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Case report

Pigmented Villonodular Synovitis in Children – A Case Series

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Abbreviations: PHPV: Persistent Hyperplastic Primary Vitreous; PVNS: Pigmented Villonodular Synovitis; JIA: Juvenile Idiopathic Arthritis

Case Report

Patient 1

A 11-year-old girl was referred by the Orthopaedic team with swelling of her right knee which started five months back. She had no systemic symptoms or preceding trauma or infection. She was seen by the Orthopaedic team, who aspirated the joint and injected it with steroids, once the cultures came back as negative. She had a good response, although the swelling returned after 8-10 weeks, associated with some stiffness. She was previously fit and well except for visual impairment in her left eye due to Persistent Hyperplastic Primary Vitreous (PHPV). Her examination revealed moderate effusion in her right knee. Her blood tests including inflammatory markers were normal. With a diagnosis of oligoarticular JIA, she had a further intra-articular steroid injection with good response lasting 4 months. Subsequently she required two more steroid injections in the same joint within six months, at which stage she was commenced on weekly Methotrexate injections at 15mg/m^2. Despite that, the swelling recurred, which did not respond to intra-articular steroid injection, following which MRI scan was requested. This showed diffuse synovial proliferation with relatively low signal components in some areas, suggestive of diffuse PVNS (Figure 1). Her immunosuppression was stopped, and she was referred for synovectomy.

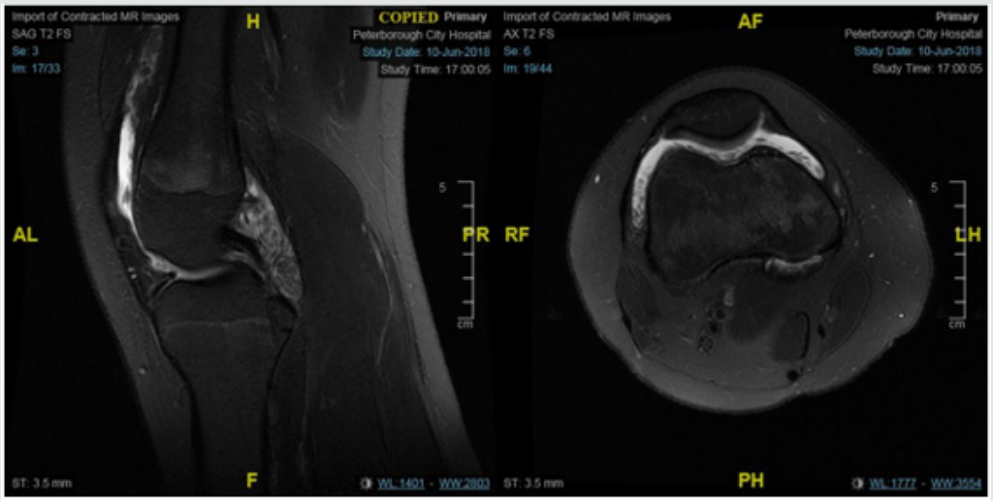

Figure 1: Diffuse synovial proliferation with relatively low signal components in some areas, suggestive of diffuse PVNS.

Patient 2

A 14-year-old boy was referred by the Oncology team with 7-month history of swelling in R knee. The onset was preceded by varicella infection. The aspiration of the knee joint carried out by the local Orthopaedic team revealed blood stained fluid, which was negative on bacterial cultures. But the swelling and the pain recurred and progressed despite regular Ibuprofen. He had no systemic symptoms including fever. His blood tests, including inflammatory markers, were normal. MRI scan of his right knee showed multi-loculated fluid collection with septas and post contrast enhancement of the collection walls with no involvement of underlying bone. He underwent synovial biopsy which ruled out sarcoma but showed synovial thickening and fibrinoid degeneration. He was then referred to pediatric rheumatology team as possible Juvenile Idiopathic Arthritis (JIA). On examination he had a large swelling of the right knee joint, more prominent in his supra patellar region with warmth and minimal restriction of movement. The rest of the joints were normal. In view of the relatively long history, the age, involvement of a single knee joint and blood-stained synovial fluid aspirate, a diagnosis of Pigmented Villonodular Synovitis (PVNS) was considered. His repeat MRI scan showed prominent and, in some places, nodular synovial proliferation around the knee and a large effusion, consistent with PVNS (Figure 2). He had poor response to intra-articular steroid injection which he had while waiting for surgery. He remains asymptomatic following open synovectomy.

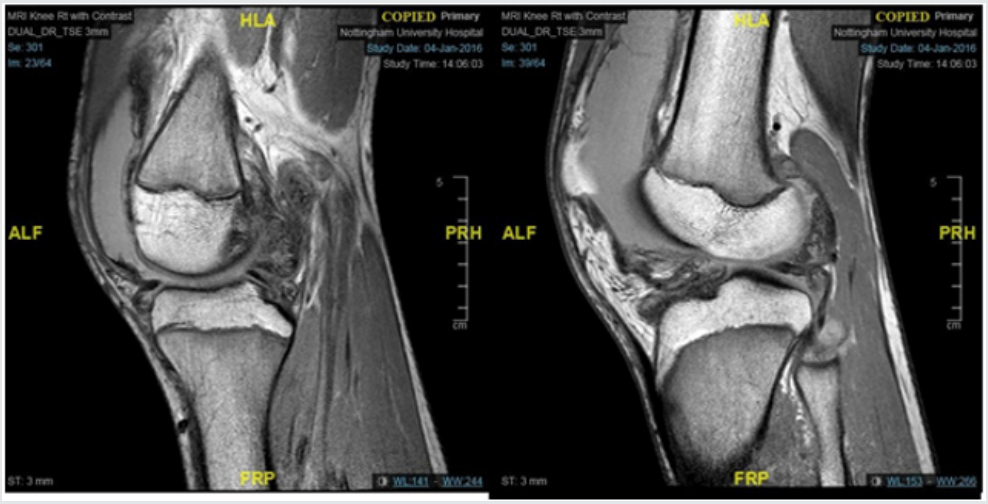

Figure 2: Prominent and in someplaces nodular synovial proliferation around the knee and a large effusion, consistent with PVNS.

Patient 3

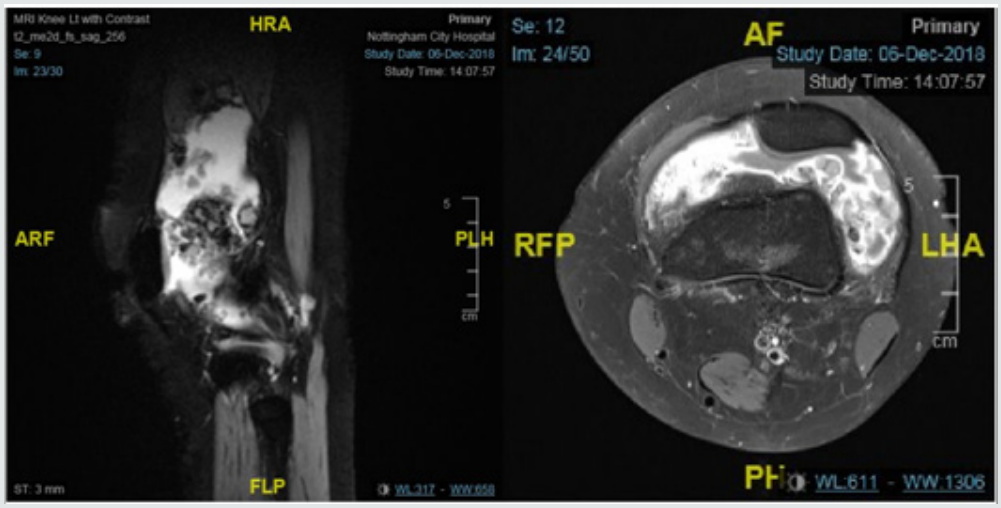

Figure 3: MRI scan of her left knee revealed large joint effusion with multiple synovial protrusions which are low signal on T2-weighted images and intermediate signal on T1-weighted images, with peripheral enhancement suggestive of PVNS.

A 17-year-old girl was referred by her GP for swelling in her left knee for 3 years. The onset was insidious with no preceding trauma. The swelling slowly increased in size, associated with stiffness and significant and painful limitation of knee flexion. Following a

failed aspiration in primary care, she had an ultrasound scan which revealed a large joint effusion. There were no concerns about any other joints or systemic symptoms. There was no significant past or family history. Her examination revealed large effusion of the left knee with restriction, with a significant suprapatellar component. Rest of the musculoskeletal examination and the systemic examination were normal. Her blood tests showed mild increase in inflammatory markers (CRP of 13 mg/L and ESR of 29 mm/hr) with negative autoantibody screen. MRI scan of her left knee revealed large joint effusion with multiple synovial protrusions which are low signal on T2-weighted images and intermediate signal on T1-weighted images, with peripheral enhancement suggestive of PVNS (Figure 3). She has been referred for consideration of synovectomy (Tables 1,2).

Table 1: When to suspect PVNS in children and young people?

When to Suspect PVNS in Children and Young People?
Older age group – > 10 years
Monoarthritic of knee
Long history
No systemic symptoms
Large effusion with significant supra-patellar component
X ray normal except for a soft tissue mass with no bony involvement
Blood stained aspirate
Hemosiderin laden macrophages on microscopy
Monoarticular JIA not responding to conventional treatment - intraarticular steroid injections and / or Disease Modifying Anti-Rheumatic Drugs (DMARDs)

Table 2: Investigations in suspected PVNS.

Investigations in Suspected PVNS
Blood tests – Full Blood Count (FBC), inflammatory markers, autoantibody screen, Borrelia serology (if history suggestive)
X ray
Synovial fluid for microscopy, culture and sensitivity (including TB if appropriate)
MRI scan with contrast – often diagnostic
Synovial biopsy [if felt appropriate following discussion at multi-disciplinary team (MDT) meeting involving the Orthopaedic, oncology, radiology, rheumatology and pathology teams]

Discussion

Pigmented Villonodular Synovitis (PVNS) is a rare condition characterized by diffuse synovial cell proliferation that affects the synovial joint, tendon sheaths, and bursa membranes [1,2]. Although a disease of young adults, it can occurs in older children, where it is usually monoarticular [3] with knee being the commonest joint involved. Pain and swelling are the usual presentation although mechanical symptoms like meniscal injury and patellar dislocation can rarely occur in adult PVNS [4]. The diagnosis is often delayed (mean of 18 months2) or mistaken for Juvenile Idiopathic Arthritis (JIA), infections like tuberculosis, hematological problems like hemophilia (due to hemarthrosis) or neoplastic processes affecting the synovium. All the above cases were referred to the Pediatric Rheumatology team as JIA by a variety of clinicians - primary care, Orthopaedic and oncology teams. The delay is often the longest in the patients who were commenced on treatment for JIA, which is demonstrated by the 2-year delay in diagnosis in patient 1.

The X ray findings are usually nonspecific - soft tissue swelling [4]. Erosive changes of bone secondary to the synovial proliferation, which can be seen in about 50% of adults with PVNS, are rare in children [5]. The synovial aspirate is often blood stained and the presence of blood-stained fluid with fragments of synovial villi in sediment and macrophages containing hemosiderin have been suggested as diagnostic criteria [6]. However, MRI findings of PVNS are characteristic (soft tissue mass with a hypo-intense signal in T1- and T2 weighted images representing hemosiderin deposits), although it is supplemented by biopsy and histopathological diagnosis in most case series. The treatment of choice is surgery, which could be open or arthroscopic synovectomy, although recurrence has been reported.

Arthroscopic and open synovectomy did not differ significantly in terms of rates of recurrence and complications [7]. Adjuvant treatment with external beam radiation therapy or intra-articular injection of radioactive material as yttrium-90 (Radio Synovectomy) [8], which has been used as an adjunct treatment modality in adults with PVNS, is hardly used in children due to risk of damage to epiphyseal growth plate and post-radiation sarcomas [2].

Conclusion

Pigmented Villonodular Synovitis is benign proliferative disorder, which can be associated with significant morbidity if left untreated. Because it is rare in children, the diagnosis is often delayed, resulting in delay in treatment. It should be suspected in chronic monoarthritic in older children without any systemic symptoms or patients with monoarticular JIA who are not responding to standard treatment. MRI scan, often in combination with synovial biopsy, confirms the diagnosis. Synovectomy seems to be the consensus treatment of choice.

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