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Case Report

Pigmented villonodular synovitis of the flexor hallucis longus tendon: A rare cause of leg pain in a 12-year-old girl ☆

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ABSTRACT

Pigmented villonodular synovitis is an uncommon benign neoplastic proliferation associated with the synovium, bursa, or tendon sheaths; most commonly occurring in the third to fourth decade of life. It is rare in children and may be painful or painless. Magnetic resonance imaging is the diagnostic study of choice. In this report, the radiologic, ultrasound, and magnetic resonance imaging findings of pigmented villonodular synovitis of the flexor hallucis longus in a 12-year-old girl are discussed. We briefly review the surgical findings as well. To our knowledge, this is the first case report that simultaneously synthesizes the imaging findings of 3 diagnostic imaging modalities for optimal visualization and is the youngest reported case of pigmented villonodular synovitis of the flexor hallucis longus tendon.

Introduction

Pigmented villonodular synovitis (PVNS) is a rare benign neoplastic proliferative disorder of the synovial joint, bursa, or tendon sheath. PVNS originates secondary to chromosomal translocations that lead to overexpression of colony-stimulating factor 1 (CSF-1), which leads to clusters of aberrant cells that create focal areas of soft tissue hyperplasia in the synovial cells lining joints or tendon sheaths. PVNS most commonly presents in the knee joint of middle-aged individuals, usually in the third or fourth decade of life, but can also be seen in the hip, ankle, shoulder, and elbow [1]. PVNS can be further classified into localized or diffuse with an intra-articular or extra-articular origin, respectively. The diffuse extra-articular subtype of PVNS is commonly

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misdiagnosed due to its slow-growing process and atypical presentation [2]. Due to the insidious onset of symptoms, diagnosis of PVNS is commonly delayed. Diagnosis often relies on physical exam, characteristic imaging findings, and histologic confirmation [1]. The lack of literature on PVNS in the pediatric population highlights the importance of documenting cases of this disease to avoid misdiagnosis and delayed treatment [4]. We present a case report including the imaging workup of PVNS of the flexor hallucis longus (FHL) tendon in a 12-year-old girl, a location which has rarely been reported in the previous literature [2]. Informed consent was obtained from the legal guardian of the patient regarding writing and publishing this article and associated images.

Case report

A 12-year-old girl presented for evaluation of right ankle and distal leg pain and swelling which had been present and increasing for 1 year. To our knowledge, the patient did not present to any other facility previously and had not undergone any extensive workup during her 1 year of symptoms. The pain was dull aching in nature and 4/10 in severity. The swelling had gradually increased in size and was localized to the posteromedial aspect of the ankle and distal leg. On examination, the swelling was about 8 × 3 cm in size, nontender, with a diffuse border, and cystic consistency. The swelling was anterior to the tendoachilles, appeared to be in the subcutaneous plane, and transillumination test was negative which aided in ruling out other cystic masses such as a ganglion cyst, prompting further investigation through imaging. The patient had no significant medical history and did not recall any antecedent trauma.

Radiographs of the tibia and fibula demonstrated an ovoid soft tissue or fluid density collection within Kager's fat pad along the posterior ankle without any osseous abnormalities (Fig. 1). This initial imaging modality provided general features of the mass to help guide further imaging. Ultrasound demonstrated a nonvascular hypoechoic fluid collection with hyperechoic layering debris (Fig. 2). The lesion measured 8.0 × 3.4 × 3.4 cm proximal to distal, anterior to posterior, and medial to lateral dimensions, respectively. The FHL tendon was seen coursing through the center of the fluid collection (Fig. 2). Given the size of the lesion, subfascial location, and intimate association with the FHL tendon, magnetic resonance imaging (MRI) was obtained. The MRI demonstrated an ovoid T2 hyperintense cystic lesion with the FHL tendon again seen coursing through the cystic lesion (Fig. 3). Axial T1 imaging demonstrated hypointensity of the lesion (Fig. 4). Postcontrast MRI demonstrated no internal enhancement with only thin peripheral enhancement of the cyst wall most consistent with a benign cystic lesion (Fig. 5). These MRI findings further supported suspicion of PVNS and assisted in preoperative planning.

The patient underwent surgical excision biopsy of the lesion through a posteromedial approach to the ankle and hindfoot, demonstrating a cystic lesion with a brown capsule and yellow-brown granules (Fig. 6). The lesion surrounded the FHL tendon and was excised en masse along with a small part of the FHL tendon proximal to the medial malleolus. The FHL tendon was attached to the flexor digitorum longus tendon proximal and distal to the lesion to avoid any functional deficits. The pathology report confirmed the lesion to be PVNS. The patient followed a normal course of recovery with no strength or motion deficits and did not have any complications. There has been no recurrence of the lesion at 1 year postoperative.

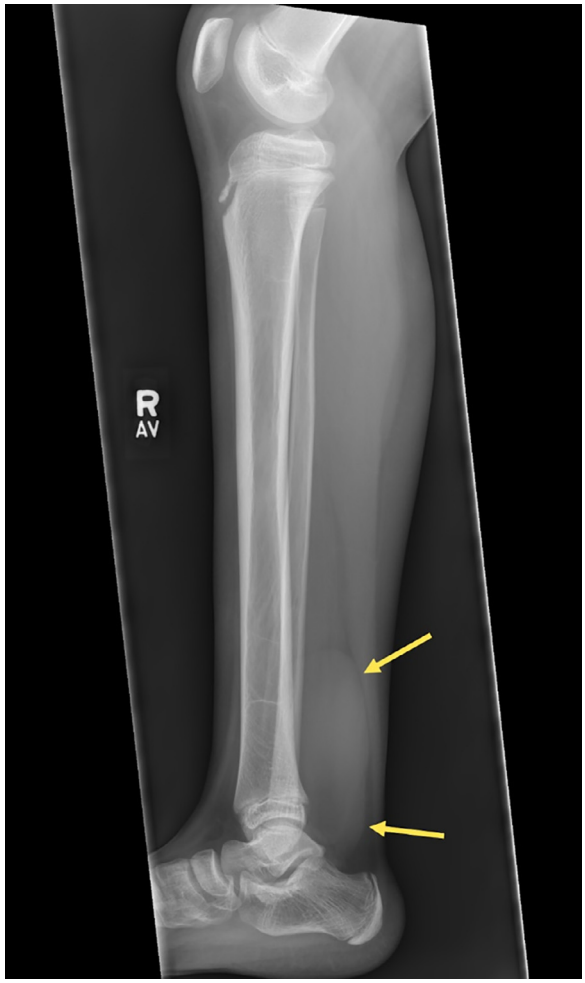

Fig. 1 – Lateral radiograph of the tibia and fibula demonstrating a fluid density lesion in Kager's fat pad along the posterior ankle (yellow arrows).

Discussion

In this report, we describe the imaging findings of PVNS of the FHL tendon in a 12-year-old girl presenting with a painful swelling of the leg. PVNS is a rare benign proliferative disorder that primarily affects the synovium of joints, bursa, or tendon sheaths [1,2]. PVNS may be categorized as localized or diffuse with the knee being the most common location [1]. PVNS is typically seen in patients 20-50 years of age and is less commonly diagnosed in the pediatric population. The ankle has

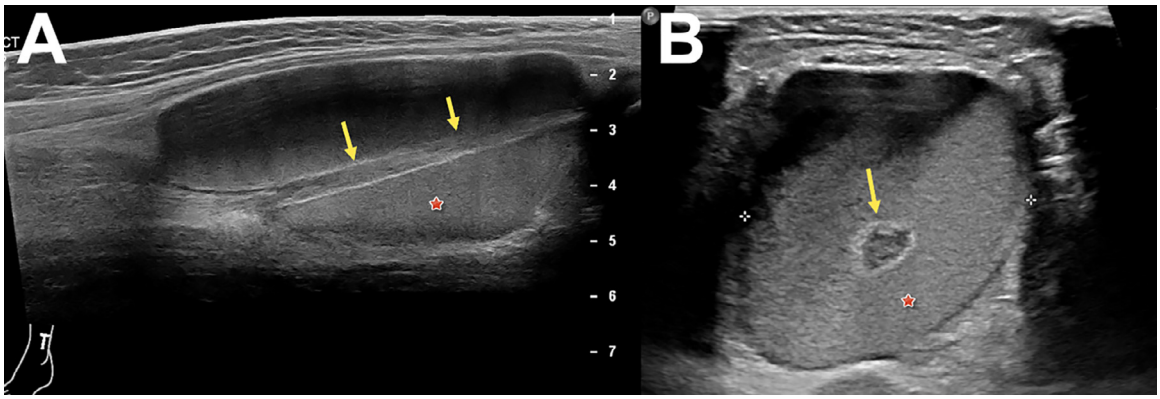

Fig. 2 – Sagittal (A) axial and (B) ultrasound images of the ankle lesion demonstrating a hypoechoic nonvascular fluid collection with layering debris (red star) and the flexor hallucis longus tendon coursing through the center of the lesion (yellow arrows).

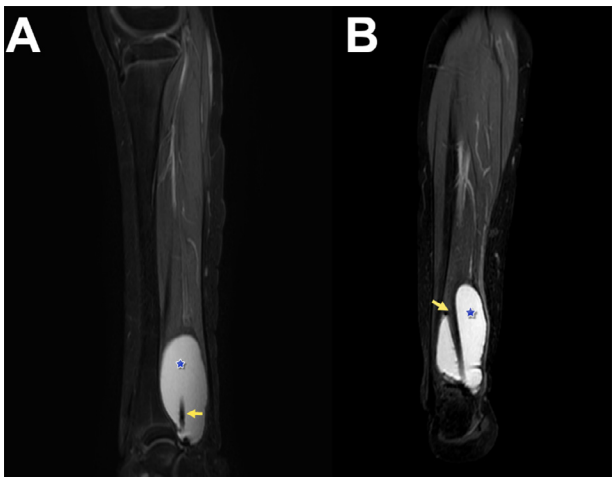

Fig. 3 – Sagittal (A) coronal T2 fat-saturation and (B) magnetic resonance imaging cuts demonstrating an ovoid T2 hyperintense cystic lesion (blue star) with the flexor hallucis longus coursing through the cystic area (yellow arrow).

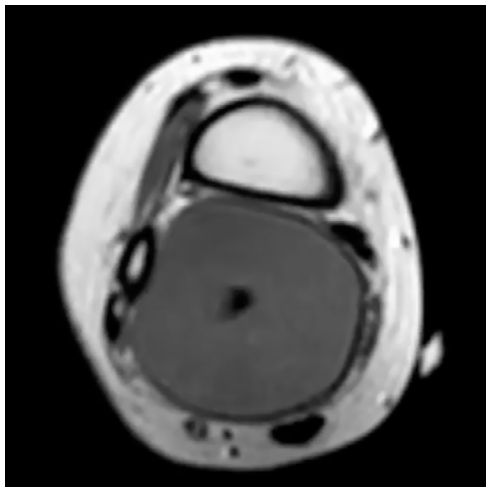

Fig. 4 – Axial T1 magnetic resonance imaging cut demonstrating a T1 hypointense cystic lesion with the flexor hallucis longus coursing through the cystic area.

been reported in adults and children with both localized and diffuse PVNS [1–4]. Although radiographs often appear normal and are not considered diagnostic, some will demonstrate degenerative changes on both sides of the joint in cases of diffuse PVNS and pressure erosions in localized PVNS [1]. Ultrasound can demonstrate a nonvascular hypoechoic cystic mass with layering hyperechoic debris as we saw with the patient discussed in this case report. MRI is the most sensitive imaging modality for PVNS which typically demonstrates a pathognomonic low signal intensity on T2 weighted sequences along with signal loss on gradient echo sequences [1,5]. This signal loss is likely due to iron in the hemosiderin deposition resulting in disruption of the local magnetic field and demonstrating susceptibility artifacts [1,6]. In general, MRI will typically yield a well-circumscribed mass for localized PVNS and ill-defined soft tissue masses for diffuse PVNS. These various imaging modalities are crucial for diagnosis and management of PVNS.

There is currently no gold standard for treating PVNS lesions and treatment should be patient-centered due to the variable location and extent of the lesions. The wide spectrum of treatment ranges from observation to open vs arthroscopic or endoscopic surgical excision and synovectomy depending on if the lesion is localized to the joint or if there is extra-articular extension into adjacent soft tissues [1]. Surgery alone has demonstrated a recurrence rate of up to 50% mostly attributable to diffuse extra-articular PVNS [7]. Guo et al. [3] reported good to excellent outcomes in patients with localized intra-articular PVNS of the ankle with no recurrence following arthroscopic synovectomy. There was recurrence following arthroscopic and open synovectomy in their cohort of

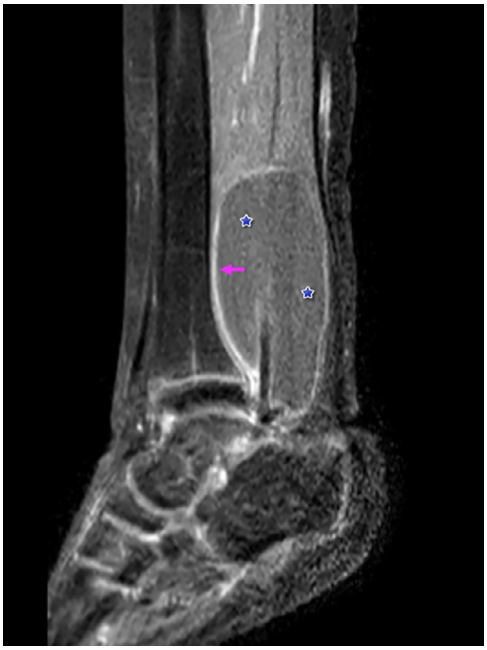

Fig. 5 – Sagittal T1 fat-saturation post-contrast magnetic resonance imaging cut demonstrating no internal enhancement of the cystic lesion (blue star) with only thin peripheral enhancement of the cyst wall (purple arrow).

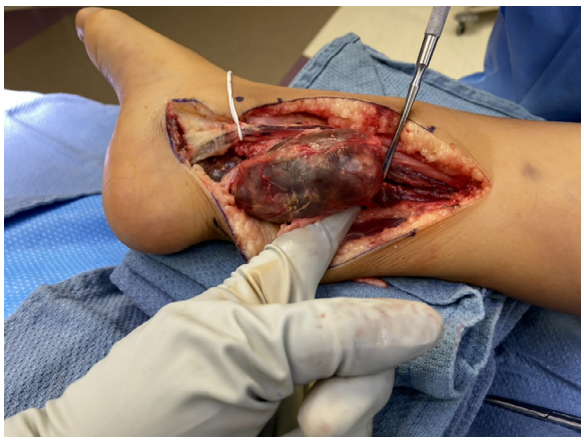

Fig. 6 – Intraoperative photograph demonstrating a brown cystic swelling encasing the flexor hallucis longus tendon within the posteromedial soft tissues of the ankle with a white vessel loop gently retracting the posterior tibial nerve, artery, and vein.

extra-articular lesions with adjacent soft tissue extension. Radiotherapy alone or in an adjuvant fashion has been previously used with good outcomes compared to surgical excision alone. Radiation therapy has fallen out of favor due to negative side-effects and recent onset of targeted immunotherapy [1,8]. Recent treatment modalities have targeted the CSF-1 pathway via monoclonal antibodies or tyrosine kinase inhibitors [7].

Ultimately, determining treatment of PVNS relies heavily on adequate imaging modalities to allow for complete excision of the lesion and provide the best clinical outcomes for patients [1]. In our case specifically, MRI was not only used for diagnostic purposes but to determine the extent of the lesion and aid in preoperative planning. We believe that an open excision of the lesion was deemed necessary to decrease the risk of recurrence due to the size and location of the lesion demonstrated on MRI. Removal of part of the FHL tendon was performed due to adherence of the mass to the tendon. The FHL tendon was transferred to the flexor digitorum longus tendon to preserve its function and decrease the risk of complete FHL deficit. The patient in our case report has not had a recurrence of the lesion upon follow-up and has not had any reported functional deficits of the foot and ankle.

Conclusion

Although PVNS is a rare condition seen in the pediatric population, it should not be excluded from differential diagnosis in young patients presenting with vague joint pain or swelling. With an insidious onset of symptoms, prompt diagnosis of PVNS can be achieved through clinical exams and appropriate imaging. MRI is the gold standard imaging modality for this disease as it not only allows for diagnosis but also guides the course of treatment and further management.

Patient consent

Informed consent was obtained from legal guardians for publication of this study and associated images in addition with an approval of ethics.

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