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Case report

PVNS of the knee simulating lipoma arborescens on MR imaging

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Abstract

A case of pigmented villonodular synovitis of the knee, simulating lipoma arborescens, is presented. The MR imaging findings are correlated with histology with emphasis to the presence of fatty tissue in the lesion that could lead to misdiagnosis.

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1. Introduction

Pigmented villonodular synovitis (PVNS) is a rare benign disorder characterized by a villous proliferation of the synovium of unknown cause, which may occur either in focal or, more frequently, in diffuse forms [1,2]. PVNS usually affects patients in the second and third decades of life, with no sex predominance. The MR imaging characteristics of PVNS are considered not pathognomonic but strongly suggestive of the diagnosis [2–4], since the presence of hemosiderin, which is paramagnetic, causes signal loss. The overall signal intensity of the lesion depends on the proportions of hemosiderin, fat, and fibrovascular elements. Very rarely, predominant areas with fat signal may be present (3). Such a case, with histological proof of abundant fatty tissue, is presented and the MR imaging findings that could lead to misdiagnosis are discussed.

2. Case report

A 35-year-old female, presented with a 10-year history of mild pain and swelling in the right knee. The pain and swelling was getting worse at the standing position. The clinical examination revealed swelling along with a moderate degree of limitation of flexion but no warmth.

An aspiration of the joint revealed a yellowish fluid. The plain X-ray (not shown) showed only soft tissue swelling. A MR imaging on the first evaluation (Fig. 1), showed intraarticular fluid and frond-like projections simulating lipoma arborescens. Few low-signal intensity strands were thought to correspond to chemical shift artefact or to hemorrhagic foci within the lesion. There were no erosions or cysts in the bone, the joint space was intact and the bone marrow had normal signal intensity. The patient refused surgery and after clinical deterioration came back two years later for a repeat evaluation and possible surgical operation. A new MR imaging was asked which showed increased dimensions of the lesion along with identification of small low-signal intensity nodules and strongly enhancing synovium (Fig. 2). The patient was operated via a middle anterior incision. A poorly defined mass was evident and an incomplete excision of the mass and synovium was performed. The gross pathologic analysis showed that the involved joint capsule was thickened and the resected synovium measured $13 \times 12 \times 2.7$ cm³. The synovium was studded with nodules and irregular papillary projections that ranged from rubbery gray-white tan to brown and to soft yellow. Between the nodules, adipose tissue was obvious (Fig. 3a). On cross sections, the cut surface revealed large areas with chocolate-brown hemosiderin deposition. Prominent villous projections ranging from slender papillae to thicker irregular nodules were also noticed. The yellowish regions corresponded to lipid deposition in foamy histiocytes.

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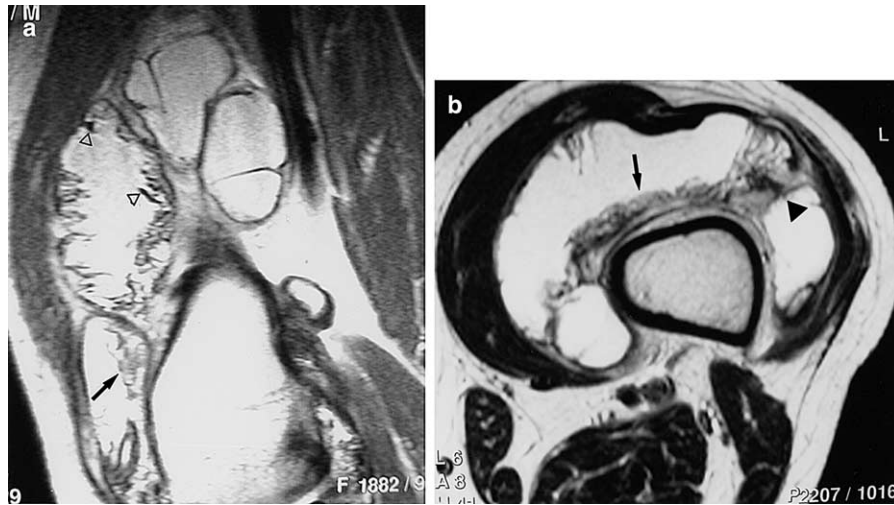


Fig. 1. Pigmented villonodular synovitis of the knee: MR imaging. (a) Sagittal Proton density Fast Spin Echo (TR/TE:4000/22 ms) and (b) T2-weighted Fast Spin Echo (TR/TE:4000/70 ms) demonstrate the intraarticular effusion and the synovial fatty elements (arrows). The low-signal intensity frond-like projections (open arrowheads) were erroneously attributed to chemical shift artifact. The low-signal intensity lesion (arrowhead) in (b) was not originally evaluated.

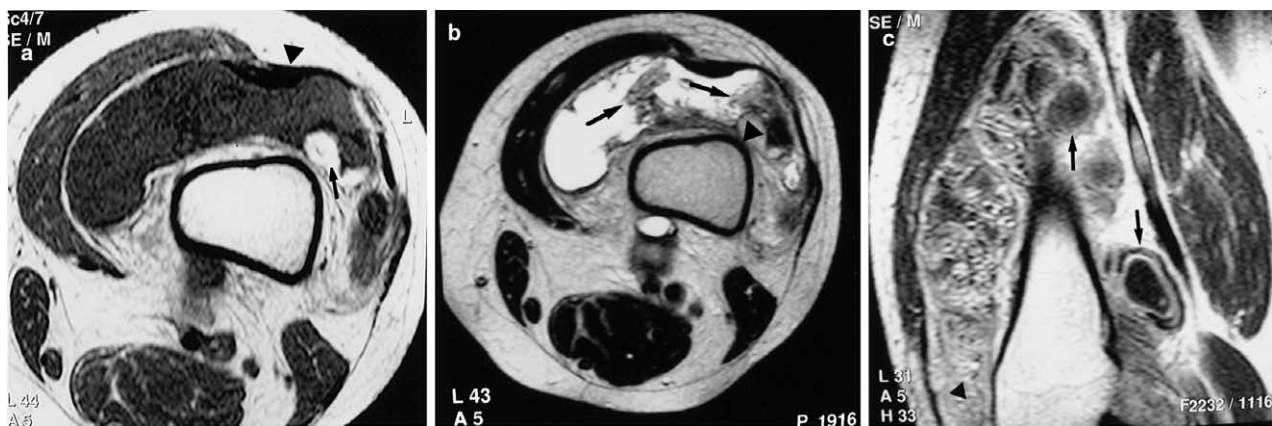


Fig. 2. Pigmented villonodular synovitis of the knee: MR imaging 2 years after Fig. 1. (a) Axial T1-weighted Spin Echo image (600/15), shows the intraarticular effusion, fatty tissue of synovial origin (arrow) and a low-signal intensity lesion anteriorly (arrowhead). (b) Axial T2-weighted Fast Spin Echo (6000/95) shows the fatty elements (arrows) and the low-signal intensity PVNS lesions (arrowhead), (c) sagittal contrast enhanced T1-weighted Spin Echo (600/15) image shows the synovial enhancement (arrowhead) along with the non-enhancing intraarticular low-signal intensity PVNS lesions (arrows).

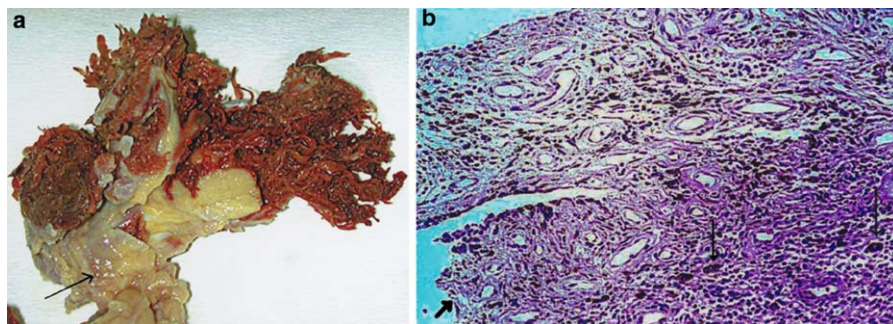


Fig. 3. (a) Macroscopic view of the lesion demonstrates the yellowish areas (arrow) that correspond to lipid deposition ($\times 80$). (b) In this nodular area of the tumor, sheets of histiocytes, foamy macrophages, chronic inflammatory cells and scattered multinucleated giant cells (thin arrows) are blended; focally denuded synovium (thick arrow) in this particular tumor permits a direct contact of the cellular infiltrate with synovial fluid (H + EX230).

The microscopic examination of the lesion showed large areas of mononuclear histiocytic cells intermingled with chronic inflammatory cells and a few, irregularly distributed multinucleated giant cells. Hemosiderin deposits were prominent. A variable number of foamy histiocytes laden either with hemosiderin or lipid, were distributed throughout the lesion individually or forming clusters. Adipose tissue was also noticed between the nodules or villous projections (Fig. 3b). Mitotic figures were few and typical. The villous structures were irregular and mostly covered by a thickened synovial layer that merged with the underlying cellular infiltrate, while in several foci the synovium was denuded, the cellular infiltrate being in direct contact with the synovial fluid. The final diagnosis was intraarticular, diffuse PVNS of the left knee.

3. Discussion

PVNS of the knee joint is a difficult entity to define and characterize. This disorder most often appears in the young adult knee with nonspecific clinical features, including a painful limitation of motion and rarely a sensation of locking. Detection and diagnosis of this localized soft-tissue mass are difficult because plain roentgenograms may be totally within normal limits. In cases of PVNS, MR imaging is the method of choice since the hemosiderin deposition leads to signal loss on both the T1- and T2-weighted images, particularly on gradient echo sequences [4–7]. Advantages of MR imaging include high-contrast multiplanar images that depict cortical bone, marrow, ligaments and tendons, fat, menisci, and articular cartilage in one image. In addition, it is noninvasive and requires no ionized radiation. MR imaging is an excellent clinical tool for the evaluation of intraarticular tumors of the knee joint [4]. Microscopically, considerable variability is seen in the histologic pattern. Therefore, there are cases of PVNS that hemosiderin is intermixed with adipose tissue and depending upon the proportions of each the characteristic imaging findings may be absent [3].

The present case of a 35-year-old woman with poor clinical findings, illustrates a limitation of MR imaging to define and characterize PVNS in cases that adipose tissue predominates over hemosiderin deposition. The first MR imaging examination (Fig. 1), revealed a gross effusion along with frond-like projections, simulating thus a lipoma arborescens. Few low-signal intensity strands were thought to correspond to chemical shift artifact or to hemorrhagic foci within the lesion. There was no joint space loss, bony erosions and cysts, and low-signal intensity nodules. In the second MR imaging, a poorly circumscribed low-signal intensity area within the fatty elements (Fig. 2), was evaluated as a possible PVNS.

Lipoma arborescens is a rare disorder characterized by villous lipomatous proliferation of the synovial tissue. This condition is more common in men than in women, and most affected patients are in the fifth to sixth decades of life. It is usually monoarticular, occurring most frequently in the knee, particularly in the suprapatellar pouch [8–11]. Affected patients usually report a long-standing, slowly progressive swelling of the joint with recurrent effusions, and variable pain and limitation of motion [8–11]. Erosive bone changes at articular margins are absent or minimal. All the above can exist in PVNS and are non-specific. MR imaging characteristics of lipoma arborescens include: villous synovial proliferation with a signal intensity similar to that of fat, mass-like subsynovial fat deposit, joint effusion, potential chemical shift artifacts at the interface of the synovial lesion and the effusion, and no evidence of hemosiderin deposition [9,12–14]. The differential diagnosis must include PVNS, synovial hemangioma, and synovial osteochondromatosis.

4. Summary

A case of diffuse PVNS of the knee, simulating lipoma arborescens, is presented. In PVNS, MR imaging is the method of choice since the hemosiderin deposition leads to signal loss on both the T1- and T2-weighted images, particularly on gradient echo sequences. Microscopically however, considerable variability is seen in the histologic pattern. Therefore, there are cases of PVNS that hemosiderin is intermixed with adipose tissue and depending upon the proportions of each the characteristic imaging findings may be absent. The MR imaging findings of such a case are presented and are correlated with histology with emphasis given to the presence of fatty tissue in the lesion that could lead to misdiagnosis.

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