CASE REPORT

# Haemophilic Arthropathy

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**Abstract**

Bleeding into joints and soft tissues are the hallmark of haemophilia. The severity of manifestations co-relates with deficiency of the factor. The resorption of intra-articular blood induces reactive synovitis and causes cartilage damage, finally causing complete destruction of it. We hereby report a similar case and highlight various rheumatic manifestations of haemophilia and its management.

**Key Words**

Haemophilia, Arthropathy, Factor VIII.

## Introduction

Haemophilia is a heredity disorder in which bleeding is due to deficiency of coagulation factor VIII. Haemophilia A is the second most common inherited coagulation disorder. Estimation of its incidence ranges from 1 in 20,000 to as high as 1 in 10,000 people. It is a classic example of x-linked recessive trait (1). The frequency and severity generally are related to the blood level of factor VIII C (2). Those with less than 1% of normal activity develop severe disease, levels between 2% and 5% of normal are associated with moderate disease and patient with 6-50% of activity develop mild disease (3). In haemophilia, arthropathy is secondary to recurrent halmarthrosis and chronic synovitis (4). The chronic arthropathy affects only a few joints, most notably the knees and elbows and involvement of other joints like shoulders, hips and ankles is less common(5). The ankle joint causes the maximum limitation of motion, even though synovitis caused by it is the least common - 2.2%(6).While advent of replacement therapy has dramatically changed the course of treatment and prognosis for patients with haemophilia, authors argue that economic burden of treating these patients is still very high (7). We also present a case of hemophilic arthropathy with involvement of various joints causing deformities and limitation of movement.

## Case Report

A 32 year male was admitted to the Government Medical College Hospital, Jammu in the month of February, 2004 with history of passage of black tarry stools 3-4 times per day, started five days prior to admission with no history of NSAID intake. Patient had history of pain and swelling of the knee, ankle, and elbow joints on both sides off and on for the last 7 years. Each episode used to last for one to two weeks and patient had one or two episodes per year. He was diagnosed as a case of haemophilia at the age of 2 years and was admitted with cutaneous haemorrhages at Paediatric Hospital, Jammu. Patient was admitted thrice, once at the age of 18 years with haemetemesis, 20 years with syncopal attack and then at 22 years with abdominal pain and on all the occasions he had received blood transfusion. During hospital stay, patient received 4 units of fresh frozen plasma (FFP) and 1000 units of Factor VIII which was given daily for 2 days. Patient had shown improvement and started passing normal coloured stools.

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Patient was of average built with visible knee joint and elbow joint deformity. Rest of the general physical examination and other systems were found to be clinically normal. On local examination, patient was found to have cubitus varus deformity of the elbow joints and fixed flexion deformity with restricted range of movement of 45º-100º. Patient also had genu varum deformity with intercondylar increased distance. Routine laboratory tests including total and differential leucocyte counts, renal function tests, liver function test and blood sugar were within normal limits. X-ray knee joints AP view showed intercondylar notch widening with a rectangular patella with erosion of the bony margins of femoral condyles. Lateral aspect of both knees reveal markedly reduced joint space and multiple sized subchondral cyst. X-ray elbow joints AP view showed enlarged, deformed radial head with juxta-articular cyst in the olecranon process and sclerosis, flattening and deformity of articulating bones. X-ray ankle joint showed soft tissue swelling with marginal osseous erosions and irregular articulating surfaces which appear widened with decreased joint space.

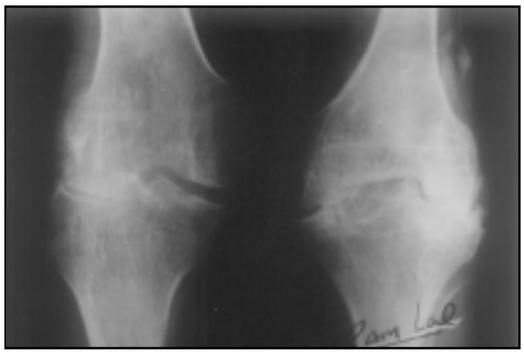

**Fig 1. X-ray knee AP view showing intercondylar notch widening with erosions of bony margins.**

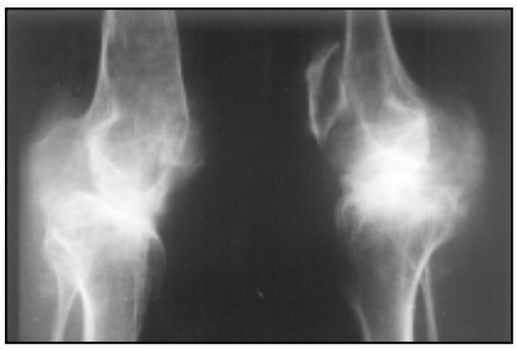

**Fig 2. X-ray knee lateral view showing markedly decreased joint space and presence of subchondral cysts.**

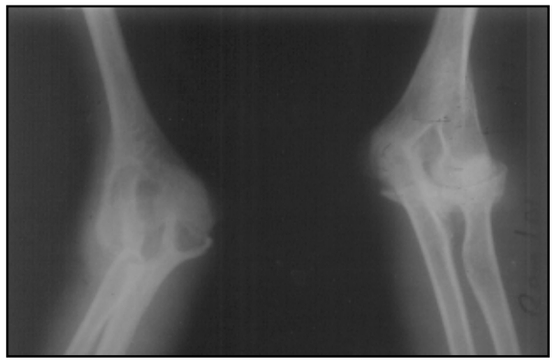

**Fig 3. X-ray elbow showing enlarged and deformed radial head with juxta - articular cyst in the olecranon process.**

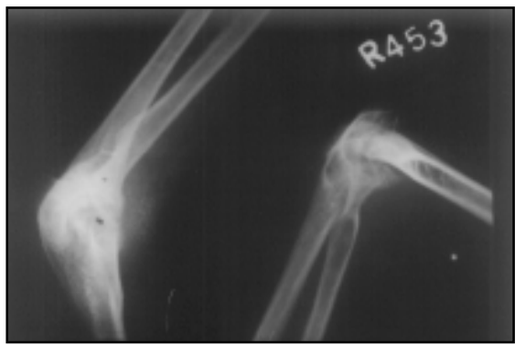


**Fig 4. X-ray elbow lateral view showing decreased and flatened joint space with radiodense effusions.**

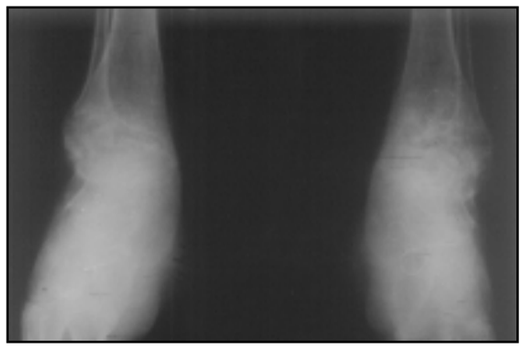

**Fig 5. X-ray ankle joint AP view showing soft tissue swelling with multiple central and marginal erosions and decreased joint space.**

## Discussion

The most common clinical manifestation of haemophilia is arthropathy secondary to recurrent hemarthrosis and

chronic synovitis with involvement of knee, elbow, ankle, hip and shoulder joints commonly (4). Intraarticular bleed occur spontaneously secondary to trauma in Haemophilia A/B and resorption of blood induces reactive haemophilic synovitis with hyperplastic changes which recur again and again leading to chronic synovitis (8, 9). The location of growth plate in vicinity of joint and effect of chronic synovitis and impaired blood supply on the plate induces deformities in joint with more younger patients more severely affected (10). The knee joints are most commonly affected almost invariably with bilateral involvement, intercondylar notch becomes wide, deep and patella develops a rectangular shape. The epiphysis are enlarged, become abnormal in shape and fuse prematurely (11). This was appreciated in our case also.

The obliteration of capitello-lateral epicondyle groove with wounding off of the lateral aspect of the distal humerus, lipping of medial trochlear surface with enlargement of radial head and juxta articular cystic lesions near humeral epiphysis (12) are seen in X-ray elbow joint, which was quite similiar to the findings in our case. X-ray ankle joint shows osseous erosions, joint space narrowing and bony ankylosis of multiple joints which was seen in our patient also. The standard care for patients with a target is to give factor VIII concentrate prophylaxis and to start 2 week course of prednisolone 1 mg/kg per day for first week and 0.5 mg/kg per day for the second week which decreases bulk of the synovium reducing risk for impingement (13). NSAIDS, local ice application, soft heel and semi-rigid bandages have the same role as above (8, 14).

Physiotherapy reduces swelling and pain, promotes full range of motion, protects joint from further injury and bleed and resolves muscle strength (15).

Newer modalities including arthroscopic synovectomy using five to six 0.5 mm incisions reduces hospital stay (16).

Chemical synovectomy has also been tried in which a substance is injected into joint which will affect only synovium without harming cartilage, bone or ligaments e.g. Rifanpin intra-articular injections are given thrice a week. Other materials used are osmic acid, thioteps, D-penicillamine and oxytetracycline (17). Synovial ablation may be accomplished by intra-articular injection of a radio active agent which concentrates in inflamed tissue, for example Gold (198 Au), Yttrium (90 g) Phosphorus (32 P) and dysprosyum (160D) (18). Other surgical procedures tried in cases of complications are joint debridement, fusion and joint arthroplasty (4).

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