with clonal chromosome aberrations, specifically rearrangement of chromosome 11q12. This finding further supports the original designation for this entity as a 'neoplasm' versus the possibility of a reactive process.

Adequate treatment of desmoplastic fibroblastomas is achieved with simple conservative excision, with no reported recurrences in the literature. Dermatologists and pathologists should beware of the existence of this tumor so that proper diagnosis and treatment can be employed, thereby avoiding unnecessarily aggressive surgery.

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