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Case Report

# Shadows of the bleeding joint: A radiologic exploration ☆

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ABSTRACT

Hemophilia is a genetic disorder that manifests due to a deficiency of clotting factors. One of the most common complications is hemophilic arthropathy which results in bleeding into large joints causing hemosiderin deposition. Recurrent hemarthrosis causes joint destruction resulting in pain and a decreased range of movements. Radiological imaging is essential for staging, prognostication, and treatment protocol. X-rays show the extent of joint destruction and narrowing of joint space. Ultrasonography helps in visualizing tendons and joint effusion. However magnetic resonance imaging is the gold standard for staging hemophilic arthropathy. Management involves both medical and surgical intervention as per the level of joint involvement.



## Background

A lack of coagulation factor VIII causes hemostasis to be compromised in hemophilia A, an X-linked recessive genetic disease [1]. Hemophilia is divided into hemophilia A and hemophilia B based on the type of coagulation factor the patient lacks. The results of factor VIII and factor IX measurements are used to categorize the disease's severity [2]. Hemophilia may manifest in many ways depending on the activity of clotting factors. The long-term result of recurring hemarthrosis is hemophilic arthropathy (HA), a chronic joint disease. About half of hemophiliacs will get severe arthropathy [3].

Hemophilia is characterized by a tendency to bleed, especially in synovial joints like the elbows, knees, and ankles [4]. The fourth most commonly reported source of bleeding is the shoulder joint [5]. A patient's quality of life can be seriously

*Abbreviations:* HA, hemophilic arthropathy; MRI, magnetic resonance imaging; PDFS, proton density fat saturation; SWI, susceptibility weighted image.

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impaired by shoulder pain and decreased joint movements. The vicious cycle of chronic synovial inflammation and thickening, neo-angiogenesis, rebleeding, remodeling, and finally osteochondral surface damage and joint space narrowing can be initiated by a single episode of hemarthrosis, either clinically overt or undiagnosed subclinical bleeding [6]. Patients usually present with symptoms of stiffness, redness of surrounding skin, and restriction of the mobility of the joint involved.

In these situations, radiological diagnostic techniques are crucial. It facilitates the division of the joint degenerative process into different stages of pathology and clinical manifestation [7]. It also helps with orthopedic exploration and radiological scoring systems [8]. The most often utilized methods are magnetic resonance imaging (MRI), X-rays, and ultrasonography. Despite their drawbacks, X-rays are still the most popular imaging technique. Its limited sensitivity for identifying anomalies in cartilage and synovial membranes is its primary flaw. The gold standard for examining joint alterations in hemophiliac patients is magnetic resonance imaging (MRI). In the early phases of hemophilic arthropathy, it is the most effective instrument for analyzing soft tissues and osteochondral changes. However, its use and periodic follow-ups to ascertain the evolution of acute and chronic processes are limited by its high prices, limited accessibility, evaluation time, and challenging applicability in youngsters [9]. The Denver scale and the European scale are 2 parameters used in MRI joint injury assessment.

Compressive bandages, analgesics, and the administration of bleeding factors are typically used as treatments. However, depending on the kind of joint and degree of damage, surgical intervention, such as synovectomy, joint debridement, arthrodesis, and arthroplasty, is frequently necessary when nonsurgical therapy fails in patients with chronic arthropathy progression [10]. In cases of severe end-stage degenerative joint degeneration, joint arthroplasty is recommended.

### *Case presentation*

We report the case of a 40-year-old male who is a known case of hemophilia A and presented with complaints of pain in his right shoulder for 2 months. He was unable to abduct his right shoulder. The patient initially experienced dull aching, persistent pain which was associated with swelling in his right shoulder. His symptoms were progressive. His range of movements in his right shoulder was decreased and was painful. He was initially able to do his daily activities but now due to decreased movements and excessive pain, he was not able to perform his daily activities like shaving or dressing. On admission, his vitals were stable with a pulse of 82 beats per minute, blood pressure of 110/70 mm Hg, and respiratory rate of 16 breaths per minute. He had a history of recurrent bleeding episodes following minor trauma. He had been transfused blood multiple times in the past. He had no history of major accidents, drug allergies, or any infectious disease. The patient's mother is a known case of hemophilia A and has a history of recurrent nasal bleeds and easy bruising. Three years back patient had a similar episode of inflammation and pain in the left knee which was diagnosed as hemarthrosis of the knee joint which caused a pathological fracture in his left femur shaft and was operated upon. An interlocking nail was used to fix the fracture. The patient had no previous surgical interventions. The patient also complained of new-onset pain in his right knee but there was no restriction of mobility. Laboratory investigations are shown in Table 1.

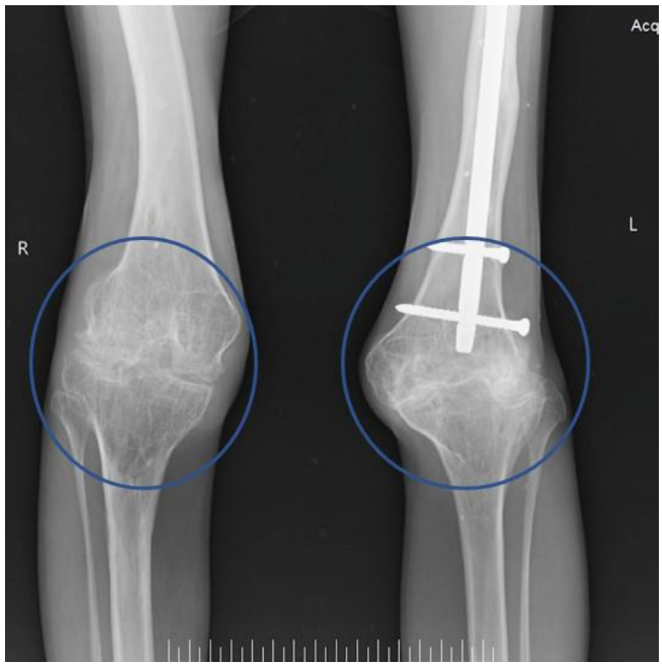


**Fig. 1 – X-ray anteroposterior view left- knee joint shows an interlocking nail in situ. X-ray anteroposterior bilateral knee shows changes of osteoarthritis (left > right) in the form of marginal osteophytes, loss of joint space, and subchondral sclerosis. There was a widening of the inter-condylar notch and flattened condylar surface (blue circles).**

The patient underwent radiological evaluation for the condition of his various joints. X-ray anteroposterior view of his left knee joint showed an interlocking nail in situ. X-ray anteroposterior bilateral knee showed changes of osteoarthritis (left>right) in the form of marginal osteophytes, loss of joint space, and subchondral sclerosis. There was a widening of the inter-condylar notch and flattened condylar surface (Fig. 1).

X-ray anteroposterior of the right shoulder was suggestive of erosive destruction of the right humeral head with loss of contour, subchondral cyst, and erosions of the visualized part including greater and lesser tuberosity and glenoid margins. X-ray of the left shoulder was unremarkable (Figs. 2 and 3).

MRI of the right shoulder was done which showed loss of articular cartilage appearing hypointense on T1/T2/PDFS (Proton Density Fat Saturation) and blooming on Susceptibility Weighted Image (SWI) sequence suggestive of chronic hemarthrosis. The tendon of the supraspinatus, infraspinatus, teres minor, and long head of biceps at the insertion site appeared hypointense on the T1/T2/PDFS sequence. There was erosive destruction of the right humeral head with loss of con-

**Table 1 – Original table depicting the laboratory investigations.**

| Parameters | Values | Normal range |
|---|---|---|
| Hemoglobin | 9.8 gm% | 13-17 gm% |
| Total leukocyte count | 6500 cells/cu mm | 4,000-10,000 cells/cu mm |
| Total platelet count | 1.76 lakh/cu mm | 1.5-4.1 lakh/cu mm |
| Mean corpuscular volume | 71.1 fL | 83-101 fL |
| Urea | 38 mg/dl | 19-43 mg/dl |
| Creatinine | 0.9 mg/dl | 0.2-1.3 mg/dl |
| Serum Sodium | 137 mm/L | 135-145 mm/L |
| Serum Potassium | 3.9 mm/L | 3.5-5.1 mm/L |
| INR | 1.5 | 1-1.3 |
| Alkaline phosphatase | 62 U/L | 38-126 U/L |
| Alanine aminotransferases | 33 U/L | <50 U/L |
| Aspartate aminotransferase | 44 U/L | 17-59 U/L |
| Albumin | 3.9 g/dL | 3.5-5 g/dL |
| Total bilirubin | 1.2 mg/dl | 0.2-1.3 mg/dl |
| Magnesium | 1.9 mg/dl | 1.6-2.2 mg/dl |
| Phosphorous | 2.1 mg/dl | 2.5-4.5 mg/dL |
| Calcium | 7.7mg/dl | 8.4-10.2 mg/dl |

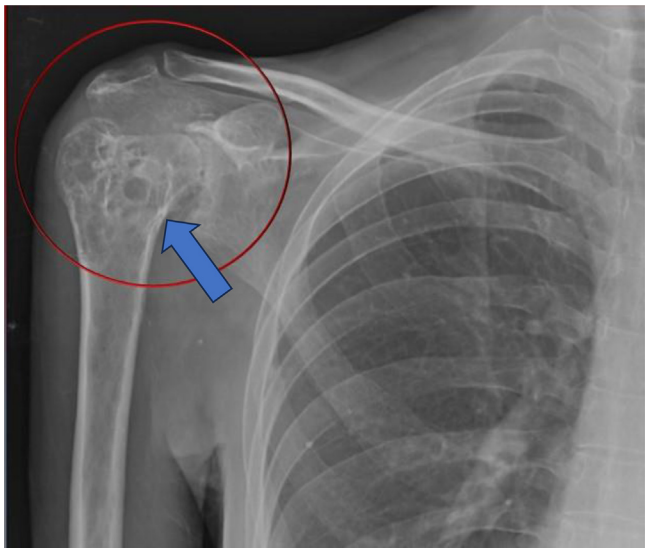

**Fig. 2 – X-ray of right shoulder joint anteroposterior view shows erosive destruction of right humeral head with loss of contour, subchondral cysts, and erosions of visualized part including greater and lesser tuberosity and glenoid margins (blue arrow).**

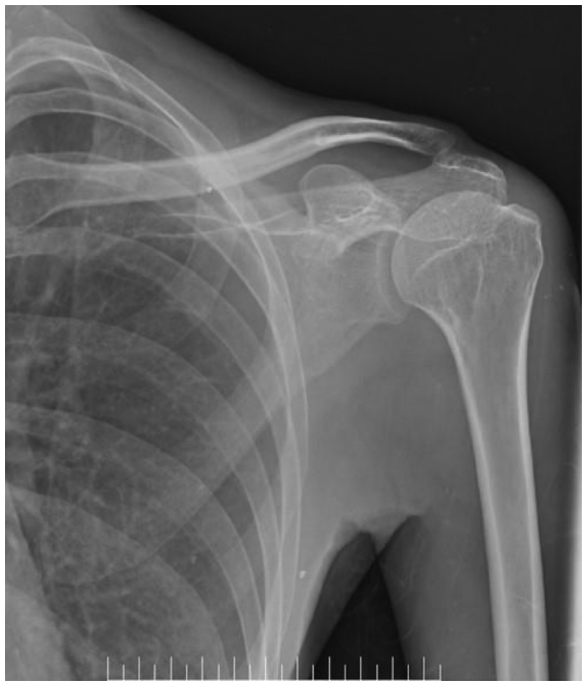

**Fig. 3 – X-ray left shoulder joint anteroposterior view appears normal.**

tour and erosions including greater and lesser tuberosity, and glenoid margins. Joint effusion was also evident (Figs. 4-7).

The patient was managed conservatively and given analgesics for pain and intra-articular injections of corticosteroids. The patient was given injections of clotting factors and RICE regime (rest, ice, compression and elevation). The patient's symptoms gradually improved over a period of 2 weeks and was discharged. Upon follow up patient was symptomatically better and was advised for regular injections of clotting factors followed by physical exercises to further improve the range of movements.

## Discussion

A clotting factor deficiency or absence results in hemophilia, an inherited bleeding condition [11]. Hemophilia is typically inherited by a mutated FVIII or FIX gene on an X chromosome. Nevertheless, both genes are susceptible to novel mutations, and spontaneous genetic changes account for around 30% of all instances. More than half of newly diagnosed people with severe hemophilia have no family history of the condition,

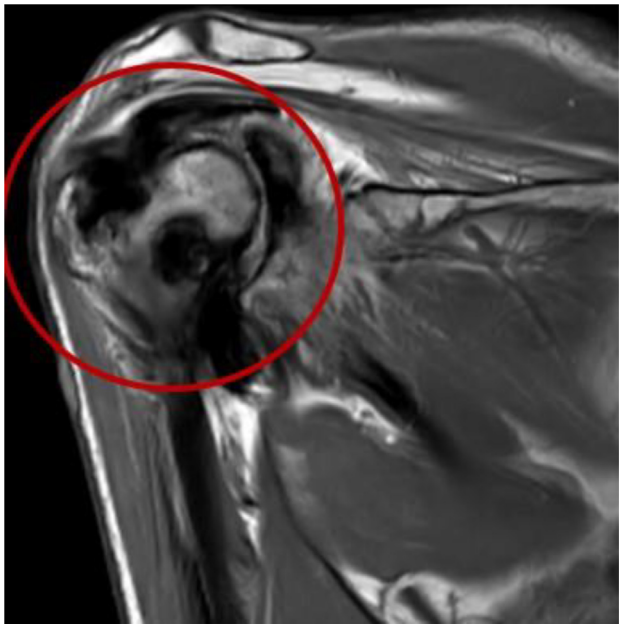

**Fig. 4 – MRI of right shoulder joint T1 sequence shows erosive destruction of the right humeral head with loss of contour, articular cartilage, and erosions of the visualized part including greater and lesser tuberosity and glenoid margins (red circle).**

according to prospective research [12]. It can be categorized as hemophilia A (factor VIII) or hemophilia B (factor IX) based on the missing clotting factor. Based on the level of factor clotting activity, hemophilia is categorized as mild (5%-30%), moderate (1%-5%), and severe (<1%). While moderate hemophiliacs only experience bleeding episodes following trauma, surgery, or tooth extraction, severe hemophiliacs may experience regular spontaneous bleeding episodes [13].

Studies examining hemophilic arthropathy of the shoulder are quite rare. In their series of 41 adult hemophiliac patients, MacDonald et al. found that 15 (37%) of them experienced shoulder complaints, with rotator cuff tears being the most prevalent anomaly (found in half of the symptomatic shoulders) [14]. In a retrospective study of 822 hemophiliac patients' medical records, Cahlon et al. [15] discovered that 93 patients (11%) reported shoulder symptoms. Our patient presented with complaints of pain and decreased range of movements in his right shoulder. He had a previous history of pathological fracture in his left femur with hemophilic arthropathy of the bilateral knee.

Hemophilia's hallmark clinical feature is bleeding into soft tissues and joints. Secondary osteoarthritis results from cartilage sloughing caused by synovitis and ferritin deposition in the hyaline cartilage. Patients experience persistent abnormalities in one or more joints that resemble rheumatoid arthritis both clinically and roentgenographically [16]. Intraarticular blood resorption results in reactive synovitis, which in turn leads to recurrent hyperplastic synovitis. In addition to impairing the development plate's blood supply, chronic synovitis causes cartilage degradation. The expansion of the epiphyses comes next. Fibrosis-related joint contractures and related joint effusions are also present [3].

Since they are inexpensive and offer a good picture of the joint, X-rays are typically the first imaging modality used to evaluate hemophilic arthropathy. X-rays are particularly useful for detecting moderate-to-severe arthropathic changes, such as subchondral cysts, uneven subchondral bone, and narrowing of the joint space brought on by cartilage abnormalities [17]. In addition to physical examination and X-rays, ultrasound can be used to assess HA. Compared to MRI, ultrasound is more accessible, less expensive, and comparatively faster. When evaluating synovial enlargement, ultrasound is a reliable method [18]. The most sensitive technique for assessing alterations in soft tissue and cartilage is magnetic resonance imaging.

A magnetic field is used in MRI imaging acquisition. Since hemosiderin deposits cause the magnetic field to become nonuniform and the MRI signal to be lost, they can be identified [19]. In our case, an MRI of the right shoulder showed hemophilic arthropathy with hemosiderin deposition and erosive destruction of the humeral head with loss of contour, articular cartilage, and joint effusion as evident from blooming on SWI and hypointensity on T1/T2/ Proton density

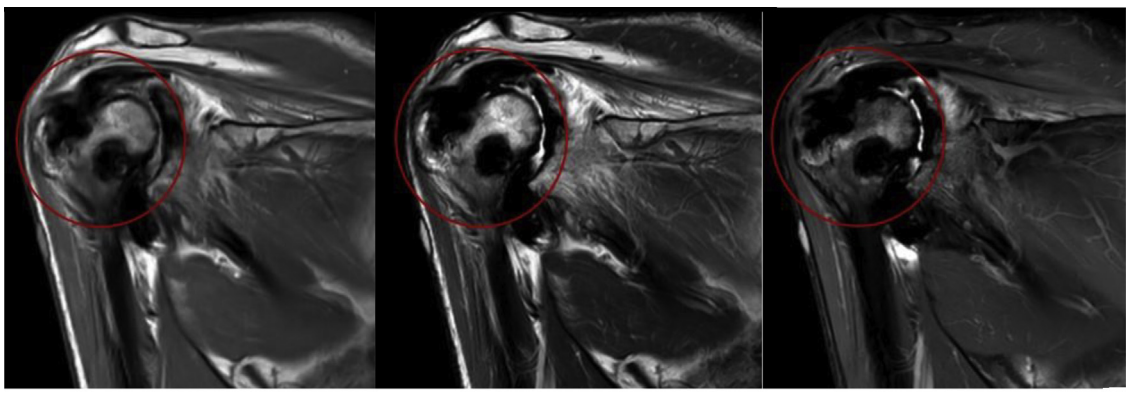


**Fig. 5 – MRI right shoulder Coronal section (A) T1, (B) T2, (C) PDFS shows: hypointensity on T1/T2/PDFS in the tendon of supraspinatus, subscapularis and long head of biceps (red circles).**

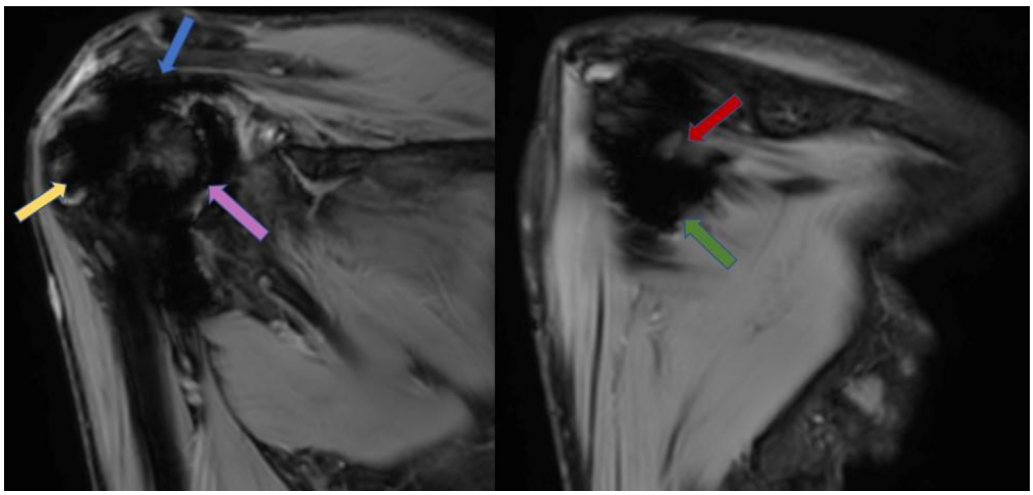

**Fig. 6 – MRI right shoulder joint, Coronal sections SWI sequence shows: the tendon of supraspinatus (blue arrow), subscapularis (pink arrow), long head of biceps (yellow arrow), infraspinatus (red arrow) and teres minor (green arrow) at the insertion site showing blooming on SWI suggestive of hemosiderin deposition.**

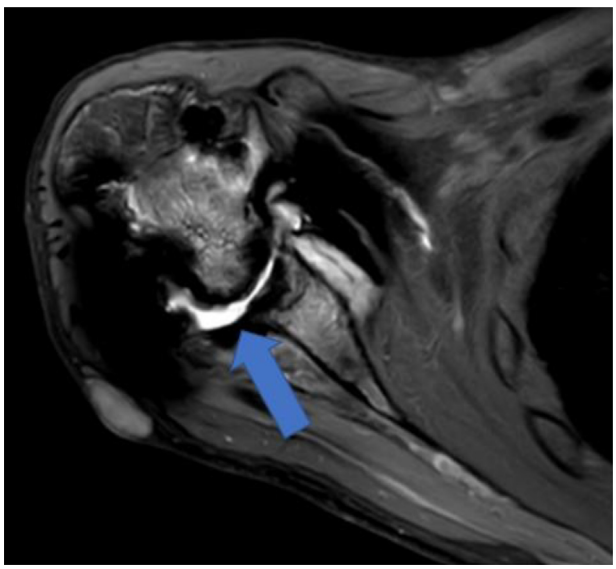

**Fig. 7 – MRI right shoulder axial section T2 sequence shows joint effusion (blue arrow).**

fat saturation sequence. X-ray of the bilateral knee joint suggested osteoarthritic changes with marginal osteophytes, decreased joint space, subchondral sclerosis, and flattened condylar surface.

Management of early hemophilic arthropathy consists of compressive dressings, analgesics, and physical therapy to prevent the development of contracture. Operative intervention like synovectomy for recurrent hemarthroses and arthroplasties for end-stage arthropathy is required. The prognosis of hemophilic arthropathy depends upon the degree of factor deficiency, presence of clotting factor inhibitors, age of the patient, and degree of joint destruction.

It is typically not required to distinguish hemophilia from other juvenile arthropathies on imaging due to the traditional history of hemophilia, which involves numerous bleeding episodes. Since some of the alterations observed are also secondary to synovial proliferation, juvenile idiopathic arthropathy shares several characteristics with HA on plain radiography. Haemosiderin deposition is not a noticeable characteristic on MRI in juvenile arthropathies, however thicker and inflammatory synovium is evident [20]. The knee joint is the most prevalent location for pigmented villonodular synovitis, a primary proliferative condition of the synovium. Often, plain radiography shows only soft-tissue edema or is normal. Unlike HA, bone density and joint space are frequently maintained until the end of the illness. The synovium appears mass-like and lobulate on MRI.

## Conclusion

Hemophilic arthropathy is commonly seen in patients suffering from hemophilia. Typically large joints like the knee, elbow, and shoulders are involved. Recurrent bleeding into the joints leads to hemarthrosis. In our case patient had involvement of bilateral knee joints previously. A multipronged approach is necessary for the prevention of hemarthrosis and the development of arthropathy. Radiological modalities play an important role in staging and diagnosing arthropathy and thus help in shaping the treatment protocol. Gene therapy and other novel modalities are being tried and various researches are going on to reduce the complications of hemophilia. Genetic testing and premarital counseling in carriers of hemophilic genes is a primordial prevention that is now gaining attention.

## Ethics approval

In compliance with ethical and legal regulations.

## Patient consent

Written informed consent was obtained from the patient for the publication of this case report.

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