FIBROMA OF THE TENDON SHEATH – A RARE HAND TUMOR

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Fibroma of the tendon sheath (FTS) is a rare, benign, soft tissue lesion. Clinically, FTS presents similarly to the more common giant cell tumor of the tendon sheath. It is distinguished histologically by the lack of giant cells, foamy histiocytes and synovial cells.

We presented a case of FTS involving the common tendon sheath surrounding the flexor tendons leading to the third metacarpal. A 63-year-old man presented with a 3-month history of a painless mass in his right palm that had recently tripled in size. Examination demonstrated a 5x4 cm firm, nodular, superficial mass that was adherent to the overlying skin. Radiographs of the hand revealed a soft tissue mass without bony abnormality. Ultrasound demonstrated a solid, heterogeneous and hypoechoic mass and computed tomography demonstrated that the mass centered predominantly at the mid and distal portions of the third metacarpal. The patient underwent excisional biopsy of the lesion and a palmar, longitudinal incision was made from the wrist to the third metacarpal. Submitted histologic sections revealed a well-circumscribed lesion closely resembling hyalinized collagen. Neither vascular proliferations, necrosis, nor mitoses were observed. Similarly, multinucleated giant cells, pigment-laden macrophages, and inflammatory cells were also not identified. A diagnosis of FTS was rendered. We provided an additional rare case to the literature of a FTS and highlight the need to consider this entity in the differential diagnosis for any soft tissue lesion in the hand. Three months post surgery the patient demonstrated full range of motion of the hand.

Key words: fibroma, tendon sheath, FTS, soft tissue tumor, hand lesion

Fibroma of the tendon sheath (FTS) is a rare, benign, soft tissue lesion. It was first described in the literature by Buxton in 1923 (1), further defined by Geschickter and Cope- land in 1949 (2), and Chung and Enzinger in 1979 (3). The most recent series of data was reported by Millon et al in 1994 when they noted that of tumors seen in the hand, it represents 2-3% of lesions (4, 5). Clinically, FTS presents similarly to the more common giant cell tumor of the tendon sheath. It is distinguished histologically by the lack of giant cells, foamy histiocytes and synovial cells (6). This report describes a case of fibroma of the tendon sheath involving the common tendon sheath surrounding the flexor tendons leading to the third metacarpal.

CASE REPORT

A 63-year-old right hand dominant man presented with a 3-month history of a painless enlarging mass in his right palm. His main complaint was of pruritis overlying the lesion and that it had tripled in size in recent months. His medical history was significant for diabetes, gout and end stage renal disease requiring dialysis. There was no history of trauma or cancer. Examination demonstrated a 5x4 cm,
firm, nodular, superficial mass in the palm of the right hand that was adherent to the overlying skin (fig. 1). There was neither venous thrill nor Tinel’s sign, and there was a normal Allen’s test.

Radiographs of the hand revealed a soft tissue mass without bony abnormality (fig. 2). Ultrasound demonstrated a solid, heterogeneous and hypoechoic mass with slight internal vascularity.

Computed tomography of the mass was performed noting the mass centered predominantly at the mid and distal portions of the third metacarpal (fig. 3). It was isodense with the surrounding musculature and contained septations. The mass completely encased the flexor tendon group at the level of the third metacarpal, however the flexor tendons remained intact. Lastly, it extended distally into the web space between the third and fourth metacarpals. An MRI was not obtained due to claustrophobia.

Ultrasound guided fine needle aspiration of the lesion demonstrated dense fibrovascular connective tissue consistent with palmar fibromatosis. No cytologic atypia, granulomas, giant cells, or inflammatory cells were observed.

The patient underwent excisional biopsy of the lesion. A palmar, longitudinal incision was made, and the lesion was easily visualized (fig. 4). The mass was firm, white colored, irregularly shaped and well encapsulated. (fig. 5A). It was carefully dissected off of the flexor tendons and neurovascular structures leading to the third digit (fig. 5B).

Fig. 1. External appearance of the lesion

Fig. 2. PA radiograph shows soft tissue density overlying long finger metacarpal

Fig. 3. 2.5 mm collimated axial post contrast CT image with soft tissue windowing demonstrates isodense mass with thin septal areas of enhancement encompassing flexor tendons and displacing neurovascular bundles

Fig. 4. Initial visualization of the mass demonstrating its lobular composition
Gross and microscopic pathologic findings

The external surface of the mass was smooth and glistening without gross abnormality or disruption that measured 5.5 x 5 x 3 cm. Sectioning revealed a homogenous, smooth, white, minimally whorled cut surface without areas of myxoid change or necrosis.

Submitted histologic sections revealed a well-circumscribed lesion composed of virtually acellular, dense, eosinophilic, fibrous connective tissue that closely resembled hyalinized collagen (fig. 6 and 7). Within this collagen-like stroma were haphazardly spaced, unremarkable, spindle- and stellate-shaped cells (fig. 8) with amphophilic cytoplasm, elongated nuclei, fine nuclear chromatin, and small nucleoli.
Haphazardly arranged cleft-like spaces were identified around the periphery of the lesion. Neither vascular proliferations, necrosis, nor mitoses were observed. Similarly, multinucleated giant cells, pigment-laden macrophages, and inflammatory cells were also not identified. A diagnosis of fibroma of tendon sheath was rendered.

The postoperative course was uneventful. At his 3 month follow-up, the patient demonstrated full active range of motion of the hand with a well healed incision (fig. 10). Despite removal of the lesion he continued experiencing palmar pruritis.

DISCUSSION

Fibromas of tendon sheath are slow-growing, firm, benign tumors that occur throughout the body. As reported by Chung and Enzinger, 98% of these originate in the extremities with 81.8% of these occurring in the fingers, hands or wrists. Of their series, 21% or 29/138 occurred in the hand. They noted that the tumors were more likely to be located on the flexor surface of the hand as well as to be located in the right hand. There was a male predominance and the most likely period of incidence was between the third and fifth decades of life. Within the subset of hand cases they studied, 27 of the 29 cases involved lesions on the palmar side. They found a recurrence rate of 24% for all lesions involved in their study.

Millon et al. were the first to focus on FTS within the hand in 1994 (4). They found that of all hand tumors, FTS had an incidence of 3% in their practice with 0% recurrence after marginal excision. They related the lack of recurrence to more aggressive resection of lesions including occasionally removing neurovascular structures adhered to the tumors. An important point raised in their report was of the possibility that these lesions are caused from some form of inciting event or trauma to the region. They had one patient who had a fibroma of tendon sheath develop at the same pulley where she had previously had surgery for a trigger release. The rest of their patients, however, did not have reported trauma to the region. In our case the patient did not have any history of trauma to the region.

The ultrasound appearance of FTS has only been reported once. Bertolotto et al. described the lesion as a “solid, hypoechoic, flattened mass” (7). In this case the lesion’s ultrasound features were once again solid and hypoechoic, but also demonstrated some vascularity and heterogenous composition.

With the aforementioned features, giant cell tumor of the tendon sheath, synovial sarcoma and neurofibroma were considered in the radiologic differential diagnosis.

Given the patient’s history of gout and dialysis dependence, an atypical presentation of a gouty tophus or an amyloidoma was also considered. Gouty involvement of the hand may include the extensor tendons at the dorsal retinaculum (8), a localized painful mass in the mid-palm (9), and tophi over the dorsal aspect of the IP or MP joints (10, 11). Initial signs of flexor tendon involvement include marked pain, erythema, acute swelling, and...
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warmth suggestive of acute suppurative tenosynovitis (12). Deposition in the hand occurs relatively late in the disease and is uncommon with good medical management (8). Amyloidosis is a condition resulting from the deposition of low-molecular weight protein, B2 microglobulin, in the bones and soft tissues that commonly occurs in patients with renal failure who are undergoing dialysis (13). Hand involvement in amyloidosis is usually characterized by cystic lesions in the carpal bones and destructive arthropathy involving the wrists and IP joints (14, 15, 16). In patients with amyloidosis, tenosynovitis is common (13, 17). Although the possibility of gout and amyloidosis existed, histological findings were definitively negative for involvement by either a tophus or amyloid.

As with this case, most fibromas of the tendon sheath are fairly smooth and well-circumscribed with a multi-lobulated appearance. More often than not attachment to a tendon or tendon sheath is noted at the time of surgical resection. Sectioning usually reveals a uniform cut surface with an opaque to gray color. Areas of cyst formation and myxoid change are also common.

Classically, fibromas of the tendon sheath exhibit spindle- or stellate-shaped cells admixed diffusely within densely collagenized stroma. These cells are cytologically unremarkable with fusiform nuclei, smooth chromatin, and small nucleoli. The majority of lesions are hypocellular with variable cellularity present amongst different regions of the tumor. Areas of increased cellularity, arranged in patterns resembling nodular fasciitis, may also be present. Elongated cleft-like spaces lined by flattened epithelial cells resembling tenosynovial spaces, but which stain with von Willebrand factor (suggesting vascular etiology), are another common feature (18). Some lesions have been described which display nuclear atypia and pleomorphism and have been given the designation ‘pleomorphic fibroma of the tendon sheath’ (19).

Immunohistochemistry is not typically needed for confirmation of diagnosis, however, lesional cells express vimentin and muscle markers (muscle-specific actin and smooth muscle actin) and do not express desmin (20).

Treatment for FTS consists of marginal excision of the lesion. As noted by Millon et al, this sometimes requires removal of adjacent structures which may decrease function of the extremity post-surgically (4). This was not required for our patient as the mass was able to be resected from around the tendons and was not adherent. Another important distinction of these tumors is that they do not pose any risk of malignant degeneration (21). This may make conservative resection, with a 76% cure rate, preferred by some patients over more aggressive margins. Treatment by marginal excision allows for extremely low recurrence rates and excellent, rapid return of function to the affected extremity.

It is important to include FTS in the differential diagnosis for any soft tissue lesion in the hand. They are slow growing, benign lesions and are more likely to occur in the fingers, hand and wrist than anywhere in the body. The primary differential consideration is a giant cell tumor of the tendon sheath. Upon clinical, radiographical, and even gross pathologic examination, fibroma of the tendon sheath and giant cell tumor of the tendon sheath are difficult to distinguish from one another. Both arise in similar locations, possess similar radiographic signals, and have firm, well-circumscribed, multi-lobulated gray-white appearances (22). However, microscopically the typical fibroma of the tendon sheath is not a difficult diagnosis and differs significantly from giant cell tumor of the tendon sheath. The features of a paucicellular proliferation of collagenized tissue with scattered bland spindle cells is characteristic and not diagnostic of other entities (3). No atypical mitotic figures are present and necrosis is not identified. More cellular lesions may be confused with nodular fasciitis or fibrous histiocytoma (23). Giant cell tumor of the tendon sheath (top of the differential clinically) is a predominantly round mononuclear cell proliferation with multinucleated giant cells, hemosiderin-laden macrophages, and inflammatory cells set in a dense fibrous stroma. Mitoses are common and often readily apparent in stark contrast to fibroma of the tendon sheath.
REFERENCES


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