RECENT TRENDS IN MANAGEMENT OF PIGMENTED VILLONODULAR SYNOVITIS OF THE KNEE

Essay Submitted for the partial fulfillment of Master Degree in Orthopedics

Presented by

Islam Mohamed Ibrahim El-Desouky
M.B.,B.Ch. (Cairo University)

Supervised by

Prof. Dr. Hazem Abd El-Azeem

Professor of Orthopedics
Faculty of Medicine
Cairo University

Prof. Dr. Hisham Mesbah

Assistant Professor of Orthopedics
Faculty of Medicine
Cairo University

Dr. Ahmed El-Ghoneimy

Lecturer of Orthopedics
Faculty of Medicine
Cairo University

Faculty of Medicine
Cairo University
2009

ACKNOWLEDGEMENT

First of all thanks to God who helped me all through my life and helped me to fulfill this work in this way.

Thanks to professor Doctor Hazem Abdelazeem whom I was honoured to have the chance to do this work under his supervision.

Thanks to professor Doctor Hisham Mesbah for his sincere help in choosing this interesting issue and for guiding me althrough.

Thanks to Doctor Ahmed El Ghoneimy for his great help in collecting this data and presenting it in this way.

Special thanks to my parents and my sweet wife who helped me alot through this work.

ABSTRACT

Pigmented villonodular synovitis **(PVNS)**, is a locally destructive fibrohistiocytic proliferation that is characterized by many villous and nodular synovial protrusions , which affects joints, bursae, and tendon sheaths .

The etiology is unknown and is often controversial , some investigators have suggested an autoimmune pathogenesis, trauma, disturbance in lipid metabolism, an inflammatory response to an unknown agent, or benign neoplasms.

PVNS is a benign condition but its problem lies with: difficult to diagnose early as symptoms are nonspecific and present late, difficult to differentiate from other conditions such as Rheumatoid arthritis, Osteoarthritis, inflammatory, neoplastic diseases of the synovium and high incidence of recurrence after surgical treatment.

Imaging plays an important role in the diagnosis of PVNS ; Plain X- Rays reveal a soft tissue density in the affected joint, erosion of subchondral bone with a sclerotic margin. MRI is extremely useful in making a diagnosis because on T2 – weighted images the intraarticular masses demonstrate a combination of a high-signal-intensity areas, representing fluid and congested synovium, interspersed with areas of intermediate to low signal intensity, secondary to random distribution of hemosiderin in the synovium .

Treatment options may include: Synovectomy which may be open or arthroscopic also some authors included physical adjuvants in their treatment protocols in order to decrease the rate of recurrence. Radiation synovectomy/radiosynoviorthesis.

Recently α- blockade with infliximab has been reported as an effective therapy also there might be a role for specific inhibitors of RANKL proteins in the treatment of PVNS.

Key words:

Pigmented villonodular synovitis (PVNS), synovectomy, radiotherapy, arthroscopy, RANKL *(**R**eceptor **A**ctivator of **N**uclear factor **K**appa β **L**igand)*, TNFα- blockade.

List of Contents

List of Figures

16	Axial T1 -weighted magnetic resonance imaging (MRI) scans show tissue areas with abnormally low and high signals anteriorly in the suprapatellar pouch and, posteriorly, deep and superficial to the medial head of the gastrocnemius**.**	27
17	Sagittal MR images of the knee in a male, 30 years old. Lateral X-ray (a-e)	28
18	Lipoma arborescens: MRI (A): Sagittal proton density MRI shows numerous structures with suprapatellar bursa exhibiting signal intensity consistent with fat.Coronal (B) and sagittal (C) T2- weighted fat suppressed images demonstrate high signal intensity joint effusion.	31,32
19	Histopathology of chondroblastoma- like pigmented villonodular synovitis A,B,C.	34
20	T1-weighted MRI of a 31-year-old male with recurrent diffuse PVNS of the knee.	37
21	Surgical technique. Operative photos of anterior synovectomy.A & B.	38
22	Surgical technique. A schematic of the skin incision of the posterior approach.	39
23	Surgical technique. A schematic of the exposure of the popliteal space.	40
24	A schematic of popliteal fossa dissection.	40
25	Surgical technique. Posterior resection of extra and intra-articular PVNS A and B.	41,42
26	Surgical technique.A schematic of the posterior capsule reconstruction. Macroscopic examination of the synovium before (A) and after (B) 20 weeks of treatment with anti-TNF α.	42
27	Macroscopic examination of the synovium before (A) and after (B) 20 weeks of treatment with anti-TNF α.	52
28	Two pathological specimens of pigmented villonodular synovitis before and after 20 weeks of treatment with anti-TNF α.	53
29	Frontal view in a X-ray without any radiologic mass.	58
30	Lateral view with posterior bone erosion.	59
31	Sagittal spin-echo T1-weighted MR image**.** Internal condyle erosion and PVNS in the posterior area of the knee, above the femoral Condyle	59
32	Sagittal FE T2-weighted MR image.	60
33	Axial FE T2-weighted MR image.	60
34	Sagittal spin-echo T1-weighted MR image, 7 years postoperatively, with PMMA filling the bone defect.	61
35	Axial FSE FATSAT T1-weighted MR image**,** 7 years postoperatively with PMMA filling the bone defect.	62
36	Coronal FSE FATSAT T1-weighted MR image, 7 year postoperatively, with PMMA filling the bone defect.	62

List of Abbreviations

- PVNS Pigmented Villonodular Synovitis
- CT Computed Tomography
- MRI Magnetic Resonance Imaging
- FFE Gradient-recalled echo sequence
- STIR Short Timed Interval Recovery sequence
- Y-90 Yttrium -90
- TNF Tumor Necrosis Factor
- RT Radiation Therapy
- ACL Anterior Cruciate Ligament
- PCL Posterior Cruciate Ligament
- RANKL Receptor Activator of Nuclear factor Kappa β Ligand
- OPG Osteoprotegerin
- PMMA Polymethylmethacrylate

INTRODUCTION

Pigmented villonodular synovitis **(PVNS)**, is a locally destructive fibrohistiocytic proliferation that was first described by ***Jaffe***, ***Lichtenstein***, and ***Sutro*** [1] in 1941 and it is characterized by many villous and nodular synovial protrusions ,which affects joints, bursae, and tendon sheaths. [2,3,4] This name describes the lesion as it is of a yellow-brown, villous, and nodular appearance. The yellow-brown pigmentation is due to excessive deposits of lipid and hemosiderin. This condition can be diffuse or localized. When the entire synovium of the joint is affected and there is a major villous component, the condition is referred to as diffuse PVNS. When a discrete intraarticular mass is present, the condition is called localized PVNS [5,6,7] or pigmented giant cell tumor of articulations. When the process affects the tendon sheaths, it is called localized giant cell tumor of the tendon sheaths or nodular tenosynovitis. [8,9,10,11,12,13]

The diffuse form usually occurs in the knee, hip, elbow, or wrist. The localized nodular form is often regarded as a separate entity. It consists of a single polypoid mass attached to the synovium. Nodular tenosynovitis is most often seen in the fingers and is the most common soft tissue tumor of the hand, exceeded only by the ganglion[14]. In the new revised classification of soft tissue tumors (2002), the WHO classifies localized intra- and extraarticular lesion as giant cell tumor of sheath[15] , whereas diffuse intra- and extraarticular forms are categorized as diffuse- type giant cell tumor (keeping PVNS as a synonym).[16]

One of the most characteristic findings in PVNS is the ability of the hyperplastic synovium to invade the subchondral bone, producing cysts and erosions.[17]

Although a few “malignant” PVNS have been reported in the literature[18,19] this diagnosis is still debatable. Recently, attention has been drawn to the extraarticlar form of diffuse PVNS, also referred to as diffuse-type giant cell tumor. This condition is characterized by the presence of an infiltrative, extrarticular mass with or without involvement of the adjacent joint. [20]

This presentation of PVNS creates a real diagnostic challenge for both radiologist and pathologist because its extraarticular location, invasion of the bones, and more varied histologic infiltrative pattern may suggest malignancy. [20,21]

ANATOMY

Anatomy of the knee joint[22]

The knee-joint was formerly described as a ginglymus or hinge-joint, but is really of a much more complicated character. It must be regarded as consisting of three articulations in one: two condyloid joints, one between each condyle of the femur and the corresponding meniscus and condyle of the tibia; and a third between the patella and the femur, partly arthrodial, but not completely so, since the articular surfaces are not mutually adapted to each other, so that the movement is not a simple gliding one.

This view of the construction of the knee-joint receives confirmation from the study of the articulation in some of the lower mammals, where, corresponding to these three subdivisions, three synovial cavities are sometimes found, either entirely distinct or only connected together by small communications. This view is further rendered probable by the existence in the middle of the joint of the two cruciate ligaments, which must be regarded as the collateral ligaments of the medial and lateral joints.

The existence of the patellar fold of synovial membrane would further indicate a tendency to separation of the synovial cavity into two minor sacs, one corresponding to the lateral and the other to the medial joint.The bones are connected together by the following ligaments:

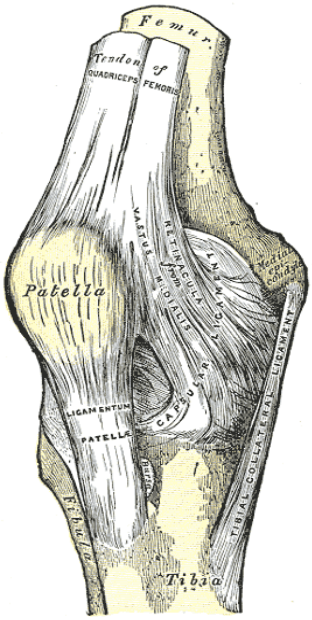
The Articular Capsule.	The Anterior Cruciate.
The Ligamentum Patellæ.	The Posterior Cruciate.
The Oblique Popliteal.	The Medial and Lateral Menisci.
The Tibial Collateral.	The Transverse ligament.
The Fibular Collateral.	The Coronary ligament.

The Articular Capsule (*capsular ligament*): *(Fig.1)*

The articular capsule consists of a thin, but strong, fibrous membrane which is strengthened in almost its entire extent by bands inseparably connected with it. Above and in front, beneath the tendon of the Quadriceps femoris, it is represented only by the synovial membrane. Its chief strengthening bands are derived from the fascia lata and from the tendons surrounding the joint. In front, expansions from the Vasti and from the fascia lata and its iliotibial band fill in the intervals between the anterior and collateral ligaments, constituting the **medial** and **lateral patellar retinacula.** Behind the capsule consists of vertical fibers which arise from the condyles and from the sides of the intercondyloid fossa of the femur; the posterior part of the capsule is therefore situated on the sides of and in front of the cruciate ligaments, which are thus excluded from the joint cavity. Behind the cruciate ligaments is the oblique popliteal ligament which is augmented by fibers derived from the tendon of the Semimembranosus. Laterally, a prolongation from the iliotibial band fills in the interval between the oblique popliteal and the fibular collateral ligaments, and partly covers the latter. Medially, expansions from the Sartorius and Semimembranosus pass upward to the tibial collateral ligament and strengthen the capsule.

The Ligamentum Patellæ (*anterior ligament*): **(*Fig.1*)**

The ligamentum patellæ is the central portion of the common tendon of the Quadriceps femoris, which is continued from the patella to the tuberosity of the tibia. It is a strong, flat, ligamentous band, about 8 cm. in length, attached *above* to the apex and adjoining margins of the patella and the rough depression on its posterior surface; *below,* to the tuberosity of the tibia; its superficial fibers are continuous over the front of the patella with those of the tendon of the Quadriceps femoris. The medial and lateral portions of the tendon of the Quadriceps pass down on either side of the patella, to be inserted into the upper extremity of the tibia on either side of the tuberosity; these portions merge into the capsule, as stated above, forming the medial and lateral patellar retinacula. The posterior surface of the ligamentum patellæ is separated from the synovial membrane of the joint by a large infrapatellar pad of fat, and from the tibia by a bursa.



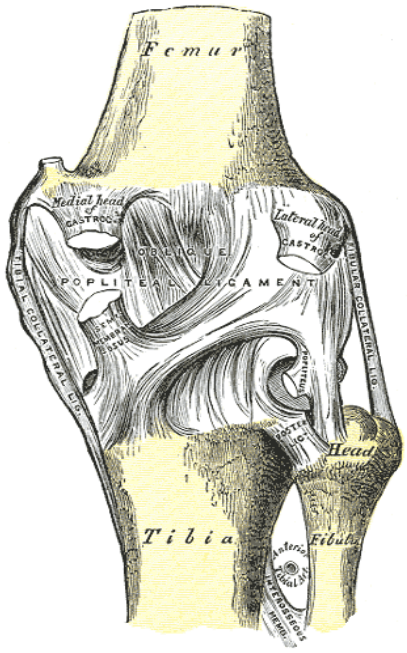
(*Fig.1*) Right knee-joint. Anterior view.[22]

The Oblique Popliteal Ligament (*posterior ligament*): (*Fig.2*)

This ligament is a broad, flat, fibrous band, formed of fasciculi separated from one another by apertures for the passage of vessels and nerves. It is attached above to the upper margin of the intercondyloid fossa and posterior surface of the femur close to the articular margins of the condyles, and below to the posterior margin of the head of the tibia. Superficial to the main part of the ligament is a strong fasciculus, derived from the tendon of the Semimembranosus and passing from the back part of the medial condyle of the tibia obliquely upward and lateralward to the back part of the lateral condyle of the femur. The oblique popliteal ligament forms part of the floor of the popliteal fossa, and the popliteal artery rests upon it.

The Tibial Collateral Ligament (*ligamentum collaterale tibiale; internal lateral ligament*): (*Fig.1*)

The tibial collateral is a broad, flat, membranous band, situated nearer to the back than to the front of the joint. It is attached, *above,* to the medial condyle of the femur immediately below the adductor tubercle; *below,* to the medial condyle and medial surface of the body of the tibia. The fibers of the posterior part of the ligament are short and incline backward as they descend; they are inserted into the tibia above the groove for the Semimembranosus. The anterior part of the ligament is a flattened band, about 10 cm. long, which inclines forward as it descends. It is inserted into the medial surface of the body of the tibia about 2.5 cm. below the level of the condyle. It is crossed, at its lower part, by the tendons of the Sartorius, Gracilis, and Semitendinosus, a bursa being interposed. Its deep surface covers the inferior medial genicular vessels and nerve and the anterior portion of the tendon of the Semimembranosus, with which it is connected by a few fibers; it is intimately adherent to the medial meniscus.



(*Fig.2*)– Right knee-joint. Posterior view.[22]

The Fibular Collateral Ligament (*ligamentum collaterale fibulare; external lateral or long external lateral ligament*): (Fig. 3).

The fibular collateral is a strong, rounded, fibrous cord, attached, *above,* to the back part of the lateral condyle of the femur, immediately above the groove for the tendon of the Popliteus; *below,* to the lateral side of the head of the fibula, in front of the styloid process. The greater part of its lateral surface is covered by the tendon of the Biceps femoris; the tendon, however, divides at its insertion into two parts, which are separated by the ligament. Deep to the ligament are the tendon of the Popliteus, and the inferior lateral genicular vessels and nerve. The ligament has no attachment to the lateral meniscus.An inconstant bundle of fibers, the **short fibular collateral ligament,** is placed behind and parallel with the preceding, attached, *above,* to the lower and back part of the lateral condyle of the femur; *below,* to the summit of the styloid process of the fibula. Passing deep to it are the tendon of the Popliteus, and the inferior lateral genicular vessels and nerve.

The Cruciate Ligaments (*ligamenta cruciata genu; crucial ligaments*):

The cruciate ligaments are of considerable strength, situated in the middle of the joint, nearer to its posterior than to its anterior surface. They are called *cruciate* because they cross each other somewhat like the lines of the letter X; and have received the names **anterior** and **posterior,** from the position of their attachments to the tibia. **The Anterior Cruciate Ligament** (*ligamentum cruciatum anterius; external crucial ligament*) **(Fig. 4)** is attached to the depression in front of the intercondyloid eminence of the tibia, being blended with the anterior extremity of the lateral meniscus; it passes upward, backward, and lateralward, and is fixed into the medial and back part of the lateral condyle of the femur.