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An unusual cause of recurrent joint effusions: nonhemophilic hemosiderotic synovitis of the knee

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A 20-year-old female presented with a painful swelling in the right knee and snapping sensation on joint motion that appeared without trauma and recurred several times. She had no history of a bleeding disease or trauma. Physical examination showed no signs of rash or temperature change or systemic or local findings of an infection. The knee was tender and knee motion was painful, with 90 degrees of flexion and full extension. The ballottement test was positive. All laboratory tests including rheumatologic and bleeding parameters were normal. Joint effusion analysis was normal except for its rusty-brown color. Magnetic resonance imaging showed synovial hypertrophy and grade 2 degeneration in the medial meniscus. During diagnostic and surgical arthroscopy, rust-colored synovial hypertrophy was noted in the suprapatellar pouch accompanied by patchy villi and nodules and cystic changes. The gross appearance of the synovium mimicked that of pigmented villonodular synovitis. Biopsy specimens were obtained from different parts of the synovium and a subtotal synovectomy was performed. The histopathologic diagnosis was reported as hemosiderotic synovitis. During a three-year follow-up, she had no pain, snapping sensation, or limitation of motion. There were no recurrent effusions.

Key words: Arthroscopy; diagnosis, differential; hemosiderosis/complications; knee joint/pathology; synovitis/surgery.

Joint effusions are seen with considerable frequency in orthopedic practice. In particular, the knee is the most affected joint. Many factors such as trauma, infections, inflammatory arthritis, osteoarthritis, and synovial pathologies may cause effusion in a joint. Synovial pathologies may present as acute or chronic synovitis. Chronic synovitis refers to a persistent, nonspecific, proliferative lesion of the synovium, usually monoarticular, with little or no involvement of the articular cartilage or bone, and without clear evidence for any other primary pathological process.[1] Correct etiological diagnosis of joint effusions needs a detailed analysis of both laboratory and radiodiagnostic tests. However, these tests may not always be helpful to diagnose some rare types such as hemosiderotic synovitis.

In this report, we present a case of hemosiderotic synovitis with diagnostic procedures and treatment.

Case report

A 20-year-old female presented with a swelling in the right knee that appeared without any evident trauma two years before and recurred several times. She gave a history of a bulky mass on the lateral side of the knee, being painful at times and giving a sensation of snapping on joint motion. The swelling recurred three times during this period and responded well to conservative treatment within seven to ten days. However, the last episode continued for two weeks with no improvement. She had no history of a bleeding disease or trauma, nor a specific family history. Physical examination showed no signs of rash or temperature

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change or systemic or local findings of an infection. The knee was tender and knee motion was painful, with 90 degrees of flexion and full extension. The ballottement test was positive. All the laboratory tests including rheumatologic and bleeding parameters were normal. Joint effusion analysis was normal except for its rusty-brown color. Roentgenograms were not suggestive of a pathologic condition. Magnetic resonance imaging showed synovial hypertrophy and grade 2 degeneration in the medial meniscus.

Diagnostic and surgical arthroscopy was performed. Rust-colored synovial hypertrophy was noted in the suprapatellar pouch accompanied by villi and nodules in patches and cystic changes (Fig. 1). The gross appearance of the synovium mimicked that of pigmented villonodular synovitis (PVNS). The joint cartilage, ligaments, and menisci were normal. Biopsy specimens were obtained from different parts of the synovium and a subtotal synovectomy was performed. Knee exercises and full weight bearing were started on the first day of treatment. The pathologic diagnosis was reported as hemosiderotic synovitis (Fig. 2). During three years of follow-up, she had no pain, snapping sensation, or limitation of motion. The patient completely regained her daily activities including sports. There were no recurrent effusions.

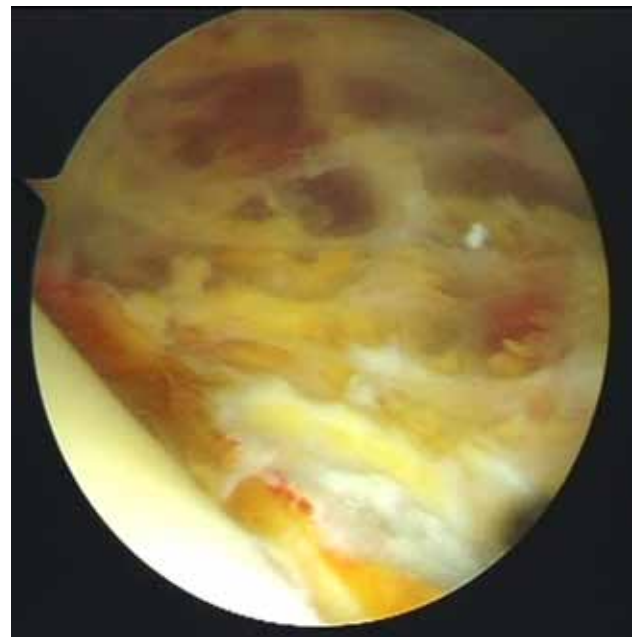

Fig. 1. Arthroscopic appearance of the rusty brown suprapatellar synovia during synovectomy.

Discussion

The most affected joint from bleeding is the knee whatever the etiology is. The gross, light-microscopic, and ultrastructural reactive changes in the synovial membrane secondary to bleeding has been well documented in the literature. A hyperplastic, vascular tissue is formed in the synovial membrane within a few days of bleeding, and with the breakdown of hemoglobin, iron-containing hemosiderin is released and stored. This accumulation causes rusty-brown discoloration of the tissue. More bleeding episodes result in more deposition of hemosiderin and darker colorization. This deposition was proposed to trigger enzymatic activities responsible for the degradation of hemarthrosis.[2]

Hemosiderotic synovitis develops with recurrent hemorrhages in the joint. The most common cause

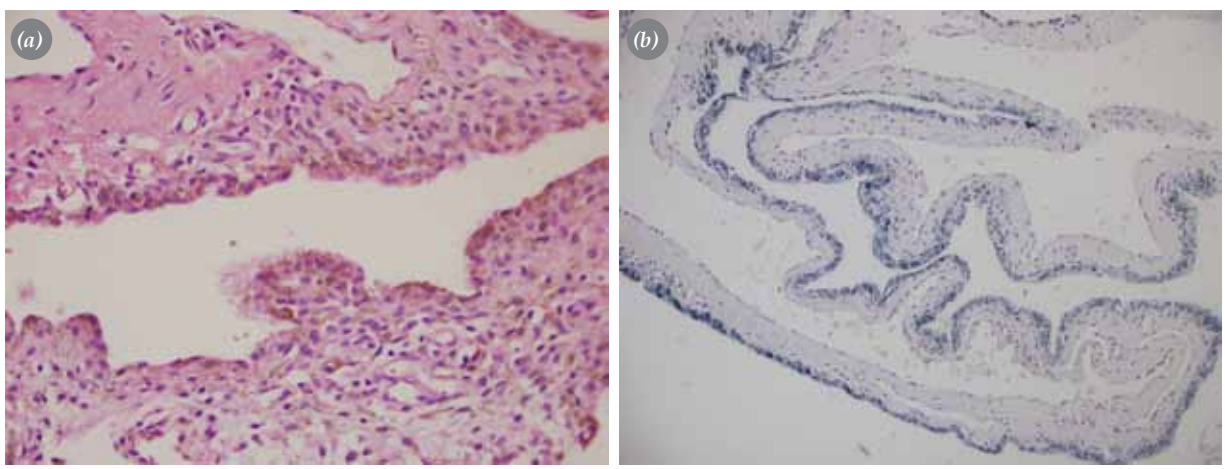

Fig. 2. **(a)** Surface synovial cells and stained hemosiderin granules in macrophages (H-E x 400). **(b)** A specially stained microscopic view demonstrating accumulation of iron localized in the superficial synovia (Prussian blue x 200).

of intra-articular hemorrhage is hereditary clotting factor deficiency diseases such as hemophilia. Hemophilic knees result in early, severe arthritis with rapid, progressive bone and cartilage destruction. However, the triggering cause of degeneration or the effect of the quantity and frequency of bleeding episodes remain unclear. Causes of hemosiderotic synovitis other than hemophilia include the following: oral anticoagulant use, trauma, rheumatoid or psoriatic arthritis, osteoarthrosis, collagen vascular diseases, PVNS, hemochromatosis, scurvy, sickle cell anemia, synovial hemangioma, and myeloproliferative diseases.[2] There was no history of drug use or trauma in our patient and the other causes were excluded both clinically and with laboratory tests.

Patients with hemosiderotic synovitis complain of pain and stiffness caused by effusion of the involved joint. The disease is diagnosed by exclusion of other causes (infections, arthritis, rheumatological diseases, osteonecrosis, cartilage lesions, and synovial pathologies such as synovial chondromatosis and crystal-induced synovitis) by history, physical examination, laboratory and radiodiagnostic tests. If the cause of joint effusion cannot be determined by these methods, diagnostic arthroscopy and pathologic examination of the synovium will provide the definite diagnosis.

Hemosiderotic synovitis, PVNS, and villous lipomatous proliferation of the synovial membrane are rare and inadequately defined synovial proliferative disorders that cause joint effusion, pain and joint motion limitation. The gross appearance of the synovium is almost similar in hemosiderotic synovitis and PVNS. Both appear in rusty-brown color with loss of the normal glistening translucency of the synovial membrane. With recurrent bleeding episodes, the synovium becomes thickened and opaque due to intra- and subsynovial fibrous scarring.[3] Clinical and radiological findings may not yield a definitive diagnosis, which may only possible by histopathologic examination.[4]

In hemosiderotic synovitis, surface synovial cells consist of hemosiderin granules lining on the flat synovial surfaces and the villous processes. In addition, subsynovial connective tissue and small blood vessels contain hemosiderin-laden macrophages. Sheets of mononuclear synovial cells, lipid-laden cells, or multinucleated osteoclast-like giant cells are not seen in hemosiderotic synovitis, whereas in PVNS, mononuclear rounded and epithelioid cells, multinucleated osteoclast-like giant cells, and lipid-rich cells form a lobular and sheet-like composition beneath the surface synovial cells, and most of the hemosiderin is deposited in these cells, in particular the mononuclear cells having large hemosiderin granules.[1]

Abrahams et al.[5] reported concentric joint space narrowing due to hemosiderotic synovitis in five hips of five patients without inflammatory arthritis or hemophilia.[5] Eichhorn and Rosenberg[6] reported a 16-year-old patient who developed hemosiderotic synovitis following intra-articular painful bleeding due to papillary endothelial hyperplasia involving the synovium and reactive synovial changes. Krebs[7] stated that patients with hemosiderotic synovitis might benefit from arthroscopic intervention as patients with synovial chondromatosis, PVNS, inflammatory arthritis, and septic arthritis, and that arthroscopy and improvements in clinical characterization led to a better understanding of the association between synovial abnormalities and degeneration.

Although synovial abnormalities are associated with increased joint degeneration, the pathologic mechanisms remain to be elucidated. It is still unclear at what point the hemorrhage in the joint starts the degenerative process and when the process becomes irreversible. This indicates the importance of early recognition and intervention. Although the development of hemosiderotic synovitis follows a series of distinct stages, its etiology affects the treatment and the prognosis. Arthroscopy is a simple, reliable, less morbid, and minimally invasive technique in both visualization of the pathology and assessment of its effect on the bone and cartilage, during which specimens from the pathologic tissue, the synovium, can be obtained for accurate diagnosis.

In conclusion, when more frequent etiologies are excluded in recurrent joint effusions, hemosiderotic synovitis should be considered among rare causes. Differential diagnosis with PVNS should be made with histopathological examination. Early and appropriate treatment of hemosiderotic synovitis prevents progressive joint degeneration, improving the prognosis.

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