Diffuse-Type Tenosynovial Giant Cell Tumor (Pigmented Villonodular Synovitis) of the Knee: A Case Report.

Abstract

Background: Tenosynovial giant cell tumor (TGCT), formerly known as pigmented villonodular synovitis (PVNS), is a rare, benign but locally aggressive proliferative disorder of the synovial membrane. Its diffuse type is more aggressive than the localized type and carries a substantially higher risk of recurrence.

Case presentation: We report the case of a 25-year-old man with no relevant medical history who presented with mechanical pain of the right knee evolving over 4 years. Clinical examination found a mildly swollen knee, tender on palpation and mobilization, with flexion limited to 110°, full extension, mild quadriceps amyotrophy, and normal ligamentous testing, without patellar tap sign or local inflammatory signs. Standard radiographs and inflammatory markers were normal. MRI showed diffuse-type TGCT involving both compartments of the knee, without extra-articular extension.

Management and outcome: Given the diffuse, bicompartmental nature of the disease, the patient underwent total open synovectomy without adjuvant treatment. Histopathological examination confirmed the diagnosis, showing villonodular hyperplasia with hemosiderin deposits, multinucleated giant cells, and xanthomatous foam cells. At three months, pain had markedly improved and weight-bearing had resumed, with no clinical sign of recurrence; however, this short follow-up remains insufficient to draw conclusions regarding long-term recurrence, which is typically assessed over several years.

Conclusion: This case highlights the importance of considering diffuse-type TGCT in young patients presenting with unexplained chronic knee monoarthropathy, and underscores the need for prolonged clinical and radiological surveillance after surgical treatment.

Keywords: Tenosynovial giant cell tumor, pigmented villonodular synovitis, knee, synovectomy, diffuse type, case report.

Introduction

Tenosynovial giant cell tumor (TGCT), formerly known as pigmented villonodular synovitis (PVNS), is a rare proliferative disorder of the synovial membrane, tendon sheaths, and bursae. The knee is by far its most common joint location, accounting for approximately 63-75% of cases [1,2]. Two anatomo-clinical forms are described: a localized (nodular) type, associated with a good prognosis and a low recurrence rate after simple surgical treatment, and a diffuse type, more aggressive and pseudo tumoral in nature, exposing the patient to a high risk of recurrence — reported between approximately 25% and 50% across series — and usually requiring a more radical surgical approach, often combined with adjuvant treatment [2,3].

The clinical presentation is nonspecific, combining mechanical pain and slowly progressive joint swelling, which frequently leads to diagnostic delay. The diagnosis is most often first suggested on MRI before being confirmed by histopathological examination [2]. Standard radiographs remain useful in the initial evaluation, although they are frequently normal or show only nonspecific findings [1].

Diffuse-type TGCT of the knee remains rare, and its management continues to raise questions, particularly regarding the extent of synovectomy and the indication for adjuvant treatment in bicompartmental disease without extra-articular extension. We report the case of a young patient with such a presentation, aiming to illustrate the diagnostic pathway and the therapeutic reasoning involved in this specific clinical scenario.

Case presentation

Patient information: A 25-year-old man, with no relevant medical history, presented with mechanical pain of the right knee evolving over 4 years.

Clinical findings: On examination, the right knee was mildly swollen. Tenderness was elicited on palpation and mobilization. There was no patellar tap sign and no local inflammatory signs. Range of motion was limited in flexion to 110°, with full extension. Mild quadriceps amyotrophy was noted, and ligamentous testing was normal, without any sign of instability.

Diagnostic assessment: Standard radiographs of the knee were unremarkable, without bone erosion or subchondral cyst. Inflammatory markers were within normal limits. MRI showed diffuse, lobulated synovial hypertrophy with a heterogeneous signal, filling the anterior and peri-condylar recesses and involving both the medial and lateral femorotibial compartments, with extension into the suprapatellar and posterior (popliteal) recesses on sagittal sequences. The heterogeneous appearance, combining areas of low signal intensity with more hyperintense foci, was consistent with the characteristic hemosiderin-related pattern of tenosynovial giant cell tumor (TGCT). No extra-articular extension was identified. These findings were consistent with diffuse-type TGCT involving both compartments of the knee.

Therapeutic intervention: Given the diffuse, bicompartmental nature of the disease, surgical treatment was decided. The patient underwent total synovectomy through an open anterior approach, without adjuvant treatment. Intraoperative exploration revealed citrine-yellow, turbid joint fluid and a hypertrophied, hemorrhagic synovium with villonodular proliferation (Figures 1-2). The surgical specimen was sent for histopathological examination, which confirmed the diagnosis, showing villonodular hyperplasia with hemosiderin deposits, multinucleated giant cells, and xanthomatous foam cells, a typical appearance of TGCT (Figure 3). After completion of the synovectomy, the articular surfaces were free of residual pathological synovium (Figures 4-5).

Postoperative care and rehabilitation: Postoperative course was uneventful. Early mobilization and progressive weight-bearing were encouraged, and the patient was referred for physiotherapy focusing on recovery of range of motion and quadriceps strengthening.

Follow-up and outcomes: At three months postoperatively, the patient reported marked improvement in pain and had resumed weight-bearing, with no clinical sign of recurrence at this stage.

Timeline.

The patient's symptoms of mechanical right knee pain evolved over 4 years before presentation. At presentation, clinical examination was notable for mild swelling and flexion limited to 110°, while standard radiographs and inflammatory markers were normal. MRI performed at this stage showed diffuse-type TGCT with bicompartmental involvement, without extra-articular extension. The patient then underwent total open synovectomy through an anterior approach; intraoperative exploration revealed citrine-yellow, turbid joint fluid and a hypertrophied, hemorrhagic villonodular synovium, and no adjuvant

treatment was administered. Histopathological examination performed postoperatively confirmed the diagnosis. At the three-month follow-up, pain had markedly improved, weight-bearing had resumed, and no clinical sign of recurrence was noted.

Discussion

Tenosynovial giant cell tumor (TGCT) predominantly affects young adults between the third and fourth decades of life, most often as a monoarticular disease, with the knee representing the most frequent location, followed by the ankle and the hip [2,4]. Our patient, aged 25 years, falls within this classically reported age range.

MRI is the key diagnostic examination. It typically shows a low signal on both T1- and T2-weighted sequences related to hemosiderin deposits, and it allows precise assessment of the extent of disease (localized versus diffuse) and of any extra-articular involvement, thereby guiding surgical strategy [2,3]. This pattern was well illustrated in our patient, whose MRI showed a heterogeneous synovial signal with low-intensity foci consistent with hemosiderin deposition, confirming the diffuse, bicompartmental nature of the disease before surgery. Diagnostic delay remains frequent, as the initial symptoms - mechanical pain and slowly progressive swelling - are nonspecific and may be misattributed to other, more common causes of knee pain [3]. In our patient, the four-year history of symptoms before diagnosis illustrates this well-documented diagnostic delay.

Localized-type TGCT is treated by simple surgical excision, with an excellent prognosis and a low, sometimes negligible, recurrence rate [1]. In contrast, the diffuse type, owing to its pseudotumoral and locally aggressive behavior, requires more extensive surgery - most often total synovectomy, performed arthroscopically, through an open approach, or through a combination of anterior and posterior approaches depending on the extent of the disease [1,2]. Arthroscopic synovectomy alone has been associated with a higher recurrence rate than open or combined synovectomy [4,7]. In our patient, the open anterior approach was chosen given the diffuse, bicompartmental extent of the disease and the associated risk of recurrence, and was further favored by the limited local availability of knee arthroscopy. Adjuvant treatment, particularly radiosynoviorthesis, is often proposed in severe or recurrent diffuse forms, typically four to eight weeks after the initial surgery [1].

The recurrence rate of diffuse-type TGCT remains high, reported between approximately 25% and 50% across series [4,7]. This risk justifies prolonged surveillance, with annual MRI recommended during the first four years after treatment [1]. In our patient, the synovectomy was judged macroscopically complete at the end of the procedure, with articular surfaces free of residual pathological synovium; on this basis, no adjuvant treatment was administered, consistent with practices reported for bicompartmental diffuse disease without extra-articular extension when resection is deemed complete. Nevertheless, this decision warrants close and prolonged clinical and radiological follow-up, and the three-month follow-up reported here remains too short to draw firm conclusions regarding recurrence.

Recent advances in understanding the role of the CSF1/CSF1R signaling pathway in the pathogenesis of TGCT have led to the development of targeted systemic therapies, such as CSF1R inhibitors, which may be of particular interest in recurrent or surgically inaccessible diffuse disease [5,6]. These treatments currently remain reserved for selected cases, and surgery continues to represent the mainstay of management for localized and most diffuse forms [2,5].

Limitations

This report has several limitations. It describes a single case with a short follow-up of three months, which does not allow assessment of the long-term risk of recurrence, particularly relevant given the diffuse nature of the disease and the absence of adjuvant treatment. Outcome was assessed clinically, without a validated functional score or postoperative MRI to confirm the absence of residual or recurrent disease, and the preoperative MRI findings were not quantitatively assessed (e.g., lesion volume or extent), limiting comparison with future follow-up imaging. In addition, the choice of an open surgical approach was partly influenced by the limited local availability of knee arthroscopy, a contextual factor that may limit the generalizability of this therapeutic choice to settings with broader access to arthroscopic surgery. Longer follow-up with systematic imaging will be required to confirm the durability of this result.

Conclusion

Diffuse-type tenosynovial giant cell tumor (TGCT) of the knee is a rare condition whose diagnosis, often delayed because of nonspecific symptoms, relies on MRI and is confirmed by histopathological examination. Its surgical management, more extensive than that of the localized type, carries a substantial risk of recurrence that justifies prolonged clinical and radiological follow-up, particularly when, as in our patient, a macroscopically complete resection allows adjuvant treatment to be withheld. Open synovectomy remains a valid option in this setting, especially where arthroscopic resources are limited. This case highlights the importance of considering this diagnosis in young patients presenting with unexplained chronic knee monoarthropathy, and underscores that a short-term favorable outcome does not preclude the need for extended surveillance.

Declarations

Conflicts of interest

The authors declare no conflicts of interest.

Authors' contributions: Data collection: AA and MK. Manuscript drafting: DZ, LH. Critical revision of the manuscript: AA, JR, KL, AM and FZ.

All authors read and approved the final version of the manuscript.

Informed consent: Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Patient perspective: From the patient's standpoint, the reduction in pain and the resumption of daily activities were regarded as a satisfactory early outcome.

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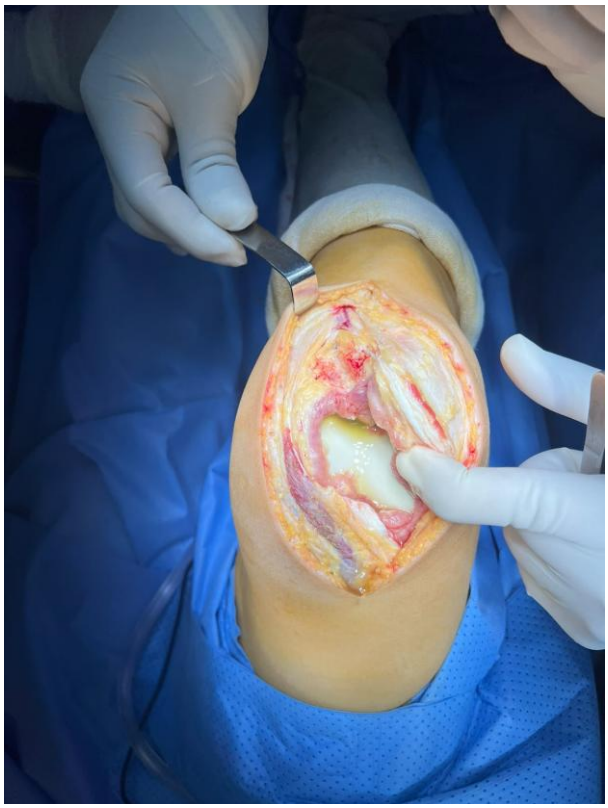
Figures:



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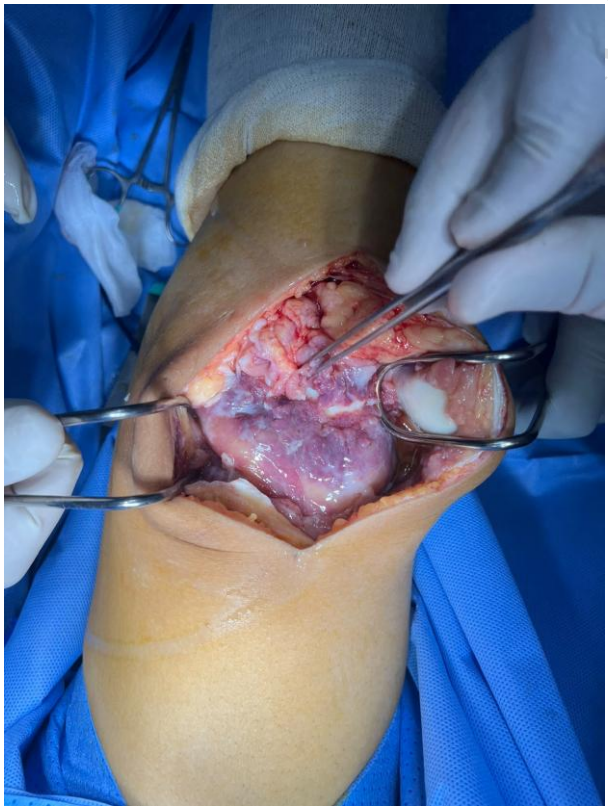

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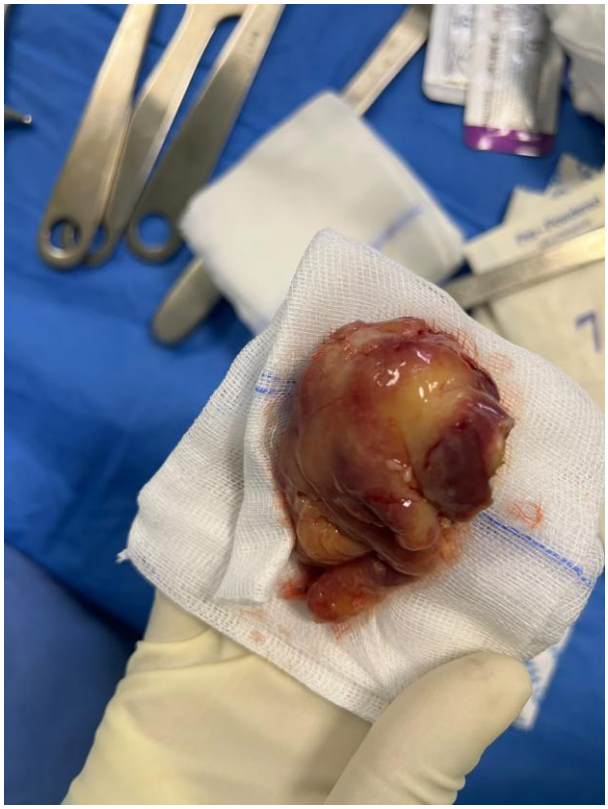
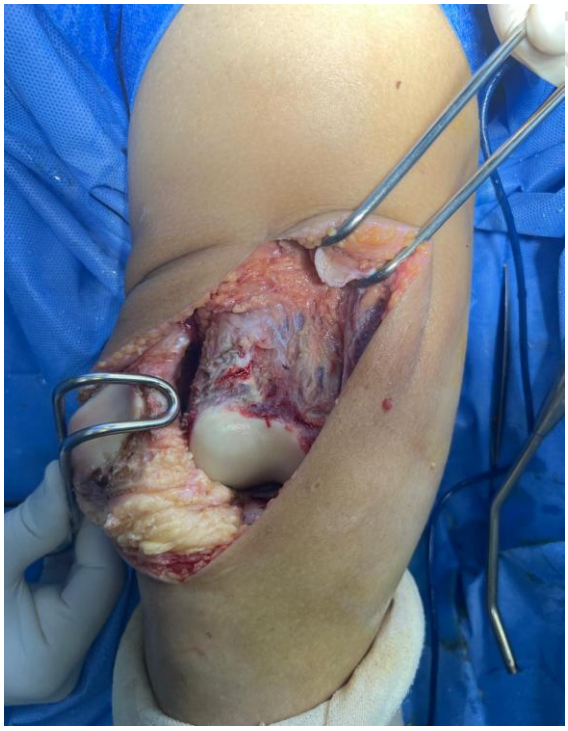


Figure 3: Surgical specimen showing a villonodular mass with a characteristic brown-yellow coloration.



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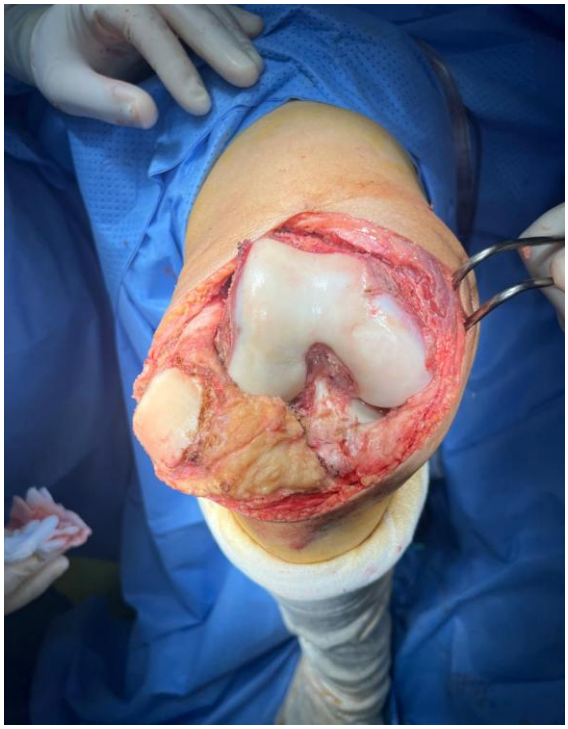

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