Pigmented villonodular synovitis of the knee confused with juvenile rheumatoid arthritis in a 3-year-old child: A case report

3 yaşındaki bir çocukta juvenil romatoid artrit olarak karışan dizde pigmente villonodüler sinovit: Olgu sunumu

Abstract
Pigmented villonodular synovitis (PVNS) is rare and characterized by diffuse synovial cell proliferation with the development of villi, or round/ovoid lobulated nodules of varied sizes. It usually presents in adults between the ages of 30 and 40 years and is uncommon in the pediatric population. We herein report a 3-year-old female patient with PVNS causing a femoral subchondral lesion in the right knee. Her symptoms started at the age of one and she had gotten treatment for juvenile rheumatoid arthritis for 2 years. The patient was operated, and the tumor was excised. Arthrotomy was performed for diffuse PVNS and diagnosis was made histopathologically. She was followed-up for 18 months with clinical examinations and MRI, as necessary. She had no pain or limitation in range of motion. She is currently fully functional and ambulatory.

Keywords: PVNS, Pediatric, Knee, Juvenile rheumatoid arthritis

Introduction
Pigmented villonodular synovitis (PVNS), a rare pathology, was first described by Jaffe [1] in 1941. It is characterized by diffuse synovial cell proliferation and concomitant development of villi, or round/ovoid lobulated nodules in varied sizes. It generally occurs in adults between 30-40 years of age [2]. As PVNS is uncommon in children, diagnosis is often delayed, insidious, unclear, and difficult to figure out from the initial radiographic findings. Differential diagnosis is also challenging: Careful distinction should be made between PVNS and rheumatoid arthritis, arthropathies related to coagulopathies, tuberculosis, other inflammatory and synovial processes [3]. There are reports in the literature stating that initial age of knee involvement in the pediatric population varies from 12 months to 15 years of age [3-5].

PVNS occurs in two forms: Localized or diffuse. Hemosiderin, macrophages, and giant cells are located in synovium. The joint, tendon sheath or bursa are the affected areas. When a single mass is present in the synovium, affecting a smaller portion, it is called localized PVNS, but mostly, the entire synovium is affected, which indicates diffuse form [2].

It is very uncommon to see PVNS and related bone erosion in children. We herein report these two uncommon conditions in one patient with diffuse PVNS, who was followed up for 18 months with clinical examinations and MRIs.
Case presentation

A 3-year-old girl presented with swelling and pain in her right knee for the last 2.5 years. Her family reported that the swelling and pain began after a rotational trauma. Joint fluid aspiration was studied, yielding a normal result. The patient was followed up with cast and anti-inflammatory drugs. The symptoms did not improve in 5 months, after which ultrasonographic examination showed fluid collection in the knee, which was evaluated as synovitis. MRI showed a diffuse lesion of high T2 signal intensity at the suprapatellar bursa (Figure 1).

The patient was referred to the Pediatric Rheumatology Clinic with the diagnosis of mono-articular juvenile idiopathic arthritis. The treatment was held at the Pediatric Rheumatology Department with subcutaneous Methotrexate 10 mg/kg and Ibuprofen for 2 years, nevertheless, knee swelling, and pain did not regress. Rheumatological markers, tuberculosis and brucellosis tests were all normal. Repeat MRI was comparatively examined with the one obtained 2 years ago, which showed an increase in the size and signal intensity of the mass in the knee (Figure 2).

The last MRI showed that there was a vascular malformation, and a large hemangioma was suspected. The patient was then referred to our clinic for further evaluation. Physical examination revealed a diffuse swelling over the knee with a palpable smooth mass all around the knee without any distinct borders (Figure 3).

There was no sign of inflammation. The range of motion (ROM) was normal with little limitation in extension, approximately 10 degrees. No fluid was found inside the knee. Muscle strength was normal and gait was a little antalgic. Plain radiography of the knee showed no abnormality (Figure 4). The laboratory examinations and rheumatologic tests were normal.

Almost complete excision of the lesion was performed through arthrotomy with medial parapatellar approach with the tourniquet inflated under general anesthesia. Macroscopically, the knee joint was covered by hypertrophic villous synovium, which was diffuse, dense, and reddish-brown pigmented (Figure 5). Bone erosions were seen in both femoral condyles, especially medially (Figure 6). Total synovectomy was performed and almost all brown pigment on the cartilage was debrided (46 x 27 x 13 mm) (Figure 7). The final histopathologic diagnosis was PVNS with brown pigment accumulation on synovial epithelium (Figure 8).
PVNS of the knee in a 3-year-old child

After skin closure, elastic dressing was applied and immediate postoperative rehabilitation was started with weight-bearing and early knee range of motion.

At eighteen months of follow-up, the patient is asymptomatic with no signs of recurrence. Control MRI showed a dramatic decrease in effusion and regression of almost all hemosiderin-containing pigments (Figure 9). Inspection of the knee showed no swelling or palpable mass (Figure 10). There was no pain and the right knee had full range of motion.

Discussion

Pigmented villonodular synovitis (PVNS) is a benign but locally aggressive proliferative lesion of the synovial membrane. The etiology of PVNS is not certain, but genetic factors, chronic inflammation, some pathologic conditions with chromosomal abnormalities that induce hemorrhagic disorders, are thought to be potential causes [3]. There is also a relationship with traumatic events. The most frequent representation is monoarticular involvement of the knee and mostly affects young adults [3]. This pathology is rare in children [5], therefore the diagnosis is often late. Presentation is mostly unspecific like swelling and effusion with or without pain. Differential diagnosis is also challenging: Careful distinction should be made between PVNS and rheumatoid arthritis, arthropathies related to coagulopathies, tuberculosis, other inflammatory and synovial processes [3].

The patient in this report had only one affected joint and her symptoms were similar to those of arthritis. She got treatment for JRA for 2.5 years. Baroni et al. [3] reported that there was an interval between the onset of the symptoms (pain, swelling, change of posture and decreasing of the range of motion) and diagnosis, it can take from 3 to 48 months (mean: 18 months). In their study, most cases were first diagnosed with pathologies other than PVNS, JRA being the most common. Like the presented case, they were followed for at least two years. Eventually, each patient was examined by histopathologic tissue sampling, which confirmed PVNS [4]. In adults, the interval between the onset of clinical symptoms to definitive diagnosis was 18 months. In this case, symptoms had started at 12 months of age. The delay in diagnosis is close to the average duration reported in the literature.

MRI is useful for diagnosing PVNS, as X-ray findings are negative in most of cases. X-ray yields positive findings in only 33% of pediatric PVNS [3] patients. Our patient also had no significant radiographic signs other than soft tissue swelling sign at the anterior aspect of the knee (Figure 2). On MRI, the pathognomonic finding of PVNS is low to intermediate intensity signal in both T1 and T2 sequences, which is compatible with hemosiderin-laden tissue. In this case, MRI demonstrated increased signal intensity at the suprapatellar area in T2 images, which was evaluated as effusion and synovitis. There was no sign associated with hemosiderin deposits. The last MRI demonstrated a mass under skin over patella and the patients was recommended for evaluation in terms of hemangioma. It was
expected, because there were three cases in the literature, in which vascular proliferation in the synovial membrane mimicked PVNS [6].

PVNS can induce synovial hyperplasia, invade the joint capsule and synovium. It erodes the cartilage and bones [7]. In this case, diffuse synovial hyperplasia caused bone erosions, and defects on the femoral condyles were demonstrated. Bone erosion is not a common entity in the pediatric population.

Surgical treatment aims to resect all abnormal tissue in the knee joint to reduce the degree of bone erosion, relieve pain and prevent recurrences. There are a lot of therapeutic options described but the best option has not yet been defined. For diffuse forms, the most popular and accepted treatment is subtotal open or arthroscopic synovectomy [8]. It is still controversial which technique is better for lowering recurrence and morbidity. In a literature review, Nakahara et al. [9] found a 23.2 % recurrence rate with open synovectomy and a 39 % recurrence rate with all-arthroscopic synovectomy. Colman et al. [10] reported that the overall recurrence rate of the combined open and arthroscopic techniques was lower compared with all-arthroscopic or open groups (9 vs. 62 vs. 64%, respectively).

Combined open and arthroscopic synovectomy is a comprehensive approach associated with low recurrence and postoperative complication rates. In this case, the patient underwent open resection via medial parapatellar approach, because the lesion was diffuse and total resection was possible with open approach only. For the presented case, we thought that arthroscopic technique had some disadvantages, such as possibility of inadequate resection of the mass, which would increase the risk of recurrence.

Due to the high rate of recurrence reported in this type of PVNS, radiotherapy was suggested as an adjuvant treatment method. However, the use of radiotherapy in children is controversial due to epiphyseal growth plate damage and occurrence of post-radiation sarcomas [4]. Because of these reasons, we did not use radiotherapy. The patient had good functional motion and no recurrence at the end of 18 months.

**Conclusion**

If such a pediatric patient with long term joint swelling is non-responsive to treatment, PVNS must be considered. This suspicion must be confirmed with MRI and joint tissue sample examination. Our case is the second earliest reported knee joint PVNS after Jawadi et al. [5], who reported a 12-month-old child with PVNS of knee.

**References**


This paper has been checked for language accuracy by JOSAM editors. The National Library of Medicine (NLM) citation style guide has been used in this paper.