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Giant Cell Tumor of the Patella Tendon Sheath Presenting as a Painful Locked Knee

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 26
Final Diagnosis: Giant cell tumor of the patella tendon sheath
Symptoms: Effusion • locking knee • pain
Medication: None
Clinical Procedure: Arthroscopy and open resection of the tumor
Specialty: Orthopedics and Traumatology

Objective: Rare disease

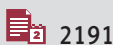
Background: The giant cell tumor of the tendon sheath (GCT-TS) is a benign proliferative synovial tumor manifesting as an intra-articular solitary nodule. When it involves the infrapatellar fat pad it can present acutely as a painful locked knee.

Case Report: A 26-year-old white male presented with a 2-week history of painful locking in his right knee. Clinical examination revealed lack of extension by approximately 20°. To help establish the diagnosis, an MRI scan of the right knee was performed, showing a large (5×4×2 cm), oval, well-circumscribed mass with a low-intensity homogeneous signal. The size of the mass prohibited the removal by arthroscopy and we therefore proceeded with an open arthrotomy. Histological examination showed a tendosynovial giant cell tumor of the patella tendon sheath. At the latest follow-up, 2 years postoperatively, there was no local tumor recurrence.

Conclusions: These rare tumorous lesions should be included in the differential diagnosis of painful locking knee, especially in the absence of definite traumatic history.

MeSH Keywords: Arthroscopy • Giant Cell Tumor of Tendon Sheath • Knee Injuries • Knee Joint

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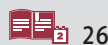
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Background

An acutely “locked” knee can be defined as a knee that cannot be passively fully straightened after an injury. This usually implies an intra-articular disorder blocking full extension, resulting in a knee held in flexion, which is painful on attempting full extension. Most cases of knee locking and giving-way are caused by mechanical block to full extension due to meniscal tears, loose bodies, ACL tears, or chondral lesions [1]. In rare cases, intra-articular benign tumors or tumor-like lesions present with symptoms that suggest acute mechanical derangement. Özalay et al. [2] reported on 33 patients with benign intra-articular masses in the knee joint; 19 of these individuals had sought medical attention for mechanical symptoms of catching or locking and 15 (79%) suffered from localized pigmented villonodular synovitis (PVNS). Loriaut et al. [3] recently reported the clinical outcome of 30 consecutive patients with local PVNS who underwent arthroscopic synovectomy and found that 15 (50%) of them had presented with some mechanical symptoms of locking.

The natural history of PVNS begins with the first report by Jaffe et al. [4] in 1941, and remained for several decades without clear histological description and with many classification controversies. Clarification has been given by the systematic approach of the World Health Organization in 2002 with the very useful demarcation of giant cell rich soft tissue tumors according to their origin: bone, soft tissue, synovium, and tendon sheath. Diffuse PVNS was renamed diffuse-type giant cell tumor (Dt-GCT) to signify the extensive involvement of the whole synovial membrane and capsule, whereas local PVNS was renamed giant cell tumor of tendon sheath (GCT-TS), confined to a distinct area of synovium [5]. Dt-GCT is a benign proliferative lesion that develops in the synovial lining of joints, tendon sheaths, and bursae, while malignant diffuse-type tenosynovial giant cell tumor (D-TSGCT) is an unusual sarcoma with considerable morphologic variability, metastatic propensity, and lethality.

The rate of occurrence of intra-articular GCTs in the adult knee is approximately 1.8 per 1 million populations, but this includes both the localized form (GCTTS) and the more diffuse intra-articular form (PVNS) [6], in contrast to the commonest finger localization of almost 85% of the cases. The localized form in the knee joint makes up 15–25% of PVNS cases, but a recent report found it was 7 times more common than the diffuse form [7]. It is usually characterized by focal involvement of the synovium with nodules of pathologic tissue. Some affected patients exhibit local warmth, swelling, and stiffness [2]. GCT-TS have been found in several locations within the knee joint: meniscus-synovium junction, intercondylar notch, lateral and medial femoral cortex, patella tendon, quadriceps tendon, posterior compartment, and ACL and PCL ligaments [7–10]. With

slow-growing tumors of the synovium it is also important to exclude other diagnoses prior to surgical intervention: e.g., synovial hemangioma, synovial chondromatosis, and synovial sarcoma [11]. Conventional plain radiographs are often not diagnostic. With advanced disease there may be evidence of soft-tissue swelling, loss of joint space, and peri-articular erosion of bone; the latter are more notable in joints with a tight capsule, such as the hips and elbows [7]. MRI helps with the diagnosis of diffuse forms as it detects areas of inflammation with hemosiderin deposits, and is the top diagnostic imaging modality for evaluating soft-tissue tumors [12]. But the localized forms are more challenging. Although the appearance can be characteristic, image-guided percutaneous needle biopsy for histopathological examination may be indicated, particularly if there is doubt about the diagnosis. A heterogeneous soft-tissue mass with T1 and T2 hyposignal appears on MRI. The signal is intermediate if only small hemosiderin deposits are present and becomes more visible with gradient echo sequence. The lesion could be undetected on MRI, despite being quite large (4 cm) in size. Bouguennec et al. [13] were still unable to detect the localized PVNS lesion despite their detailed preoperative paraclinical assessment including X-rays, CT arthrography and MRI; the lesion had mimicked a lateral meniscus injury and was only confirmed during arthroscopy and subsequent biopsy. The principal treatment of GCT-TS and PVNS is surgical excision, especially in the knee and foot [14]. Open procedures are commonly used to manage the diffuse forms of PVNS and may be combined with adjuvant radiation therapy [7,15]. Arthroscopic approaches have gained increasing popularity because they are minimally invasive procedures and reduce the risk of complications. For patients with localized pigmented villonodular synovitis, the best indication is arthroscopic treatment; however, even diffuse forms can be treated effectively by an experienced arthroscopic surgeon, with the use of both anterior and posterior portals [15,16].

We present an unusual case of a giant cell tumor arising from the patella tendon sheath causing painful locking of the knee joint. Our purpose is to caution the surgeon from attributing these features to a meniscal tear – the most common cause of mechanical intermittent knee locking – and to proceed with further investigation, including laboratory analysis, accurate physical tests, and, for the most, specific imaging by MRI.

Case Report

A 26-year-old white male presented to the Emergency Department of our university hospital with a 2-week history of painful locking in his right knee. There was no history of antecedent trauma except for some mild disturbances during soccer playing on recreation level 1 and 3 months before. Pain was exacerbated the last 48 hours and was the main cause of



Figure 1. Normal anteroposterior and lateral X-ray of the right knee.

inability to bear weight. Extended anamnesis was possible by specific questions to his mother who was accompanying him, showing no family history of hereditary diseases or personal history of genetic abnormalities. Clinical examination revealed moderate swelling, medial joint tenderness, and lack of extension by approximately 20°. No effusion was visible and there was no palpable mass across the joint line. The color of the skin was normal without any redness or nettle rash and the temperature, tested by the examiners hands, felt the same on both knees. No fever at the time of examination was present and no other inflammatory symptomatology or clinical inflammatory manifestation during the last 2 months was reported. The most usual knee functional diagnostic tests were used, but results were unreliable because of the limited range of motion and the partial collaboration of the patient, who showed signs of pain during the maneuvers. There was no evidence of quadriceps muscles atrophy or wasting. Simple isometric exercise was performed at both thighs, with normal and symmetrical response. Right patellar reflex was impossible to test objectively. Neurological examination was normal, without sensibility deficit below L1 level. Plain radiography by anteroposterior and lateral view were without evidence of pathology (Figure 1). Blood analysis parameters and basic metabolic panel results were within normal range. Afterwards, synovial fluid was obtained by arthrocentesis to perform macroscopic, microscopic, and biochemical examination. Macroscopical analysis revealed a slightly opaque fluid with a slight rose color, with pH 7 and specific weight 1010. Synovial fluid analysis showed a total of 2570 cells, of which 20 were RBC, 78 were WBC, 13 were LBC, and 9 were large mononuclear cells. Synovial fluid glucose level was 78 mg/dl and blood serum glucose level was 92 mg/dl, making the diagnosis of septic

arthritis unlikely. This was further confirmed by negative CRP and ESR levels as well as negative Gram stain. Microscopy results were also negative for presence of crystals such monosodium urate and calcium pyrophosphate.

To help establish the diagnosis, an MRI scan of the right knee was performed. Coronal and sagittal T1-weighted and T2-weighted images (Figure 2) revealed a large (5×4×2 cm), oval, well-circumscribed mass with a low-intensity homogenous signal. The extension margins were in close proximity with the patellar tendon, occupying the posterior aspect of it. The mass projection was to an intra-articular direction.

Once the diagnostic approach was terminated and the pre-operative examination found no contraindications, the patient was taken to the surgical theater. Examination under general anesthesia revealed full flexion of the knee and full extension except from terminal degrees where a soft block could be felt without accompanying effusion or palpable mass. Subsequent knee arthroscopy was performed by typical arthroscopic portals. A unique, large, intraarticular mass was identified, originating posteriorly to the patellar tendon (Figure 3). The size of the mass prohibited removal by arthroscopy and we therefore proceeded with an open arthrotomy. In this way, via medial parapatellar incision, a complete mass excision was achieved and the lesion was sent for histological interpretation. Gross macroscopic examination revealed a well-circumscribed, firm, homogenous, yellowish-to-brown mass measuring 5×4×1.8 cm. Microscopically, the lesion was moderately cellular, with the presence of ovoid-to-epithelioid mononuclear cells and scattered multinucleated giant cells with a varying number of nuclei, ranging from 3 to approximately 50 (Figure 4). The mononuclear

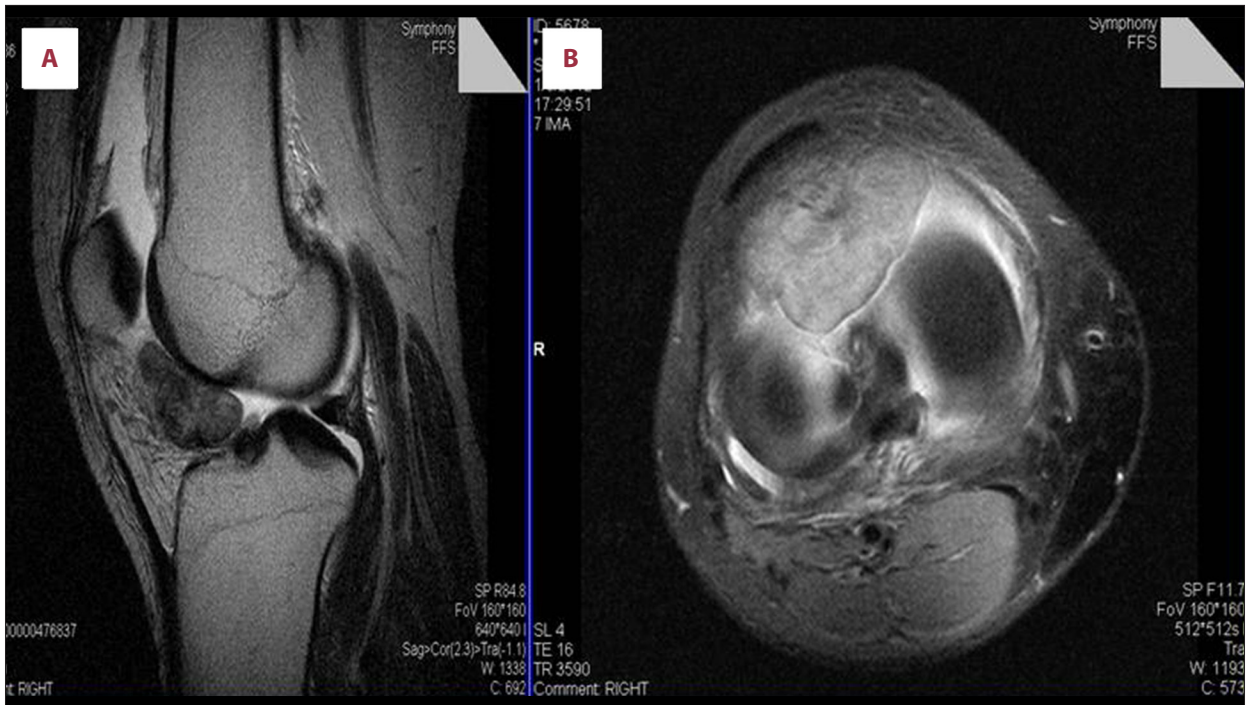


Figure 2. Magnetic resonance images of the right knee joint. T2-weighted fat-saturated, proton density sagittal (A) and axial (B) images showing a lobulated mass in the posterior aspect of the infrapatellar fat pad. The mass had hypointense signal intensity and extended into the intercondylar notch.

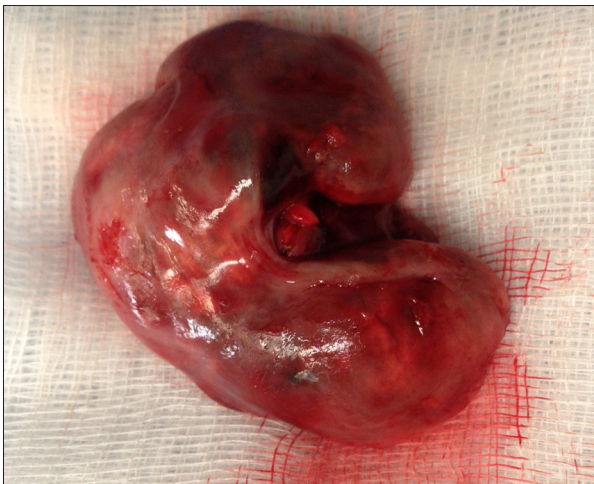


Figure 3. The mass is encapsulated by fibrous tissue and the diameter is about 5 cm. There are no villous fronds or evidence of cystic component. Signs of acute hemorrhage were also noted.

cells displayed minimal atypia, moderate amount of cytoplasm, and large, round nucleus. The cells were embedded in dense collagenous stroma. Very low mitotic activity (1–2 mitotic figures/10 HPF) was observed.

Immunohistochemical analysis revealed that both the mononucleated and the multinucleated cells displayed strong

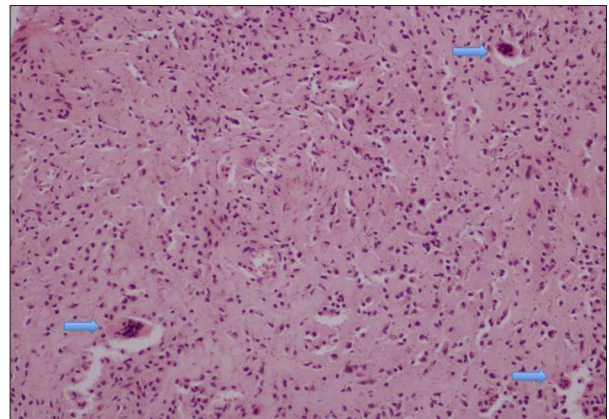


Figure 4. Histological features of localized-type tenosynovial giant cell tumor. Note the presence of round-to-ovoid mononuclear cells embedded within collagenous stroma. A few multinucleated giant cells, with varying number of nuclei, are also present (arrows). There is no significant cellular atypia.

immunoreactivity for the macrophage marker CD68 and focally positive for the mesenchymal marker Vimentin. On the contrary, they were negative for the epithelial markers keratins AE1/AE3 and the endothelial markers CD31 and CD34. The proliferation index Ki67 was very low.

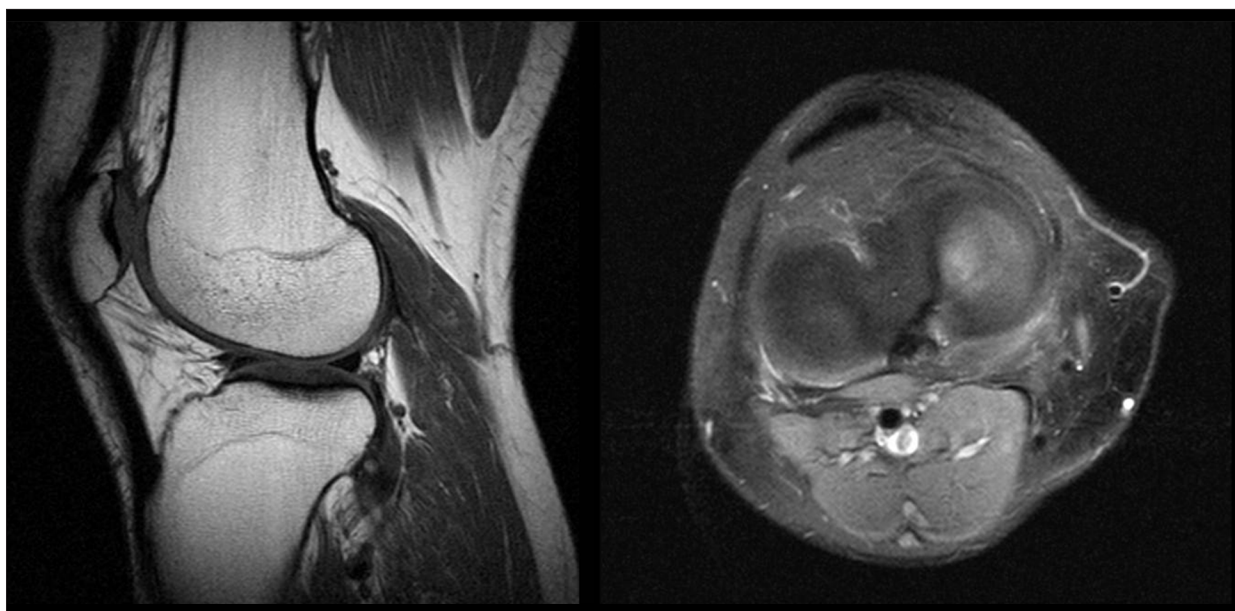


Figure 5. Magnetic resonance imaging at 2 years postoperatively. There is no evidence of recurrence in T1-weighted sagittal and axial sequences.

Based on the history, physical examination, laboratory studies, imaging studies, and histologic picture, a diagnosis of giant cell tumor of the tendon sheath (GCT-TS) was confirmed. Clinical follow-up was performed at 3, 6, 12, and 24 months, during which the patient remained totally asymptomatic. Two years post-operatively an MRI confirmed the disease-free condition (Figure 5).

Discussion

In most patients with the diffuse form of PVNS, the diagnosis is straight-forward given the suggestive clinical picture of pain, repeated hemarthrosis, stiffness, formation of bone cysts, and cortical erosion. In the localized form, however, the observation of non-specific symptoms makes diagnosis quite challenging. The average age of patients with PVNS is typically from 30 to 50 years old [17], which corresponds to the patient described here. Occurrence of the lesion in younger patients has also been reported [18].

Clinical presentation of GCT-TS is variable: discomfort is always present but it can be accompanied by locking, effusion, diffuse pain, decreased range of motion, palpable mass, or joint line tenderness suggesting a meniscus injury [3,7,13,19]. The clinical presentation can also suggest the presence of an intra-articular floating foreign body, non-specific patellofemoral joint tenderness, and symptoms related to cartilage injury [20–22]. A preoperative clinical diagnosis of meniscus injury was made on 30% of localized PVNS cases [19] and the PVNS diagnosis was made in only 25% of localized form. In our case the main

complaint was painful lack of knee extension without significant history of previous trauma. A similar case has been recently reported by Napier and McCormack [23] in a 39 year-old patient who presented with a 12-month history of painless locking in his left knee. He was referred by his general practitioner who suspected a meniscal tear. Without MRI scanning, the surgeons proceed directly to arthroscopic evaluation where a large intra-articular lesion (40×35×15 m) was identified originating posterior to the medial patella. The lesion was finally excised via a medial parapatellar incision. Rao et al. [24] reported on 3 cases of GCT-TS presenting as locked knee with lack of full extension. The tumors originated from the lateral meniscus (2.2 cm), suprapatellar pouch (2 cm), and medial meniscus (3 cm); age at presentation was 35, 33, and 28 years old, respectively. Resection was performed arthroscopically in the first 2 cases and open in the last case. Williams and Myers [25] reported on a 34-year-old woman who presented with a history of recurrent episodes of knee locking and swelling. Arthroscopy revealed localized PVNS in the intercondylar notch region causing interference with joint motion. The lesion was resected arthroscopically with prompt resolution of symptoms. Finally, Yotsumoto et al. [26] reported on a 26-year-old patient with acute locking knee due to a GCT-TS in the intercondylar notch associated with a lateral meniscus tear. The patient had similar symptoms 2 years before and was treated as having a meniscal injury. The tumor was located between the ACL and PCL and contributed to blocking, together with a meniscus flap. Both were resected arthroscopically.

Our case presented in the emergency department with a painful right knee, lack of extension, and inability to bear weight.

MRI revealed a large intraarticular mass in contiguity with the patellar tendon and intra-articular extension. Arthroscopic confirmation of the lesion was followed by total excision utilizing a mini-arthrotomy. The histopathological examination confirmed the giant cell tumor arising from the patellar tendon sheath. Volume, size, and intraarticular propagation induced lack of extension in the knee, which was manifested by locking symptom-like condition. Clinical follow-up was performed at 3, 6, 12, and 24 months, with the patient remaining totally asymptomatic. Two years post-operatively, MRI confirmed the disease-free condition.

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Conclusions

In conclusion, GCTTS in the knee is rare, especially when presented as painful locked knee. Our purpose is to caution the surgeon from attributing these features to a meniscal tear – the most common cause of mechanical intermittent knee locking – and to proceed with further investigation, including laboratory analysis, accurate physical tests, and, for most, specific imaging by MRI.

Consent

The patient has given his informed consent for the case report to be published.