Case Report

Extraarticular tenosynovial giant cell tumor of the ring finger: case report and literature review

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Abstract

Introduction: Tenosynovial Giant Cell Tumors (TSGCT) are common benign tumors, most commonly involving the hands and feet. Most common site of origin is from the flexor-extensor tendon sheath lining. However, it has been reported to arise extraarticularly from the joint capsule or manifest intraarticularly as pigmented villonodular synovitis. Here we report a case of an extraarticular TSGCT arising from the ring finger proximal interphalangeal joint capsule, sparing the tendon sheath without bone or joint involvement.

Case Report: Forty year old Caucasian female presented with a gradually progressive swelling over the left ring finger proximal interphalangeal joint, of 3 months duration, causing cosmetic deformity and functional debility. Radiographs showed a well defined homogenous radio-opaque lesion. Magnetic resonance imaging demonstrated an enhancing lesion arising from the joint capsule with intact flexor-extensor tendons. Fine needle aspiration cytology showed spindle cells, plasmacytoid cells and multinucleated giant cells (with 3-50 nuclei); confirming the diagnosis of Giant Cell Tumor. En Bloc resection of the tumor was performed through the stalk of the mass with closure of the capsular defect. At one year follow up, patient was painless with full range of motion without signs of recurrence of the disease, with no evidence of joint arthritis.

Conclusion: Correct assessment of the origin of the TSGCT directs appropriate surgical technique of tumor excision and soft tissue reconstruction. Tumors arising from the joint surface require meticulous capsular repair to prevent secondary joint seeding or joint degeneration. Simple en mass excision remains the gold standard of treatment of extraarticular TSGCT.

Keywords: Tenosynovial giant cell tumor; extraarticular tumor; tumor excision; benign hand tumor.
involvement.

Case Report
Forty year old, Caucasian female, presented with a gradually progressive, 1 x 1 cm, painless, firm, nonmobile, swelling over the left ring finger proximal interphalangeal joint, of three months duration. The patient complained of cosmetic deformity and interference with activities of daily living. The patient gave no history of any similar swelling in the past or in any other region of the body, without any systemic complaints. The differential diagnoses based on clinical examination included giant cell tumor of tendon sheath, ganglion cyst, fibroma of tendon sheath and the rare possibility of a synovial sarcoma.

Radiograph (Fig. 1) showed a well defined homogenous radio-opaque lesion overlying the volar aspect of the proximal interphalangeal joint of the ring finger. There was no evidence of adjacent joint erosion or degeneration. Magnetic resonance imaging (MRI) (Fig. 2) showed a well defined, 11 x 8 x 8 mm homogenously enhancing T1 and T2 hypointense, STIR hyperintense lesion with no flow voids within, without any intraarticular extension with normal adjacent bony cortices.

En Bloc resection of the tumor mass was performed, through a Littler’s Zigzag incision over the volar aspect of the PIP joint (Fig. 3). The stalk of the mass was delineated, traced up to its origin onto the joint capsule, separated from the surrounding structures and removed en mass (Fig. 4). The contiguity of the flexor tendon sheath was ensured and the rent in the capsular sheath was sutured to close the defect. Active range of motion of the finger was encouraged postoperatively. At one year follow up patient was painless with full range of motion and no signs of recurrence of the disease.

Discussion
Tenosynovial Giant Cell Tumor (TSGCT) was first described in 1852 by Chassaignac who called it, “malignant tumor of the tendon sheaths”[6]. The patient was a 40 year old female. TSGCT’s occur in all age groups, with peak incidence in the third to fifth decades[6]. The tumor has a predilection for the older age group with a female preponderance[4,7]. Women are affected twice as commonly as men[8]. However as per Cassier PA et al[9], there is no sex predilection in intra-articular disease, and a slight female predominance in extra-articular disease. TSGCT’s are most commonly located on the palmar aspect of the fingers of the hands, involving the first three fingers, with the index being most often involved[7]. Other rare sites of presentation are wrists, ankle and knees[8,10]. The tumor classically presents as a solitary, slow growing, lobulated mass. Most cases are asymptomatic, others may rarely present with pain, decreased mobility, triggering of the affected digit, and rarely numbness in the fingertip[10]. Lesions involving the toes or knees are more likely to be symptomatic[11]. Common differential diagnoses would include tendinous xanthomas, ganglion cyst, synovial chondromatosis, synovial hemangioma, epithelioid sarcomas and fibroma of tendon sheath[1,6,12]. Tenosynovial GCT is broad term which encompasses all the tumors arising from the tendon sheath, the bursae and from the synovial lining of the joint space. TSGCT can be classified based on the site of origin as (Table 1): Extraarticular (arising from
Tendon sheath / Bursa) and Intraarticular (also known as Pigmented Villonodular Synovitis (PVNS)[13]. Extraarticular TSGCT can be further divided into Localised GCT and Diffuse-type GCT[13]. Extraarticular Localised GCT is the most common type[14,15]. Al-Qattan outlined an alternate classification of TSGCT’s based on their operative appearance[16]: Type 1 included tumors enveloped in a pseudocapsule, further subclassified depending on capsule thickness and multilobulation; Type 2 tumors were not encapsulated and were subclassified by the presence of satellite, diffuse-type, or multicentric lesions. All recurrences in their series were in type 2 lesions[16]. This classification serves an important guideline while deciding the plan of treatment, as Type 1 tumors can be managed by excision alone (as evidenced in our case) while Type 2 tumors would need adjuvant therapy to prevent recurrence. Histologically, TSGCT lesion is lobulated and circumscribed, pseudoencapsulated in a collagenous stroma, with a medley of different cell types: eosinophilic round cells, osteoclast-like multinucleate giant cells, xanthoma cells, histiocytes, lymphocytes with hemosiderin deposition[5,8]. Most tumors appear to be clonal, neoplastic proliferations driven by Colony Stimulating Factor-1 (CSF1) production of the neoplastic cells[5]. However, the neoplastic cells constitute a minor component within the tumor, accounting for only 2- 16% of the cells[5]. Most cells are non-neoplastic, inflammatory cells recruited and activated by CSF1 produced by neoplastic cells, a phenomenon called tumor landscaping[5]. CSF1 Receptor (CSF1R), which is highly expressed in TSGCT, is a group III receptor tyrosine kinase that shows structural homology to KIT[5]. Radiographs demonstrate a well circumscribed soft tissue shadow, only rarely accompanied by degenerative changes in the adjacent joint or osseous damage due pressure necrosis. Ultrasound demonstrates a homogeneous, hypo- or hyperechoic solid lesion or, rarely, a heterogeneous lesion, with increased vascularity on doppler study, and in relation to the tendon sheath, with which it does not move[10,17,18]. MRI is considered to be the most accurate imaging test for diagnosis[17,18,19]. The mass shows intermediate signal intensity on T1 and T2 images, high intensity on short tau inversion recovery (STIR) or T2 with fat suppression images with post-contrast enhancement. The treatment of Extraarticular Localized GCT is primarily complete en mass excision of the tumor with clear margins under direct vision, with adequate soft tissue reconstruction[7]. Radiation therapy has been reported to be advocated as an adjuvant measure in cases with high risk or recurrence like high mitotic rate, bone involvement or incomplete resection[4,7]. A lower rate of recurrence should be expected when magnifying glasses or microscope, are used at the time of mass resection[5]. Imatinib mesylate, which has been shown to inhibit CSF1R tyrosine kinase, has been found to be effective in the treatment of locally advanced and metastatic TSGCT and PVNS as an adjuvant to surgical treatment[3]. Emactuzumab, a monoclonal antibody that inhibits CSF1R activation, has been shown to be effective in treatment of locally advanced TSGCT[9]. Recurrence rate ranging from 7 – 45 % has been reported[14,15,16,20]. As per Williams et al.[21] extension of the primary into the extensor tendons, flexor tendons and joint capsule is associated with recurrence. Other factors which have been reported to be associated with a higher incidence of recurrence include intraosseous invasion[22,23] and degenerative joint disease[14]. Recurrences are treated with re-excision along with use of adjuvant treatment if deemed necessary.

**Table 1:** Classification of Tenosynovial Giant Cell Tumors [13]

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<thead>
<tr>
<th>Site of Origin</th>
<th>Localized</th>
<th>Diffuse</th>
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<tbody>
<tr>
<td>Extra-articular (Tendon Sheath / Bursa)</td>
<td>GCT of Tendon Sheath</td>
<td>Diffuse-type GCT</td>
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<tr>
<td>Intra-articular</td>
<td>Localized PVNS</td>
<td>Conventional PVNS</td>
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**Figure 4.** Surgical steps of en mass excision 4a Flexor tendon sheath identified, mass delineated. 4b Stalk of the mass dissected off the proximal interphalangeal joint capsule. 4c Capsular continuity restored and tendon sheath contiguity ensured. 4d Mass measuring 13 x 13 mm excised.
References


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