

Case Study



35 year old man twisted his ankle while playing and was surgically for treated for ankle sprain for instability symptoms 2 years. Increasing pain around the ankle and pain present all the time more on weight bearing.

Your Diagnosis?

Diagnosis PIGMENTED VILLONODULAR SYNOVITIS

Pigmented villonodular synovitis is a proliferative condition of the synovium.

Monoarticular involvement, the most common process, occurs in two forms: localized and diffuse.

The localized form is characterized by focal involvement of the synovium, with either nodular or pedunculated masses;

The diffuse form affects virtually the entire synovium.

The localized form has an excellent prognosis and a low recurrence rate when managed surgically.

The more common diffuse form has a reported recurrence rate of up to 46%.

Although the condition can present in any joint, the knee is the most commonly affected site.

Pigmented villonodular synovitis is often aggressive, with marked extra-articular extension.

Open synovectomy is the standard method of management. Arthroscopic synovectomy, which has gained popularity,

It has several advantages over the open technique, but it is associated with higher recurrence rates in diffuse pigmented villonodular synovitis.

Synovectomy by any approach, however, may prevent secondary osteoarthritis and subsequent joint arthroplasty.

Radiation-induced synovectomy has shown mixed results.

Combined surgical and nonsurgical approaches may be necessary, and in some patients, total joint arthroplasty may be the only effective treatment.

The use of adjuvant radiation treatment is still debated. The literature is sparse; however, there may be a limited role for intra-articular radiation treatment or external beam radiation treatment as an adjuvant to surgery.

Pigmented villonodular synovitis [Jaffe] presented the pathologic (PVNS) is a condition of the synovitis, thereby shift synovial membrane that is characterizing the focus from a neoplasm.

Pathogenesis

1. Injury history may be present in 1/3rd cases. But this condition is not due bleeding.

Pathology from chronic bleeders of Hemophilic joint is different from PVNS.

Hemophilic synovial pathology differs from PVNS, however, in that it lacks lipid-laden histiocytes and giant cells, which are considered classic indications of PVNS.

2. Abnormal local metabolic activity also has been suggested as an inciting event for the inflammation seen in PVNS, but not proved.

3. There is still some support in the literature to suggest that PVNS is a neoplastic process. The presence of trisomy 7 and clonal DNA rearrangements have been reported by several authors.

4. Oehler found in their analysis of cell populations in PVNS strong support for its being a chronic inflammatory process.

Currently, there are no conclusive data to substantiate the etiology of PVNS as either an oncologic or an inflammatory process.

Natural History

I **The localized form** [LPVNS] of the disease is characterized by a pedunculated, lobular lesion localized to one area of the synovium.

In the knee, LPVNS lesions occur most commonly in the anterior compartment. Most of these lesions arise at the meniscocapsular junction. The synovium in the region of the anterior horn of the medial meniscus is the most common site of involvement.

Patients with lesions in this location often present with signs and symptoms suspicious of meniscal pathology.

Pedunculated appearance of this discrete lesion in the infrapatellar fat pad, suprapatellar pouch, intercondylar notch been reported. Has a favorable prognosis. No studies have examined the long-term outcomes of patients left untreated for LPVNS, likely because LPVNS has a lower recurrence rate and is frequently more easily treated.

2. **The diffuse PVNS [DPVNS]** is characterized by involvement of most or all of the joint synovium [Cf trauma]. It is the more common form of PVNS.

Despite treatment, the recurrence rate for DPVNS is reportedly high.

Early reports of recurrence rates after treatment of PVNS were as high as 46%.

With careful and thorough surgical excision, however, the recurrence rate has been reported to be as low as 8%.

Figure 2

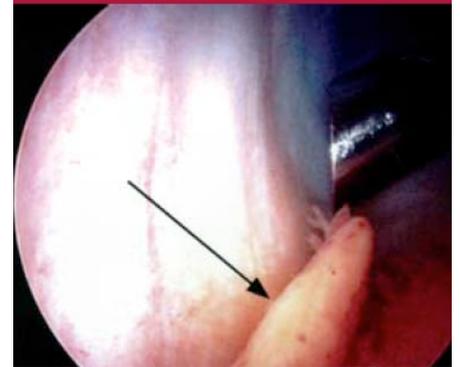


Figure 3



Early clinical experience proved that the natural history of marked by continued pain, swelling, and decreased range of motion of the affected joint. The continued inflammation and joint erosions that develop lead to articular cartilage destruction and subsequent osteoarthritis. The end result can be the need for total joint arthroplasty.

Although no radiographic changes were found at 2-year follow-up in one patient who refused treatment after biopsy proved the presence of PVNS, five of the seven patients required a total hip replacement before age 40 years.

Although a seemingly clear-cut distinction has been made between these 2 types, the two entities exist along a continuum of one disease process. It is important to note, however, that there are patients who present initially with Local PVNS who are later found to have more extensive disease involvement.

Clinical Presentation

The estimated incidence of PVNS is 1.8 per million

Slow and insidious onset of pain, swelling, and stiffness in the involved joint.

PVNS frequently has a delayed diagnosis or is misdiagnosed as early osteoarthritis, rheumatoid arthritis, a meniscal tear, or a ligamentous injury.

Local PVNS also presents with symptoms of pain and swelling, but it more commonly presents with locking, catching, and instability. Symptoms are often intermittent.

PVNS is typically a monoarticular process that often involves the large joints.

The knee is the most commonly involved joint, but others include the hip, ankle, shoulder, and elbow.

PVNS often appears in the third and fourth decades of life.

No sex-based predilection.

Diffuse PVNS: Tends to have a more rapidly destructive course and, as a result, a poorer prognosis.

Extra-articular PVNS can encroach on major neurovascular structures, making surgical excision more challenging and complete excision difficult.

Diagnosis

Only 17% of patients were appropriately diagnosed with PVNS before referral.

The majority of cases have no plain radiographic findings.

Plain radiographs can be helpful, particularly in the hip, elbow, and ankle.

They may show periarticular erosions, with a thin rim of reactive bone.

Reciprocal bony lesions on opposite sides of the joint, despite articular preservation, are highly suggestive of PVNS but also can be seen in other conditions.

A late finding of joint space narrowing on plain radiograph indicates articular cartilage loss, which can be difficult to distinguish from primary osteoarthritis.

The plain radiographic findings may be seen in as few as 30% of patients and in even fewer patients with knee involvement.



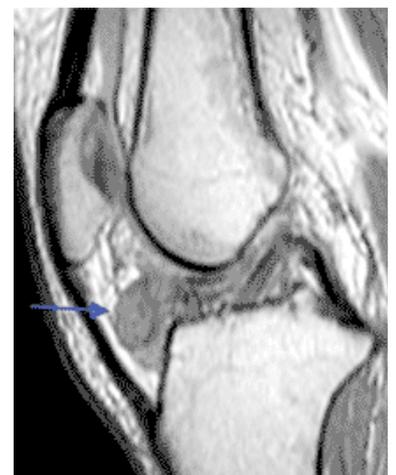
MRI

MRI is noninvasive and, with newer sequences, can be highly sensitive and specific.

It also can be helpful in determining the extent of disease involvement.

The high hemosiderin content causes the mass to appear as either a spotty or an extensive low signal on T1- and T2-weighted images. The signal is similar on both T1 and T2 sequences.

T1-weighted coronal magnetic resonance image of diffuse pigmented villonodular synovitis. Note the periarticular erosions on both sides of the joint line (arrows). Spotty low signal areas, representing hemosiderin deposition.



A localized pigmented villonodular synovitis lesion in the anterior Compartment.

3. Synovial fluid aspiration is a commonly reported technique. Brownish-stained bloody fluid is indicative of PVNS.

4. **Tl-201** Mackie reported six cases of biopsy-proven PVNS showing increased uptake of Tl-201 in both early and delayed images may be useful in recurrent cases.

5. **CT** for assessment of bone destruction

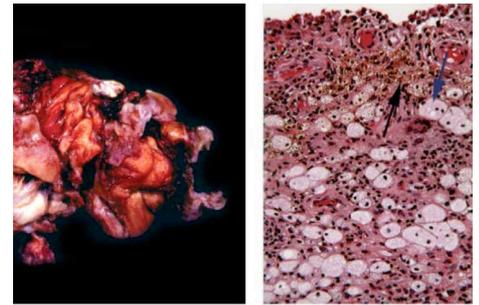
6. Histology

Diffuse, nodular pigmented villonodular synovitis lesion

No cellular atypia nor abnormal mitotic level

Characteristic presence of lipid-laden macrophages, multinucleated giant cells, hemosiderin deposition, and fibroblast cell proliferation.

Malignant conversion: Is rare. When occurs, the mortality rate was 50%.



B

Treatment

Goal: is to eradicate all abnormal synovial tissue, thus removing the source of pain and reducing the risk of joint destruction and recurrence. Patients presenting with recurrent disease often have much more extensive involvement and a poorer likelihood of success. A combination of surgical and nonsurgical approaches may be necessary; in some patients, total joint arthroplasty may be the only effective treatment.

Radiation Therapy

Radiation has been used for many years as an alternative to surgical synovectomy in patients with synovitis. Radiation-induced synovectomy for the treatment of PVNS has evolved, but results have been mixed.

Serious potential complications are associated with external beam radiation, however, including skin reactions, poor wound healing, joint stiffness, and sarcomatous transformation. No significant advantage has been reported to using adjuvant external beam radiation compared with surgical synovectomy alone.

O'Sullivan noted that external beam radiation therapy can be highly useful in managing refractory cases of PVNS or in those with extensive extra-articular involvement. In at least two patients who had failed repeated open and arthroscopic surgery, radiation therapy enabled limb amputation to be avoided.

More recently, interest has shifted from external beam radiation to intra-articular radiation synovectomy. Intra-articular radiation therapy has been used in Europe since the 1960s to manage rheumatoid arthritis and has been experimentally extended to the treatment of PVNS. Most reports regard it as an adjuvant treatment modality.

Chin: 300 mCi (using dysprosium Dy-165) after combined open anterior and posterior synovectomy. They reported a recurrence rate of 17% with radiation, compared with a 0% recurrence rate for open synovectomy alone. These results suggest no advantage to using intra-articular radiation for the treatment of PVNS.

One concern associated with the use of intra-articular radiation therapy is radionecrosis of the soft tissues.

Arthroscopic Treatment of PVNS

Familiarity on the part of most orthopedic surgeons with arthroscopic techniques has made the use of arthroscopy in the management of PVNS particularly attractive. Arthroscopy has been associated with better functional results and lower rates of postoperative stiffness than have open techniques. Conversely, improper application of this technology has been associated with unacceptable recurrence rates in some instances. Reports of arthroscopic synovectomy for PVNS have been almost exclusively in regard to the knee. Arthroscopic synovectomy in the knee offers excellent visualization of the anterior compartment as well as the medial and lateral recesses.

Arthroscopic synovectomy for PVNS in other joints, such as the shoulder and ankle, has been described but is uncommon.

Localized PVNS

Arthroscopic partial synovectomy is our preferred surgical option for LPVNS. Limited or partial synovectomy necessitates débridement of the PVNS mass along with a rim of surrounding healthy synovium. In most modern series, recurrence is rare after limited local treatment of LPVNS lesions.

Diffuse PVNS

DPVNS is considerably more common.

For DPVNS the posterior compartment is typically involved and requires a surgeon who is comfortable with the placement of access through posterior arthroscopic portals as well as the use of both 30° and 70° angled arthroscopes.

Ogilvie was among the first to report on arthroscopic management of DPVNS. They described 20 cases of DPVNS treated solely with arthroscopic synovectomy. Eleven patients were treated with complete synovectomy and nine with partial synovectomy. A thorough anterior synovectomy was performed in the partial group; however, the posterior compartment was not addressed. The patients undergoing complete synovectomy had a significantly lower risk of recurrence than did the patients undergoing partial synovectomy. The complete synovectomy group had only 1 recurrence out of 11, whereas the partial group had 5 failures out of 9 cases. The authors concluded that thorough, complete synovectomy is the treatment of choice for DPVNS.

Zvijac performed complete arthroscopic synovectomy on 12 patients with DPVNS and reported a 14% overall recurrence rate.

Open Surgical Management

Good success rates have been reported for LPVNS with open resection of the lesion.

In the knee, this is performed through a combined anterior and posterior approach. Our preferred anterior approach is through a midline incision and medial parapatellar arthrotomy.

After the anterior approach is performed, the patient is turned prone for the posterior approach. Use a “lazy S-shaped” incision posteriorly. The neurovascular bundle is carefully dissected, and both the medial and lateral heads of the gastrocnemius are detached. This should allow full exposure of the posterior joint capsule, in which an H-shaped capsulotomy is performed. The neurovascular bundle is shifted either medially or laterally to gain access to either side of the posterior aspect of the joint. Similarly, extensive open approaches can be performed for other joints afflicted with PVNS; however, because of the rarity of this condition, most series have focused on treatment of PVNS in the knee.

Early series of radical synovectomy in the knee with DPVNS reported excessively high recurrence rates.^{20,23} In 1968, Byers et al² reported a recurrence rate of 46% after open complete synovectomy.

Flandry reported a series of 25 knees with biopsy-proven DPVNS; open anterior and posterior complete synovectomy was done, with two anterior parapatellar incisions and one posterior medial incision. The authors reported a recurrence rate of 8% at average follow-up of 58 months.

Compared with arthroscopic synovectomy, open synovectomy is associated with a longer hospital stay and longer rehabilitation period. One major criticism of the open technique for PVNS in the knee is postoperative stiffness, which often requires manipulation to achieve a long-term decreased range of motion. The rate of postoperative stiffness was 24%.

Factors for recurrence of PVNS

- _ Delay in instituting treatment
- _ Intralesional primary treatment
- _ Intraoperative hesitation in thoroughly debriding the lytic lesions from the articular surface.
- _ Arthroscopic treatment for diffuse form of PVNS affecting large joints.

References

1. Flandry Diagnostic features of diffuse pigmented villonodular synovitis of the knee. CORR 1994;298:212-220.
2. Jaffe HL Arch Pathol 1941;31:731-765.
3. Chin J Bone Joint Surg Am 2002;84:2192-2202.
4. Pigmented Villonodular Synovitis J Am Acad Orthop Surg 2006;14:376385
5. Johansson CORR; 1982;163:159-166.
6. Ogilvie-: J Bone Joint Surg Am 1992;74:119-123.