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Our clinical outcomes in patients operated with the diagnosis of pigmented villonodular synovitis

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Abstract

The aim of this study was to evaluate the clinical and surgical results of 21 cases treated with pigmented Villonodular Synovitis in our clinic between 2010 and 2018 and to share these results with the literature. The 5-year follow-up results of 21 cases with pigmented Villonodular Synovitis were evaluated retrospectively. Only patients undergoing surgical treatment were included in the study. The patients were investigated in terms of age, gender, anatomical region, size of the tumor, a surgical technique applied and recurrence. The involved joints are in turn; seven knees, five ankles, four feet, two hands, two hips, and one shoulder joints. The patients' ages ranged from 9 to 81 years (mean 45). 11 of them were male, and 10 were female. Clinically; Pain and swelling in the involved joints were the most common complaints, and the most common symptom was the compression effect of the mass. The most commonly involved anatomic region was the knee joint. The majority of patients had no history of trauma. Recurrence was found in 23.8% of patients who underwent surgical treatment. Pigmented Villonodular Synovitis is a middle-aged disease that causes severe morbidity and compromises the quality of life. Arthroscopic and open surgical synovectomy can be performed successfully in the treatment of this condition.

Keywords: Joint, pigmented villonodular synovitis, effusion, debridement, synovectomy

Introduction

Pigmented villonodular synovitis (PVNS); is a benign, proliferative, inflammatory disease of the synovial tissue of an unknown etiology [1-6]. It was first described by Jaffe. The disease presents itself in two forms, the local form, and the diffuse type. The localized form is a focal proliferation of the synovial tissue characterized by nodules. In the diffuse form, all synovial tissue in the joint is affected. It is a rare disease. Two new patients are involved each year in every one million [2,3]. It usually holds a single joint. In less than 1% of cases, multiple joint involvements is detected. It can be seen in all synovial joints, most commonly in the knee joint. The incidence is 2 / 1.000.000 [1,2]. PVNS can be seen in all age groups, but the most frequently affected age group is between 20-50 years of age. It affects males and females equally. The patients are most commonly admitted to the hospital with complaints of pain and swelling in the related extremity. There may be frequent recurrent effusion attacks in the knee joint. It may also originate from the synovial of extraarticular structures such as fascia and bursa and the tendon sheath [1-5]. The macroscopic appearance is While yellowish-brown synovial mass with synovial thickening and finger-like villous projections in the synovium. polymorphonuclear neutrophils, histiocytic giant cells, fibroblasts, hemosiderin, and lipid-loaded foamy cells can be seen in the pathology.

Plain radiographs show an increase in the soft tissue density around the associated extremity. According to the degree of the disease, degenerative changes in the bone structure and subchondral cysts may be observed in the involved joint. Magnetic resonance imaging can show low signal intensity related to hemosiderin increase in nodule content and high signal intensity related to the increase in fat tissue [7,8].

PVNS treatment depends on the degree of injury in the joint and the age of the patient. Total synovectomy is the most favorable treatment modality in patients without significant joint damage and bone erosion [9]. It can be done by open or arthroscopic methods (Figure 4). In cases where total excision cannot be performed, adjuvant radiotherapy may be

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applied postoperatively. In the advanced stages of the disease, arthroplasty or arthrodesis are other surgical treatment options.

Material and Methods

Patients who underwent surgery for PVNS between 2010 and 2018 were examined retrospectively after the necessary approval (No: 2019/2-2) was obtained from the Ethics Department of our University. Patients with a clinical and radiological diagnosis of PVNS who underwent surgical treatment and whose diagnosis was confirmed histologically were included in our study. Patients receiving adjuvant or neoadjuvant radiotherapy or systemic therapy were excluded from the study. The demographic distribution was determined according to age and gender. Patients were evaluated according to the anatomical region of involvement, the tissue from which the anatomical localization originates (joint, tendon sheath, fascia, bursa), the presence of trauma history in this anatomical region, size of the tumor, a surgical technique applied, number of surgeries performed and local recurrence.

Patients who were admitted to our orthopedics and traumatology outpatient clinic with joint and / or extremity pain and swelling were evaluated with X-ray and MRI imaging. Following the clinical and the radiological evaluation of the patients, the treatment plan was performed. The surgical procedure and the approach to each patient were decided according to the patient's clinic and radiological features. By MR imaging, the anatomic localization, the form of the disease (diffuse / localized) and dimensions of the lesion were determined.

Following the surgery, the patients were called to the outpatient clinics every 3-6 months and followed up clinically. According to the clinical status and complaints of the patient, MRI was performed at 6-12 month intervals, and radiological follow-up was performed.

Results

Between 2010 and 2018, 21 patients with PVNS were treated surgically and a total of 28 surgical procedures were performed in our clinic. The diagnosis of all patients was confirmed pathologically. 11 of our patients were male, and 10 were female, and the mean age was 45 (9-81).

The rate of involvement of anatomical region by frequency order ; the knee (7), ankle (5), foot (4), hand (2), hip (2), shoulder (1). In nine of 21 patients, the tumor was originating from the joint and in the remainder 12 patients the origin was from the tendon sheath. When we questioned the patients' stories, six patients described a history of trauma to the involved area, and 15 patients had no history of trauma. The size of the tumor in our preoperative MR images was below 5 cm in six patients, 5-10 cm in eight patients, 10-15 cm in five patients, and above 15 cm in two patients. Treatment success was achieved in 17 patients in a single surgical session. Two of our patients had a second session, and two patients had their second and third session. Recurrence was observed in 5 patients (23.8%) during the 5-year follow-up period (Table 1).

Table 1. Patients who were admitted to our orthopedics and traumatology with pigmented villonodular synovitis

FEATURES	NUMBER OF PATIENTS
SEX	
MALE	11
FEMALE	10
MEAN AGE	45
ANATOMICAL LOCATION	
KNEE	7
ANKLE	5
FOOT	4
HAND	2
HIP	2
SHOULDER	1
ORIGIN	
JOINT	9
TENDON SHEATH	12
HISTORY OF TRAUMA	
PRESENT	6
NOT PRESENT	15
TUMOR SIZE (cm)	
<5	6
5-10	8
10-15	5
>15	2
NUMBER OF SURGERIES	
1	17
2	2
3	2
LOCAL RECURRENCE	5

The most common anatomical site of involvement is the knee joint. Seven of the 21 patients (33.3%) had knee joint involvement. Four of these patients had localized nodular type, and 3 had a diffuse lesion. Knee joint intra-articular involvement areas; three patients had an anterior injury, three patients had suprapatellar region involvement, and one patient had the posterior area injury. Only one patient had extraarticular involvement. As surgical treatment; three patients underwent open total resection and synovectomy, two patients underwent arthroplasty following complete synovectomy, one patient underwent arthroscopic resection and synovectomy, and one patient underwent open + arthroscopic combined resection with synovectomy operations. Recurrence was observed in the follow-up of two patients. The first was a male patient who underwent an arthroscopic / open combined treatment with a lesion in front of the knee. Recurrence was detected 48 months after his first surgery, and he was treated with open resection / debridement. The other patient with recurrence was a 57-year-old male with a lesion behind the knee and underwent open resection. Recurrence was observed 18 months after surgery. As the patient's complaints were not excessive, reoperation was not performed, and symptomatic treatment was administered.

The number of patients with ankle involvement is five. All of these were associated with the tibiotalar joint as well as non-articular tissues involvement. Anterior / posterior / medial /

lateral approaches were performed according to the region, and all patients underwent open surgical procedure. Two patients had recurrence twice. Both patients underwent reoperation two times in addition to the primary surgery.

All four patients with foot involvement had lesions originating from the synovial tissues around the tendon. Three of the four patients had an injury in the dorsal aspect of the foot, and two patients had sinus tarsi-related lesions. One patient had isolated toe involvement. The injury of this patient was originating from extensor tendon in the dorsal aspect of the 1st toe. All patients underwent open surgical debridement according to the lesion site. No recurrence was seen in any of them.

We had three patients with upper extremity involvement. Two of them had extraarticular lesions in the phalanx, and other patient had to shoulder the articular participation. The first of the patients with hand lesions had an injury in the first finger and the second patient had an injury involving the second phalanx. Both patients underwent successful open debridement and no evidence of recurrence. The patient with shoulder joint involvement was a 40-year-old male patient. He had complaints of left shoulder pain and severe limitation of movement. Radiological findings revealed widespread PVNS lesions and joint destruction in the shoulder joint. The patient underwent open surgical debridement. During the follow-up period, the patient's experienced worsening pain in the shoulder. Radiological findings revealed local recurrence of tumor lesions progressive joint destruction for which proximal humerus resection was performed.

We have two patients with hip involvement, the first one was an elderly male patient and the second one was a female patient. In addition to PVNS lesions, severe joint destruction was observed for which arthroplasty was decided. Both patients underwent a primary total hip arthroplasty with the lateral approach and no complications and recurrences were observed during follow-up.

The demographic and clinical characteristics of our patients are summarized in Table 2.

Table 2. The demographic and clinical characteristics of pigmented villonodular synovitis patients

						SURGICAL INTERVENTION			
PATIENT	**AGE**	**SEX M: MALE F: FEMALE**	**FOLLOW UP PERIOD**	**INVOLVED REGION**	**EXTRA-ARTICULAR LESION**	**PRIMARY TREATMENT**	**APPROACH A: ANTERIOR P: POSTEİİOR M: MEDIAL L: LATERAL**	**TREATMENT FOR RECURRENCE**	**RECURRENCE (PERIOD IN MONTHS)**
		M	9 YEARS	LEFT KNEE ANTERIOR	+	ARTHROSCOPIC +OPEN	A	OPEN	+ (48)
2	42	F	5 YEARS	RIGHT KNEE ANTERIOR	-	OPEN	A	-	-
3	68	M	3 YEARS	LEFT KNEE ANTERIOR	-	TKA	A	-	-
4	81	F	6 YEARS	RIGHT KNEE SUPRAPATELLAR	-	TKA	A	-	-
5	56	M	5 YEARS	RIGHT KNEE SUPRAPATELLAR	-	ARTHROSCOPIC	A	-	-
6	37	M	2 YEARS	RIGHT KNEE SUPRAPATELLAR	-	OPEN	A	-	-
7	57	M	8 YEARS	LEFT KNEE POSTERİOR	+	OPEN	P	FOLLOW UP	+ (18)
8	39	F	5 YEARS	RIGHT ANKLE	-	OPEN	P	-	-
9	38	M	3 YEARS	RIGHT ANKLE	+	OPEN	P + L	OPEN	+ (24),(36)
10	10	M	3 YEARS	LEFT ANKLE	+	OPEN	M	-	-
11	38	M	3 YEARS	LEFT ANKLE	+	OPEN	M + L	OPEN	+ (7), (7)
12	20	M	1 MONTH	LEFT ANKLE	+	OPEN	A + P	-	-
13	33	F	1 YEARS	LEFT FOOT 1ST TOE	+	OPEN		-	-
14	49	M	6 MONTHS	LEFT FOOT DORSAL	+	OPEN		-	-
15	36	F	5 MONTHS	LEFT FOOT DORSAL	+	OPEN		-	-
16	46	F	2 MONTHS	LEFT FOOT DORSAL	+	OPEN		-	-
17	50	F	3 MONTHS	RIGHT HAND 1ST PHALANX	+	OPEN		-	-
18	9	F	3 YEARS	RIGHT HAND 2ND PHALANX	+	OPEN		-	-
19	70	F	4 YEARS	RIGHT HIP	-	THA	L	-	-
20	75	M	2 YEARS	LEFT HIP	-	THA	L	-	-
21	40	F	5 YEARS	LEFT SHOULDER	-	OPEN	A	PROXIMAL HUMERUS RESECTION	+ (4)

Discussion

PVNS; In its historical development, with its symptomatology and clinical course, it has been named as many tumoral lesions and got different names. Chassaignac in 1852; defined the lesion as the nodular mass originating from the flexor tendon sheath of the middle and index fingers. In the literature, xanthoma or giant cell tumor (Targett 1897), myeloxantoma (Dar), villous arthritis (Dowd) and benign synovioma (Stevvart 1948) names were used. In 1941, Jaffe, Lichtenstein, and Sutro first used the term pigmented villonodular synovitis [6].

Fisk in 1952; suggested that PVNS results from hemarthrosis and synovial hypertrophy after minor traumas. In later years, trauma and recurrent intra-articular bleeding were thought to play a role in the etiology. Young and Hudacek in 1954; stated that similar synovial macroscopy to PVNS occurred due to blood injections to dog knees but hemosiderin loaded with multinuclear giant cells, characterized by PVNS'in does not happen. Hoagland et al. In 1967 compared the synovial changes in the knees of hemophilia patients to PVNS but found that the histopathological examination was not compatible [6]. Therefore, the effect of trauma on PVNS has not been proven yet. Six of the patients in our series had a significant history of injury. This suggests that some factors other than injury play a role in the primary etiology. The common idea of most researchers is that this pathological condition is an inflammatory response to an unknown stimulus.

PVNS; is a middle-aged disease with insidious onset usually characterized by a single joint involvement. It is rarely seen in children and older ages (1-5,13,14). In our patient group; There are two adolescents / children under 18 years old. The number of patients older than 65 years is four. The mean age in the series given in the literature is consistent with the mean age of our patient group (45 years). The gender difference is not very clear. Of the 21 patients we operated, 11 were male, and 10 were female.

Multiple joint involvements is rarely reported in the literature [2,3,13]. All of the cases we treated included a single anatomical region. There was no multiple involvements.

It is reported that knee joint is involved in three-quarters of the cases. Then, respectively, hip, ankle, shoulder and elbow involvement is seen [1-5]. In our series, the most common joint was the knee joint. However, compared to the literature, the rate of knee joint involvement was lower (33.3%). Jaffe [1] stated that the knee was ten times more commonly affected than hips and reported two hip and one ankle and calcaneocuboid joint involvement in response to 20 knee involvement in his series including 25 cases.

In recent years, the most detailed study on PVNS was performed by Flandry et al. They reported 25 knee results in 23 patients (15 males, eight females) with a mean age of 39 (1 7-82) in their study and reported two new patients with a PVNS incidence of one million per year. 28% of patients had excellent, 64% had good, and 8% had poor results. Two patients with poor outcome experienced a relapse. They found that the trauma history was only 1/3 of the patients, but all patients had pain, swelling, and limitation of motion. In 16% of patients, there was no macroscopic pigmentation suggestive of typical PVNS, but they were observed to have an iron deposition in all pathological examinations. Typical, bloody synovial fluid was detected in 44% of cases [2,3]. This rate was 69% in the series of Myers et al.

Schajovvicz; In a study of 80 cases, stated that the age of the patients was between 15-52 years and that PVNS affected both sex equally. He reported that 52 of the patients had knee and eight had foot and ankle, and five had hip involvement. Hand, wrist, and shoulders had a lower rate of engagement. Schajovvicz states that malignant change is infrequent and only one of his cases developed malignant histiocytic proliferation [4]. Kalil, on the other hand, reported that a 21-year-old patient with diagnosed PVNS developed malignant transformation at the age of 85 [15].

Ogilvie-Harris et al .; reported that PVNS occurred between 17-80 years of age (mean 38) and total arthroscopic synovectomy had a lower recurrence rate than partial synovectomy in 25 disease series [7].

Atmure et al. , Spjut et al.; They suggested that iron deposition, hyalinization, and fibrosis degree increased the severity of PVNS as in other inflammatory diseases. In contrast, Rao and Vigorita; they reported that there was no connection between them and that the number of mitosis and relapse were related [1,8].

Schwartz et al.; reported recurrence in 25 of 99 patients on average 14 years (1-47) of follow up [16]. Zvijak et al. found a recurrence in two of 14 patients (14%) following arthroscopic resection with a mean follow up period of 42 months (8-83) (11). Rarely, relapse occurs after many years of treatment [17].

Dorwart et al .; in a large series of 146 patients, reported that there were 51% bone changes, while Wu et al. in a series of 24 cases, showed that only two patients (8%) observed narrowing of the joint space and bone erosion [18,19]. Bone changes reported by Rao and Vigorita by 48%, by Ogilvie-Harris et al. 16%, and by Schajovvicz by 30% were determined in eight of our patients (38%). In our series, we observed bone changes in four of seven patients with hip involvement; this creates the idea that the diagnosis in the hip joint done at later stages. That patient either admitted to the hospital at a late time or could not benefit from MRI in the early stages. Therefore; PVNS should always be considered in patients with long-term pain in the joint, especially in the hip.

There are few publications on PVNS in our country. Erginer et al. In 1987, presented the results of six patients (two males and four females). The mean age was 36.5 (14-55). Four of the patients had knee, one had hip and on had ankle involvement. No recurrence was observed in any of the patients followed for an average of 4.8 years [20]. Yel et al. reported 10 patients (eight men, two women) with PVNS who treated arthroscopically, their age ranged from 21 to 58 years (mean age 34 years), and two patients (20%) had recurrence [21]. Yıldız et al. In their study in 2001, in their study on 21 patients (14.3%) who were operated with the diagnosis of pigmented villonodular synovitis reported recurrence [22-23].

Conclusion

Lysosomal acid lipase deficiency; in patients with high LDL and / or low HDL levels, hepatomegaly and / or high transaminase levels without obesity or metabolic syndrome should be considered.

In our study, the use of lipid electrophoresis in the diagnosis of

primary hyperlipidemia is one of the weaknesses of our study.

In our study, we could not find any relationship between these two diseases. As a result of our study, LAL deficiency was not detected in patients with primary hyperlipidemia. However, because the incidence of LAL deficiency is very low, large-volume clinical studies are needed to evaluate the frequency of patients with primary hyperlipidemia.

Competing interests
The authors declare that they have no competing interest.

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Ethical approval
Ethics committee approval was obtained.

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