GIANT CELL TUMORS OF TENDON SHEATH

Keith A. Glowacki, MD, and Arnold-Peter C. Weiss, MD

Giant cell tumor of tendon sheath is a well-recognized histopathologic entity. It is the second most common tumor of the hand, and although it can occur in other locations of the body, such occurrences are quite uncommon. Several authors have examined the clinical and histologic parameters of this tumor, and in fact there is no general agreement as to the nomenclature of the lesion. In addition, the cause of these lesions as well as the optimal treatment regime is poorly understood at the present time.

Jaffe has termed these lesions a localized pigmented villonodular synovitis based on the histologic similarity to the diffuse form of pigmented villonodular synovitis involving larger joints. He believed these to be variants of the same process with a localized form found more predominantly in the hand and upper extremity and the diffuse form seen more commonly as a monoarticular arthritis in the lower extremity. Anatomically, the lesion is less frequently directly associated with joints when it presents in the upper extremity and is always associated with the intracapsular region when presenting in the lower extremity.

Moore et al prefer the term “localized nodular tenosynovitis” to try to more accurately describe the clinical appearance of the tumor when it is noted in the hand. Other descriptive names that have been used to describe these lesions include fibrous xanthoma, xanthoma of the synovium, benign synovialoma, and sclerosing hemangioma. Certainly the lack of definitive nomenclature for this particular tumor demonstrates that the exact pathologic nature is unknown. This tumor is one that is frequently misdiagnosed and thought to represent a cyst. Commonly, it is only after presentation to and evaluation by a hand surgeon that the definitive diagnosis of this lesion is made. This scenario appears to be more due to the lack of widespread knowledge on this particular condition than inappropriate examination. Although surgical excision is the most common route, reported recurrence rates of 5% to 50% of primary excisions suggest that for several reasons the present surgical treatment is not always adequate.

CLINICAL ISSUES

Although giant cell tumor of tendon sheath may occur at any age, it is most common between the ages of 30 and 50 years with a peak incidence in the fifth decade. A 3:2 predominance is noted towards women. Typically the lesion occurs in the hands and fingers; it is second only to ganglions as the most common tumor of the hand. Giant cell tumors of the hand and forearm are discussed in detail elsewhere in this issue.
tumor of tendon sheath most commonly occurs adjacent to the distal interphalangeal (DIP) joint and has a propensity to occur in the index and long fingers (Figures 1A and B). The lesion is found along the volar aspect of the digit in approximately two thirds of patients who present with an easily palpable and definable lesion location. Other less common sites of involvement include the foot, ankle, knee and hip.4'6'11'15

On initial physical examination, the lesions are noted to be firm, lobulated, nontender masses firmly fixed to the deep tissues. The tumor may involve all the structures of the digit and include the tendon sheath, volar plate, capsular ligaments and joints. When they are located dorsally in the digits, they frequently involve the joint itself or the tendinous attachments to bone. The tumor is slow-growing, increasing in size only gradually for a long period of time, and can remain dormant as to size increase for several years at a time. It is not uncommon to see a tumor envelop the flexor or extensor tendons or even the neurovascular bundles. A large review of 115 cases demonstrates joint involvement by the tumor in only one fifth of all patients.11 The giant cell tumor often extends into the flexor sheath to the vinculae and, rarely, can erode the underlying bone or joint.

Cartilage invasion and cystic bone disruption have not been generally described with this lesion. These two changes are characteristically noted in pigmented villonodular synovitis involving the toes, knee and hip joints.6 The lesion does not appear to be related to trauma despite its common location at the DIP joint of the digit. A strong association between the presence of giant cell tumor and rheumatoid arthritis has been reported.10 No documented case of degeneration of a benign giant cell tumor of tendon sheath into a malignant form has been reported. Nevertheless, the lesion can be quite aggressive and behave in a low-grade malignant fashion: recurrence after surgical removal, often requiring several further procedures for eventual eradication, is reported in many patients.

On clinical presentation, patients frequently note nodular swelling in the region of the tumor. They commonly describe the lesion as slowly growing in size; very rarely, pain is associated with the growth. Numbness at the distal aspect of the finger is occasionally noted but if present is usually mild. Patients note occasional decreased range of motion, especially if the lesion is in a palmar location, and occasional snapping of unknown cause is also noted during digital flexion. Duration of symptoms prior to pre-
sentation for treatment ranges from several weeks to 30 years, with an average of approximately 2 years in most reported studies.\textsuperscript{3, 9, 11}

**GROSS AND HISTOLOGIC APPEARANCE**

Giant cell tumor of tendon sheath most frequently presents as a multilobular mass that is fairly well circumscribed and subcutaneous. The villous structure and deep brown hemosiderin pigmentation often associated with diffuse pigmented villonodular synovitis is seldom found in a localized multilobular form of giant cell tumor of tendon sheath.\textsuperscript{8, 10} The color of these lesions varies significantly but is generally a fairly bright yellow with areas of discoloration turning to a brown hue. Tumors vary in color from entirely greyish brown to entirely yellow-orange. The color of the lesions is affected by the degree of hemosiderin and collagen and the quantity of histiocytes present in the lesion.\textsuperscript{7} The nodules range in size from 0.5 to 5.0 cm with a variable degree of encapsulation from the surrounding tissue. Giant cell tumor of tendon sheath that occurs in association with large joints is more difficult to diagnose because lesion location is more frequently intra-articular and symptoms are generally nonspecific as far as localization is concerned. Ushijima et al. have recognized the clinical and pathologic differences between digital and large-joint forms of this tumor.\textsuperscript{14}

Giant cell tumor of tendon sheath in the digits generally present as relatively small, firm and regular-appearing lesions. Lesions elsewhere, including the feet, are generally large. Tumors associated with the digits are generally surrounded by a thin fibrous capsule with very little invasion of the capsule into the lesion itself. Those lesions found in larger joints are frequently covered by layers of synovial cells. The lesions when isolated in the digits do not appear to always have a defined association with the flexor or extensor tendon sheath. Nevertheless, there are frequently small "stalks" of the tumor that extend to the flexor tendon or extensor tendon sheath region, and often if the mass is relatively large, several little contiguous satellite lesions will be noted extending into the tendon sheath and synovium. These satellite lesions tend to be smaller in size than the main central lesion, which is usually located in an eccentric location.

On microscopic examination, the lesions consist of variable portions of collagenized stroma, hemosiderin pigmentation, multinucleated giant cells, and the characteristic polyhedral histiocyte (Fig. 2). Both diffuse and localized forms of this tumor contain actively proliferating histiocytes and the large multinucleated giant cells. The origin and differentiation of these tumors has been extensively debated; recent histochemical studies demonstrate that the mononuclear cells and the multinucleated giant cells represent osteoclasts and exhibit a phenotype consistent with the bone-marrow-derived monocytes and macrophages.\textsuperscript{1} Jaffe et al. have pointed out an evolution of these nodules from more cellular immature lesions to more advanced acellular lesions with a hyalinized stroma.\textsuperscript{6} A recent study by Abdul-Karim et al. compared the localized form of giant cell tumor of tendon sheath to a more diffuse form of this condition and pigmented villonodular synovitis.\textsuperscript{1} They concluded that all three lesions were essentially similar from a histopathologic basis but formed clinically distinct lesions. They proposed a spectrum ranging from a localized, benign form to a more destructive diffuse form. A measure termed the "proliferative index" may assist in distinguishing between aggressive and benign lesions. The diffuse form of this condition shows a more rapid proliferation and less controlled biologic behavior which should be considered when managing and counseling the patient about the possibility of recurrence.

On rare occasion, the histologic features may be confused with those of other soft tissue tumor such as synovial sarcoma, fibroma of tendon sheath and rhabdomyosarcoma. The nodular growth pattern, occasional presence of mitotic figures and the propensity for recurrence in certain lesions after inadequate removal should all suggest not an inflammatory process but a neoplastic one. The histologic pattern of the lesion and its size cannot necessarily be correlated with recurrence rate. Wright et al. stated that the more immature and cellular lesions demonstrated a higher recurrence rate.\textsuperscript{15} Recurrence rate is generally also associated with forms of the tumor that are more destructive to both soft and hard tissue.

**RADIOGRAPHIC APPEARANCE**

Plain radiographic examination demonstrates a typical lesion of only a soft tissue...
mass in 50% of the cases. The lesion itself does not appear to have a characteristic radiographic appearance. When the lesion is located in direct contact with the bony cortex, a pressure phenomenon on the cortex can be seen with some cases demonstrating indenting of the involved bone. No particular clinical diagnostic dilemma occurs with regards to benign versus malignant determination when the tumor appears in the hands and feet without bony erosion. The difficulty arises when radiographic changes are atypical. Fortunately this is quite uncommon. On rare occasion, however, a reaction may mimic changes seen with a periosteal chondroma. Intralesional calcifications may be present and mimic synovial chondromatosis, periosteal chondroma or calcific tendinitis. Radiographically, the differential diagnosis of digital lesions should include fibroma or chondroma of tendon sheath, synovial chondromatosis, synovial hemangioma, foreign body granuloma, chronic tophaceous gout and periosteal chondroma.

These lesions can also be evaluated by MR imaging. MR scanning using both T-1 and T-2 weighted images shows that giant cell tumors of tendon sheath have a signal intensity similar to that of pigmented villonodular synovitis. There is a decreased signal on both the T-1 and T-2 weighted images, which is an uncommon appearance for extra-articular soft tissue masses, particularly when they occur in the hands or feet. These findings may suggest the diagnosis of giant cell tumor of tendon sheath. Pigmented villonodular synovitis does show a higher tendency towards intralesional bleeding and greater deposition of hemosiderin often resulting in a more inhomogeneous appearance. Despite these findings, diagnosis is generally still made based on clinical examination grounds.

TREATMENT

The optimal treatment for giant cell tumor of tendon sheath is unknown. Based on patients who have had the lesions for a long time, it appears that growth is relatively slow, although progressive, in the vast majority. Patients frequently present with distal symptoms of neuropathy secondary to a compressive phenomenon or with concerns regarding the cosmetic appearance of the lesion itself. Rarely do functional consequences play a role in electing treatment but occasionally this is noted with finger flexion. Excision of the lesion is generally considered the treatment of choice; conservative management does not resolve the condition. Excision can be tedious because the presence of the mass is often within the flexor tendon sheath or the synovial joint. It is not uncommon that a partial excision of the sheath or joint capsule is required to ensure complete removal of the
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Although the lesion tends not to invade the soft tissues and can be shelled out, it tends to have frequent satellites which make careful dissection and exploration mandatory. Recurrence of giant cell tumor of tendon sheath is common despite meticulous removal; however, malignant transformation of the lesion has not been reported even after multiple recurrences. An association with the development of arthritis at the distal interphalangeal joint has been noted by Jones et al. Occasionally with removal of the tumor, debridement or fusion of the DIP joint is required in order to afford complete excision of the lesion itself. On rare occasion the skin is also involved in the lesion and is best taken with excision of the tumor, requiring secondary skin grafting.

OPERATIVE TECHNIQUE

In general,-operative approach to the lesion is directed towards the area of the major mass effect. For lesions along the volar aspect of the hand, a Brunner-type incision is most commonly performed, whereas lesions occurring in an eccentric fashion can often be excised using a mid-lateral approach (Fig. 3). Lesions that occur along the dorsal aspect of the digit are usually approached through a longitudinal incision, or if relatively localized, a transverse incision. Once the pseudocapsule is localized about the lesion, careful dissection in the soft tissues, taking care not to directly probe the lesion, is important to try to establish the boundaries of the tumor. As careful dissection of the surrounding tendons and soft tissues are undertaken, a Freer elevator or other blunt probe is useful in trying to manipulate the lesion without actually puncturing its surface to see if one can “tease” any satellites from underneath surrounding tendons or other structures. It is frequently possible to completely remove these satellites using this “teasing” technique rather than direct exposure of the entire site. A generous soft tissue pathway is required prior to trying to tease the lesion out from these nooks and crannies or the surgeon risks leaving small fragments behind. In general, it is wise to look around any adjacent tendons since satellite lesions are sometimes seen that do not appear to be contiguous with the main tumor. These are often fairly small in size, only several millimeters. For lesions exposed along the mid-lateral or volar aspect, careful dissection of the digital neurovascular structure should be undertaken prior to excision, and the tumor can completely surround this most important structure (Fig. 4).

For lesions that involve some bony invasion, careful curettage after teasing the tumor out of these areas is required. If any doubt is present, it is better to undertake an extensive debridement of the bone using a curette and rongeur to ensure that no tumor is left behind. On rare occasions, reconstruction of an extensor or flexor tendon may be required if the lesion has invaded these structures (Fig. 5A to F). This finding is more frequently seen secondary to attenuation due to a pressure phenomenon rather than aggressive invasion. The patient should be warned postoperatively that lesions that are noted to have both bony and tendinous invasion have a higher incidence of recurrence.

DISCUSSION

Giant cell tumor of tendon sheath, also known as localized nodular tenosynovitis, has long been included in the general category of pigmented villonodular synovitis. Both are considered benign growths of polygonal or round histiocyte-like cells associated with multinuclear giant, foam, and hemosiderin-laden cells. Most lesions produce one or more discrete soft-tissue nodules, usually along the tendon sheath or small joints of the fingers and toes. Pigmented villonodular synovitis is more commonly identified in a diffusely proliferate synovial membrane and has a darker pigmented appearance with or without nodular formation. Pigmented villonodular synovitis also more frequently involves the larger joints of the knee and hip rather than the digits and hand.

Giant cell tumor of tendon sheath is a benign condition and does not metastasize. Although there is a high incidence of joint involvement in lesions involving the digits of the hand, this is not universally the case. Various theories of pathogenesis and etiology have been proposed but none codified. These lesions are characterized by some as inflammatory in nature and by others as representing a neoplastic source. The key elements of treatment involve a generous surgical exposure in appropriate tissue planes to allow protection of the neurovascular bundles and flexor and extensor tendons. Careful exposure working more on the adjacent tissues
For volar lesions, a Brunner incision is classically used to allow access to both digital neurovascular bundles before excision of the tumor.

When giant cell tumor of tendon sheath is localized along the volar aspect of the distal finger, direct and circumferential involvement of both digital neurovascular structures is common. Vessel loops are placed around both digital neurovascular structures demonstrating the close proximity of these two structures to the tumor itself.
Figure 5. A lateral radiograph of the right thumb in this 32-year-old man, who has had recurrence of his tumor, shows cortical erosion at the base of the proximal phalanx dorsally (arrow) (A). During surgical exploration of this locally aggressive giant cell tumor of tendon sheath, the main lesion can be seen to extend both on the radial and ulnar aspects of the extensor tendon with attenuation of both the tendon and the sagittal hood (*, extensor tendon) (B). Because of the extensive involvement of the tumor in this particular patient, transection of the extensor tendon was required to gain exposure to all the satellite lesions as well as the bony intra-articular extensions (C). With the extensor tendon retracted distally, carefully piecemeal excision of the tumor down to its invasion of the bony cortex was accomplished.

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Figure 5 (Continued). (D). Careful curetage of all the small cavernous sinuses invaded by the tumor in the dorsal aspect of both the metacarpal and proximal phalanx was undertaken with removal of any tissue that appeared to be hemosiderin stained (E). A gross photograph of this locally aggressive giant cell tumor of tendon sheath shows the multiloculated contour of the tumor as well as the variations and hemosiderin deposition throughout the tumor itself (F).
than on the lesion itself, thereby avoiding puncture and possible seeding of the tumor cells to other soft-tissue structures, is vital. A method of "teasing" the lesion from its various interstices to identify and remove all satellite lesions is quite helpful in its overall excision. Recurrence rates in the reported literature range from 5% to 50%, but generally less than 10%.

References


Address reprint requests to
Arnold-Peter C. Weiss, MD
University Orthopedics, Inc.
Medical Office Center
2nd Floor Suite
2W Dudley Street
Providence, RI 02905