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Giant Cell Tumors of the Synovial Sheaths of Tendons: A Case Report

Kamli Abderrahim[1*], A. Boutahar[1], N. Chetahi[1], M. Mimouni[1], M. Tazi, H. Abdellaoui[1], K. Atarraf[1], M. Afifi[1]

[1]Department of Pediatric Surgery, University Hospital Hassan II, Fes, Morocco

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***Corresponding author:** Kamli Abderrahim
Department of Pediatric Surgery, University Hospital Hassan II, Fes, Morocco

Abstract | **Case Report**

***Background*:** Tenosynovial giant cell tumors (TGCT) are benign proliferative lesions arising from the synovial lining of tendon sheaths, joints, and bursae. While they represent the most common softtissue tumors of the hand and fingers in adults, TGCT remain exceptionally rare in the pediatric population. Their etiopathogenesis is poorly understood, and the combination of a slow clinical course with low clinical suspicion frequently results in diagnostic delay in young patients. ***Case Presentation*:** We report the case of a 13-year-old boy presenting with a progressively enlarging, painful palmar swelling of the right middle finger, evolving over four years. Clinical examination, ultrasonography, and MRI were performed. Surgical excision was carried out under general anesthesia, and histological examination confirmed the diagnosis of tenosynovial giant cell tumor (WHO Grade 0). ***Conclusion*:** TGCT should be included in the differential diagnosis of any firm, long-standing digital mass, regardless of patient age. MRI is the imaging modality of choice, and complete surgical excision remains the standard treatment. Long-term follow-up is mandatory given the significant risk of local recurrence. This pediatric case highlights the diagnostic challenges and the importance of early surgical management to prevent functional impairment.

Keywords: Tenosynovial Giant Cell Tumor, Tendon Sheath, Pigmented Villonodular Synovitis, Hand Tumor, Pediatrics, Surgical Excision.

INTRODUCTION

Giant cell tumors of the synovial sheaths of tendons, also referred to as tenosynovial giant cell tumors (TGCT) or pigmented villonodular synovitis, are benign proliferative lesions arising from the synovial lining of tendon sheaths, joints, and bursae. First described by Chassaignac in 1852 [1], they represent the most common soft-tissue tumors of the hand and fingers in the general adult population. While this designation reflects their relative frequency among hand tumors, they nonetheless remain an uncommon entity when considered in the broader context of all soft-tissue neoplasms.

In the pediatric population, TGCT are exceptionally rare. The vast majority of reported cases affect adults, with a peak incidence between the third and fifth decades of life, and a slight female predominance [4]. Occurrence in children and adolescents is infrequent and accounts for only a small proportion of published cases [6]. This rarity, combined with the often slow and insidious clinical course, frequently leads to diagnostic delay in young patients, as lesions may be present for months or even years before medical attention is sought.

From a diagnostic standpoint, the workup relies on a combination of clinical examination, imaging, and histopathology. Clinically, TGCT typically presents as a firm, slowly enlarging, painless or mildly painful mass on the palmar aspect of a finger. Ultrasonography serves as a useful first-line modality, demonstrating a well-defined, vascularized soft-tissue formation [2]. Magnetic resonance imaging (MRI) is the gold standard for pre-operative characterization, offering superior soft-tissue contrast and revealing the pathognomonic low-signal areas on T1- and T2-weighted sequences, corresponding to hemosiderin deposition [3]. Definitive diagnosis, however, rests on histopathological examination, which demonstrates a characteristic proliferation of mononuclear synovial cells, multinucleated osteoclast-like giant cells, foamy histiocytes, and hemosiderin-laden macrophages [2, 3].

Therapeutically, complete surgical excision remains the cornerstone of treatment. Despite its benign nature, TGCT displays locally aggressive behavior, with reported local recurrence rates ranging from 10 to 44% following surgery [4, 5]. Incomplete resection and tumor multifocality are the main risk factors for recurrence. In recent years, targeted molecular therapy with

pexidartinib, a CSF1R inhibitor, has emerged as a promising option for adult patients with advanced or unresectable disease [7], though its use in the pediatric setting remains to be established.

We herein report the case of a 13-year-old boy presenting with a tenosynovial giant cell tumor of the right middle finger, managed at the Department of Pediatric Surgery of the University Hospital Hassan II, Fes, Morocco, and discuss the diagnostic and therapeutic specificities of this rare pediatric entity.

Case Report

A 13-year-old boy was referred to our department for a painful swelling of the palmar aspect of the right middle finger, which had been progressively increasing in volume over four years. The patient denied any history of trauma. Personal and family histories were unremarkable.

Physical examination revealed an oval mass with a bilobed appearance, measuring approximately 3 cm in its greatest dimension — notably large for a localized TGCT — located on the palmar surface of the right middle finger. The mass was firm in consistency, adherent to the deep plane, suggesting close relation to the underlying tendon sheath, with no inflammatory signs on the overlying skin. Digital flexion was moderately limited. Regional lymph nodes were not enlarged.

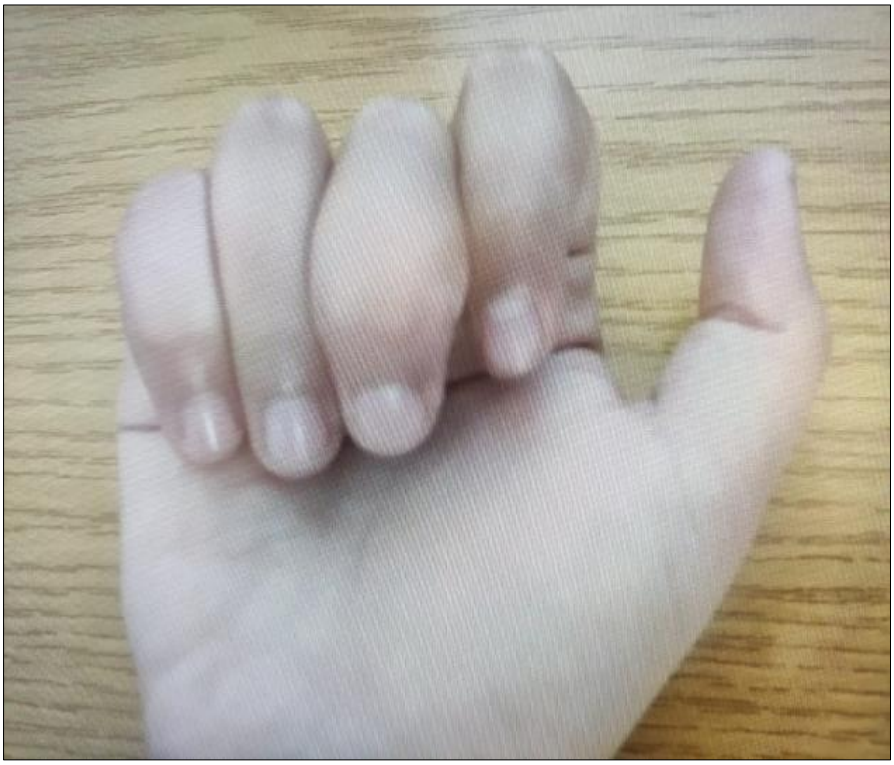

Figure 1: Clinical presentation: bilobed palmar swelling of the right middle finger

Ultrasonography of the soft tissues demonstrated an oval, polylobulated formation with well-defined margins and a longitudinal major axis, showing marked vascularity on Doppler examination. Magnetic resonance imaging (MRI) of the right hand revealed a well-circumscribed mass with heterogeneous signal intensity. On T1-weighted sequences, the mass demonstrated predominantly intermediate signal with focal hypointense areas. On T2-weighted sequences, heterogeneous signal was noted with characteristic areas of marked hypointensity on both T1 and T2, corresponding to hemosiderin deposition — a pathognomonic feature of TGCT. Post-gadolinium sequences demonstrated marked and heterogeneous enhancement. The relationship of the mass to the flexor tendon sheaths was clearly delineated, with no bone erosion or articular involvement.

Plain radiographs of the right hand showed no calcifications, bone lysis, or periosteal reaction.

The patient underwent surgical excision under general anesthesia with a pneumatic tourniquet. Intraoperatively, the mass was found to be intimately adherent to the sheaths of the flexor digitorum superficialis and profundus tendons, without macroscopic tendon invasion. A complete en-bloc excision was performed, including the affected portion of the synovial sheath, with preservation of the flexor tendons and neurovascular structures.

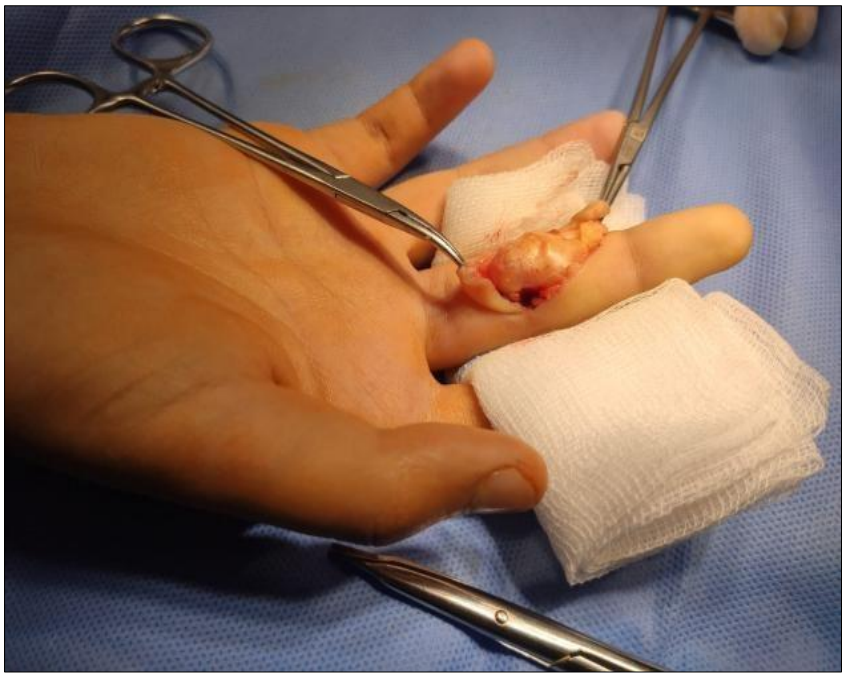

Figure 2: Intraoperative view: en-bloc excision of the tenosynovial giant cell tumor from the right middle finger

Histological examination confirmed the diagnosis of tenosynovial giant cell tumor. The surgical specimen showed a proliferation of mononuclear synovial-type cells admixed with multinucleated osteoclast-like giant cells, foamy histiocytes, hemosiderin-laden macrophages, and chronic inflammatory infiltrates. No mitotic figures, necrosis, or features suggestive of malignant transformation were identified (WHO Grade 0) [2].

The immediate post-operative course was uneventful. Hand physiotherapy was initiated two weeks after surgery. At the six-month follow-up, the patient reported complete resolution of pre-operative pain with no clinical evidence of local recurrence, and near-complete restoration of digital mobility and grip strength. Given the well-documented risk of local recurrence, which ranges from 10 to 44% [4, 5], the patient has been enrolled in a structured long-term surveillance program with annual clinical and ultrasonographic assessments.

DISCUSSION

Giant cell tumor of the tendon sheath is the localized form of TGCT and represents the second most common benign soft-tissue tumor of the hand, after ganglion cysts. It predominantly affects the palmar aspect of the fingers, particularly the index and middle fingers, and occurs most frequently in women between the third and fifth decades of life [4]. Pediatric cases, such as ours, are comparatively rare and may be associated with significant diagnostic delays, as illustrated by the four-year evolution prior to presentation in our patient [6]. Furthermore, the tumor size in our patient (3 cm) was notably larger than typically described in the literature, likely reflecting the prolonged diagnostic delay inherent to pediatric presentations of this entity.

The clinical presentation of GCT-TS is typically insidious, with lesions growing slowly over months to years before prompting medical consultation. Clinically, it presents as a firm, mildly painful mass that may progressively restrict digital motion. The differential diagnosis of a firm digital mass includes ganglion cyst, lipoma, fibroma of the tendon sheath, epidermal inclusion cyst, glomus tumor, and — less commonly — malignant soft-tissue tumors such as synovial sarcoma. Among these, fibroma of the tendon sheath deserves particular attention as it is the most important histological differential diagnosis, sharing similar clinical features but distinguished by its dense, collagenous stroma and absence of giant cells on pathological examination [3].

From an imaging standpoint, ultrasonography serves as a useful first-line modality, providing dynamic morphological assessment and Doppler characterization of lesion vascularity, as demonstrated in our case. MRI remains the gold standard for pre-operative characterization, offering superior soft-tissue contrast and enabling identification of the pathognomonic low-signal areas on T1- and T2-weighted sequences, corresponding to hemosiderin deposition within the tumor [3]. These imaging features, combined with the clinical presentation, strongly orient the diagnosis prior to histological confirmation.

Complete surgical excision is the definitive treatment for localized GCT-TS. The primary surgical objective is to achieve negative resection margins while preserving the integrity of the flexor tendons and

neurovascular structures, as performed in our patient. Despite its benign classification, TGCT demonstrates locally aggressive behavior, and local recurrence rates ranging from 10 to 44% have been reported following surgical resection, attributed to microscopic residual disease, tumor multifocality, or intraoperative cell dissemination [4, 5]. These figures underscore the importance of meticulous surgical technique and thorough inspection of the entire synovial sheath during excision.

Regarding emerging systemic therapies, pexidartinib, a selective inhibitor of the colony-stimulating factor 1 receptor (CSF1R), was approved by the US Food and Drug Administration (FDA) in 2019 for adult patients with symptomatic, advanced TGCT not amenable to surgical resection [7]. However, its clinical application is currently limited by significant hepatotoxicity, which has been observed in a subset of patients and requires rigorous liver function monitoring throughout treatment. Although not applicable to this pediatric case, this targeted therapy represents a meaningful paradigm shift in the management of diffuse and recurrent forms of the disease, and may hold future relevance for refractory pediatric cases as evidence accumulates.

Conclusion

This case underscores the importance of considering tenosynovial giant cell tumor in the differential diagnosis of any firm, long-standing digital mass in a child or adolescent. The pediatric presentation is particularly challenging given the rarity of the entity and the insidious clinical course, which frequently result in prolonged diagnostic delay and, consequently, larger tumor size at the time of diagnosis. MRI is the imaging modality of choice for pre-operative assessment, while histopathological examination remains the gold standard for definitive diagnosis. Complete surgical excision with preservation of adjacent neurovascular structures is the treatment of choice. However, the non-negligible risk of local recurrence necessitates strict and prolonged follow-up, extending well beyond the early post-operative period. Emerging targeted therapies such as pexidartinib offer promising perspectives for advanced or refractory forms, although their safety and efficacy profile in the pediatric population remain to be established.

Patient Consent

Written informed consent was obtained from the patient's legal guardian for the publication of this case report and any accompanying images. A copy of the written consent is available from the corresponding author upon request.

Conflicts of Interest: The authors declare no conflicts of interest relevant to this publication.

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