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Case Report

A Rare Case of Pigmented Villonodular Synovitis of the Knee Joint in a Pediatric Patient with Morphological Abnormality

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Abstract

A 13-year-old female with morphological features refers to an uncommon genetic syndrome. The patient has a history of congenital heart disease (pulmonary valve stenosis) and presented with right knee swelling that began 2 years after the trauma. The swelling has gradually increased in size, movement limitation, and pain. Blood investigations showed iron-deficiency anemia, a normal white cell count, and a platelet level. The C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) levels were slightly elevated later; both CRP and WBC were normal. A biopsy and MRI reported suspicion of a giant cell tumor. Genetic investigations for pediatric and adult patients might be essential for further understanding of this condition. Radiotherapy, when justified, could be a treatment option for certain patients who have undergone incomplete surgical resection. Early detection is necessary to avoid joint damage. MRI is the golden diagnostic; biopsy is not necessary with MRI availability.

Keywords: Knee Joint, Pediatric, Pigmented Villonodular synovitis, Synovial tumors.

حالة نادرة من التهاب الغشاء الزليلي الخشبي المصطبغ في مفصل الركبة لدى مريض أطفال يعاني من خلل مورفولوجي

الخلاصة

الحالة تخص أنثى تبلغ من العمر 13 عاما ذات سمات مورفولوجية تشير إلى متلازمة وراثية غير شائعة. المريضة لديها تاريخ من أمراض القلب الخلقية (تضيق الصمام الرئوي) مع تورم في الركبة اليمنى الذي بدأ بعد عامين من ظهور الأعراض. زاد التورم تدريجيا في الحجم مع تقييد الحركة والألم. أظهرت فحوصات الدم فقر الدم الناجم عن نقص الحديد، وعدد الخلايا البيضاء الطبيعي، ومستوى الصفائح الدموية. ارتفعت مستويات البروتين التفاعلي C (CRP) ومعدل ترسيب كريات الدم الحمراء (ESR) قليلا في وقت لاحق. كان كل من CRP و WBC طبيعيين. أفادت الخزعة والتصوير بالرنين المغناطيسي عن الاشتباه في وجود ورم خلية عملاقة. قد تكون الفحوصات الجينية للمرضى الأطفال والبالغين ضرورية لمزيد من فهم هذه الحالة. يمكن أن يكون العلاج الإشعاعي، عندما يكون مبررا، خيارا علاجيا لبعض المرضى الذين خضعوا لعملية استئصال جراحية غير مكتملة. الكشف المبكر ضروري لتجنب تلف المفاصل. التصوير بالرنين المغناطيسي هو التشخيص الذهبي. الخزعة ليست ضرورية مع توفر التصوير بالرنين المغناطيسي.

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INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a rare, non-cancerous condition that causes the lining of the joints to grow too much, mostly affecting the large joints in adults [1]. The main characteristic feature of PVNS is the synovial hyperplasia and deposition of hemosiderin [2]. The common clinical features of PVNS are pain, joint swelling, and limited movement [2]. However, the occurrence of PVNS in children is rare and might correlate with other morphological abnormalities. There is no accurate estimated incidence of PVNS in pediatrics. However, the overall incidence of PVNS is less than 2/1,000,000, with more than 60% involvement of the knee joint and fewer than 50 cases of PVNS involvement in pediatrics [1-3]. Pathology could be easily misdiagnosed with other joint pathologies such as idiopathic juvenile arthropathy and septic arthritis [3, 4]. The etiology of PVNS is still unknown [2-4]. The presentation of PVNS might be in the form of the localized type or more joints in the form of the diffuse type. It is more commonly presented as a pedunculated nodule [4]. Early diagnosis and treatment are important to avoid progressive destructive joint damage in otherwise healthy joints [5]. Most children were misinterpreted initially as having septic arthritis or juvenile rheumatoid arthritis [4,6]. Reports addressing PVNS in resource-constrained settings, such as Iraq, are notably scarce. This absence of comprehensive data represents a critical gap in knowledge, particularly for those with atypical presentations or associated abnormalities [3,7]. The main objective of this case presentation is documentation of a rare case of PVNS in a female pediatric age group with features of morphological abnormality. In addition to that, what are the challenges in diagnosing PVNS in low-resource settings, and what are the potential implications of rare morphological abnormalities on the clinical course and management of PVNS in pediatric patients? To our

knowledge, this is the first published case of PVNS around the knee in a female child with a facial appearance abnormality related to a congenital syndrome in Iraq. This case report is based on clinical assessment, radiological and histopathological examinations, and supported by a review of related articles, primarily case reports. These reports were evaluated using the CARE 2013 checklist for case reports. The literature search was limited to the last 10 years (2014–2025). Articles that describe PVNS in the knee in patients younger than 17 years old (school age) and those describing cases with congenital or chromosomal abnormalities were included. Articles reporting PVNS in other joints were excluded.

Case Presentation

Patient information and clinical finding

A 13-year-old female with morphological features refers to an uncommon genetic syndrome! With a history of congenital heart disease (pulmonary valve stenosis). The first facial morphological sign referred to Noonan syndrome presented with right thigh and knee swelling for 2 years after trauma (Figure 1).

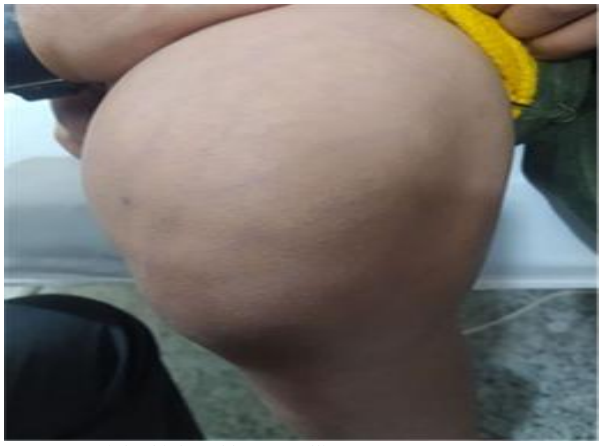

Figure 1: Right thigh and knee swelling.

The swelling has gradually increased in size, movement limitation, and pain. Because of these features, the patient unfortunately left school because of students bullying! The patient history revealed suspicion of a mandibular tumor, which gave the patient these features. Unfortunately, there was a lack of her clinical data and a sort of family neglect! The family lives in a rural area with a low socio-economic level. Which could be the main reason for their bad follow-up.

Diagnostic assessment

Blood investigations showed iron-deficiency anemia, a normal white cell count, and a platelet level. The C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) levels were slightly elevated later; both CRP and WBC were normal. A biopsy done by a radiologist from the anterior aspect of the right thigh reported suspicion of a giant cell tumor of the tendon sheath. MRI reported. The hypoechoic collections were located anterior to the distal femoral shaft, suprapatellar region, and medial aspect of the knee with projections and features of hemosiderin deposition (Figures 2 and 3).

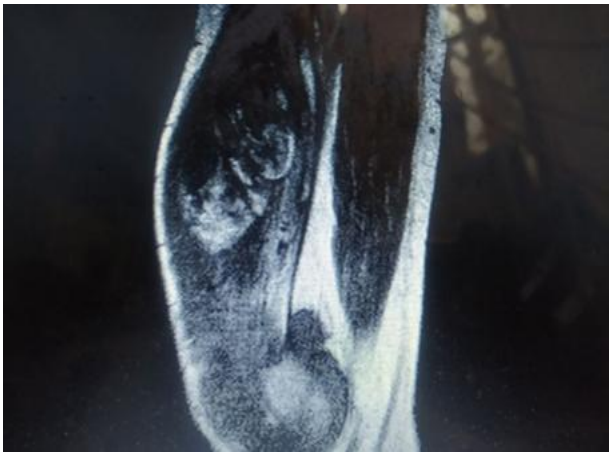

Figure 2: MRI image of the Rt thigh.

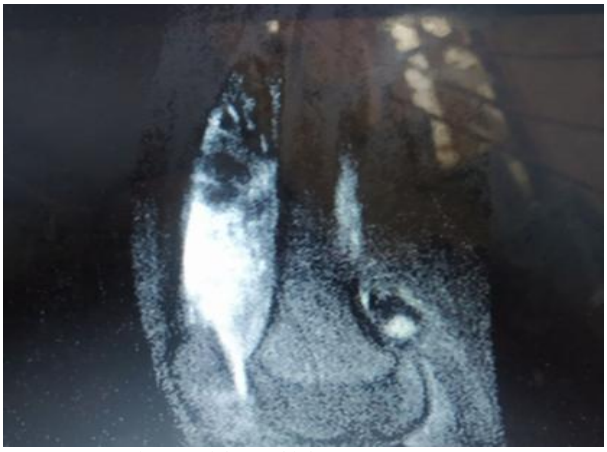

Figure 3: MRI image of the Rt thigh.

The macroscopic features of the postoperative excisional Lump Showed a yellow to brown surface irregular Three masses about 3x8 cm (Figure 4).

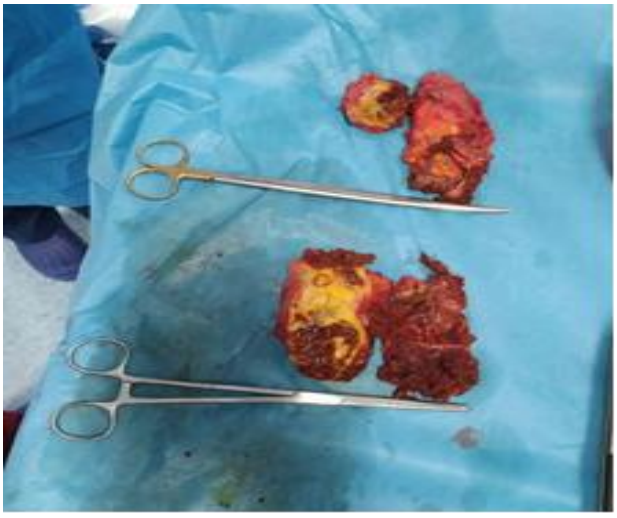

Figure 4: The macroscopic features of the PVNS.

Microscopic features show villous-like projections lined by synovial cells, chronic inflammatory cell infiltration (lymphocytes, histiocytes), hemosiderin-laden histiocytes, multinucleated giant cells, and no atypical or malignant changes were seen.

Therapeutic intervention and follow-up

For this case, partial surgical excision (ant. approach) open arthrotomy has been done (Figure 5).

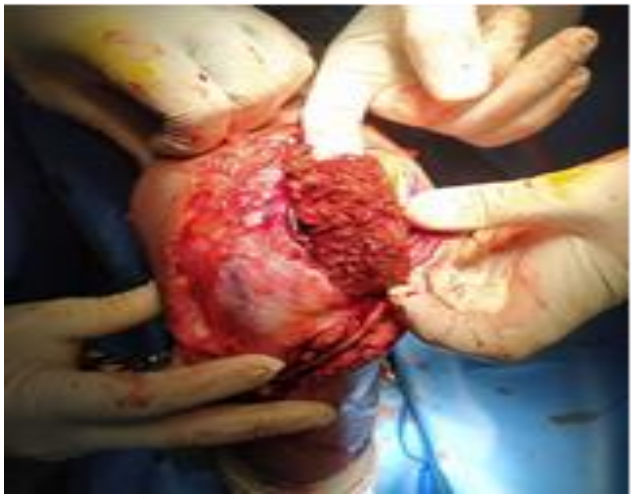

Figure 5: Anterio approach arthrotomy.

There was a plan for a second surgical intervention to the posterior knee compartment. After complete removal of the tumor at the anterior compartment and the suprapatellar area, cartilage damage in the medial and lateral femoral condyles has been noticed (Figure 6).

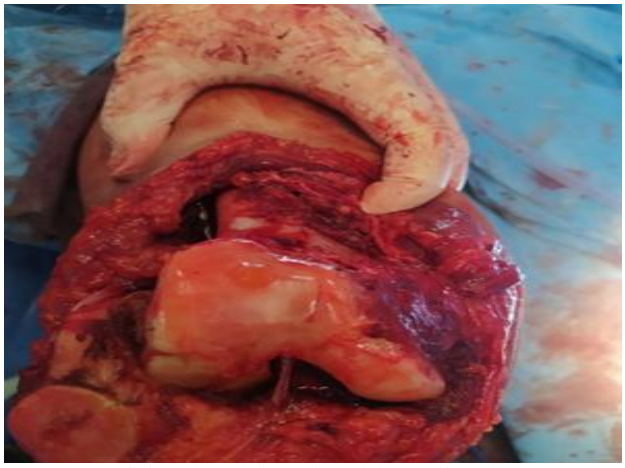

Figure 6: Medial and lateral femoral condyles cartilage damage.

Because of the patient's medical condition and the family's wish, the second surgical intervention to the posterior knee has not been done. The radiotherapy team suggested radiotherapy in order to avoid recurrence at the anterior compartment and as an attempt to regress the growth of the tumor in the knee's posterior aspect. The patient received radiotherapy of 13 fractionated doses. Postoperatively, there was early physiotherapy, and there was a significant increase in the range of motion without any pain or limitation. The patient presented after 3 years. Examination of the right knee revealed a full range of motion with no signs of swelling, pain, or tenderness. There was a shortening of less than half a centimeter in the right lower limb.

DISCUSSION

This case report is about a 13-year-old female with a dysmorphic abnormality, which led the pediatricians to suspect Noonan syndrome based on her facial appearance. Due to the family's low economic status and certain obstacles, genetic analysis was difficult to obtain. Among all the reported cases reviewed, PVNS has not been documented as being linked to Noonan syndrome or any other congenital anomaly. Most available literature reports primary affection in the adult age group [1]. Several studies have reported a potential genetic link to PVNS [8-10]. However, Noonan syndrome has never been mentioned in relation to PVNS. This case highlights the impact of socioeconomic factors and limited healthcare access on delayed diagnosis, which, in contrast to existing studies, is often misdiagnosed as juvenile idiopathic arthritis or septic arthritis [4,7]. MRI is the diagnostic tool of choice [1]. It is very beneficial in diagnosing the condition and can also determine the extent of the tumor for the purpose of surgical excision. Due to the deposition of hemosiderin, there will be low-intensity signals on both T1 and T2 images. Additionally, enhanced signals may be observed around the tumor due to lipid congestion, effusion, and inflamed synovium. For this reason, MRI with contrast is recommended [4]. Surgery may lead to early-onset secondary osteoarthritis, an unstable knee joint, a stiff joint, and even injury to the growth plate [2,4]. Due to the family's refusal for further surgical intervention, we opted for radiotherapy. However, radiotherapy is a debated strategy in literature due to the risk of growth plate damage [4]. An incidental shortening of less than 0.5 cm was discovered when the patient visited me with her mother after three years. At this point, the patient was nearly 17 years old. While PVNS is generally characterized by a recurrence rate of more than 30% [5], this patient remained symptom-free without recurrence or further growth at the posterior aspect after three years. A limitation of this case report was the family's refusal for genetic investigation. Further genetic research could help clarify potential syndromic associations. Additionally, follow-up was challenging due to the family's social, economic, and cultural issues.

Conclusion

Genetic investigation for pediatrics and adults might be essential for further understanding of this condition. Radiotherapy with justification could be a choice of treatment in certain patients with incomplete surgical resection. Early detection is necessary to avoid joint damage. MRI is the golden diagnostic method; biopsy is not necessary with MRI availability.

Conflict of interests

The authors declared no conflict of interest.

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Data sharing statement

Supplementary data can be shared with the corresponding author upon reasonable request.

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