ACTA
ORTHOPAEDICA
et
TRAUMATOLOGICA
TURCICA

ORIGINAL ARTICLE

Acta Orthop Traumatol Turc 2011;45(3):149-155
doi:10.3944/AOTT.2011.2442

Long-term results of surgical treatment of pigmented villonodular synovitis of the knee

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Objectives: The aim of this study was to evaluate the long-term results of total synovectomy in pigmented villonodular synovitis of the knee (PVNS).

Methods: Open total synovectomy was performed for 19 patients (9 men, 10 women; mean age: 42.8 years) with PVNS. Of these patients, 15 had diffuse and 4 localized PVNS. The patients were followed for an average of 80.2 months and the average time between the onset of complaints and surgery was 23 months. In 4 patients, PVNS was identified during total knee replacement (TKR) performed due to gonarthrosis. Radiotherapy was performed as an adjuvant treatment in one patient with recurrence. Puncture was performed in 11 patients due to effusion and 8 to 70 cc of fluid was aspirated. Diagnosis was made during the exposure for TKR in 4 patients, by a biopsy in 2 and based on joint puncture and MRI findings in the rest.

Results: Recurrence occurred in 5 patients. A second total synovectomy was performed in 4 patients. Radiotherapy was used for the remaining one patient. Two patients were operated three times. During the follow-up, TKR was performed in 7 of the 19 patients. None of the patients developed infection and hemarthrosis requiring puncture nor required amputation or arthrodesis. Three patients had a postoperative knee joint stiffness of 10 to 25 degrees. The patients were evaluated according to the Knee Society Score and 8 (42.2%) had perfect, 9 (47.3%) good and 2 (10.5%) bad results.

Conclusion: PVNS is a disease with a high risk of recurrence. No individual or combined treatment method can offer a definitive solution. Open or arthroscopic radical synovectomy is still considered as the gold standard. If necessary, adjuvant intraarticular or extraarticular radiotherapy can be added to the treatment.

Key words: Knee; pigmented villonodular synovitis; total synovectomy.

Pigmented villonodular synovitis (PVNS) is a proliferative and inflammatory disease of benign course, first described by Jaffe, Lichtenstein and Sutro in 1941. This rare disease is common betweenthe ages of 20 and 60. Its yearly incidence is around 1.8 per 1.000,000 people. It usually involves a single joint, most commonly the knee or hip. PVNS is categorized as extraarticular and intraarticular. Extraarticular PVNS is also known as giant-cell tumor of the tendon sheath. The intraarticular PVNS has its localized and diffuse forms.[1-7]

The macroscopic appearance of PVNS is a nodular mass with a color that varies between dirty yellow and dark brown. Apparent synovial thickening and finger-like villous protrusions in the synovium are seen (Fig. 1). Microscopically, proliferation is observed in the synovial cells. Polymorphonuclear neutrophils, histiocytic giant cells, fibroblasts and

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Submitted: March 5, 2010 **Accepted:** July 27, 2010

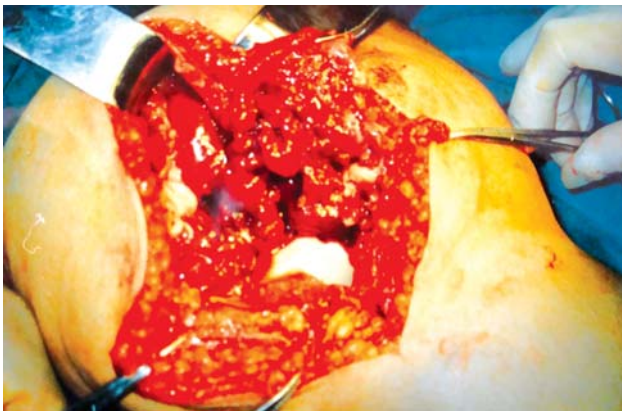

Fig. 1. Pigmented villonodular synovitis of the knee joint. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

foam cells laden with hemosiderin and lipid deposits are abundantly observed.

However, the most efficient screening device that helps in reaching a diagnosis is the MRI.[4,8-11] While conventional radiography reveals soft tissue swelling in earlier stages, subchondral cysts and erosive alterations may be observed in later stages.[2-12] In the earlier stages of the disease, the treatment of choice is arthroscopic or open total synovectomy. In the advanced stages of the disease, more invasive produres such as arthrodesis and arthroplasty may be necessary.[13]

The aim of the present study was to evaluate the long-term results of patients diagnosed with PVNS and treated with various methods.

Patients and methods

19 patients (9 men, 10 women; mean age: 42.8, range: 15 to 62 years) who underwent surgical treatment for PVNS between 1996 and 2009 were included in the study. Fifteen patients had diffuse and 4 patients had localized PVNS. In 11 patients (58%), the right knee and in 8 (43%), the left knee was involved. The mean follow-up time was 80.2 (range: 15 to 156) months. .The mean time between the onset of complaints and the surgery was 23 (range: 3 to 52) months. The results of the blood tests were in normal ranges. All patients had tolerable pain and swelling, and limping was present in 3 patients. In 9 patients, a limitation of movement in the knee joint (ranging between 15 and 30 degrees) was observed.

A history of trauma was present in only two patients.

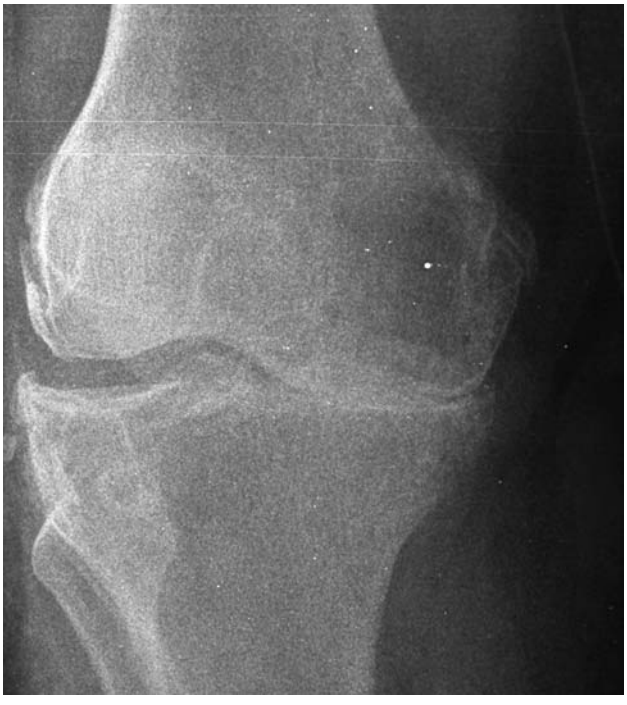

Fig. 2. Degenerative changes in the bone structure in anteroposterior radiography.

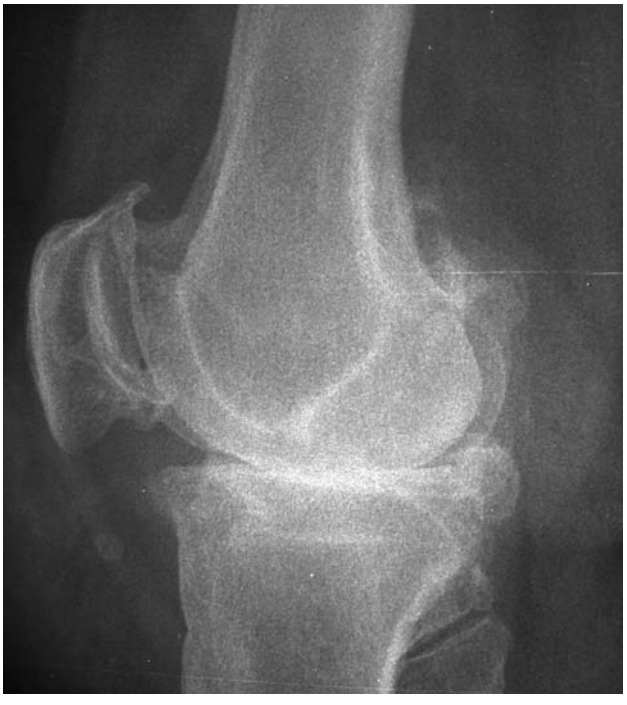

Fig. 3. Degenerative changes in the bone structure in lateral radiography.

All patient had radiographic evaluation, which was normal in earlier stages. In the 8 patients with advanced stages of the disease, pathological changes in bony structures were observed (Figs. 2 and 3). During knee puncture, 8 to 77 cc of synovial fluid was aspirated from the 11 patients (58%). The aspirated fluid varied from dirty yellow to dark brown. Three patients had reoccurence of the effusion. The examination of the synovial fluid did not reveal any differential characteristics of PVNS. Two suspect ed patients were scheduled to undergo surgery after biopsy. Fifteen patients underwent MRI and CT.

As in total knee replacement, the knee was exposed through an anterior midline incision and total synovectomy was performed. The diagnoses of the patients were confirmed by postoperative histopathology reports.

The diagnosis of 4 patients was made during total knee replacement, performed due to advanced gonarthrosis. Following osteotomy, radical synovectomy was performed on these patients. One patient had a partial anterior cruciate ligament rupture, one had a meniscus rupture and one had a parameniscal cyst. These lesions were repaired during the synovectomy. We preferred open synovectomy over arthroscopic synovectomy because the former allows for a more complete synovectomy.

Results

In 13 patients, lesions were located in the anterior region, while in 3, 2 and 1 patients they were located in the anterolateral, medial and posterior regions, respectively.

All patients were followed via MRI/CT and conventional radiography. Recurrence occurred in 5 (26%) patients. The recurrences developed between 2 to 5 years following surgery. Open synovectomy was performed for the second time in four patients. However, gonarthrosis developed in two of these patients despite the repeated synovectomy, and total knee replacement had to be performed on two patients one and two years after the surgery. No signs of recurrence were found in the remaining two patients. During the 9 year follow-up period of the TKR patients, septic or aseptic loosening of the knee prostheses was not observed. The fifth was referred to the radiation oncology department due to the development of recurrence in the posterior region (Fig. 4). One patient underwent total knee replacement due to gonarthrosis, six years after the surgery (Figs. 5 and 6). Collectively, during the follow-up of the 19 patients, 7 underwent total knee replacement and 2 underwent surgery thrice. All patients who underwent total knee replacement had diffuse PVNS. Detailed information on the 19 patients is given in Table 1.

The preoperative movement limitation of 9 patients, ranging between 15 and 30 degrees, remained in only 3 patients. These were the patients with the longest time lapse between the onset of complaints and surgery. None of the patients required arthrodesis or amputation. The patients were evaluated according to the Knee Society Score and the results were perfect, good and bad in 8 (42.2%), 9 (47.3%) and 2 (10.5%) patients, respectively.

Discussion

PVNS is most commonly seen between the ages of 20 and 60. Localized PVNS is more common in the 5th and 6th decades, while diffuse PVNS is more prevalent in the 3rd and 4th decades of life.[14] Mean age of our patients was 42.8 years, which is consistent with the literature. The ratio between men and

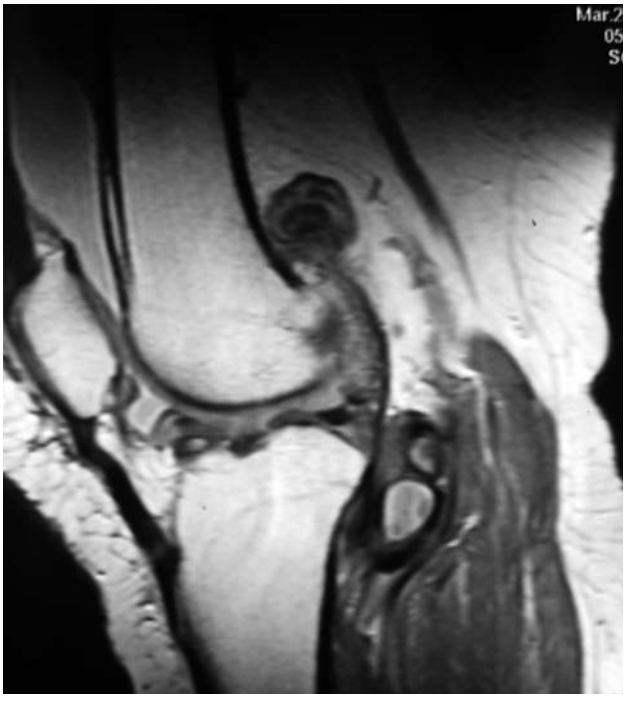

Fig. 4. PVNS recurrence in the posterior compartment.

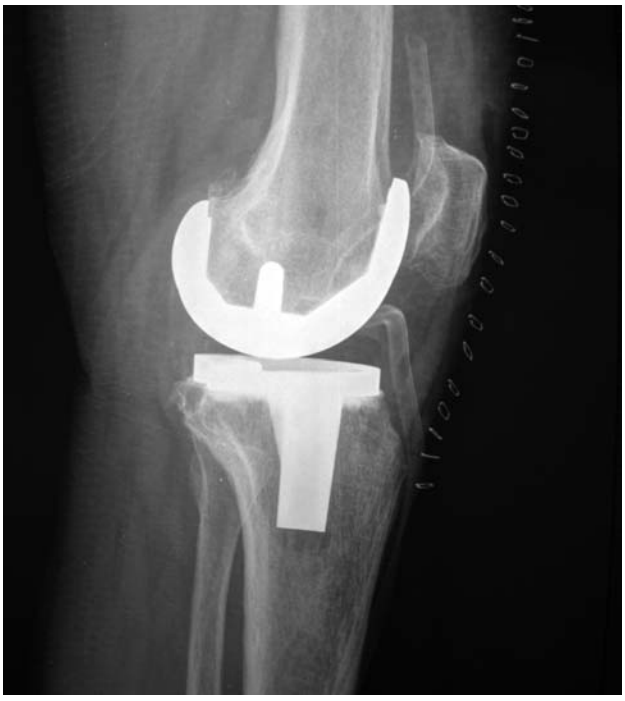

Fig. 5. Lateral radiograph of a patient who had total knee arthroplasty.

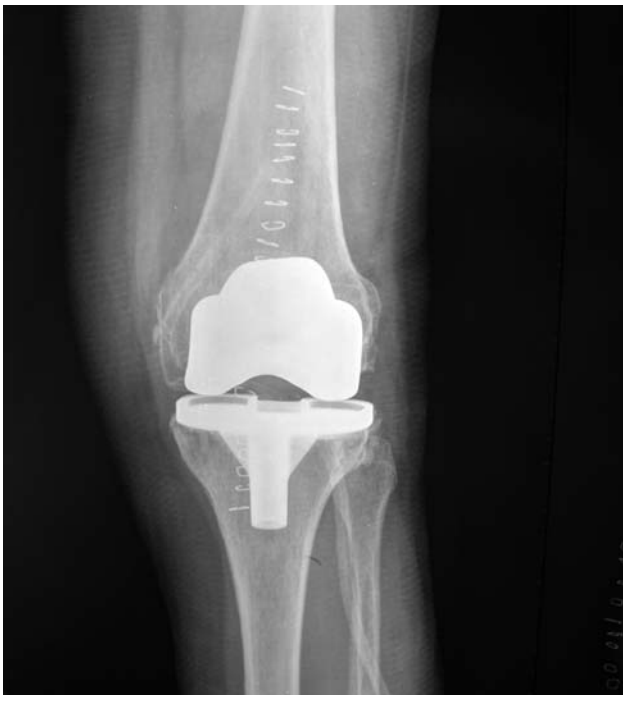

Fig. 6. Anteroposterior radiograph of a patient who had total knee arthroplasty.

women is almost equal, with a slightly higher prevalence among men.[4,5,8,9,13,15] In the present study, 47% of the patients were male and 53% were female which was inconsistent with ratios reported in the literature.[4,8,9,15]

Common complaints with PVNS are pain, swelling and contracture. Consistently, all patients in the present study had tolerable pain and swelling. Three limped and 9 had a contracture. Limping and contracture were not present in any of the patients with localized PNVS. A reactive process that develops due to chronic inflammation, direct trauma to the joint, repeated hemarthroses, overuse of the joint and disruption of the extensor mechanism is thought to be responsible for PVNS.[6,16-19] However, these etiologies are not confirmed. According to a recent theory, PVNS originates from chromosome anomalies and monoclonality.[15] In the present case series of 19 patients, only two patients presented with a history of trauma. This result indicates that factors other than trauma play a role in the etiology of PVNS.

Commonly, PVNS is considered to have two types: localized and diffuse. However, Beguin et al. introduced a third, a mixed form of PVNS, which is a variety of the diffuse form and contains pedicled-bulky tumors associated with diffuse synovitis.[20] Rydholm, on the other hand, introduced three types of PVNS; the extraarticular type commonly involving the hand, the solitary intraarticular nodular type commonly involving the knee and a diffuse villous, pigmented process involving the entire synovial membrane of large joints.[21]

The diffuse type of PVNS has a high risk of recurrence. The results are better in localized PVNS than in diffuse PVNS, regardless of whether arthroscopic or open synovectomy is applied.[2,4,6,10,22] Zvijak et al. reported a recurrence rate of 14% while the rate reported by Chin et al. in their case series of 40 patients was 18%.[4,23] In the study of Kotwal et al., it was noted that the factors contributing to the risk of recurrence were incomplete excision, mitotic activity and bone involvement.[24] O'Sullivan et al. interpreted bone involvement accompanied by muscle, skin and tendon involvement and extension to the neurovascular bundle as a sign of poor prognosis.[18] During our follow-up period of 80.2 months, recurrence was seen in 5 (26%) of the patients, all of whom had diffuse PVNS. Recurrence and loosening of the knee prosthesis did not occur in the patients who underwent total knee replacement. We believe

Table 1. Surgical treatment in pigmented villonodular synovitis of the knee.

No	Age/ Sex	Side	Complaint	Type	X-ray findings	Preop ROM limit.	Type of surgery	Follow-up period	Recurrence	Conclusion
1	36/M	Right	Pain, swelling	Diffuse	Soft tissue swelling	18°	Total synovectomy	156 months	_	
2	29/M	Left	Pain, swelling, limping	Diffuse	Normal	_	Total synovectomy	144 months	_	
3	48/F	Left	Mild pain, swelling	Diffuse	Normal	_	Total synovectomy	108 months	5 years later	Recovery
4	52/F	Right	Swelling with pain	Diffuse	Grade IV gonarthrosis	20°	TKA + T. synovectomy	108 months	_	
5	55/F	Right	Pain, swelling	Diffuse	Gonarthrosis	15°	TKA + T. synovectomy	96 months	3.5 years later	Total synovectomy
6	15/M	Right	Mass with pain	Local	Normal	_	Local synovectomy	96 months	_	
7	18/M	Right	Pain, swelling	Diffuse	Soft tissue swelling	_	Total synovectomy	84 months		
8	60/K	Left	Pain, swelling	Diffuse	Grade IV gonarthrosis	30°	TKA + T. synovectomy	60 months	_	
9	58/M	Left	Pain, swelling	Diffuse	Gonarthrosis	15°	Total synovectomy	48 months	4 years later	Total synovectomy
10	31/M	Right	Isolated mass, pain	Local	Normal	_	Local synovectomy	48 months	_	
11	47/F	Right	Swelling with pain	Diffuse	Early gonarthrosis	_	Total synovectomy	72 months		
12	23/F	Right	Lump with pain	Local	Normal	_	Total synovectomy	36 months	_	
13	59/M	Left	Pain, swelling, limping	Diffuse	Gonarthrosis	15°	Total sinovyektomi	60 months		
14	53/M	Right	Pain, swelling	Diffuse	Soft tissue swelling	_	Total synovectomy	96 months	2 years later	Recovery
15	17/M	Left	Pain, swelling	Local	Normal	_	Local synovectomy	120 months		
16	62/F	Right	Pain, swelling, limping	Diffuse	Gonarthrosis	17°	Local synovectomy	132 months	_	
17	37/F	Left	Swelling, takılama	Diffuse	Normal	_	Total synovectomy	24 months	3 years later	Radiotherapy
18	52/F	Left	Pain, swelling	Diffuse	Grade IV gonarthrosis	25°	TKA + T. synovectomy	18 months	_	
19	51/F	Right	Pain, swelling	Diffuse	Grade IV gonarthrosis	22°	TKA + T. synovectomy	15 months	_	

that this is the result of a complete synovectomy performed after osteotomy.

There is no consensus on the treatment of choice for PVNS. The general approach is surgery. Marginal excision for localized PVNS and total synovectomy for diffuse PVNS are recommended.[5,9] Open or arthroscopic synovectomy, external radiotherapy and intraarticular radiation synovectomy (yttrium, dysprosium, colloid chromic radiophosphate P32) are used for the treatment of diffuse PVNS in many medical centers. TNF-α antagonists are used against resistant PVNS. The efficiency of surgical or medical synovectomy, used alone or combined, has been shown in the literature.[1,2,6,7,22] Along with these methods, arthrodesis, bone grafting and primary arthroplasty are also used in cases with diffuse form of PVNS.[22] However, recurrence increases if synovectomy is not performed, and it is not always possible to perform using the arthroscopic method in cases with diffuse PVNS.

Malignant transformation is rarely seen in PVNS. Noting that malignant transformation is rare in PVNS, Schajowicz reported that histiocytic proliferation developed in only one patient in his case series of 80 patients.[25] Kalil and Unni, on the other hand, reported that malignant transformation developed in an 85 years old patient who had been diagnosed with PVNS at the age of 21.[26] We did not observe malignant transformations in any of our 19 patients during follow-up.

One of the largest problems with PVNS is that diagnosis may be delayed due to the insidious course of the disease. The time between the onset of complaints and the diagnosis in some cases may exceed months or even years. This may cause the focal diease to become gradually more aggressive and result in bone, muscle and tendon invasion. Schwartz et al. reported an average time lapse of 4 years between the onset of symptoms and the diagnosis of PVNS.[27] The similarities between PVNS and other intraarticular pathologies may also delay the diagnosis, resulting in the progression of the disease.[28] In our case series, the mean time between the onset of complaints and diagnosis was 23 months. As PVNS was inactive in 4 patients (31%), the diagnosis of the disease could not be made despite the long lasting symptoms. Diagnosis could only be made during the total knee replacement performed due to gonarthrosis.

In conclusion, PVNS is a rare disease of which the diagnosis is often delayed. This may result in the extension of the disease and involvement of the adjacent soft and bone tissues. The disease may become gradually more aggressive and recurrence, especially in diffuse PVNS, occurs frequently. PVNS must be considered in patients who present with joint effusion and develop contracture, and their examinations should be performed accordingly. The path to follow in treatment depends on the damage to the joint, the age of the patient and whether recurrence is present. There is no definitive treatment method. If possible, the probability of recurrence should be minimized through aggressive radical synovectomy, followed by internal or external radiotherapy, especially in diffuse PVNS.

Conflicts of Interest: No conflicts declared.

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