SUMMARY AND CONCLUSION

Pigmented villonodular synovitis (PVNS) is a "benign proliferative disorder of uncertain etiology that affects synovium lining joints, bursae, and tendon sheaths".

Pigmented villonodular synovitis and Giant Cell Tumourof the Tendon Sheath (GCTTS) are closely related benign neoplasms that develop in the synovial lining of joints, tendon sheaths, and bursae of diarthrodial joints.

Two primary forms are described, including a diffuse form that affects the entire synovial lining of a joint, bursa, or tendon sheath and a rare focal or localized form. Both are sub-typed according to the site into intra- versus extra-articular lesion.

The incidence of pigmented villonodular synovitis is 1.8 cases per 1 million people per year, with no environmental, genetic, ethnic or occupational predilection. Approximately, 75% of all patients have the diffuse form.

Pigmented villonodular synovitis generally occurs in patients between the ages of 20 and 45 years, but it has been found in patients as young as 11 years and as old as 70 years.

Etiology remains controversial; however, several theories are based on the histologic appearance and cellular components of the lesion. Some authors have suggested the development of pigmented villonodular synovitis is caused by partial disturbance in lipid metabolism, inflammation, or a benign neoplastic process. The most widely held theory is that the disease is an inflammatory reaction of the synovium. However, some evidence exists that it is a benign neoplastic process.

Macroscopic features of PVNS include thick synovium comprising matted masses of villi and synovial folds or pedunculated nodules. This

accounts for the diffuse nodular distribution evident on scintigraphy. Coarse papillae have been described to have a "mossy," "straggly beard," or "shaggy carpet" appearance, while fine villi are described as "fern-like".

Most lesions contain considerable numbers of pigment-laden cells staining positively with Prussian blue reaction for ferric iron.

Recurrence is the major concern with PVNS with 9-46% recurrence rates reported for the diffuse-type PVNS. Nodular PVNS has reported lower recurrence rates of 5–29%.

There are two factors associated with a greater incidence of recurrence: the diffuse type of PVNS and the location in large joints. However, despite recurrence, surgical treatment of PVNS leads to good functional results. Preoperative MRI is successful in providing the correct preoperative diagnosis. Magnetic resonance imaging, also, facilitates detection of recurrence especially in patients who are asymptomatic.

Thus, from a practical point of view, pigmented villonodular synovitis should be regarded as locally aggressive, non-metastasizing lesions. Therapy should be based on a desire to remove the tumour as completely as possible without producing sever disability to the patients.

Pigmented villonodular synovitis refers to a synovial proliferative disease that usually occurs in the large joints. PVN presents in a nonspecific and insidious manner and remains a diagnostic challenge.

Although the basic radiographic features of pigmented villonodular synovitis have been discussed in orthopaedic and radiographic literature, there is a paucity of literature discussing the clinical presentation of patients with synovial proliferative disorders.

It is usually slow-growing with subtle presentations. The average duration of symptoms before diagnosis had been ranged from 2-72 months.

Patients with pigmented villonodular synovitis often have vague complaints and physical findings, but there are clues that should raise a clinician's suspicion for pigmented villonodular synovitis and direct additional examination.

☒ The intermittent nature of pain, complaints of extreme pain, monoarticular involvement, young age of the patient at presentation, and positional relief should raise suspicion for pigmented villonodular synovitis.

☒ Certain radiographic clues also can increase a physician's suspicion for pigmented villonodular synovitis such as bony erosions appear as cyst-like structures on anteroposterior (AP) radiographs. Sclerosis may be present around the erosion, and the joint space usually is preserved until late in the disease.

☒ Joint aspiration reveals blood stained synovial fluid, which in the absence of a history of trauma, is highly suggestive of pigmented villonodular synovitis. Confirmation of the diagnosis can be made with biopsy of the synovium.

☒ Up to 90 percent of patients complain of mild to moderate tenderness, mainly over the medial patellofemoral area.

☒ In addition, arthrocentesis yields blood-tinged synovial fluid in 44 to 69 percent of patients.

Periarticular soft tissue density is one of the characteristic findings. Only few exhibit erosive and cystic changes. Calcifications are a very rare occurrence. Fifty-one percent of patients with PVNS have osteocartilagenous abnormalities, which include cortical pressure erosions and delayed joint space narrowing.

Magnetic resonance imaging (MRI) is a noninvasive, highly accurate diagnostic modality in characterizing PVNS. Apart from diagnosis, MRI also is useful in defining the extent of disease because the

ligaments, tendons, menisci, and cartilage are visualized in addition to the lesion.

The gold standard for diagnosis is biopsy of the suspected lesion. Synovial biopsy usually reveals the definitive diagnosis of pigmented villonodular synovitis. The biopsy can be performed either arthroscopically or via open arthrotomy, depending on the location and extent of involvement. PVNS is rarely definitively diagnosed preoperatively.

Local excision is sufficient in localized PVNS. Total or subtotal synovectomy is the treatment of choice in diffuse PVNS.

It was reported that the limited combined anterior and posterior approach has provided inadequate access to all posterior sites of pigmented villonodular synovitis in the knee, especially when there is advanced disease with extra-articular extension. The presence of extra-articular disease in their patients prompted them to pursue a more radical debridement without sacrificing the capsule or major ligaments of the knee. It is unlikely that arthroscopic or limited open synovectomy will provide adequate visualization for resection. Because of the higher risk of recurrence after initial operative procedures and incomplete synovectomy, a more extensile approach to the posterior structures of the knee was developed for patients presenting with advanced primary disease or recurrent disease with extra-articular extension.

Arthroscopic total synovectomy has lower operative morbidity, decreased risk of joint stiffness, lower risk of wound complications, and more rapid rehabilitation when compared with open synovectomy. Arthroscopic total synovectomy for diffuse PVNS of the knee must be meticulous and include close examination of the posterior compartments to minimize recurrence.

However, arthroscopic total synovectomy is technically demanding due to difficulty visualizing the posterior synovium, with proper technique and appropriate use of posteromedial, posterolateral, and posterior trans-septal portals, adequate visualization and debridement of the posterior synovium can be achieved via arthroscopy. Using this technique, total 5-compartment arthroscopic synovectomy can be an effective primary treatment modality for diffuse pigmented villonodular synovitis of the knee.

This method involves creating an aperture through the posterior septum to allow passage of the arthroscope from the posteromedial to posterolateral compartments and vice versa.

The posterior transeptal portal may be used in any patient in whom the posterior joint spaces must be visualized completely and accessed for an arthroscopic procedure. The surgeon may plan to use the transeptal portal preoperatively by identification or confirmation of these pathologies through magnetic resonance imaging (MRI), radiography, or ultrasound. Also, however, the need for a transeptal portal may be unforeseen preoperatively but created intra-operatively to address posterior joint space pathology detected on routine anterior portal arthroscopy.

Advantages of arthroscopic treatment include lower operative morbidity, ability to treat other knee pathology, decreased risk of joint stiffness, lower risk of wound complications, and more rapid rehabilitation.

The complications of synovectomy are similar to these of arthroscopy in general. These include major complication such as hemarthrosis, infection, thrombosis, neurologic injury, recurrence, reflex sympathetic dystrophy, and adhesion, and minor complication affecting wound healing, such as erythema and blisters.

High relapse rates of PVNS led to increasing interest in radiation synovectomy as a possible alternative or even supplementary treatment approach. Highly significant clinical improvement and diminished blood-pool activity of three-phase bone scan images were reported after combined surgical and radiation synovectomy in 11 patients, lasting for at least one year.

De-bulking surgery using a traditional open or arthroscopic synovectomy (subtotal synovectomy) in selected patients with primary or recurrent extensive diffuse PVNS, leaving behind a microscopic amount of disease, followed by intraarticular Y^{90} injection is a good and safe solution as a joint-saving procedure. In this way we avoid the disadvantages of more aggressive total synovectomy.

Y^{90} injection is a reliable adjuvant for surgery in the management of diffused PVNS. Local tumor control and good function, associated with only mild morbidity are achieved in the majority of the patients.

Total knee arthroplasty, which is often superior for the treatment of degenerative joint disease, also is a viable option for patients who have diffuse pigmented villonodular synovitis of the knee with destructive joint changes. Total knee replacement can provide a pain-free, well functioning extremity in a patient who would otherwise have stiffness and pain.