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Unravelling the Mystery and Taming the Giant - A Case of Tenosynovial Giant Cell Tumor

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ABSTRACT

INTRODUCTION: Tenosynovial giant cell tumors are classified into subtypes by growth pattern - localized-type vs diffuse-type and location - intra-articular vs extra-articular, they have a wide clinical spectrum effecting patients of all ages.
CASE REPORT: A 36Y old female presented with the complaints of swelling over left little finger, palm and distal end of forearm above the wrist.
ON EXAMINATION: Multiple swellings felt extending from medial aspect of left distal forearm, palm and little finger with nodular surface, non-tender and well-defined borders.
CE MRI: F/s/o likely tendon sheath tumor.
PROCEDURE: Tumor excision.
INTRAOPERATIVE FINDINGS: Tumor engulfed flexor digitorum superficialis and pro fundus tendons of little finger and extended to palmar aspect of hand below superficial palmar arch up to lumbricals.
CONCLUSION: Teno synovial giant cell tumors are a group of fibro histiocytic tumors, which are usually rare, locally aggressive, typically benign neoplasms arising from synovium of joints, bursae and tendon sheaths. Etiology is unknown. Surgery is the mainstay of treatment.

Keywords: Tenosynovial giant cell tumor, tendon sheath, tumor, excision.

1. INTRODUCTION

Tenosynovial giant cell tumors (TGCTs) are uncommon, generally benign neoplasms arising from the synovial lining of joints, bursae, and tendon sheaths. They are characterized histologically by a variable mixture of mononuclear cells, multinucleated osteoclast-like giant cells, macrophages, and hemosiderin deposition. Despite their benign histological nature, these tumors may demonstrate significant local aggressiveness, with infiltration of surrounding soft tissues, tendons, joint capsules, and occasionally adjacent bone.
TGCTs are broadly classified into localized and diffuse types according to their growth pattern. The localized type generally presents as a well-circumscribed nodular lesion and is frequently encountered in the tendon sheaths of the hand and foot. The diffuse type is characterized by more extensive involvement of the synovial lining and tends to have a more infiltrative behavior, greater functional morbidity, and a higher risk of recurrence. The term giant cell tumor of tendon sheath has traditionally been used for localized lesions involving the tendon sheath of the hand.
Clinically, TGCTs commonly present as a slowly enlarging, firm, painless or mildly painful swelling. Because of their nonspecific clinical presentation, lesions involving the hand may be mistaken for more common conditions such as ganglion cysts, fibroma, lipoma, or other benign soft-

tissue tumors. The diagnosis may therefore be delayed, particularly when the lesion is small or lacks characteristic clinical features. Magnetic resonance imaging (MRI) plays an important role in diagnosis and preoperative planning. It provides accurate delineation of the tumor and its relationship with the tendon sheath, flexor and extensor tendons, joints, neurovascular structures, and adjacent bone. The presence of hemosiderin typically produces low signal intensity on both T1- and T2-weighted sequences, with susceptibility or blooming on gradient-echo sequences, and may help differentiate TGCT from other soft-tissue tumors.

The pathogenesis of TGCT has been increasingly associated with abnormalities involving the colony-stimulating factor 1 (CSF1)/CSF1 receptor pathway. This has provided a molecular basis for the development of targeted systemic therapies, particularly for diffuse or unresectable disease. Nevertheless, surgical excision remains the principal treatment for localized and surgically resectable lesions.

Although TGCTs commonly occur in the hand, extensive lesions involving multiple flexor tendons with proximal extension from the little finger through the palm into the distal forearm are uncommon. Such lesions may pose considerable surgical challenges because complete excision may require sacrifice of involved tendons and joint structures, potentially resulting in significant functional impairment.

We report a case of an extensive tenosynovial giant cell tumor involving the flexor tendon sheath of the left little finger, with extensive involvement of the flexor digitorum superficialis and profundus tendons and proximal extension into the palm and distal forearm. The case highlights the diagnostic difficulty, locally aggressive behavior, and surgical challenges associated with extensive TGCT of the hand and forearm.

2. CASE REORT

2.1 Case history: A 36-yr old female, home maker, presented to OPD with history of swelling over left little finger, palm and distal end of forearm above the wrist.

2.2 Clinical examination:

General examination: The patient was conscious, oriented. Vital signs on presentation were within normal limits.

Local examination: Multiple swellings felt extending from the medial aspect of the left distal forearm, palm and little finger, with nodular surface, well defined borders, non-tender and firm in consistency along with a healed scar over the forearm from previous excision 6 months ago.

Patient was clinically diagnosed as ganglion cyst.

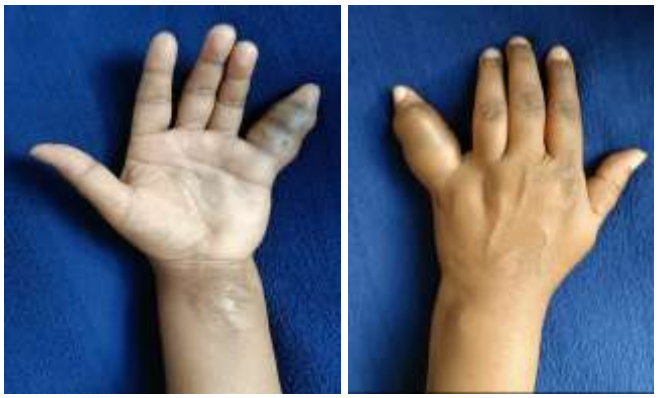


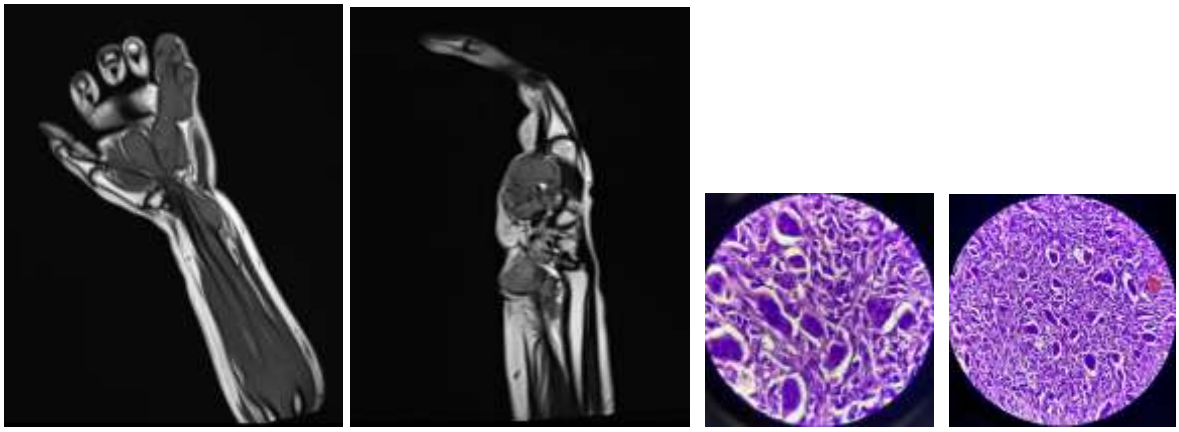
Figure 1 and 2: Clinical image of the tumor.

2.3 Investigations:

USG of left forearm and hand showed - Multiple ill-defined heterogenous lesions noted in the intramuscular plane along either side of flexor tendons in distal forearm, in flexor retinaculum involving all the muscles

within, along palmar and dorsal aspects of 5th digit, showing mild vascularity within -? plexiform neurofibromatosis/? neoplastic etiology.

CE MRI left forearm and hand showed - Large lobulated altered signal intensity mass lesion within the flexor compartment of the left distal forearm, hand and 5th digit involving the tendons with mass effects and bony erosions of 5th digit. Distal phalanx could not be made out completely.
F/s/o neoplastic etiology likely tendon sheath tumor - likely tenosynovial giant cell tumor.
FNAC showed - Histological features suggestive of Tenosynovial giant cell tumor - Tendon sheath.



Figures 3,4,5,6 show MRI findings and Histological features of the tumor.

2.4 Operative management:
A diagnosis of Tendon sheath Tenosynovial giant cell tumor was made and patient underwent tumor excision under general anesthesia along with K-wiring of metacarpophalangeal joint and interphalangeal joint of left little finger.
Intra-op findings: Tumor was found to engulf the tendons of flexor digitorum superficialis and profundus of the 5th finger extending to the palmar aspect of the hand below the superficial palmar arch up to the lumbrical muscles. Flexor digitorum superficialis was cut in the forearm since it could not be separated from the tumor. Flexor digitorum profundus was sacrificed from the A1 pulley distally. Tumor was adherent to the proximal and distal interphalangeal joint capsule; hence it was removed along with the palmar part of the capsule. K-wire fixation done to stabilize the metacarpophalangeal joint and interphalangeal joint of 5th finger.

Figures 7,8,9 and 10 show intra-op images, whole excised tumor and post-op image of the sutured hand and forearm with drain in-situ.

3. DISCUSSION

Tenosynovial giant cell tumors are uncommon soft-tissue neoplasms arising from synovial tissue and are most frequently encountered in the extremities. The hand and wrist are among the common sites of involvement, particularly for localized-type lesions arising from tendon sheaths. They usually present as a slowly growing, firm, well-circumscribed swelling and may remain asymptomatic for a considerable period. The clinical presentation, however, is nonspecific and may closely mimic other common lesions of the hand, particularly ganglion cysts.

In the present case, the patient was a 36-year-old woman who presented with multiple nodular swellings involving the left little finger, palm, and distal forearm. The presence of a previous surgical scar following excision six months earlier suggested recurrent disease. Despite the characteristic firm and nodular nature of the swellings, the initial clinical diagnosis was a ganglion cyst. This emphasizes the difficulty in establishing a clinical diagnosis of TGCT based solely on physical examination.

Imaging was particularly important in the present case because of the extensive anatomical involvement. Ultrasonography demonstrated multiple heterogeneous lesions along the flexor tendons with internal vascularity. The differential diagnosis included plexiform neurofibromatosis and other neoplastic lesions. Contrast-enhanced MRI subsequently demonstrated a large lobulated mass involving the flexor compartment of the distal forearm, hand, and fifth digit, with involvement of the flexor tendons and associated bony erosion. The extensive distribution of the lesion provided important information for operative planning.

MRI is considered the imaging modality of choice for defining the local extent of TGCT. The characteristic low signal intensity produced by hemosiderin deposition can be an important diagnostic clue. More importantly in extensive disease, MRI enables assessment of the relationship between the tumor and adjacent tendons, joints, neurovascular structures, and bone. In the present case, MRI demonstrated involvement extending beyond the expected confines of a localized tendon sheath lesion, thereby indicating the need for extensive surgical exploration.

The cytological findings in the present case were suggestive of TGCT. Histopathologically, TGCTs typically demonstrate mononuclear stromal cells, multinucleated giant cells, foamy macrophages, and hemosiderin deposition. Histopathological examination of the excised specimen remains important for definitive diagnosis and for excluding other giant-cell-rich lesions of the hand.

One of the important features of the present case was the extensive involvement of the flexor tendon apparatus. Intraoperatively, the tumor was found to engulf both the flexor digitorum superficialis and flexor digitorum profundus tendons of the fifth digit and to extend proximally into the palm below the superficial palmar arch and up to the lumbrical muscles. The lesion was also adherent to the proximal and distal interphalangeal joint capsules and was associated with bony erosion of the fifth digit. Such extensive involvement demonstrates that, despite its generally benign biological nature, TGCT can produce considerable local tissue destruction and functional compromise.

Complete surgical excision is considered the treatment of choice for resectable TGCT. The primary objective is complete removal of the tumor while preserving normal tendons, neurovascular structures, and joint function whenever feasible. However, extensive or recurrent disease may make preservation of involved structures impossible. In the present case, the flexor digitorum superficialis could not be separated from the tumor and therefore had to be divided. The flexor digitorum profundus was also sacrificed distal to the A1 pulley because of extensive tumor involvement. The involved portion of the

joint capsule was excised to achieve adequate tumor clearance.

Following extensive excision, stabilization of the involved joints was required, and K-wire fixation of the metacarpophalangeal and interphalangeal joints was performed. This highlights an important surgical consideration in extensive TGCT: achieving adequate tumor clearance may sometimes require sacrificing functionally important structures, and reconstruction or stabilization may consequently be necessary.

Recurrence is an important concern following treatment of TGCT. Localized lesions generally have a lower recurrence rate following complete excision, whereas diffuse-type lesions have a greater tendency for recurrence because of their infiltrative nature and difficulty in achieving complete resection. The previous history of excision in our patient followed by development of extensive lesions raises the possibility of residual or recurrent disease. Long-term follow-up is therefore essential, particularly in patients with extensive or recurrent tumors.

The molecular understanding of TGCT has also evolved considerably. Dysregulation of the CSF1/CSF1R signaling pathway plays an important role in tumor development and has led to the use of CSF1R-targeted therapies in selected patients with symptomatic, unresectable, or recurrent disease. These treatments may be particularly relevant when complete surgical excision would result in unacceptable functional morbidity. However, in surgically resectable disease, complete excision remains the preferred treatment.

The present case is noteworthy because of the unusual extent of tumor involvement, extending from the little finger through the palm into the distal forearm and involving both major flexor tendons. It also demonstrates the potential for TGCT to mimic more common benign lesions clinically and the importance of MRI in determining the true extent of disease. Extensive lesions of the hand and forearm require careful preoperative assessment and meticulous surgical planning, with particular attention to preservation of neurovascular structures and assessment of tendon and joint involvement.

Thus, this case emphasizes that TGCT should be included in the differential diagnosis of persistent, recurrent, or nodular swellings of the hand and wrist, particularly when they are closely related to tendon sheaths. Early recognition and appropriate imaging may facilitate complete excision before extensive involvement of functionally important structures occurs.

4. CONCLUSION

Tenosynovial giant cell tumor is a rare, benign but locally aggressive tumor that can occasionally present with extensive involvement of multiple tendon sheaths and adjacent structures. This case highlights the diagnostic challenge and surgical complexity of an extensive lesion involving the flexor tendons of the little finger with extension into the palm and distal forearm. MRI is valuable for defining tumor extent and surgical planning. Complete surgical excision remains the mainstay of treatment, with long-term follow-up essential because of the risk of recurrence.

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