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Update Article

Diffuse pigmented villonodular synovitis in knee joint: diagnosis and treatment☆

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ABSTRACT

Pigmented villonodular synovitis is a rare proliferative condition of the synovium. Although the condition can present in any joint, the knee is the most commonly affected site. Despite being a benign condition, pigmented villonodular synovitis is often aggressive, with marked extra-articular extension in some cases. Monoarticular involvement occurs in two forms: localized and diffuse. The latter is more common, with a high recurrence rate. There is no standard method of management of this lesion. Open surgery is a classical and effective method for treatment. Arthroscopic synovectomy, however, has gained popularity, and has several advantages over the open technique particularly in exclusively articular cases. The combined approach is suggested in cases with extra-articular involvement. Synovectomy through any approach may prevent secondary osteoarthritis and subsequent joint arthroplasty. Internal irradiation or external beam radiation as an adjuvant treatment to surgical synovectomy appears to decrease the rate of local recurrence in diffuse cases. The authors observed a great heterogeneity in reporting of functional results, and specific conclusions should not be drawn. Each patient should be managed in accordance with his/her particular condition.

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Sinovite vilonodular pigmentada difusa no joelho: diagnóstico e tratamento

RESUMO

A sinovite vilonodular pigmentada é uma rara condição proliferativa da membrana sinovial. Apesar de a doença poder estar presente em qualquer articulação, o joelho é o local mais frequentemente afetado. Ainda que doença benigna, geralmente tem comportamento agressivo, pode ter extensão extra-articular em alguns casos. O acometimento monoarticular ocorre em duas formas: localizada ou difusa. A forma difusa é mais comum e tem alta taxa de recorrência. Não há método padronizado para o manejo dessa lesão. O tratamento cirúrgico aberto é o método clássico e efetivo. A sinovectomia artroscópica, entretanto, tem ganhado popularidade e tem diversas vantagens sobre a técnica aberta, principalmente em casos exclusivamente articulares. A abordagem combinada é sugerida em casos com envolvimento extra-articular. A sinovectomia pode prevenir a osteoartrose secundária e o subsequente tratamento reconstrutivo. A radioterapia usada como tratamento adjuvante à sinovectomia parece diminuir a taxa de recorrência local na forma difusa da doença. Os autores encontraram grande heterogeneidade na forma como os resultados funcionais foram reportados e não se deve chegar a conclusões específicas. Cada paciente deve ser manejado de acordo com suas particularidades.

Palavras-chave:
Sinovite pigmentada vilonodular
Joelho
Radioterapia

Introduction

Pigmented villonodular synovitis (PVNS) is a rare proliferative process that affects the synovial joints, tendon sheaths, and bursas. In 1852, Chassaignac[1] reported the first case of a lesion in the flexor tendon sheath of the second and third fingers; this was subsequently reported in other joints. In 1941, Jaffe et al.[2] coined the term "pigmented villonodular synovitis"; subsequently, Granowitz et al.[3] expanded the terminology, distinguishing the localized (LPVNS) and diffuse (DPVNS) forms from other synovial lesions. Recently, the World Health Organization has defined PVNS and giant cell tumor to be equivalent terms.[4,5]

The estimated incidence of PVNS ranges around 1.8 per million.[6,7] It is usually monoarticular, affecting large joints. The knee is the most affected site (28%–70%), but cases in the hip, ankle, shoulder, and elbow are often observed.[5,6,8] The disease presents in two forms, localized or diffuse, and both types have similar appearance: a synovial membrane characterized by inflammation and presence of hemosiderin deposits.[3,9] Microscopically, it is characterized by the presence of lipid-laden macrophages, multinucleated giant cells, hemosiderin deposits, and proliferation of fibroblasts and stromal cells. LPVNS is characterized by discrete or pedunculated nodular lesions. In turn, DPVNS is the most common presentation, involving intra-articular tissues; it may have extra-articular extension, behaving as a chronic process.[10–12]

In the last 100 years, little progress has been made regarding treatment. The goal of PVNS treatment is to remove all synovial tissue in order to relieve pain, decrease the risk of joint destruction, and prevent local recurrence. Several treatment options have been proposed for this disease in cases of genicular involvement, ranging from observation and radical local surgery to total knee arthroplasty (Fig. 1).[4,5,8,13,14]

Etiology and pathophysiology

The etiology of PVNS is still unknown. Some authors suggest that the disease occurs as a result of baseline trauma and subsequent bleeding in the affected joint.[13–20] This theory is supported by the fact that patients with hemophilia present progressive destruction of the cartilage during the natural course of the disease. However, studies that produced PVNS-like histological findings by injecting iron or blood into the joint were unable to replicate the classic lipid-laden histiocytes and giant cells.[15] However, most studies reported a history of trauma in less than one-third of patients. Abnormal metabolic activity has also been indicated as an adjuvant event in the inflammation observed in PVNS, but this is an inconsistent finding.[16] There are also reports in the literature that PVNS may be a neoplastic process. Several authors suggested the presence of chromosome 7 trisomy and clonal rearrangements as a cause.[11,17] Some reports, albeit rare, indicate the occurrence of malignant transformation and metastases in patients initially diagnosed with PVNS.[17,18] Despite the case reports of malignant PVNS and aneuploidy, there is evidence against the theory that PVNS is a neoplastic process. In their analysis, Oehler et al.[9] observed a strong evidence of chronic inflammation. Their findings were based on the presence of a cellular marker of inflammation with a heterogeneous population of mononuclear cells. They also postulated that the presence of large amounts of iron in the lesion would stimulate synoviocytes and fibroblasts to develop macrophage-like characteristics.

Histologically, LPVNS and DPVNS are similar. However, they differ in clinical presentation, prognosis, and response to treatment. In the knee, LPVNS is more frequently observed in the anterior compartment.[12] Flandry et al.[19] reported that most lesions begin in the meniscocapsular junction. The most

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