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Case Report

Hemophilic Arthropathy in a Child: A Case Report

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Received: January 08, 2025; **Published:** February 14, 2025

Abstract

This is a 15-year-old boy with a history of Hemophilia B, presenting with hemarthrosis in both knees. He is consulting for swelling in both knees with tenderness and a deficit in flexion and extension, which is more pronounced in the left knee, this has been evolving for 5 weeks, without any history of trauma.

Keyword: *Hemophilic Arthropathy; Hemarthrosis; IMR*

Introduction and Case Report

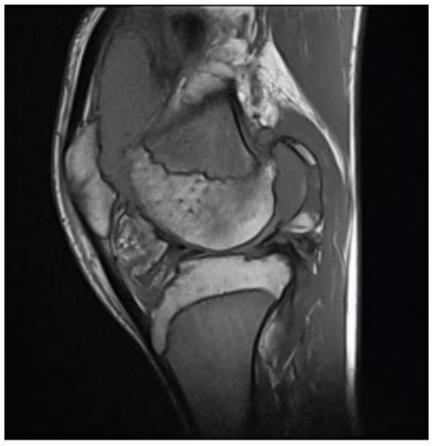
This is a 15-year-old boy with a history of Hemophilia B, presenting with hemarthrosis in both knees. He is consulting for swelling in both knees with tenderness and a deficit in flexion and extension, which is more pronounced in the left knee, this has been evolving for 5 weeks, without any history of trauma.

Our patient, who is being monitored for Hemophilia B, was admitted to the emergency department due to increased swelling in both knees, more pronounced in the left knee, with difficulty walking. His pediatrician requested an MRI of both knees, which showed the following findings.

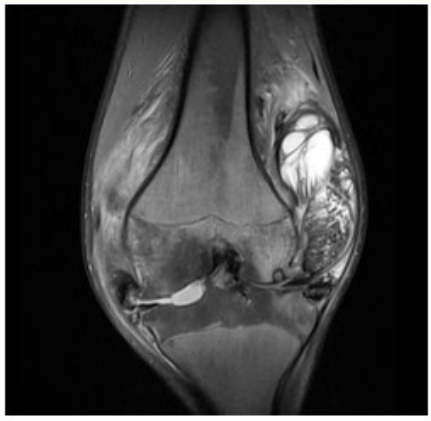
Left knee (Figure 1):

- Joint space narrowing with irregularity of the articular surfaces.
- Irregularity of the femorotibial plateaus with subchondral geodes of the lateral femoral condyle showing fibrous signal (hypointense on T1 and T2) with an area of peri-lesional bone edema, associated with osteophytic spurs on the femoral, tibial condyles, and the patellar base.
- Irregular thickening of the synovium with intermediate signal on T1 and hypointense signal with magnetic susceptibility artifacts on the GRE sequence, slightly enhanced after Gadolinium injection.
- Compartmentalized intra-articular effusion of moderate abundance with liquid signal.
- Absence of differentiation of the ACL.
- Bucket-handle fracture with extrusion of the posterior horn of the lateral meniscus, grade 3.

- Extrusion of the posterior horn of the medial meniscus.
- Cysts in the bilateral parameniscal recesses.

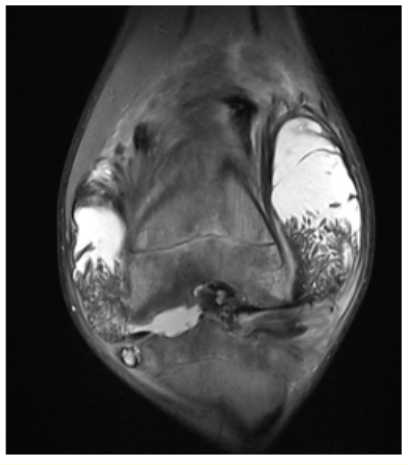


A: A sagittal T1 section of the left knee shows joint space narrowing with irregularity of the articular surfaces, associated with osteophytic spurs on the femoral and tibial condyles, as well as the base of the patella.



B: A coronal proton density image of the left knee shows:

- *Subchondral geodes of the lateral femoral condyle.*
- *Absence of delineation of the ACL.*
- *Bucket-handle tear with extrusion of the posterior horn of the lateral meniscus, grade 3.*
- *Extrusion of the posterior horn of the medial meniscus.*



C: A coronal proton density image of the left knee shows:

- *A cyst in the parameniscal recess (arrow).*
- *Compartmentalized effusion with a fluid signal of moderate abundance within the joint.*

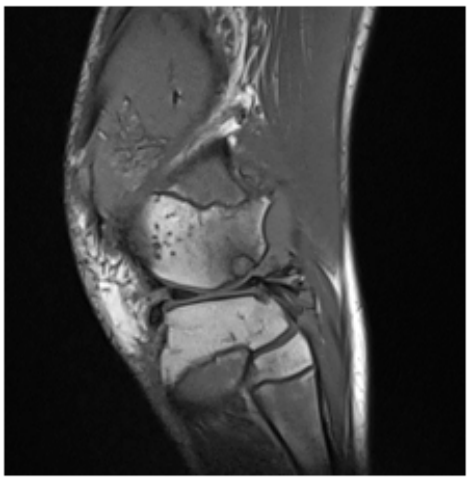
Figure 1: *Left knee.*

Right knee (Figure 2):

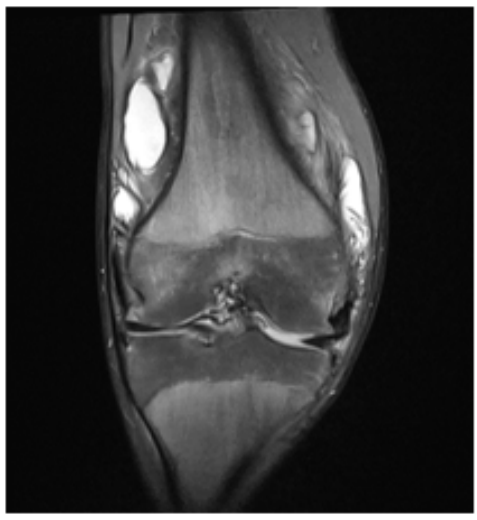
- Joint space narrowing with irregularity of the articular surfaces.
- Irregular thickening of the synovium with intermediate signal on T1 and hypointense signal with magnetic susceptibility artifacts on the GRE sequence, slightly enhanced after Gadolinium injection.
- Irregularity of the femorotibial plateaus with subchondral geodes of the lateral femoral condyle showing fibrinous signal (hypointense on T1 and T2) with an area of peri-lesional bone edema, associated with osteophytic spurs on the femoral and tibial condyles, and the patellar base.
- Intra-articular effusion of moderate abundance with liquid signal.
- Cysts in the bilateral parameniscal recesses.
- Small Baker's cyst measuring 14x15 mm.
- Intact internal and external menisci.

Discussion

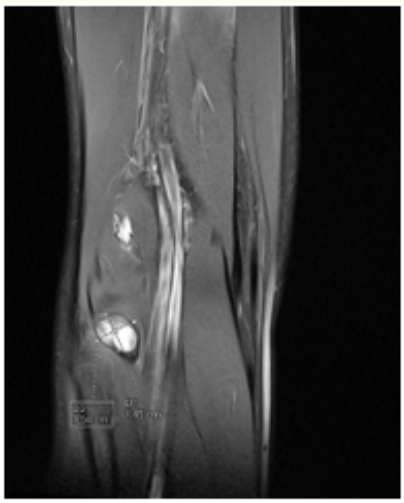
This is a 15-year-old patient being followed for hemophilia B, who presents with pain and swelling in both knees, more pronounced in the left knee.



A: A sagittal T1 section of the right knee shows joint space narrowing with irregularity of the articular surfaces, associated with osteophytic spurs on the femoral and tibial condyles, as well as the base of the patella, and subchondral geodes of the lateral femoral condyle.



B: A coronal proton density image of the right knee shows: Effusion with a fluid signal of moderate abundance within the joint.



C: A coronal proton density image of the right knee shows: A small Baker's cyst measuring 14 x 15 mm.

Figure 2: *Right knee.*

Hemophilia B is a usually severe, hereditary recessive disease linked to the X chromosome, resulting from a deficiency in factor IX. It primarily affects boys rather than girls. It most often occurs before the age of two years, with large joints such as the knee, elbow, ankle, hip, and shoulder being the most commonly affected. Clinically, patients with hemophilia B experience bleeding and excessive hemorrhages following minor trauma or spontaneously without any triggering factor. Hemophilic arthropathy is the leading cause of mortality in children with hemophilia. It presents as pain in the affected joint, along with other manifestations that typically resemble rheumatoid arthritis [1,2].

The most typical manifestation of hemophilia is hemarthrosis. During hemarthrosis, synovial deposits of hemosiderin lead to the release of local inflammatory mediators. Synovial villous hyperplasia, infiltration by inflammatory cells, and increased fibroblastic activity are the substrates of this synovitis [3]. The neoangiogenesis associated with this synovial inflammation contributes to the creation of a hemorrhagic vicious circle due to increased capillary permeability, promoting new episodes of hemarthrosis [4]. Over time, the highly vascularized inflammatory synovial hypertrophy becomes fibrous and acellular.

Independently and simultaneously, there is a direct deleterious effect of the presence of intra-articular blood on the cartilage [4-6]. Reactive oxygen species, formed from the iron in intra-articular red blood cells, contribute to chondrocyte apoptosis [4].

Finally, it should be noted that there is an imbalance in bone remodeling, whether solely due to cartilage destruction or related to independent factors. This is accompanied by changes in the subchondral bone [5], including osteoporosis, formation of subchondral cysts, erosions, and the development of osteophytes.

Standard radiography of the affected joint shows: joint effusion, periarticular osteoporosis, as well as epiphyseal hypertrophy consistent with juvenile rheumatoid arthritis and paralysis. There are several classifications, notably the Arnold-Hilgartner classification on a radiograph centered on the affected knee.

Ultrasound is an easy and quick examination due to its low cost and availability. It allows for the evaluation and monitoring of hemophilic arthropathy, especially in cases of acute hemarthrosis by revealing a fluid effusion of variable echogenicity.

CT scan does not have significant interest and is mainly indicated pre-operatively.

MRI of the affected joint remains the key examination for diagnosing and early detection of joint involvement. It can also show abnormalities that are not detectable on a normal radiograph, including: hemarthrosis, effusion, synovial hypertrophy, hemosiderin deposits, and focal cartilage defects [7]. Radiographs can be normal in patients being followed for chronic hemophilic arthropathy. The abnormalities found on MRI are mainly represented by [7]:

- Hemarthrosis: Variable signal depending on the stage; if it is acute, it appears heterogeneous with areas that are hyperintense on T1 and hypointense on T2. Otherwise, it is hyperintense on both T1 and T2 if it is subacute. It is recommended to perform the MRI away from both stages.
- Synovial hypertrophy: Often intermediate signal on T1 and T2. If active, it appears of the same signal as the fluid.
- Synovial hypervascularization: By injecting contrast agent (gadolinium), it helps to distinguish between active synovitis and fibrous synovitis [8].
- Hemosiderin synovial deposits: Appearing as focal areas or extensive hypointense regions on T1 and T2. Gradient-echo sequences optimize the detection of these deposits due to their magnetic susceptibility. They then appear strongly hypointense. These sequences are particularly useful in cases of very low hemosiderin accumulation.
- Cartilage lesions: Signal abnormalities and erosions.
- Subchondral lesions: Erosions and geodes of variable size presenting a liquid signal (hypointense on T1 and hyperintense on T2), fibrinous (hypointense on T1 and T2), hemorrhagic (hyperintense on T1 and T2), or most often combining these different signals. Subchondral edema indicates advanced chondropathy.

Differential diagnosis

- Hemophilic arthropathy
- Hemophilic arthropathy and hemarthrosis
- Juvenile idiopathic arthritis
- Pigmented villonodular synovitis.

The management of patients with hemophilia B is multidisciplinary, requiring the integration of several specialties, particularly radiology. The etiological treatment of hemophilia is based on replacement therapy with deficient coagulation factor concentrates (via intravenous infusion). If these injections are administered as early as possible, especially before the age of two, it reduces the risk of joint involvement, which is the leading cause of mortality in hemophilic children.

Conclusion

In conclusion, this case of hemophilic arthropathy in a patient with Hemophilia B highlights the importance of early and accurate diagnosis using MRI, which provides detailed insights into joint involvement. The chronic nature of hemarthrosis and its impact on joint health emphasize the need for early intervention with factor replacement therapy to prevent long-term joint damage. A multidisciplinary approach involving radiologists, hematologists, and pediatric specialists is crucial for effective management and improved quality of life in patients with hemophilia.

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Volume 8 Issue 3 March 2025