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Rare case of fibroma of the tendon sheath in a pediatric patient

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Abstract
Fibroma of tendon sheath (FTS) is a benign uncommon soft tissue tumor that presents as a solitary, firm, slow-growing painless nodule. This tumor arises from the synovium of the tendon sheath that occurs mostly around small joints such as the fingers, hands, and wrist. This soft tissue tumor rarely arises around large joints such as the knees, shoulders, elbows, and ankles with intra-articular or extra-articular involvement. It most commonly presents as a painless, slow-growing tumor with small nodular swelling and is often seen in adults 20-50. Our report reviews a unique case of fibroma of the tendon sheath in a pediatric patient. A 12-year old right-hand dominant female presented to the clinic with a mass located on the volar region of the proximal phalanx of the right long finger. This mass was present for 6 months and progressively increased in size. It caused intermittent pain, with prolonged activity. An ultrasound displayed a 1.3 cm mass around the proximal portion of the right long finger, which was consistent with a giant cell tumor versus a neuroma. Surgical dissection was performed and the mass was about 1 cm in diameter and emanated deep to the flexor tendons in the flexor sheath. When the patient followed up in the clinic postoperatively, there was no tenderness of the hand and mild tenderness over the volar portion of the right long finger. There was no palpable mass or angular deformity noted and the patient was neurovascularly intact. Clinical presentation, surgical management, recurrence, differential diagnosis, progression, and growth of the mass make this case unique. Typically, this is seen in the young and middle-aged demographic and reported in a 3:1 male to female ratio. The differential diagnosis for FTS includes giant cell tumor, mucinous cyst, lipoma, epidermal cyst and leiomyoma. Future studies should investigate other forms of management for Fibroma of the tendon sheath.

Keywords: Fibroma, tendon sheath, tumor, benign, pediatric, giant cell

Introduction
Fibroma of tendon sheath is a benign uncommon soft tissue tumor that presents as a solitary, firm, slow-growing painless nodule [1]. This tumor arises from the synovium of the tendon sheath that occurs mostly around small joints such as the fingers, hands, and wrist [2]. However, fibroma of tendon sheath (FTS) rarely arises around a large joint (knee, shoulder, elbow, and ankle) with intra-articular or extra-articular involvement. It most commonly presents as a painless, slow-growing tumor with small nodular swelling and is often seen in adults 20-50 [2]. The case reports which we reviewed examined the pediatric patients they ranged in age from 8-17.

In a case series of 138 patients, it was shown that only 20% of fibroma of tendon sheath cases were diagnosed in children and upper extremity involvement occurred in 86% of cases [1, 2]. When juvenile cases of FTS are identified, older children and adolescents are more commonly affected [2]. FTS is known to strongly adhere to the surrounding skin, tendon, tendon sheath, or neurovascular bundles, but that obvious adhesion may not be present in cases that are caught early in pediatric patients. Generally, the overlying skin is not adherent to the fibroma even if other surrounding tissues are [2]. Given the atypical findings on physical exam and imaging studies, FTS is commonly misdiagnosed as other benign soft tissue tumors or cysts. Histopathological analysis can be used to differentiate them. The clinical course of FTS in pediatric patients is usually uneventful and not associated with preceding trauma. In some cases, the palmar or digital fibromas may come to attention when interfering with sports or activities that require constant gripping [3]. The patients may complain of pain or reduced range of motion that is limited to these specific movements. The differential diagnosis of fibroma of tendon sheath includes epidermal cyst, mucinous cyst, neuroma, leiomyoma, nodular fasciitis, synovial sarcoma, and tenosynovial giant cell tumor.

The clinical features of the tenosynovial giant cell tumor are most similar to that of fibroma of the tendon sheath. Both have a high likelihood of occurring in the fingers with attachment to the tendon sheath and have similar MRI signals. Additionally, they have a firm, well-circumscribed, and multi lobulated gray-white appearance. The mainstay of treatment for fibroma of the tendon sheath is surgical excision. Surgical excision should be performed carefully as the recurrence rate is 24% in cases in the hands and fingers.

Local surgical excision with the preservation of tendons under general anesthesia is the procedure of choice in pediatric patients with FTS. Given the extremely low risk of malignant transformation or metastasis to distant sites, it has been suggested that the surrounding functional tissues remain intact when removing these masses [4]. This will help avoid any postoperative dysfunction. High recurrence rates have been associated with incomplete excision if the FTS is adhered to surrounding tendons, tendon sheaths, or nerves [4]. More aggressive approaches may be performed in adults to ensure complete excision. These methods may involve ligating digital arteries or excising palmar cutaneous nerves so they are generally avoided in pediatric cases. Postoperative courses tend to be uneventful with no major complications and those who participate in sports are typically able to return to play within the first few months [1, 3, 5]. Potential ways to reduce risk or local recurrence include performing a careful and complete resection with an air tourniquet and following up with these patients for a minimum of 2 years. This report provides a starting point for further research into the underlying significance of these abnormalities. FTS is a relatively uncommon tumor that typically presents as a painless, slow growing, firm nodule. In the case of a young pediatric patient age 12 within a 3-month time span of visiting the orthopedic physician and waiting for another appointment. Her range of motion decreased in her right hand and the swelling increased. It was confirmed with a few more tests that she needed surgery. With the proper care the FTS was removed. In her case and many other youth this procedure is not something you can prolong. It may pose significant anatomical and neurological complications if not treated in a timely manner.

Case Report

A 12-year old right-hand dominant female presented to the clinic with a mass located on the volar region of the proximal phalanx of the right long finger. This mass was present for 6 months. The patient reported that the mass was slightly increasing in size and caused intermittent pain, which occurred with prolonged activity. She had previously visited her primary care physician, who obtained radiographic images in addition to an ultrasound. The patient denied any history of trauma to the hand.

Physical examination findings revealed that the patient's skin was intact and no skin changes over the volar mass of the right ring finger. No significant tenderness was elicited when palpating over the mass around the volar aspect of the right long finger. There was full range of motion with the patient's left upper extremity. AP, lateral and oblique radiographs were obtained and reviewed, which demonstrated scalloping over the distal portion of the proximal phalanx. There was also soft tissue swelling which was noted in this area. The joint was well reduced and congruent. An ultrasound was ordered approximately two weeks later which displayed a 1.3 cm mass around the proximal portion of the right long finger, which was consistent with a giant cell tumor versus a neuroma. During the clinical encounter, a probable diagnosis was discussed with the patient and her mother with the aid of an interpreter. The orthopaedic surgeon explained that there was a mass around the right long finger, most likely in the region of the flexor tendon, most consistent with a giant cell tumor. Three months after the initial clinical encounter, the patient scheduled a follow-up appointment. It was noted that the mass has slightly enlarged and caused intermittent pain. During the physical examination, the patient lacked 25 degrees of full flexion. Gross sensation was intact and she was negative for epitrochlear, or axillary lymphadenopathy. The plan was discussed with the patient and her mother to remove the mass for definitive diagnosis. A surgical plan of care was performed, and the patient and informed consent was administered. The surgery was scheduled for a month from the informed consent.

On the date of the procedure, the Orthopaedic Surgeon counseled the family on the risks, benefits and alternative management options to the procedure. The specific risks that were mentioned included infection, bleeding, postoperative pain, as well as the major complication of paralysis and loss of major body function. Additionally, the possibility of future procedures was discussed. During the procedure, the patient was placed in a supine position, and after she was placed under general anesthesia, the right upper extremity was draped and prepped in a sterile fashion. The right upper extremity was then exsanguinated with an Esmach bandage. A mid-axial incision was made on the radial side of the finger. The skin and subcutaneous tissue was then incised, and the mass was immediately identified. Dissection also occurred around the fascial tissues and the attenuated flexor sheath. The mass a white, rubbery and glistening in appearance and was easily extruded, as there were no soft tissue attachments. The sample of the tissue was sent to Pathology for review. The mass was about 1 cm in diameter and emanated deep to the flexor tendons in the flexor sheath. Which caused limitation of motion due to the mechanical block. The incision was closed with 1 layer of absorbable suture. A digital block was injected in the base of the finger using 05. % Marcaine and sterile dressing was applied. The patient followed up in the clinic for the postoperative visit, eleven days after the procedure. On physical examination, there was no tenderness of the hand. However, there was mild tenderness over the volar portion of the right long finger. There was no palpable mass noted. In addition, the patient was neurovascularly intact. There was no angular deformity and there was proximal interphalangeal joint flexion to 100 degrees.

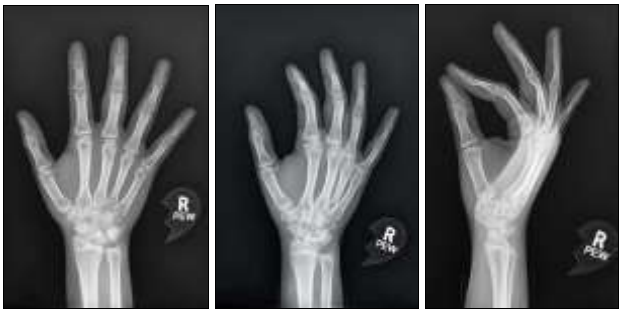

Fig 1: Preoperative anterior posterior (AP) and lateral view of the right hand displaying a fibroma of the tendon sheath of the proximal phalanx of the long finger

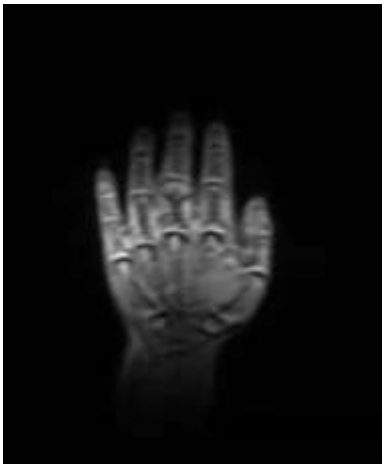

Fig 2: MRI of the right hand displays a fibroma of the tendon sheath in the proximal phalanx of the long finger

Discussion

Fibroma of the tendon sheath (FTS) is a relatively uncommon benign tumor that presents as a painless and slow-growing mass in the palm and digits. Typically this is seen in the young and middle-aged demographic and reported in a 3:1 male to female ratio [3]. The differential diagnosis for FTS includes giant cell tumor, mucinous cyst, lipoma, epidermal cyst and leiomyoma. Clinically, they most commonly behave like giant cell tumors of the tendon sheath. In this particular case, the differential diagnosis for the mass was a giant cell tumor versus a neuroma.

The differentials mentioned above can be diagnosed with the help of clinical history and examination, imaging and histopathologic features. Though tenosynovial giant cell tumors present similar to FTS, they differ in some aspects. A giant cell tumor is a benign proliferative and inflammatory lesion marked by multinucleated giant cells, lipid-laden macrophages, round stromal cells and hemosiderin, and it can arise from synovium of joints, bursae or tendon sheaths.[6] Giant cell tumor of the tendon sheath is the second most common benign tumor found in hand [7]. When local, it is most commonly found on fingers and wrist, while diffuse forms are found on large joints. Definitive diagnosis is made by histological features mentioned above or magnetic resonance imaging (MRI), which is sensitive and specific. On imaging, giant cell tumors differ from FTS in lesion morphology and intensity patterns. On proton density weighted imaging (PDWI), giant cell tumors are more likely to have uniform signaling and the low intensity pattern is mostly granular or separated, as opposed to stripe like pattern in fibroma of tendon sheath. In terms of morphology, FTS tends to be round or ovoid while giant cell tumors are lobulated or have a casting mold pattern [7]. Excision with or without radiotherapy is the treatment of choice with a recurrence rate of 45% [7].

The other differential diagnosis, a neuroma, is a tumor consisting of disorganized nerve tissue, macrophages and fibroblast forming in response to neuronal injury, which may be chronic irritation, crush injury, blunt trauma, laceration, pressure or stretch.[8] Clinical symptoms include pain, numbness, paresthesias and cold intolerance [8]. On histology, neuromas are significant for scar tissue, disorganized nerve fibers, regenerating fascicles in various stages of maturation and overlying skin or soft tissue regeneration. Conservative treatment includes physical therapy, desensitization and analgesics. If conservative methods fail after 6 months, surgical interventions, such as transposition of neuroma into muscle or vein, burying in bone, Gorkisch pinch, flap or tissue coverage, nerve stripping or re-repair of poorly repaired nerves may be considered [8].

The third differential diagnosis is a lipoma, which is a benign mobile tumor of adipose tissue located subcutaneously and encapsulated by fibrous tissue. It is typically painless and harmless, hence usually there is no need for excision, unless for cosmetic reasons [9]. Another benign soft tissue mass is a ganglion cyst, which is the most common soft tissue mass found on hands and wrist. They are fluid filled synovial cyst commonly found on the dorsal aspect of the wrist and communicate with a joint via a pedicle. Most are asymptomatic but may present with pain, tenderness, decreased range of motion or weakness. Though usually not indicated, MRI will reveal fluid fill mass with uniform intensity; on physical exam, a ganglion cyst will transilluminate. Ganglion cyst may be left alone, aspirated or surgically removed [9].

An epidermal cyst is a movable cutaneous cyst with a central punctum. Diagnosis is often clinical. On histology, the cyst wall is surrounded by stratified epithelium from the infundibulum of a hair follicle, while the cyst is filled with keratinous material [10]. Usually epidermal cysts are asymptomatic but may become inflamed. They may resolve spontaneously, but if indicated cyst may be excised [10].

This case is significant because of the rarity of fibroma of the tendon sheath and the demographic in which this case presented. Fibromas of the tendon sheath commonly present in 20 - 50 year old patients with a male to female ratio of 3:1 [3]. The patient in this case was an adolescent female. While the patient's mass was slow growing and on the volar aspect of the hand, it was significant for intermittent pain and decreased range of motion. While most fibromas of the tendon sheath are asymptomatic, about one third of cases present with tenderness and mild pain if underlying nerves are compressed [4]. As mentioned previously, fibroma of the tendon sheath is often misdiagnosed as a giant cell tumor, which was the initial diagnosis in this case. Histopathologic features usually lead to the definitive diagnosis. The treatment of choice in this case, surgical excision, is a common solution, with a recurrence rate of 25% [5]. Though this patient's postoperative course was uncomplicated, continued follow up would be necessary to note for recurrence. Future directions for research include a focus on different management and treatment modalities for Fibroma of the Tendon Sheath. It would be interesting to conduct a retrospective study of this condition and recurrence rates as well as resolution of symptomatology. It would also be valuable to research if management styles have changed over the years; based on effectiveness in extraction of this mass while keeping the complex neurovascular system intact.

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