Pigmented villonodular synovitis of the knee joint: Place of magnetic resonance imaging (MRI)

C Ayadi [1*], N Amsiguine [2], M Karouchi [3], N Rguieg [4], J El Fenni [5], H Ennouali [6]
[1-2, 5-6] Department of Radiology, Military Hospital Mohammed V, Rabat, Morocco
[3] Department of Pediatric Surgery, University Hospital Ibn Sina, Rabat, Morocco
[4] Department of Anatomopathology, Military Hospital Mohammed V, Rabat, Morocco

* Corresponding Author: **C Ayadi**

Article Info

ISSN (online): 2582-8940
Volume: 03
Issue: 03
July-September 2022
Received: 18-07-2021;
Accepted: 21-08-2021
Page No: 44-46

Abstract
Pigmented villonodular synovitis (PVNS) is a rare benign proliferative disorder of the synovium of joints, bursae, and tendon sheaths, with unknown etiopathogeny. It is a synovial hyperplasia related to the formation of villosities and nodules characterized by an intracellular hemosiderin deposit. We present the case of a 27-year-old patient, without previous history, who reported pain and swelling of the right knee. Magnetic resonance imaging evoked the diagnosis of villondular synovitis, particularly on the T2 gradient echo sequence, which was confirmed by histology after synovectomy. This pathology can be diffuse, which is the most frequent form, or local, and affects young adults between the 2nd and 4th decade of life. The most affected location is the knee joint. The diagnosis is based on a combination of radiological criteria, in particular MRI, which identifies the lesion, and histological criteria, which identify the presence of intracellular hemosiderin. The objective of this work is to show the interest and technique of MRI in the diagnosis of villonodular synovitis.

Keywords: Villonodular synovitis, MRI, gradient echo, hemosiderin, synovectomy

Introduction
PVNS is a rare benign disease, affecting 1.8 million people worldwide [1]. It is characterized by a proliferation of cells of the villous and nodular type on the synovial membrane, and generally occurs in middle-aged people [2]. Two forms are distinguished, localized, also called giant cell tumor, and diffuse, which is the more aggressive with a high risk of recurrence and malignant transformation [3]. The diagnosis is double radiological, especially MRI, and histological. The present case illustrates the clinical-radiological aspects and highlights the diagnostic difficulties of this pathology,

Presentation of case
A 27-year-old patient presented a slow-growing mass of the right knee for one year. He did not report any history of trauma or other medical or surgical history. On examination, there was a swelling of the medial aspect of the right knee that was tough, irregular and painful, estimated at 5 cm long. The patient had already performed an ultrasound (US) of the knee, which showed a significant and diffuse joint effusion within which a hypertrophied, irregular and heterogeneous synovitis was visualized.
MRI is then indicated, which objectified diffuse and irregular synovial thickening with nodular structures characterized by low signal in T1, intermediate intensity in T2, and low signal in gradient echo (GE), heterogeneously enhanced, and associated with significant joint effusion (Fig. 1). Therefore, the diagnosis of PSVN is evoked.
An open synovectomy was performed, and the anatomopathological study confirmed the diagnosis of villonodular synovitis by showing a dense polymorphic and inflammatory infiltrate, composed of histiocytes, siderophagi, and giant cells, the vessels were numerous with fibrohyaline walls (Fig 2). The clinical evolution after surgery was favorable in the short term.

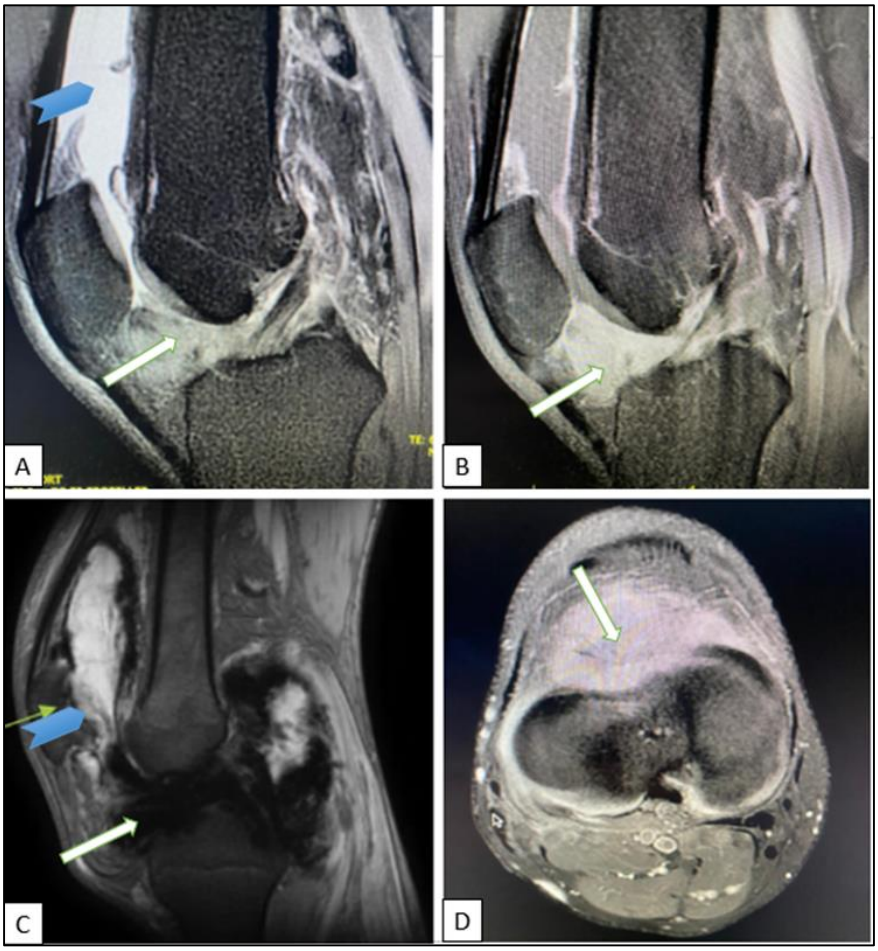

Fig 1: MRI of the right knee in sagittal T2 (A), T2 EG (C), and sagittal (B) and axial (D) gadolinium injection weighted sequences, showing an intra-articular effusion (blue arrow) in hyposignal T1, intermediate signal T2 with empty areas of signal better visible in T2 EG evoking hemosiderin deposits, with lesion enhancement (white arrow)

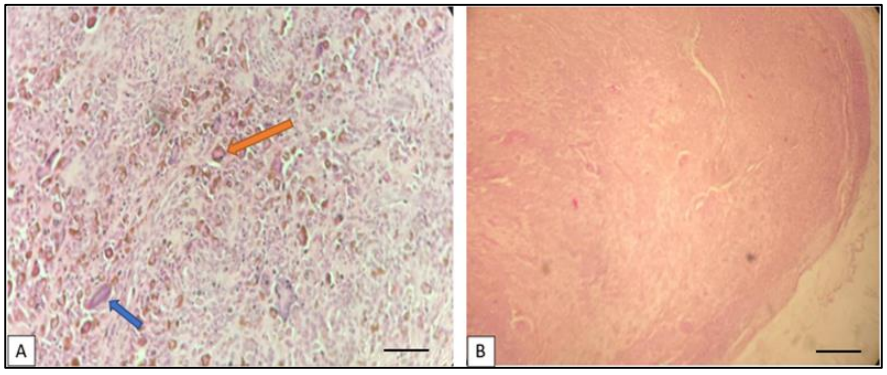

Fig 2: (A) Dense polymorphic inflammatory infiltrate made of histiocytes, siderophagous (orange arrow), and giant cells. There are many vessels with fibro hyaline walls (blue arrow) (Original magnification 100 x scale bar 50µm). (B) Polypoid synovial lining (blue arrow). (Original magnification 40 x scale bar 20µm)

Discussion

Pigmented villonodular synovitis (PVNS) is a benign inflammatory proliferation of the articular synovium that was first described in 1941 by Jaffé, and corresponds to intracellular deposition of hemosiderin, associated with nodular and villous hyperplasia and thickening of the synovium [3]. It is a rare disease with an estimated incidence rate of 1.8 cases per million inhabitants per year [4].

SVN mainly affects young adults, with a predominance of age between the second and fourth decade of life, without gender preference; the most affected joint is the knee in 66-80% of cases [4] as in our case. It then affects the ankle and hip in the remaining cases, and exceptionally the temporomandibular joint, sternoclavicular joint, the fingers or the spine [5]. Extra-articular forms are also possible and may involve the tendon sheaths or the synovial bursa.

We distinguish two types of SVN, a diffuse form that affects the entire synovial membrane called diffuse-type giant cell tumor (Dt-GCT) or pigmented villonodular synovitis (PVNS), and a localized form in a single area of the synovium, called tenosynovial giant cell tumor (TGCT), which is more rare, representing 6 to 11% of cases [6]. In fact, they are the same disease in terms of histology, but their prognosis is different. Evolution of the diffuse form is

characterized by multiple recurrences, with a high incidence rate of 21 to 85% after treatment [6]. In contrast, the localized type has a better prognosis with very low risk of recurrence after synovectomy. Malignant transformation has been rarely reported in the literature [7].

The symptomatology of PVNS is rarely characteristic, with a variable delay before consultation, which corresponds to the presented case whose diagnosis was evoked only after one year. It manifests classically by a recurrent effusion with pain that is most often chronic and moderate, functional impairment that can cause quadricipital amyotrophy. Some forms have misleading clinical manifestations, the most frequent is an isolated and chronic popliteal cyst. Therefore, the absence of any characteristic element makes a correct differential diagnosis more difficult.

The radiography is most often normal, especially if it is a localized form. Diagnosis of NVS on MRI is based on detecting nodular thickening of the articular synovium containing areas characterized by low signal intensity on T1 and T2 weighted images. Gradient echo (GE) T2 sequence shows areas of empty signal related to hemosiderin deposits that are pathognomonic of PVNS [8]. However, MRI remains non-specific in differential diagnosis between PVNS and other chronic hemorrhagic forms.

Up to now, the only way to diagnose PVNS is based on histological examination, synovial biopsy performed under open surgery or arthroscopy, makes PVNS diagnosis with certainty: it reveals hypervascularized papillary synovial tissue as well as a chronic inflammatory infiltrate composed of foamy histiocytes, mononuclear cells, giant cells, with multiple hemosiderin deposits [9].

The main differential diagnosis is Synovialosarcoma both clinically, radiologically and histological aspects. The treatment is based on total synovectomy, if possible arthroscopic or after arthrotomy, these two techniques can be complementary to ensure a total excision. In case of diffuse lesions, surgery must be more extended [10].

Conclusions

Villonodular synovitis is an uncommon pathology of the young adult, it mainly affects knee joint. Diagnosis is based on imaging in particular MRI with Gradient Echo-sequence detecting hemosiderin deposits in synovial nodules. However its confirmation remains histological. The treatment is surgical with risk of recurrence in case of diffuse form

References

1. Fałek A, Niemunis-Sawicka J, Wrona K, *et al*. Pigmented villonodular synovitis. Folia Med Cracov. 2018; 58:93-104.
2. Daoud J, Aouad D, Hassan Y, El Rassi G. Localized pigmented villonodular synovitis of the posterior knee compartment with popliteal vessel compression: A case report of arthroscopic resection using only anterior knee portals. Case Rep Orthop, 2018, 7532358.
3. Sugandhi A, Kondaveeti SKS, Sunder A. A Case of Pigmented Villonodular Synovitis. Cureus. 2022; 14(6):e25957. Doi: 10.7759/cureus.25957. PMID: 35855244; PMCID: PMC9286003.
4. Rachdi M, Feki A, Gassara Z, Bouaziz MA, Ben Jemaa S, Akrout R, *et al*. Villonodular synovitis (VNS): about a series of 11 cases. Revue Du Rhumatisme. 2020; 87:A268. doi:10.1016/j.rhum.2020.10.484.
5. Ma X, Shi G, Xia C, *et al*. Pigmented villonodular synovitis: A retrospective study of seventy five cases. Int Orthop. 2013; 37:1165-70.
6. Ottaviani S, Ayral X, Dougados M, Gossec L. Pigmented villonodular synovitis: A retrospective singlecenter study of 122 cases and review of the literature. Semin Arthritis Rheum. 2011; 40:539-46.
7. Bertoni F, Unni KK, Beabout JW, Sim FH. Malignant giant cell tumor of the tendon sheats and joints (malignant pigmented villonodular synovitis). Am J Surg Pathol. 1997; 21:153-63.
8. Warren Garner H, Ortiguera CJ, Nakhleh RE. Pigmented villonodular synovitis. Radiographics. 2008; 28(5):1519-23. PubMed.
9. Faten Frikha, *et al*. Une synovite villonodulaire localisÃ©e au coude: Ã propos dÂ´un cas. PAMJ - Clinical Medicine. 2021; 5:43. [doi: 10.11604/pamj-cm.2021.5.43.22246]
10. Dines J, Berardino T, Jason L, Dodson C, Warren F, Shindle M, *et al*. Long-term follow-up of surgically treated localized pigmented villonodular synovitis of the knee.Arthroscopy. 2007; 23(9):930-937.