Case Report

Non-Hemophilic Hemosiderotic Synovitis

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Pigmented Villonodular Synovitis (PVNS) is a locally aggressive neoplastic synovial disease characterized by joint effusions, expansion of the synovium, and bony erosions. It typically affects patients between 30 and 40 years old and presents with recurrent atraumatic knee hemarthrosis. Diagnosis involves clinical assessment, MRI imaging, arthrocentesis, and biopsy.

Hemosiderotic synovitis is a rare synovial proliferative disorder caused by recurrent haemorrhages in the joint, commonly affecting the knee. The most frequent cause is hereditary clotting factor deficiency diseases such as hemophilia. However, we report a rare case of non-hemophilic hemosiderotic synovitis of the knee, where the patient had no history of bleeding diathesis. Histopathological examination provided the definitive diagnosis. Early recognition and awareness of this subtype can lead to timely treatment, reducing joint destruction and improving patient outcomes.

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Introduction

The synovium is a specialized mesenchymal tissue that lines the inner surface of synovial joints and plays a vital role in joint lubrication and locomotion. It is susceptible to a variety of pathological conditions, including hemosiderotic synovitis, an uncommon proliferative disorder resulting from repeated intra-articular hemorrhage.[1] Hemosiderotic synovitis is most frequently associated with hereditary coagulation disorders such as hemophilia but may also occur secondary to trauma, anticoagulant therapy, synovial hemangioma, inflammatory arthritis, pigmented villonodular synovitis (PVNS), hemochromatosis, and other less common conditions.[2]

The knee is the most commonly affected joint irrespective of the underlying etiology. Recurrent bleeding leads to hemosiderin deposition within the synovium, causing chronic inflammation, synovial hypertrophy, fibrosis, and progressive cartilage degeneration.[3] Clinical presentation is often nonspecific, consisting of pain, swelling, recurrent effusions, and restricted range of motion. Radiological investigations, particularly magnetic resonance imaging (MRI), may demonstrate diffuse synovial thickening with characteristic low-signal intensity due to hemosiderin deposition; however, these findings overlap considerably with PVNS, making definitive diagnosis difficult.[1,3] Histopathological examination remains the gold standard for differentiating hemosiderotic synovitis from PVNS and other proliferative synovial disorders.[4] We report a rare case of post-traumatic non-hemophilic hemosiderotic synovitis of the knee in a young adult male,

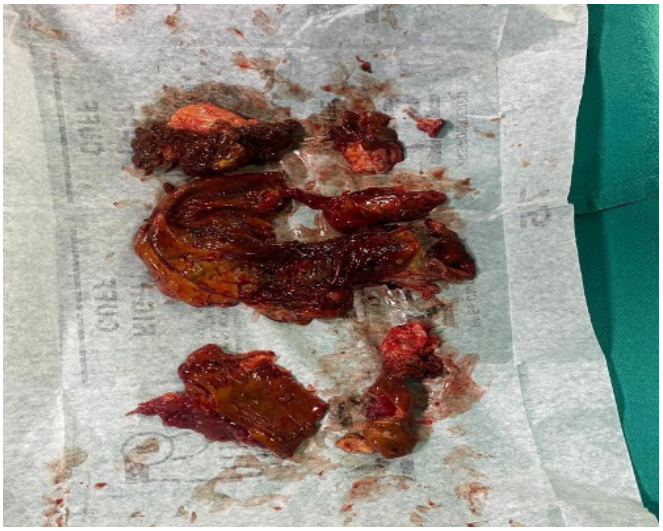

Figure 1: Synovium obtained after synovectomy

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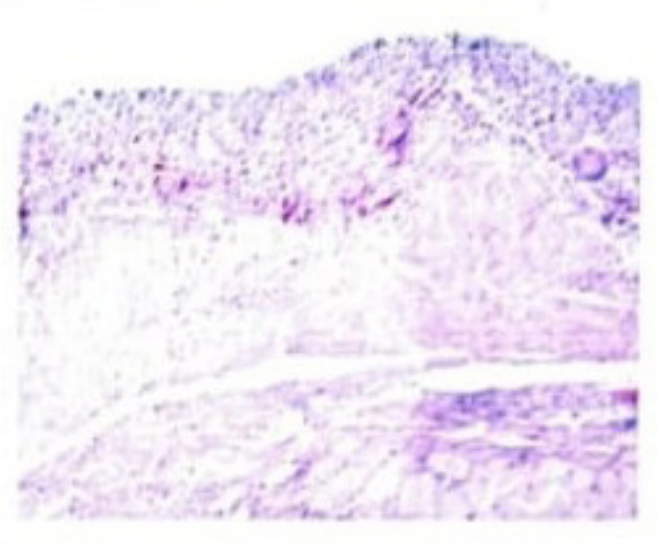

Figure 2: Microscopic section of obtained Synovium

highlighting the diagnostic challenges and importance of histopathological evaluation.

Case Report

A 22-year-old male presented to the U.C.T.H Orthopaedic outpatient department with progressive swelling and tenderness of the left knee joint for two months. The patient had a history of trauma from a self-fall while riding a motorbike. Clinical examination [Clinical pics

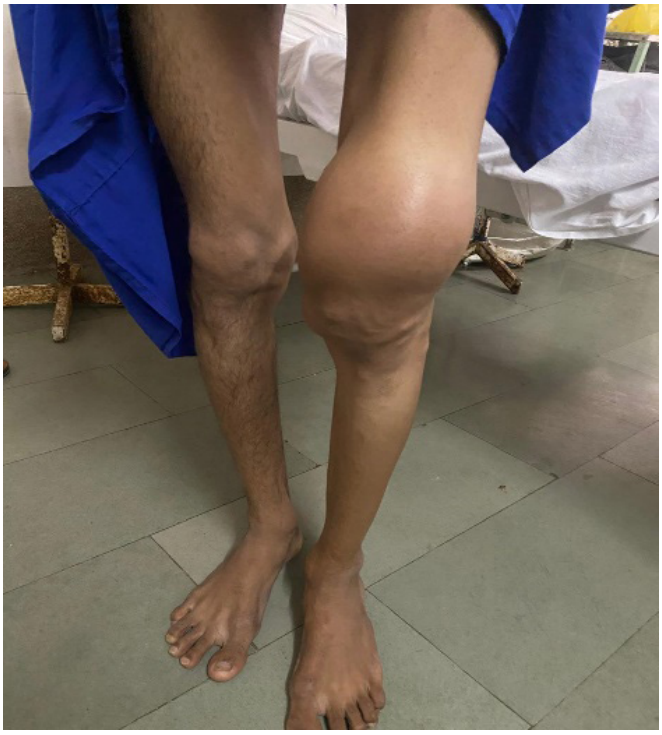

Figure 3: Pre Operative Clinical Pics

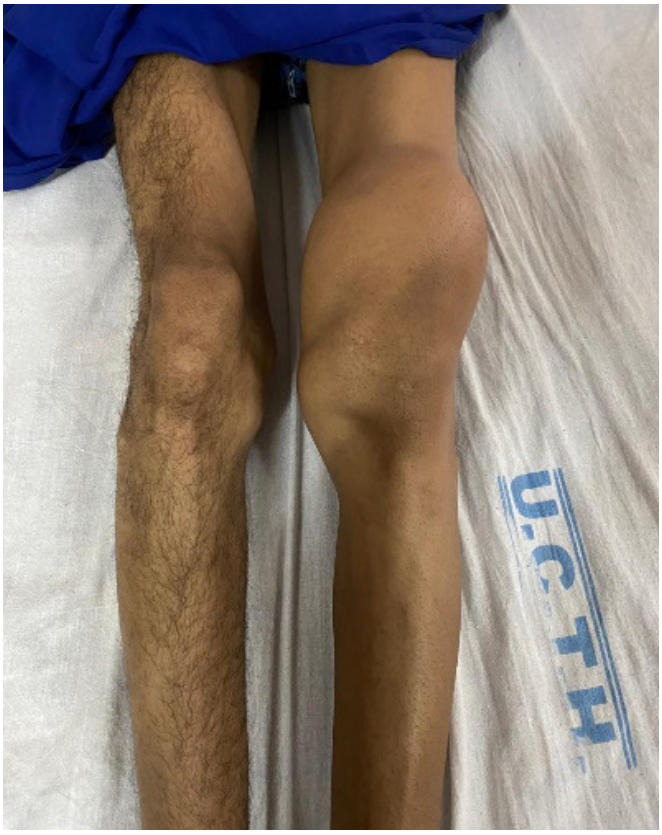

Figure 4: Pre Operative Clinical Pics

(pre-operatively) Figure-3 & 4] revealed knee swelling, tenderness, and restricted painful movement. There was no history of anticoagulant drug intake.

A synovectomy was performed, and the synovium obtained (Figure 1) from the knee was sent for histopathological examination. Microscopically, sections revealed osteoclastic giant cells surrounded by lymphocytes, hemosiderin-laden macrophages, and epithelioid-like cells. Extensive necrosis and haemorrhage were also noted in the underlying tissue(Figure 2).

Discussion

Hemosiderotic synovitis is a chronic proliferative lesion of the synovium resulting from recurrent intra-articular haemorrhage. Although the majority of reported cases occur in patients with hemophilia, non-hemophilic hemosiderotic synovitis remains exceedingly rare.[2,5]

The condition commonly presents with persistent monoarticular pain, swelling, recurrent joint effusions, and progressive restriction of movement. MRI is considered the imaging modality of choice because hemosiderin deposition produces characteristic blooming artifacts on gradient-echo sequences; however, these findings may closely resemble those observed in PVNS,

making histopathological examination indispensable for establishing the diagnosis.[1,3]

Grossly, the synovium demonstrates a brownish or rust-coloured appearance due to extensive hemosiderin accumulation. Microscopically, hemosiderotic synovitis is characterized by abundant hemosiderin-laden macrophages, chronic inflammatory infiltrates, fibrosis, and occasional multinucleated giant cells. In contrast to PVNS, there is less pronounced villous and nodular proliferation with relatively limited mononuclear stromal cell proliferation.[4]

Repeated intra-articular bleeding results in iron deposition within the synovium, stimulating inflammatory pathways and increasing the production of cytokines that inhibit cartilage matrix synthesis while promoting cartilage degradation and secondary osteoarthritis. The exact mechanism remains incompletely understood, but macrophage activation following iron phagocytosis appears to play a central role in the inflammatory cascade.[4]

In the present case, the patient developed persistent knee synovitis following trauma without any evidence of coagulation abnormalities or anticoagulant therapy. Histopathological examination established the diagnosis of non-hemophilic hemosiderotic synovitis. Similar rare cases have previously been reported by France and Gupta involving the shoulder and by Jayalakshmi et al. involving the knee, emphasizing that this condition should be considered in patients presenting with chronic monoarticular synovitis despite the absence of hemophilia.[2,5]

Jain et al. also described the characteristic MRI features of hemosiderotic synovitis, emphasizing the role of advanced imaging in narrowing the differential diagnosis while underscoring the importance of histopathological confirmation.[6] Early synovectomy remains the preferred treatment because it removes diseased synovium, relieves symptoms, decreases recurrent hemorrhage, and may prevent progression to degenerative arthritis.[2,5,6]

Recent evidence by Ichikawa et al. emphasized that recurrent traumatic hemarthrosis can produce hemosiderotic synovitis with MRI features overlapping those of PVNS, making histopathological confirmation essential.[7]

Because PVNS remains the principal differential diagnosis, recent systematic reviews recommend MRI followed by histopathological examination for definitive diagnosis.[8]

Conclusion

Non-hemophilic hemosiderotic synovitis is an extremely uncommon cause of chronic monoarticular synovitis that should be considered in patients with persistent knee swelling following trauma, even in the absence of bleeding disorders. Histopathological examination remains the definitive diagnostic modality for distinguishing this entity from PVNS. Early recognition and prompt synovectomy can prevent progressive cartilage damage, preserve joint function, and improve long-term clinical outcomes.[2,4,5]

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