Pigmented Villonodular Synovitis: A Retrospective Single-Center Study of 122 Cases and Review of the Literature

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Objectives: Pigmented villonodular synovitis (PVNS) is a rare but disabling disease. The objective was to describe the clinical presentation and outcomes of PVNS according to its localization.

Methods: Retrospective, systematic study of all cases of biopsy-proven PVNS followed in 1 tertiary-care center specialized in isotopic synoviorthesis. Cases were selected by keyword. Collected data included disease localization, therapeutic modalities, and outcomes.

Results: A total of 122 cases (mean age 33.0 ± 13.1 years, 58% female, 89% diffuse form) of histologically confirmed PVNS were analyzed with a mean follow-up of 5.8 ± 4.3 years (707 patient-years total). The main localizations were the knee (75%) and ankle (16%). Clinical presentation included joint pain (80%) and joint effusion (79%) with hemarthrosis (75% of analyzed articular fluid). The mean delay before diagnosis was 2.9 ± 3.7 years. Magnetic resonance imaging was helpful for diagnosis in 83%. Surgical synovectomy was initially performed in 98% of cases and was often associated with isotopic synoviorthesis (knee: 57%; other localizations: 74%). In patients with a diffuse form treated at first line by surgery followed by isotopic synoviorthesis, the relapse rate was 30% (knee) and 9% (other localizations), respectively, with a mean delay before relapse of 2.6 ± 2.4 and 2.4 ± 0.9 years, respectively.

Conclusions: PVNS occurs in young adults, mainly in the knee joint; joint pain and effusion with hemarthrosis are the most frequent signs. Relapse is frequent, in particular, for diffuse knee PVNS; the usefulness of isotopic synoviorthesis remains to be confirmed.

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Pigmented villonodular synovitis (PVNS) is a rare disease characterized by proliferation of synovial tissue in the joint, tendon sheath, and bursa. The annual incidence was estimated to be 1.8 patients per million population in the USA (1). The etiology is unclear. Some authors think it is caused by chronic inflammation (2). Others suggest it is a tumor-like disorder. This theory is supported by monoclonality and chromosomal abnormalities (3-5). Histology of PVNS reveals hypertrophic synovial process characterized by villous, nodular, and villonodular proliferation and pigmentation from hemosiderin. The following 2 forms are described: diffuse (when 1 compartment or the entire synovium of a given joint is affected) or localized (when a single mass is affected) (6). The diffuse intra-articular form of PVNS most frequently affects the large joints, with the knee involved in 66-80% of cases (7,8). Localized intra-articular involvement represents 6% of all cases (1). Clinical symptoms depend on the localization, ie, intra-articular or extra-articular. Extra-articular localized PVNS (tendon or bursae) most frequently leads clinically to a soft-tissue mass and pain. Common clinical symptoms associated with the intra-articular type of PVNS are pain and swelling (9). The localized intra-articular form of PVNS is not visible on radiographs in most cases. Radiographs of patients with diffuse intra-articular PVNS may appear normal or can reveal joint effusion, soft-tissue swelling, extrinsic erosion of with a rim of sclerosis (9). PVNS is locally progressive and can cause joint damage (6). Treatment is not standardized to date; the anchor is surgical synovectomy. Recurrence can occur, although there is much heterogeneity: the relapse rate has been described as

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8-56%, differing with treatment, follow-up duration, definition of relapse, and outcome evaluation (7,10-14). Complementary therapy, such as isotopic synoviorthesis (13,15) or external radiotherapy (16-19), has been proposed but not validated.

To date, clinical and radiologic characteristics of PVNS were reported only in small series, mainly for PVNS affecting the digital tendon sheath (1,20,21). Therefore, data are lacking both on the clinical history of this disease and on the efficacy of treatment modalities.

Our center is a tertiary-care rheumatology unit with access to specialized isotopic synoviorthesis. Patients with PVNS are seen, either just after the diagnosis is made or after initial treatment and relapse.

The aim of the present study was to describe clinical and radiologic findings in PVNS and to evaluate outcomes and relapse rates, in particular, after isotopic synoviorthesis in the treatment of PVNS. Furthermore, a literature review of clinical presentation of PVNS and efficacy of isotopic synoviorthesis in treatment of PVNS was performed.

PATIENTS AND METHODS

Design

The design of the present study was a systematic monocentric retrospective analysis.

Patients

The patients in the present study included all cases of biopsy-proven PVNS seen in 1 center either as outpatients or inpatients, between June 1991 and October 2008. Histology was analyzed where the biopsy was performed (outside or in our hospital).

Patient Selection

Cases were selected by the keyword "PVNS" in the computerized database. A total of 133 patients were screened. Twelve were excluded: 10 had no PVNS (5 undetermined monoarthritis, 4 psoriasis arthritis, and 1 proliferative synovitis without histology of PVNS) and 2 had no interpretable data available regarding definite diagnosis and follow-up. Data of the 121 selected patients were collected retrospectively on hospital charts in paper files and completed by telephone to obtain follow-up information where needed. As 1 patient had 2 localizations (knee and ankle) of PVNS, the total number of cases was 122 for 121 patients.

Data Collection: Clinical Data

Demographic data (age at onset, gender, ethnicity, follow-up duration, diagnostic delay), joint localization, and clinical presentation (pain, joint effusion, hemarthrosis, posterior swelling) were collected. The follow-up was obtained from hospital charts, or if necessary, by a phone call. If patients did not answer, the last date where the patient was seen was chosen for last follow-up.

Imaging

Radiologic presentation (magnetic resonance imaging (MRI) findings, nodular or diffuse form) was collected. MRI was performed in 115 of 122 cases at diagnosis. MRI was collected as performed, ie, generally with T1, T1 with gadolinium injection, and T2 sequences. Hypertrophy of synovium, synovium contrast intake, and nodules were defined according to the local radiologist. Bone lesions were defined as bone erosions or cystic bone lesions on plain radiographs or on MRI. Hemosiderin deposits on MRI were not collected because echo gradient sequence was not performed or information was unavailable.

Treatment and Outcomes

Therapeutic modalities (surgical treatment, complementary treatment) and outcomes (relapse, delay before relapse) were collected. Treatment was either surgical synovectomy alone or synovectomy followed by isotopic synoviorthesis. Surgery was performed by arthroscopy and/or arthrotomy. Isotopic synoviorthesis was performed in the 6 months following surgery. The different isotopes were Yttrium 90 (4-5 mCi) for the knee, Rhenium 186 for the ankle (2 mCi), and hip (3 mCi). Isotopic synoviorthesis was performed under radioscopic control. During the synoviorthesis, a corticosteroid (triamcinolone hexacetonide) was injected with the isotope.

In the present study, relapse was defined as recurrence of synovitis on MRI, with or without clinical symptoms. Because our center is tertiary-care, some patients were initially treated elsewhere with various therapeutic modalities and were sent to us after 1 or several relapses. Because patients who have already relapsed may be more liable to relapse again, we decided, when analyzing relapse rates and to counter this potential bias, to study only patients initially treated in our department as first-line treatment.

Statistical Analysis

Analyses were descriptive by means and standard deviations (SD) or percentages. Comparisons between groups according to the joint localization were performed by Fisher's exact test or Wilcoxon's ranked test as appropriate.

RESULTS

Patient Selection

A total of 121 patients were screened. As 1 patient had 2 localizations (knee and ankle) of PVNS, the total number of cases was 122 for 121 patients. All 122 cases of histologically confirmed PVNS were analyzed. To confirm the diagnosis of PVNS, histology was analyzed by the local pathologist after