Extra-Articular Localized Tenosynovial Giant Cell Tumor of the Deep Infrapatellar Bursa

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ABSTRACT

Tenosynovial giant cell tumor (TGCT), previously called pigmented villonodular tenosynovitis (PVNS) or giant cell tumor of tendon sheath, is a rare mesenchymal neoplasm arising from the synovium of joints and tendon sheaths. This condition has insidious and nonspecific symptoms making its diagnosis challenging. In most cases, localized tenosynovial giant cell tumors are described in the hand and are infrequent in larger joints. In the rare cases that affect the knee, the most common location is Hoffa's fat pad, followed by the suprapatellar bursa and the posterior capsule. Here, we describe a case of a histopathologically proven TGCT of the knee, found in an unusual location in the deep infrapatellar bursa, which was diagnosed by magnetic resonance imaging.

CASE REPORT

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A 17-year-old female presented with left knee pain. The patient reported intermittent knee pain and swelling for months prior which was worse with physical activity. She underwent formal physical therapy, activity modification, and bracing without significant improvement in symptoms. Her medical history was otherwise unremarkable. On physical examination, there was point tenderness at the distal aspect of the medial patellar tendon. There was trace effusion as well as some focal swelling around the medial aspect of the patellar tendon and the tibial tubercle, without surrounding warmth. The patient had normal strength and a normal range of motion (0° to 140°). The musculoskeletal examination was otherwise unremarkable.

The radiographic findings of the left knee were normal, without a visible soft-tissue mass, bone erosion, fracture, dislocation, or effusion. In addition, no calcifications were present in the deep infrapatellar bursa. A magnetic resonance imaging (MRI) study without contrast of the left knee revealed prominent fluid in the deep infrapatellar bursa which also contained nodular isointense lesion on T2 fat saturation images and low intense on T1 images. No gradient echo sequences were available. Based on these findings, a diagnosis of a loose body and localized tenosynovial giant cell tumor were favored, and operative excision was recommended.

The knee was approached via a medial parapatellar arthrotomy and a sub-patellar tendon bursectomy was performed. The lesion was identified and was rubbery in appearance, possibly consistent with LTGCT. It was sent to pathology for examination. The wound was copiously irrigated and closed in layers.

The rubbery mass sent for routine histopathological examination was 1.8 x 1.2 cm. Histologic examination at 50 power magnification revealed papillary synovial proliferation with multinucleated giant cells in a background of admixed histiocytic-like and fibroblastic-like cells. On higher-power www.RadiologyCases.com

examination (200 magnification), it was found that the tumor contained foci of multinucleated giant cells, mononuclear histiocytes, fibroblastic-like cells, hemosiderin-laden macrophages, and xanthoma cells (foamy macrophages), which are typical characteristics of L-TGCT. Routine, anaerobic, and fungal cultures revealed no growth.

In the intervening months of follow-up up to 1 year, the patient had no complications, with a pain-free and full return of knee and lower-extremity function and no clinical evidence of recurrence.

DISCUSSION

Tenosynovial giant cell tumors (TGCT's) are rare, locally aggressive, and typical benign neoplasms that derive from the synovium of joints, bursae and tendon sheaths [1]. This condition is typically diagnosed in individuals between the ages of 40 and 60, with a higher prevalence among females [2,3]. TGCTs can be classified according to their site (intraand extra-articular) and growth pattern (localized and diffuse). There is no clear histological distinction between both subtypes, and diagnosis is based on radiological features and clinical presentation [1,4].

Localized TGCT's typically present as small, solitary, pedunculated and well circumscribed lesions, and the diffuse type are more aggressive and infiltrative lesions [1]. L-TGCT's occurs in an extraarticular location in 90% of cases, involving tendon sheaths of the volar aspect of fingers (85%), followed by foot and knee (15%) [5]. Localized TGCT's in the knee joint have been previously reported in the literature at the meniscocapsular junction, intercondylar notch, anterior tibial eminence, or lateral recesse [2,6].

This study's objective is to present a rare case of localized TGCT, offering novel perspectives on distinguishing lesions occurring on the deep infrapatellar compartment of the knee joint. The differential diagnosis includes bursitis with synovial tissue proliferation, synovial chondromatosis, hemosiderotic synovitis, lipoma arborescens, and ganglia [7]. Nonspecific and insidious symptoms make the diagnosis challenging, especially in less affected joints such as the knee. Therefore, the diagnosis can only be confirmed by histopathological examination. Although a benign pathology, surgical resection is the treatment of choice due to its aggressive nature [2,8]. To our knowledge, this is the second report of arthroscopic resection of LTGCT in the deep infrapatellar bursa and the third report of such location [7,9].

In localized TGCT's, the symptoms depend on the location of the mass, ranging from mechanical blockage to diffuse pain. Howie et al. reported a series of asymptomatic cases until pedicle torsion [10,11]. Our patient presented with a lesion in the deep infrapatellar bursa of the knee, being asymptomatic until the occurrence of a knee sprain. This description can indicate a traumatic trigger, like the pathology described by Howie et al. [10]. Conventional radiography does not establish the diagnosis; however, it should be obtained to rule out calcifications, which are rarely seen in TGCT's but may be found in other potential diagnoses. Conventional radiography/ CT are usually normal or show a dense soft tissue nodule related to iron content. Osseous pressure erosion may be seen in tight joints. Ultrasound shows a well-circumscribed focal mass with heterogeneous echogenicity and increased Doppler signal.

The MRI findings for localized TGCTs usually reveal the presence of a nodule, with contrast increasing the sensitivity of the exam. MRI is also helpful in localizing the mass and evaluating for other intra-articular lesions

LTGCT's can be managed by complete marginal resection, with low recurrence. Contamination of the joint in intralesional resections is rarely reported, but a large multicenter study of L-TGCT showed a lower recurrence rate after open versus arthroscopic surgery (13 % versus 20 %) [12]. Cupp, J.S, et al. studied both sub patterns of TGCT specimens and evaluated CSF1 RNA and protein expression, CSF1 translocation, and CSF1R RNA expression and found majority (35 of 57; 61%) of the evaluable specimens demonstrated both a CSF1 translocation and high levels of CSF1 RNA expression by in situ hybridization [13]. Systemic therapies, particularly tyrosine kinase inhibitors (TKIs) targeting CSF1/CSF1 receptor (CSF1R) signaling pathways, have shown promising results as novel treatment options in TGCT patients [14].

TEACHING POINT

The current case report describes an uncommon form of a rare pathology, a localized TGCT in the deep infrapatellar compartment of the knee that should be considered in the evaluation of lesions from the deep infrapatellar compartment of the knee.

QUESTIONS

1. What is the most common location for the localized TGCT's?

- a) Fingers
- b) Knee
- c) Cranium
- d) Shoulder

Answer: (a) 85 % of Localized TCGT's are found in Fingers.

2. What is the modality of choice to diagnose TGCT's?

- a) CT
- b) MRI
- c) Plain radiograph
- d) Ultrasound

Answer: (b) Magnetic resonance imaging (MRI) is the modality of choice to diagnose TGCT and discriminate between subtypes.

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3. What is the most likely pathogenesis of sub patterns of TGCT's?

- a) CSF1 translocation resulting in CSF1 overexpression
- b) EGFR overexpression
- c) Chromosomal translocation
- d) TGF overexpression

Answer: (a) TGCT subtypes share a common underlying pathogenesis, mainly related to a Colony-Stimulating Factor 1 (*CSF1*) translocation resulting in CSF1 overexpression. CSF1 overexpression causes an increase in neoplastic cells by binding to CSF1-receptors (CSF1R) and accumulating CSF1R presenting cells.

4. What is the mainstay of treatment for Localized TGCT's?

- a) Medical management
- b) Surgical management
- c) Radiation

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d) Physical therapy

Answer: (b) Surgery is the mainstay of Localized TGCT treatment, performed either open or arthroscopically.

5. Which is not true with regards to Localized TGCT's?

a) The absence of blooming artifact on GRE images excludes tenosynovial giant cell tumor

b) Localized tenosynovial giant cell tumors present as a slow-growing, painless mass

c) Localized tenosynovial giant cell tumors are most common in the fingers

d) Periosteal reaction and calcification are uncommon

Answer: (a) The absence of blooming artifact on GRE images does not excludes tenosynovial giant cell tumor

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FIGURES



Figure 1: PD sequence in MRI knee of a 17 year old female shows an oval hypointense nodule in the deep infrapatellar bursa with mild bursal fluid.



Figure 2: Axial T2 FS sequence in MRI knee of a 17 year old female shows a small iso to hyperintense nodule in the deep infrapatellar bursa with mild bursal fluid.



Figure 3: Sagittal T2 FS sequence in MRI knee of a 17 year old female shows small iso to hyperintense nodule in the deep infrapatellar bursa with mild bursal fluid.

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