

# Giant Cell Tumor of the Tendon Sheath in the Knee of an 11-year-old Girl

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Giant cell tumors are commonly found over the flexor tendon sheath of the hand and wrist. However, giant cell tumors in the knee joint are rare, especially in children. We report an interesting case of an 11-year-old girl who presented with a painful lump on her right knee that enlarged over time. Clinically, she had fullness over the anterolateral part of her knee. Magnetic resonance imaging revealed an encapsulated mass inferior to the patella. The tumor measured  $3 \times 3.5 \times 1.5$  cm. Histopathological findings confirmed that it was a tenosynovial giant cell tumor. Because of initial mild symptoms, there was a delay of 2 years from the initial symptoms until tumor excision. Her follow-up period was 35 months, and her health to date is excellent with no recurrence. We believe that reporting this rare case will help clinicians update their knowledge on possible causes of lumps in the knee, and avoid diagnostic delay. It could also prove to be beneficial in arriving at a diagnosis in future cases. [*J Chin Med Assoc* 2010;73(1):47–51]

**Key Words:** diagnosis, giant cell tumor, histopathology, knee, MRI, pediatric

## Introduction

Giant cell tumors of the tendon sheath (GCTTS) most commonly occur in the flexor tendon sheath of the hand and wrist, followed by the ankle and foot region. GCTTS in the knee joint is rare, particularly in children. The rate of occurrence in the adult knee is approximately 1.8 in each 1 million, but this includes both the localized form (GCTTS) and the more diffuse intra-articular form known as pigmented villonodular synovitis (PVNS).<sup>1</sup>

Gholve et al<sup>2</sup> reported on 29 children with GCTTS, but only 1 male and 1 female, both aged 16 years old, had a tumor in their knee. Ushijima et al<sup>3</sup> had 207 patients, 8 with GCTTS in their knees, while Rodrigues et al<sup>4</sup> had 28 patients, 4 with GCTTS in their knees. However, neither reported whether these were adults or children.

The clinical importance of the tumor is highlighted by the fact that the symptoms of the disease are nonspecific.<sup>5</sup> Here, we highlight an interesting case of giant cell tumor (GCT) occurring in a child who presented to our outpatient department. Clinical knowledge of the presentation of a GCT may be beneficial for clinicians.

## Case Report

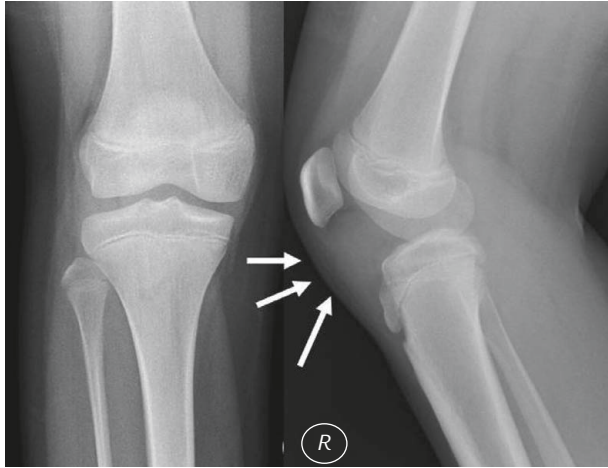
The research findings are presented with the consent of the girl's parents and ethics committee approval. An 11-year-old Malay girl presented with an insidiously growing swelling on her right knee. The lump was initially painless, but as it grew larger over a period of 2 years, it became increasingly painful. The pain was dull in nature and aggravated by strenuous activity. There was no history of trauma. The patient had no sensation of her knee giving way when walking on uneven surfaces, nor did she experience any locking of her left knee. There was no history of any constitutional symptoms such as night sweats, fever, loss of appetite or weight loss.

Clinically, the patient was an active and healthy child. Examination of her right knee revealed fullness at the anterolateral part that was more prominent on flexion. The range of motion was full, and there was no joint line tenderness. No enlarged inguinal lymph nodes were detected.

The patient's blood parameters were normal. Plain radiography showed an opacity underneath the patella tendon (Figure 1). Magnetic resonance imaging revealed an encapsulated mass inferior to the patella tendon with surrounding fluid (Figure 2).



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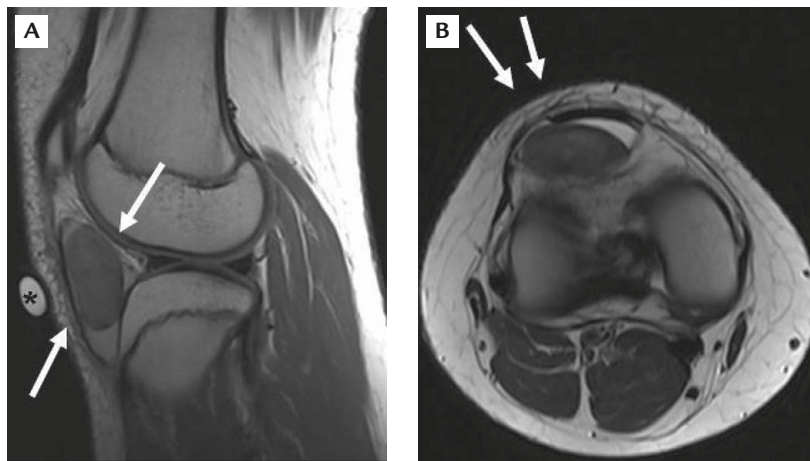


**Figure 1.** X-ray (lateral) showing opacity at the infrapatellar region (arrows).

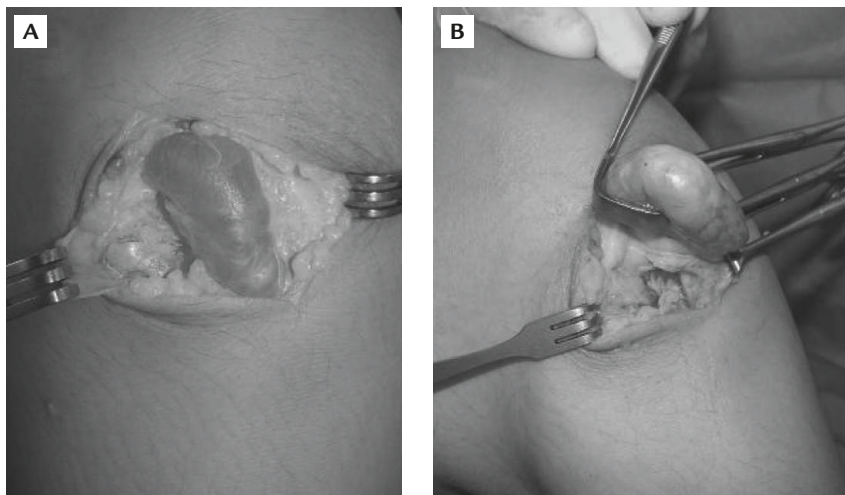
The differential diagnoses at that point were an intra-articular lipoma or GCT. The patient later underwent an excision biopsy, revealing a cyst overlying a pedunculated mass measuring  $3 \times 3.5 \times 1.5$  cm (Figures 3 and 4).

Histopathological examination showed a partially circumscribed mass consisting of cellular fibrous tissue with septa formation separating numerous fibroblast-like cells, foamy histiocytes, cholesterol clefts, siderophages, and multinucleated giant cells. No cellular atypia or other evidence of malignancy was seen (Figures 5 and 6). A final diagnosis of tenosynovial GCT was made.

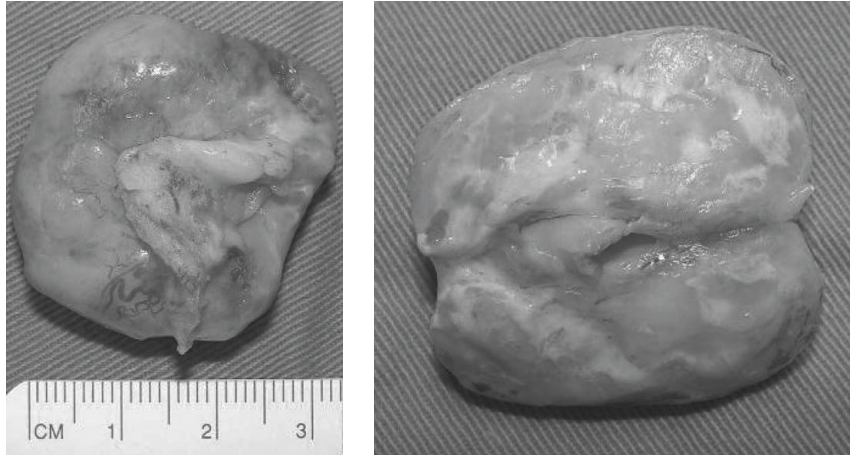
The patient recovered well without any perioperative problems. The knee pain gradually subsided. At follow-up 35 months later, her recovery was excellent, with no evidence of recurrence. She had no tenderness in the knee, a normal gait, and full range of motion of



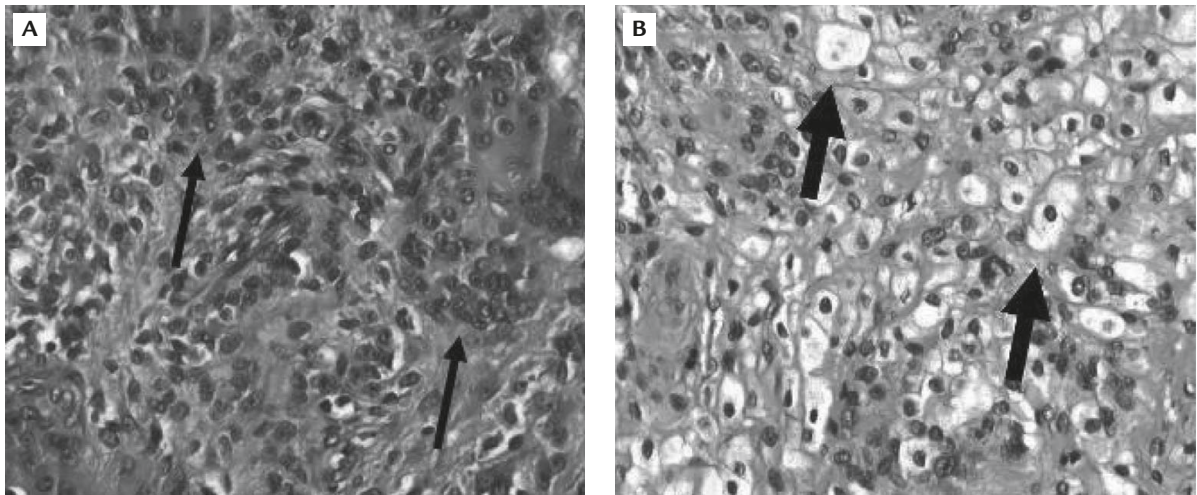
**Figure 2.** Magnetic resonance image of the knee: the encapsulated mass (arrows) opposite a radio-opaque marker (\*) is clearly seen within Hoffa's fat pad. An effusion surrounds the mass.



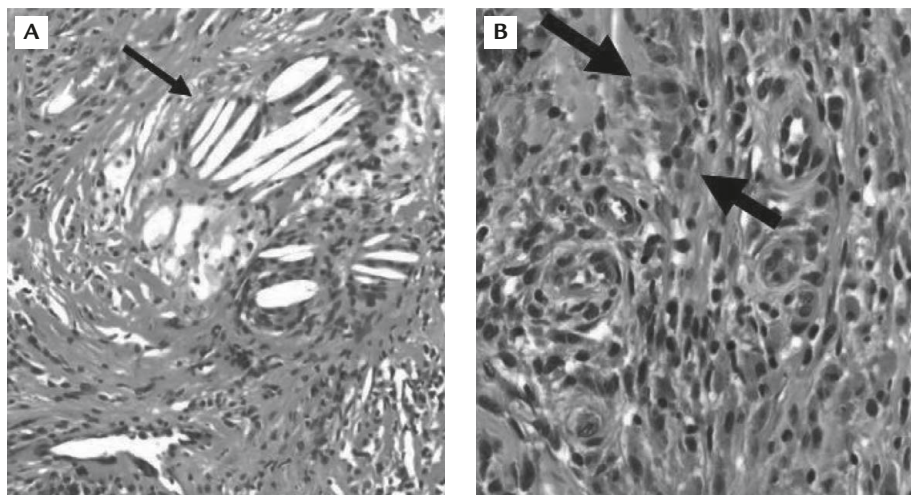
**Figure 3.** (A) Cyst present on initial dissection. (B) The pedunculated mass was easier to see once the cyst was fully exposed.



**Figure 4.** Photograph of the tumor. It was whitish in color, measured 3.0×3.5×1.5 cm, and had a smooth surface. When cut open, multiple white and yellow nodules were seen.



**Figure 5.** Histopathology (hematoxylin & eosin, 100×) shows: (A) multinucleated giant cells (arrows); and (B) foamy macrophages and histiocytes (arrows).



**Figure 6.** Histopathology (hematoxylin & eosin, 100×) shows: (A) cholesterol clefts (arrow); and (B) siderophages (arrows).

her knees. She mentioned occasional mild pain in her right knee after prolonged sitting.

## Discussion

GCTTS predominantly develops in the hand but can also be found around the foot, ankle, knee or hip. Ushijima et al reported 87.9% of cases in the digits (fingers and toes) and 12% in the large joints (knee, elbow, ankle, hip) in both adults and children.<sup>3</sup> Gholve et al described the occurrence of GCTTS in children as similar to those of adults.<sup>2</sup>

Jaffe et al describes a multiple manifestation of a single disease with 3 forms of benign synovial proliferative lesions identified: an isolated discrete lesion involving tendon sheath (GCTTS); a solitary intra-articular nodule (localized nodular synovitis); and a diffuse, often villous, pigmented process involving the synovial tissue (PVNS).<sup>6</sup> The basic histologic findings for these lesions are similar. The multinucleated giant cells are interspersed in a background of polygonal ovoid mononuclear cells. Patches of hyalinized tissue, collagen fibers, and xanthomatous foci composed of lipid-laden macrophages or foam cells are seen with rare or absent cellular atypia, or mitotic figures.

The proliferating synovial fibroblasts or primitive mesenchymal cells have a propensity for collagen production, and the histiocyte-like cells have a phagocytic function. A difference between GCTTS and PVNS is that PVNS has marked cellular hyperplasia and large hemosiderin deposits.<sup>6,7</sup>

We did not expect a diagnosis of GCT of the tendon sheath in a child's knee. One author, with cases of PVNS, had diagnostic delays up to 2 years and entertained diagnoses such as bacterial synovitis and juvenile rheumatoid arthritis.<sup>8</sup> In this case, we considered the possibility of an intra-articular knee joint lipoma.

The exact etiology of GCTTS remains unclear. Theories include inflammatory, neoplastic, traumatic, toxic, allergic, and genetic (*nm23* gene) factors.<sup>6,9</sup>

In large joints, GCTTS is difficult to diagnose because the symptoms are nonspecific and signs are few. The soft tissue mass grows and expands into areas of least resistance, often presenting itself as a mechanical derangement of the knee and/or with indistinguishable pain.<sup>7,10</sup> Mechanical symptoms such as locking, limitation of motion, or giving way might be related to the size of the mass. Sharma et al concluded that in chronic cases with a painless or painful mass in a large joint, one should have a high index of suspicion.<sup>1</sup>

Plain radiographs are often unhelpful, but might show bony erosions or occasionally a soft tissue swelling.

Magnetic resonance imaging is an important diagnostic tool. A differential diagnosis of a synovial cyst, synovial sarcoma, malignant fibrous histiocytoma, lipoma or ganglion must be considered. T1- and T2-weighted images may demonstrate a low intensity homogenous intensity signal in a GCTTS because of the presence of dense fibrous tissue. This may also be seen in PVNS but is usually less homogeneous because of a higher tendency of bleeding and greater hemosiderin deposition in PVNS.

Thus, the combination of a low intensity homogeneous signal in T1 and T2 images favors the diagnosis of a GCTTS, as exemplified in our case.<sup>11</sup> Excision of the tumor is usually sufficient, provided that all the affected tissue is removed.

A recurrence rate of 16–17% for GCTTS has been quoted for adults. Rates differ between adults and children, with children experiencing less recurrence and better outcomes.<sup>2,12,13</sup> Al-Qattan devised a classification for GCTTS in the hand to prognosticate lesions with a high chance of recurrence.<sup>14</sup> Lesions were classified into 2 types dependent on whether the entire tumor was surrounded by a pseudocapsule (type I) or not (type II), and further subdivided into single, multilobulated, diffuse or multicentric nodules. The classification was also enhanced by 5 potential factors that might lead to a high recurrence rate: poor surgical technique/incomplete excision; bony invasion; cellularity and mitotic activity; *nm23* gene negative; and type II tumors. However, these findings need to be validated by larger studies, although other authors have reported lower recurrence rates when good surgical techniques with magnification have been used.<sup>2,13,14</sup>

Using the classification above, our GCTTS falls into the type I category, without bony involvement or mitotic activity. Therefore, the probability of recurrence is low. Our child had excellent recovery, similar to the recovery of the 29 children with GCTTS reported by Gholve et al.<sup>2</sup>

In conclusion, GCTTS in the knee is rare, especially in children. We wish to draw the practitioner's attention to the possibility of GCTTS occurring in the knee of a child with a painful lump. This will allow earlier diagnosis.

## References

1. Sharma H, Jane MJ, Reid R. Pigmented villonodular synovitis: diagnostic pitfalls and management strategy. *Curr Orthop* 2005; 19:215–22.
2. Gholve P, Hosalkar HS, Kreiger PA, Dormans JP. Giant cell tumor of tendon sheath: largest single series in children. *J Pediatr Orthop* 2007;27:67–74.

3. Ushijima M, Hashimoto H, Tsuneyoshi M. Giant cell tumor of the tendon sheath (nodular tenosynovitis): a study of 207 cases to compare the large joint group with the common digit group. *Cancer* 1986;57:875–84.
4. Rodrigues C, Desai S, Chinoy R. Giant cell tumor of the tendon sheath: a retrospective study of 28 cases. *J Surg Oncol* 1998;68:100–3.
5. Bruns J, Yazigee O, Habermann CR. Pigmented villo-nodular synovitis and tenosynovial giant-cell tumors. *Z Orthop Unfall* 2008;146:663–75. [In German]
6. Jaffe HL, Lichtenstein L, Suro CJ. Pigmented villonodular synovitis, bursitis and tenosynovitis. *Arch Pathol* 1941;31:731–65.
7. Relwani J, Factor D, Khan F, Dutta A. Giant cell tumor of the patellar tendon sheath—an unusual cause of anterior knee pain: a case report. *Knee* 2003;10:145–8.
8. Neubauer P, Weber K, Miller NH, McCarthy EF. Pigmented villonodular synovitis in children: a report of six cases and review of the literature. *Iowa Orthop J* 2007;27:90–4.
9. Grover R, Grobbelaar AO, Richman PT, Smith PJ. Measurement of invasive potential provides an accurate prognostic marker for giant cell tumor of tendon sheath. *J Hand Surg Br* 1998;23:728–31.
10. Yoo JH, Yang BK, Park JM. Localized nodular synovitis of the knee presenting as anterior knee pain: a case report. *Knee* 2007;14:398–401.
11. Balsara ZN, Stainken BF, Martinez AJ. MR image of localized giant cell tumor of the tendon sheath involving the knee. *J Comput Assist Tomogr* 1989;13:159–62.
12. Jones FE, Soule EH, Coventry MB. Fibrous xanthoma of synovium (giant cell tumor of tendon sheath, pigmented nodular synovitis): a study of one hundred and eighteen cases. *J Bone Joint Surg Am* 1969;51:76–86.
13. Ozalp T, Yercan H, Okcu G, Ozdemir O, Coskunol E, Begue T, Calli I. Giant-cell tumors of the tendon sheath involving the hand or the wrist: an analysis of 141 patients. *Acta Orthop Traumatol Turc* 2004;38:1202–4. [In Turkish]
14. Al Qattan MM. Giant cell tumor of tendon sheath: classification and recurrence rate. *J Hand Surg Br* 2001;26:1:72–5.