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Diffuse pigmented villonodular synovitis: An exuberant presentation

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CASE REPORT

We present a case of a 44-year-old male, with a history of right knee swelling from insidious onset and two years of progressive evolution. There was not history of trauma or other associated symptoms (fever, pain, asthenia, weight loss, and other swollen joints).

He was referred to the orthopedic consultation by the primary health care specialist, after unspecific knee magnetic resonance imaging (MRI) and needle arthrocentesis, followed by unsuccessful physical therapy sessions.

At physical exploration it was clear a tender and voluminous swelling of the right knee, especially at the sub-quadricipital bursa, without other clear inflammatory signs (Figure 1). The patient denied other symptoms or even knee pain at exploration, although it as present on daily activities.

The X-ray images were considered normal, without bony lesions. At the MRI, there was a signal increase with heterogeneous shape, especially at sub-quadriceps and suprapatellar bursa, without bony erosion (Figure 2). However, small areas of hippo-signal were observed which determine some intra-sac septations and vegetations, aspects which may be compatible with pigmented villonodular synovitis (PVNS).

He has proposed to knee arthroscopy and needle arthrocentesis for final diagnosis and further treatment. It was performed under general anesthesia, the initial arthrocentesis revealed a pathognomonic xanthochromic synovial fluid (Figure 3) >120 cc and the arthroscopy images (Figure 4) were clear of vascularized polypus formations spread through the entire synovial cavity, with higher density in sub-quadricipital area. Tissue and liquid samples were acquired for cytochemical, histologic, and microbiological identification.

The histologic analysis revealed characteristic of chronic inflammation with, reactive hyperplasia of synovial tissue and presence of high hemosiderin deposits in the synovial stroma (Figure 5A).

At a second surgical time, after confirmation of diagnosis, the patient was submitted to total synovectomy with necessity of conversion from arthroscopic to open synovectomy (Figure 5B).

Postoperative period and rehabilitation showed with no complications. At two years of follow-up there is still no clinical or imagiological sign of recurrence, and maintains annual consultation.

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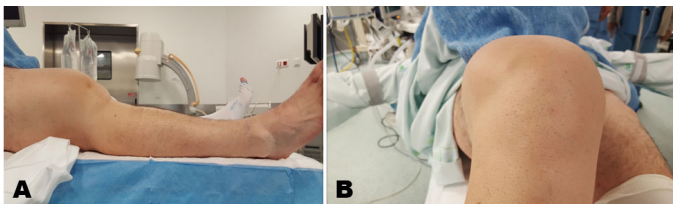

Figure 1: Preoperatory aspect of right knee. (A) Lateral view, in extension. Augmented volume of knee, mainly seen on the suprapatellar region. (B) Front view, in 90° of flexion. Supero-external volume augmentation.

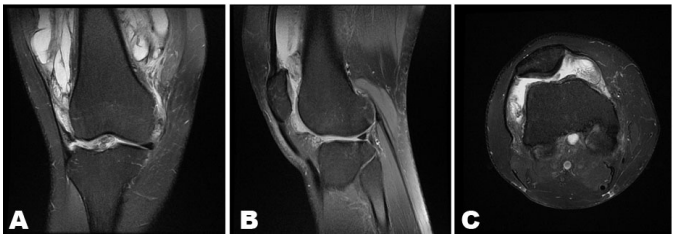

Figure 2: MRI study. (A) T2 Sagittal reconstruction, general synovial proliferation with nodular aspect. (B) T2 Coronal reconstruction, general synovial proliferation, with no bony erosion. (C) T2 Axial reconstruction, general synovial proliferation, occupying the femoropatellar space.

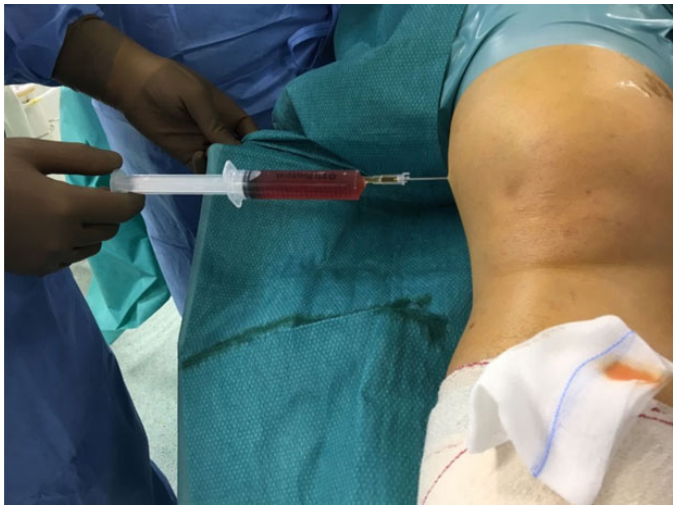

Figure 3: Intraoperatory knee aspiration from lateral patellar portal with xanthochromic synovial fluid.

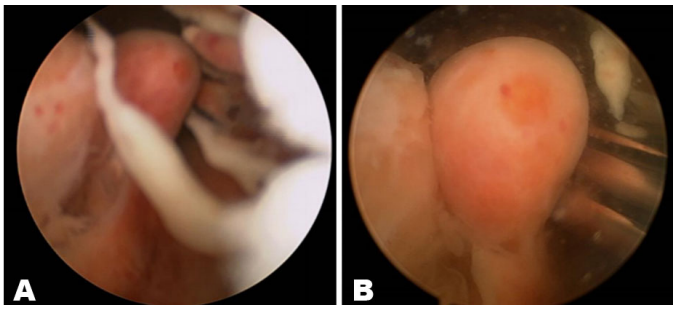

Figure 4: Arthroscopic appearance. (A) Multiple papillar highly vascularized intra-articular formations covering all aspect of synovial, from the sub-quadricipital area. (B) Intra-articular formation biopsied to histologic diagnosis.

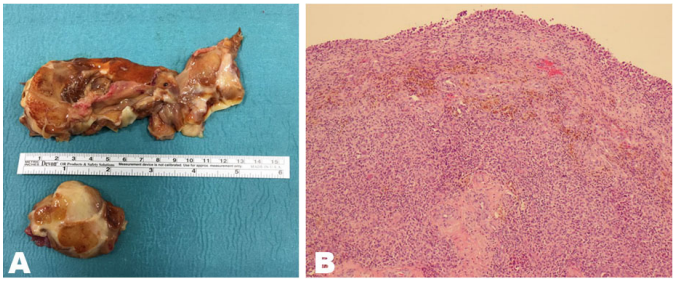

Figure 5: Recession piece: (A) Macroscopic appearance of synovial resected piece. (B) Microscopic evaluation of part of synovial hemosiderin deposits clear visible.

DISCUSSION

Pigmented villonodular synovitis is a rare, benign but potentially locally aggressive and recurrent disease, occurring 1.8 cases per million of inhabitants/year worldwide [1]. It occurs mainly in young adults, predominantly in females, between 30 and 40 years of age [2]. It is a chronic proliferative tumor that may involve joints, tendons, and bursa, characterized by World Health Organization (WHO) as giant cell tumor [3]. The diffuse intra-articular form corresponds only to 23% of these tumors and 0.9% of all benign soft tissue tumors [1].

The occurrence of malignant transformation and metastasis, although rare, is described in the literature [4]. It is typically monoarticular and affects large joints, most often in the knee (80% of cases), hip (15% of cases), and ankle (5% of cases) [2]. The etiology of PVNS is unknown, although numerous causes are pointed out potential, such as the occurrence of a chronic inflammatory process, recurrent trauma, hemarthrosis, inflammatory reaction to an unidentified agent or a local disorder of the lipid metabolism in the synovial membrane [1].

Xanthochromic or dark red joint arthrocentesis may help in diagnosis but its present in only 64% of patients. X-ray evaluation has a minor role in diagnosis workup, reserved for exclusion of other pathology. Magnetic resonance imaging findings are diagnostic in more than 95% of patients and histopathology is still recognized as the gold standard for the final diagnosis of PVNS [5, 6].

The treatment is surgical, with arthroscopic or open resection. In localized PVNS, local lesion excision by arthroscopy is advocated. In diffuse PVNS, synovectomy (total or subtotal) is recommended, but the role of arthroscopy is debatable as it affects a larger area of knee and predominantly affects the posterior compartment, the most difficult to access by arthroscopy [7].

CONCLUSION

Although benign, pigmented villonodular synovitis can result in significant morbidity if left untreated. Pain, loss of function, and joint destruction may occur. In primary treatment it is surgical, with recession via synovectomy or/and radiotherapy. Recurrence is reduced with complete resection, which is best achieved with localized form, with relapse rates from 10% to 30% to the diffuse form. It is mandatory to maintain patients with diffuse form of PVNS in follow-up for several years after surgery.

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Keywords: Diffuse, Knee, Pigmented villonodular synovitis

How to cite this article

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Author Contributions

Pedro Xavier Fernandes – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Raquel Filipa Corda Teixeira – Conception of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Hugo Filipe Marques Ribeiro – Conception of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Afonso Cevadinha Caetano – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

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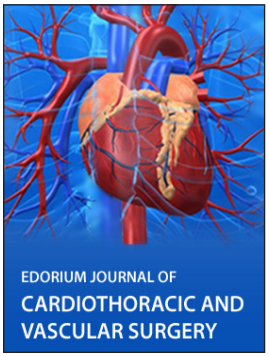
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.



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