A rare case of extra-articular pigmented villonodular synovitis of the knee in a child

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ABSTRACT

Introduction: Pigmented villonodular synovitis (PVNS) is a benign proliferative disorder that affects the synovial joint, tendon sheaths, and bursa membranes. It is a rare condition of unknown etiology, with only around 100 cases in the pediatric population reported in the literature. To the authors' knowledge, this is the first case of extra-articular PVNS of the knee in a child.

Case Report: We report a case of a 10-year-old boy, with localized extra-articular PVNS of the knee. The child presented with a 4-month history of a slow growing mass in the lateral-superior aspect of the left knee, which appeared after a minor trauma. The child had no pain, joint effusion or movement impairment. Magnetic resonance imaging (MRI) revealed a cystic lesion with multiple septae, with no specific diagnosis. Excisional biopsy was performed, and the histopathology revealed the diagnosis of PVNS.

Conclusion: Pigmented villonodular synovitis is a rare disorder but can present in children as an intra-articular or extra-articular lesion. The diagnosis can be difficult, as the clinic is unspecific, and exams may not be diagnostic. The preferred treatment is surgical excision, but recurrence can occur.

Keywords: Knee, Pediatric, Tumor, Villonodular synovitis

INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a rare, benign, proliferative neoplastic condition characterized by hypertrophy of the synovial membrane with pigmentation secondary to hemosiderin deposition[1]. Pigmented villonodular synovitis was first described by Chassaignac et al. in 1852 [2] and since then categorized in localized and diffuse forms of synovial involvement [3]. It can also be characterized in intra-articular (involves the synovium of the joint) and extra-articular (involves the bursa or tendon sheath) [1, 4], being the localized form the most common, usually extra-articular and histologically identical to a tendon sheath giant cell tumor [3]. Localized PVNS involves mostly the hands and wrist (65–89% of cases) and frequently manifests as a soft-tissue mass and pain, whereas joint dysfunction or swelling are unusual (0–4%) [4].

Pigmented villonodular synovitis has an incidence of 9.2 and 1.8 cases per 1,000,000 people in the extra-articular and intra-articular forms of the disease, respectively [4], and typically presents between the third and fourth decades of life [1]. The association with other disorders is rare, and polyarticular or multifocal involvement is unusual [3], being more frequent in immature patients, an otherwise rarely affected age group [4]. The literature regarding PVNS in the pediatric population is limited to small series [5–7] and few case
reports, with only around 100 cases in the pediatric population reported in the literature [5–12].

CASE REPORT

The authors present a case of a 10-year-old boy, previously healthy, who was directed to the emergency department due to a left knee mass with four months of evolution.

The mother reported a fall with minor trauma to the left knee in the week prior to the mass being detected and starting to grow.

The mass was located in the lateral-superior part of the knee and did not seem related with the joint as there was no joint effusion, pain, or movement impairment (Figure 1). The mass was movable, soft, and painless. There was no story of drainage. The patient underwent an MRI scan, which revealed a cystic lesion with $39 \times 31 \times 17$ mm with multiple septae (Figure 2), between the iliotibial tract, external knee retinaculum, and epidermis, with no characteristics of aggressive behavior but presenting contrast capture in the septae. There were no signs of bone erosion.

According to the MRI report, it was decided to perform an excisional biopsy. Macroscopically, the lesion appeared a brownish inflamed synovium (Figure 3), with no communication with the knee joint. The posterior aspect had continuity with the iliotibial tract/external lateral knee retinaculum and an excision biopsy was performed. The histopathology revealed a proliferation of synovial hyperplastic tissue, in villus/papillae pattern (Figure 4), composed of round to ovoid cells with central nuclei and no cellular atypia or mitotic figures, with histiocytes and hemosiderophagus, compatible with PVNS. The proliferation was Vimentin positive and smooth muscle actin negative (Figure 5).
The surgical scar healed uneventful, and the patient shows no sign of recurrence at six months post-operation. The patient maintains follow-up and the family was warned of the risk of recurrence.

DISCUSSION

The etiology of PVNS is still unknown [5, 9, 13], being probably multifactorial. History of trauma has been reported inconsistently in several studies, between less than one-third [13] to more than half [4] of patients. Our patient had a traumatic event prior to the appearance of the mass, but the association is difficult to prove, as the first observation in our center was four months after the setting of the mass.

Intra-articular PVNS may involve any joint, but usually affects large joints such as knee, hip, ankle, or shoulder [7]. Extra-articular PVNS is most frequent in the hands and wrists, being the most second common soft tissue mass in this location, with rare involvement of the knee [4]. The knee is the most involved joint in the pediatric population [7], being the cases in published literature all intra-articular PVNS, either localized or diffused [5–7, 9–12]. To the authors’ knowledge this is the first published case of extra-articular PVNS around the knee in a child.

The diagnosis of PVNS is usually delayed, due to different nonspecific clinical signs and symptoms [12]. The typical presentation of PVNS in a child is chronic recurrent joint pain or swelling, and multiple medical observations [10], making differential diagnosis with infectious or rheumatological pathologies. The initial X-rays are negative in a high percentage of cases [13]. Magnetic resonance imaging has been recognized as the best imaging method for diagnosing PVNS [14], as it allows for a differentiation between the diffuse and localized form and the evaluation of the extension of bone and soft-tissue involvement [5]. The high hemosiderin content causes the lesion to appear as a spotty or extensive low signal on T1- and T2-weighted images, classically described as “dark on dark” [1]. However, studies reveal that around 50% of the cases are not diagnosed correctly before surgery [11]. In our case, the MRI was not diagnostic, but excluded intra-articular extension.

The treatment goal of PVNS is to eradicate all abnormal synovial tissue [1], surgical excision being the preferred method of treatment for all forms of PVNS [4]. Other treatment options include radiation therapy, pharmaceutical modulation of the disease, or a combination of these approaches [4]. Recurrence after the initial surgery is a major concern, particularly in large joints and for the diffuse type of PVNS [15], with recurrence rates reaching around 46% [13]. In the localized form of PVNS, as in our case, marginal excision is usually the treatment of choice with good results [8].

CONCLUSION

Pigmented villonodular synovitis in a child is a rare condition but should be considered as a differential diagnosis for joint effusion or an isolated tender mass. Magnetic resonance imaging is the most sensible exam, but does not always provide the exact diagnosis. An excision biopsy and long-term follow-up are recommended to decrease the risk of recurrence and for early detection, in order to preserve the joint and structures involved.

REFERENCES


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Author Contributions

Raquel Teixeira – Conception of the work, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

João Pimentel – Acquisition of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Susana Norte – Acquisition of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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