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Case Report

# Tenosynovial Giant Cell Tumour of the Knee: An Approach to Treatment

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## Abstract

**Background:** Tenosynovial giant cell tumor (TSGCT) is a rare, benign tumour arising from joint synovia, bursae, and tendon sheaths. The majority of cases are localized, presenting with a localized lesion of the synovium, with only 10% of presentations being of a diffuse type synovial involvement.

**Case Report:** We present our case of 41 years old female that presented with right knee symptoms, namely instability, and locking associated with a right thigh mass of 4 months duration. Examination of the right distal thigh showed a large mass. The mass was firm and irregular, mobile with mild tenderness on deep palpation. Post-subtotal surgical resection diagnosed diffuse TSGCT of the right knee that required adjunct synoviothesis. At the last follow-up, she had improved pain, and range of movement with mild residual swelling and she was still awaiting adjunct synovial ablation.

**Conclusion:** TSGCT is a rare benign heterogeneous tumour. The pathological type at presentation guides the viable surgical treatment option. The localized type is usually addressed with arthroscopic synovectomy while the diffuse types requires combined approaches similar to our case of discussion. Biologics and novel therapies may hold potential for future cases.

***Keywords:*** *Case Report; Tenosynovial Giant Cell Tumor; Knee; Tumor; Classification*

## Introduction

Tenosynovial giant cell tumor (TSGCT) is a rare, benign tumour arising from joint synovia, bursae, and tendon sheaths [1,2]. The worldwide incidence is noted at 10/million persons/year for the localized type TSGCT and 4/million persons/year for the diffuse TSGCT (dTSGCT) type [3]. Approximately 90% of TSGCT are localized, characterized by a single lesion that is well circumscribed on the affected synovium. The residual 10% of TSGCT is diffuse and can affect one compartment or the entire synovium of a given joint. Due to its rarity and benign nature, clinical evidence and reporting are sparse, with only a few case studies focusing on uncommon tumour sites and experimental treatment options for TSGCT. Our case report discusses the presentation of TSGCT of the knee highlighting investigations and treatment options for the knee joint.

## Case Report

Ms. S.Z, a 41 years old female presented initially to the sports clinic at our hospital for right knee symptoms, namely instability, and locking associated with a right thigh mass for a duration of 4 months, with recent pain developing in the mass. She denied any significant medical or surgical history. The physical examination didn't reveal any obvious stigmata of chronic illness. Examination of the right thigh

revealed a 30 x 20 x 20 cm mass around the right distal thigh. The mass was firm and irregular, mobile with mild tenderness on deep palpation. However, the mass was non-pulsatile but associated with a massive effusion. The patient's distal neurovascular status was intact at the assessment. The laboratory work-up was also normal. Radiographs and an MRI scan are shown in figure 1a-1c. The x-rays show an increased soft tissue shadow while figure 1c shows a large supra-patellar pouch soft tissue lesion adherent to the femur with an associated massive effusion.

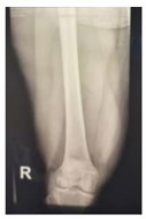


Figure 1a

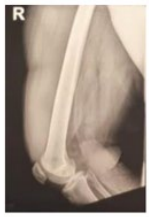


Figure 1b

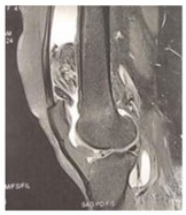

Figure 1c

A clinical diagnosis of pigmented villonodular synovitis of the right knee was entertained due to the benign nature of the clinical presentation and the radiological appearance. The diagnosis was discussed with the patient and an agreement was made for a sub-total synovectomy of the right knee with possible residual synovial ablation (synoviothesis) by the nuclear medicine department to minimize the risks of recurrence. Intra-operative images (Figure 2a-2c) show brown (haemosiderin) fluid, a large lobulated brown fleshy mass during excision, and the whole mass post-excision with its associated excised synovium.

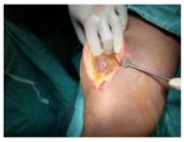

Figure 2a

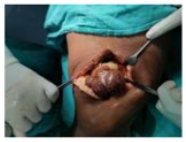

Figure 2b

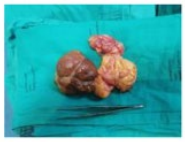

Figure 2c

The histopathologist's report diagnosed a Tenosynovial giant cell tumour of the knee, a rare benign tumour on synovial tissue. At write-up, the patient was still awaiting synovial ablation with the nuclear medicine department. However, her initial knee-related symptoms had resolved, and the surgical wounds had fully healed with slight residual swelling to report.

## Discussion

Tenosynovial giant cell tumor (TSGCT) is a rare, benign tumour of joint synovia, bursae, and tendon sheaths [1]. The worldwide incidence is noted as 10/million persons/years for the localized type and 4/ million persons/years for the diffuse type [3]. Approximately 90% of TSGCT are localized, being characterized by a single well-circumscribed lesion of the affected synovium. The residual 10% of diffuse cases variably affect one whole compartment or the entire synovium of a given joint. The latter presents mainly in large joints (> 5 cm in size) with the knee joint carrying the largest burden of presentation [2]. Gender predilection is towards female patients. The median age at presentation is 47, with the bulk of patients being primarily treated with open surgery [3]. Our case is that of a 41 years old female patient that also necessitated open sub-total resection. TSGCT is rare and not life-threatening, clinical evidence and research is sparse and most previous studies focused on uncommon tumour location as case reports and/or on the various treatment options for TSGCT

making the treatment of the knee TSGCT controversial [2,7]. Pathologically, TSGCT is generally a monoarticular inflammatory disease that can be characterized by joint bleeding, history, and examination usually including trivial trauma, discomfort repeated joint swelling and restricted range of motion. All latter symptoms can mimic a meniscal tear or haemophilic bleed in male patients [4]. Our case was initially presented to the Sports unit for meniscal tear-type symptoms. TSGCT has been recently described by immunologists as an intermediary between rheumatoid arthritis (RA) and synovial sarcoma, with clinical manifestations (joint swelling, destruction), histopathological lesions (synovitis, hyperplasia, joint cartilage destruction) and chromosomal aberrations (trisomy 5/7, p53 mutations) being cited for that proposal. Histological analysis typically reveals hemosiderin deposition and a predominance of $CD68^{+}$ cells [7].

### Investigation

Radiographic imaging usually reveals soft tissue swelling, bony erosions, and some periosteal reactions [5]. However, MRI is the gold-standard imaging modality, assisting with diagnosis, staging, and follow-up evaluation. Localized forms tend to be well-circumscribed lesions demarcated with a low signal intensity capsule due to fibrosis or haemosiderin [5]. Sizable lesions, as in our case, show heterogeneous soft tissue lesions. Macroscopically, both types are similar and either can present pedunculated, in most cases, however sessile or adherent forms are a possibility as was the case with our patient. Lesional dimensions can vary with an average lesion measuring 13 cm, our lesion was 20 cm at resection [4].

### Treatment

A treatment-directed classification system is advocated of late. TSGCT of the knee has variable presentation types which guide surgical management. One of three, either a Type 1-localized, Type 2-diffuse intra-articular, or Type 3 diffuse across the knee synovium is a possibility, our case was classified as a Type 2 [6]. All the types are further sub-categorized into two types: subtype1a - Localized intra-articular and subtype 1b - localized extra-articular, arthroscopic resection being recommended for the former and open resection for the latter, Type 2 lesions; subtype 2a - diffuse with normal bone is also treated with open visual resection, this was our treatment of choice for our case [6]. Subtype 2b has bone destruction that necessitates resection and arthroplasty reconstruction, while the dreaded Type 3 and subtypes requiring resection and adjuvant therapy and/or amputation for a subset of patients [6]. Some authors have strongly recommended a combined approach to minimize recurrence in diffuse lesions with the anterior compartment treated arthroscopically and the posterior compartment by open surgery. However, posterior to the knee is a technically challenging surgery and prone to complications. Radio-synovectomy ablation with radio-isotopes can be used as an adjunct to aid synovial ablation while radiotherapy application is now obsolete [7]. With TSGCT being described as an intermediary between RA and synovial sarcoma as described above has led to the development and research of novel systemic targeted therapies such as monoclonal antibodies or tyrosine kinase inhibitors [7]. Furthermore, other molecules that target synoviocytes are being studied for potential applicability [7]. To the latter intraarticular zinc and cadmium, both with their pro-apoptotic properties could decrease inflammation and limit joint synovial destruction minus the harmful toxicity of other treatments, more research will be required further [9]. PUMA, a pro-apoptotic gene combined with a new adenovirus-baculovirus complex vector has shown promising results in inducing synoviocytes inflammation reduction and destruction in laboratory studies, but safety for such therapies for human prescription still need validation [10].

## Conclusion

TSGCT is a rare benign tumour that presents in a heterogeneous manner. The pathological classification type at presentation dictates the surgical treatment option for that individual patient. The common (localized) type is usually addressed with arthroscopic synovectomy while the diffuse types are likely to benefit from combined approaches and the need for adjuvant synovial ablation to minimize complications and recurrence should be a standard for those affected by the latter. Biologics and novel have the potential application provided they satisfy testing and safety regulations.

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