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Original Research Article

GIANT CELL TUMOR OF THE TENDON SHEATH IN THE KNEE: AN UNCOMMON ADOLESCENT CHALLENGE

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Abstract:

Introduction: Chronic knee pain in young adults can often be challenging to diagnose due to its wide array of potential underlying conditions. This case report delves into a particularly rare and intricate case involving a18-year-old male who presented with giant cell tumor of the tendon sheath (GCTTS) within the knee joint, an atypical occurrence for this benign lesion. Typically, GCTTS originates from the synovium of joints, bursae, or tendon sheaths, with a higher prevalence in the fingers. However, its manifestation in the knee is exceptionally rare, making this case unique in terms of patient age and tumor size.

Case presentation: The patient's initial presentation consisted of non-specific symptoms, including infra-patellar pain and the development of a growing swelling, posing a diagnostic challenge. Employing magnetic resonance imaging (MRI) played a pivotal role in the identification of a well-circumscribed soft tissue mass. The diagnosis of GCTTS was established through histopathological examination following surgical excision.

Conclusion: This case report aims to raise awareness of the potential occurrence of GCTTS in young patients with knee pain and non-specific symptoms. Early and accurate diagnosis is underscored as essential for appropriate treatment and management, especially given the rarity and diagnostic intricacies and broad differential diagnoses associated with this condition. Favorable prognosis can be achieved following successful surgical excision, when GCTTS is promptly diagnosed and treated in atypical locations, such as the knee joint, demonstrating the significance of heightening awareness among healthcare providers and clinicians to consider GCTTS in patients with knee pain and swelling.

Keywords: giant cell tumor of the tendon sheath, knee pain, diagnostic challenge, young patient

Introduction:

Chronic knee pain in young adults can present a diagnostic dilemma, as it may be attributed to a wide range of conditions. In this case report, we focus on a particularly rare and challenging case of giant cell tumor of the tendon sheath (GCTTS) occurring in the knee joint of a 18year-old male. GCTTS is typically a benign lesion arising from the synovium of joints, bursae, or tendon sheaths, with a higher incidence in the fingers. However, GCTTS in the knee is exceptionally uncommon, and this case is notable for its atypical presentation both in terms of patient age and tumor size. The patient initially presented with non-specific symptoms, including infra-patellar pain and a growing swelling, which posed a diagnostic challenge. Magnetic resonance imaging (MRI) played a crucial role in identifying the well circumscribed soft tissue mass, ultimately leading to the diagnosis of GCTTS through histopathological examination. With a focus on the rarity and diagnostic challenges of this case, we aim to raise awareness about the potential of GCTTS in young patients with knee pain and non-specific symptoms, emphasizing the importance of early and accurate diagnosis for appropriate treatment and management.

Case report:

With the consent of the boy's parents, the study discusses the case of a18-year-old boy who presented with a hidden, gradually enlarging swelling and three months of persistent left knee pain. The pain was exacerbated on activity and localized around the lower front part of the left knee. Upon physical examination, there was tenderness in infrapatellar region and restricted knee range of motion. There were no general symptoms like night sweats, fever, or weight loss, and no enlarged inguinal lymph nodes were detected. Notably, the skin temperature of the left knee was higher than the right one. Blood analysis came back normal. To gain further insights, plain X-rays and magnetic resonance imaging (MRI) were conducted. The X-rays did not reveal any bony abnormality. However, the MRI showed an encapsulated soft tissue lump located in the fat pad just below the patellar tendon (Figure 1). Initially, we considered the possibility of this lump being a lipoma, synovial sarcoma, or fibroma of the tendon. To clarify, an excision biopsy was performed, resulting in the removal of a globular mass measuring approximately 3x2 cm (Figure 2), following a midline incision. Examination of the tissue sample revealed an abundance of mononuclear cells, accompanied by varying numbers of foamy histiocytes, hemosiderin-laden macrophages, and multinucleated giant cells (Figure 3). Notably, there were no signs of mitotic activity or malignant features. Subsequently, the final diagnosis was Giant Cell Tumor of the Tendon Sheath (GCTTS) as per histopathological report.

Post-surgery recovery was successful, marked by the absence of local tenderness, a normal skin temperature of the knee, and the restoration of full range of motion in the knee joint. The patient was able to return to normal life without any issues, and over the course of a 6-month follow-up, there was no evidence of the tumor returning locally (Figure 4).

Discussion:

Giant cell tumors of the tendon sheath (GCTTS) are benign soft tissue growths that typically originate from the synovium-lined tendon sheaths around joints and tendons. The exact cause of GCTTS is still uncertain and may involve various factors like inflammation, trauma, toxins, allergies, clonal chromosomal abnormalities, and aneuploidy (1–2). Jaffe et al described this condition in 1941 (3). They occur most commonly in the fingers, other sites being the knee, elbow, hip and ankle (4), affecting people between the ages of 30 and 50, with female preponderance (5). Usual presentation is nonspecific, making the diagnosis difficult. Patients may present with a painless mass. Rarely, there may also be pain, that exacerbates during movement, which can be attributed to the mechanical irritation caused by the tumor within the tendon sheath, with limitation of range of motion.

GCTTS was initially regarded as an inflammatory disease; however, the aneuploidy in certain cases along with demonstration of clonal chromosomal abnormalities strongly supports a neoplastic origin (4). Antecedent trauma being one of the causes, only 15% of cases presented with history of trauma (6). The diffuse and localized extra-articular forms of pigmented villonodular synovitis (PVNS) are presently referred to as diffuse-type giant cell tumor and giant cell tumor of the tendon sheath (GCTTS), respectively (7).

Most localized giant cell tumors are small (size, 2.0 cm) (5). mass in our case, was much larger than normal occurring size. Tumors are well-circumscribed, lobulated, white to gray in color, with yellowish and brown patches. In contrast, diffuse-type giant cell tumors infiltrate and expand more extensively. These subtypes are now classified as ‘fibriohistiocytic tumors’ in the World Health Organization system of classification of bone and soft tissue tumors (4). The soft tissue mass grows and infiltrates into areas of least resistance, such as the tendon sheath (8).

Plain radiographs are often unhelpful in diagnosing GCTTS, as they may not show bone erosions or soft tissue swelling.

Magnetic resonance imaging (MRI) is a more effective and sensitive diagnostic tool. Typically, MRI features T1- (T1WI) and T2-weighted (T2WI) images, which may present with a homogeneous low signal intensity in GCTTS. Reasoning is that tumors contain dense collagen and hemosiderin-laden macrophages (9).

The hallmark of GCTTS is the presence of multinucleated giant cells scattered within a fibrous and histiocytic stroma. This histological pattern was confirmed in the excised specimen in this case. Additionally, hemosiderin deposits were observed, which are often seen in GCTTS and are indicative of intralesional hemorrhage.

Treatment consists of careful and complete local excision and postoperative radiotherapy. Surgical removal is the treatment of choice for GCTTS. Adequate and proper initial local excision effectively limits the risk of local recurrence (10). Recently, microscopic excision has been prescribed to prevent recurrence. Ikeda et al (11) conducted a study involving 18 patients, where they employed a surgical technique known as microscopic excision to treat Giant Cell Tumor of the Tendon Sheath (GCTTS). Remarkably, out of these 18 patients, only one experienced a recurrence of the tumor. The recurrence was attributed to the fact that microscopic excision was not applied in the case of a patient with a specific subtype of

GCTTS known as the "diffuse-type."

As an additional measure to prevent the recurrence of GCTTS following surgical removal, postoperative radiotherapy was recommended and administered as an adjuvant therapy. This combination of surgical excision and postoperative radiotherapy has been shown to be effective in reducing the risk of the tumor returning. Kotwal et al (12) published a prospective study in which the recurrence rate was only 4%.

GCTTS is a benign type of tumor, but it has a high incidence of recurrence. Recurrence following resection has been a problem, and occurs in 9 to 44% of cases (13).

When evaluating knee soft tissue masses, it is essential to consider a broad differential diagnosis, including synovial sarcoma, lipoma, ganglion cyst, and other benign and malignant soft tissue tumors. Preoperative imaging and histopathological examination are crucial for distinguishing GCTTS from other entities.

Conclusion: This case report underscores the importance of raising awareness about GCTTS in less common locations such as the knee joint. Healthcare providers should consider GCTTS when evaluating patients with knee pain and swelling, especially when clinical and radiological findings suggest a soft tissue mass. Our report also emphasizes the successful surgical management and favorable prognosis associated with this rare condition when diagnosed and treated appropriately. Patient should be followed up regularly for a long time as the condition is associated with high rate of recurrence.

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Figures:

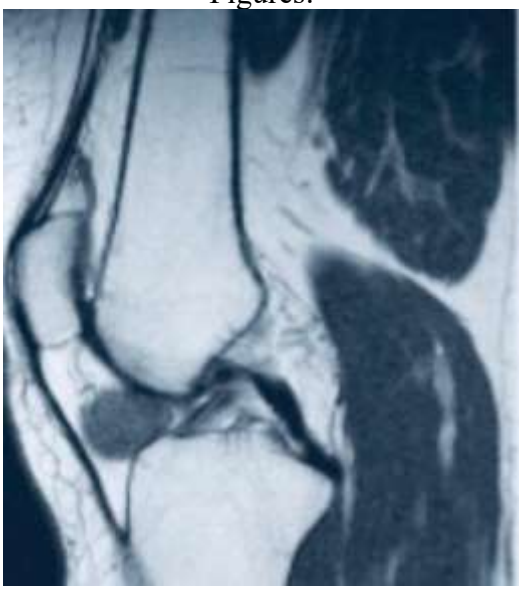

Figure 1. Preoperative MRI showing infrapatellar encapsulated mass

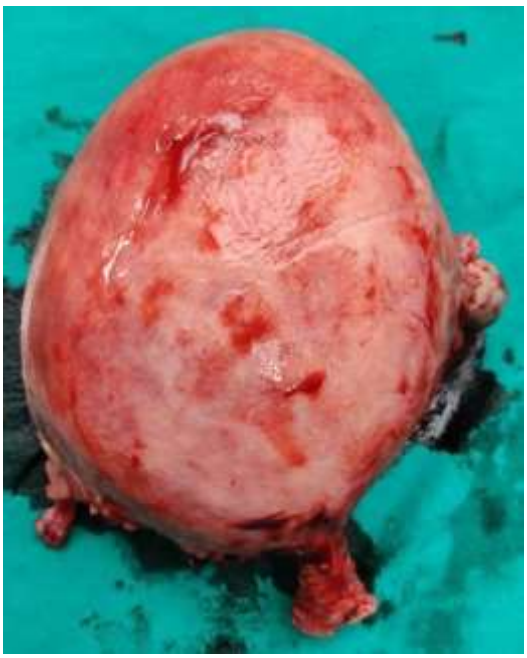

Figure 2. Intraoperative image showing excised mass

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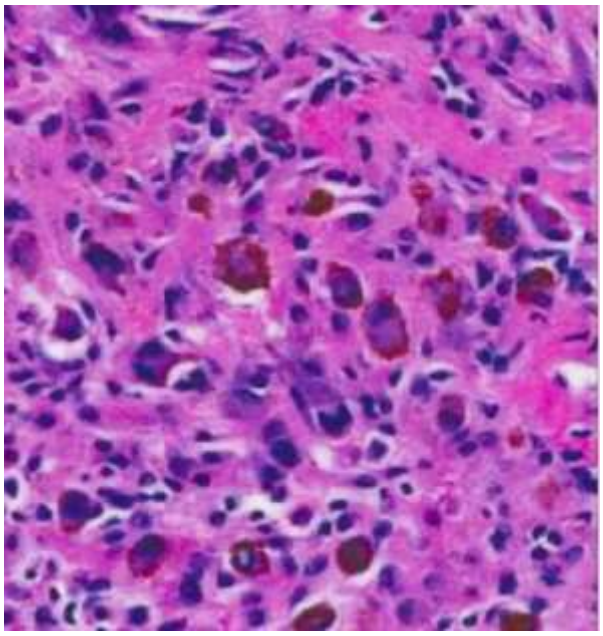

Figure 3. Histopathological image showing abundance of mononuclear cells, accompanied by varying numbers of foamy histiocytes, hemosiderin-laden macrophages, and multinucleated giant cells

Figure 4. 6 months post operative follow up showing no recurrence