SPECTRUM OF RADIOGRAPHIC FINDINGS IN HAEMOPHILIC ARTHROPATHY: A PICTORIAL ESSAY

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ABSTRACT

BACKGROUND

Haemophilia is a hereditary bleeding disorder characterised by recurrent hemarthrosis leading to arthropathy. Plain radiography has an important role in its evaluation. This pictorial essay discusses and illustrates the various musculoskeletal imaging manifestations in haemophilia using plain radiography.

KEYWORDS

Haemophilia, Haemophilic Arthropathy, Hemarthrosis, Pseudotumour.

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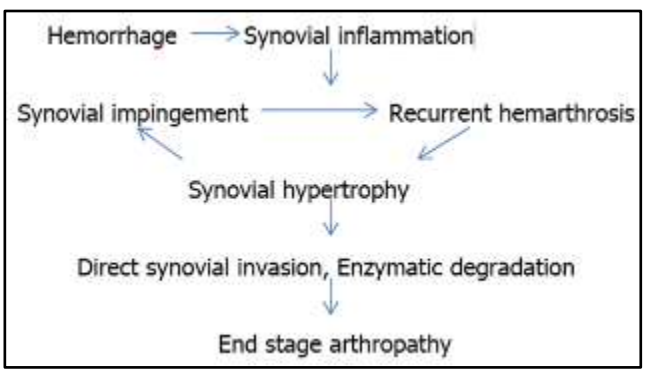
BACKGROUND

Haemophilia is an X-linked recessive bleeding disorder resulting from the deficiency of clotting factors. It occurs predominantly in males and is transmitted through females. The two common forms, haemophilia A and B are due to the deficiency of factors VIII and IX respectively. Homophilic patients are prone for developing bleeding in the musculoskeletal system and arthropathy results from recurrent bleeding into the joints (hemarthrosis). Knee, elbow and ankle are the most frequently involved joints.[1,2] This pictorial assay is aimed to demonstrate the various musculoskeletal imaging manifestations from radiographs of 150 haemophilic patients.

Pathophysiology

Haemophilic arthropathy evolves through three stages: acute hemarthrosis, chronic synovitis and degenerative arthritis. Recurrent bleeding within the joint produces hemosiderin and cause inflammation of synovium. Synovium then becomes hypertrophic and highly vascular, gets impinged between the articular surfaces further increasing the risk of hemarthrosis. Thus a vicious cycle of inflammation and bleeding results in damaging the articular cartilage, which ultimately leads to degenerative arthritis.[3]



Radiography

Various imaging modalities like X-ray, USG, CT, MRI are available for the evaluation of haemophilia. Plain radiography remains the most frequently used radiological investigation and it plays key role in the diagnosis, structural assessment and follow up. But early soft-tissue changes are not clearly delineated on plain radiographs. By the time radiographic changes appear, arthropathy is usually well advanced and irreversible.

Most radiographic findings in haemophilic arthropathy appear in sequences and are essentially the same in all joints. Recurrent hemarthrosis causing effusion and distension of the joint capsule followed by synovial hypertrophy with pannus formation and periarticular oedema usually appear as a nonspecific soft-tissue swelling (Figure 1). Hemosiderin deposits following hemarthrosis may appear as dense areas within the soft tissue swelling. Local inflammatory changes with hyperaemia cause accelerated bone maturation, overgrowth (ballooning) of the epiphyses, gracile diaphysis and periarticular osteopenia in children (Figure 2). Osteopenia may be worsened by disuse. Premature physeal fusion can result in limb shortening. Irregularity of the subchondral bone and articular cartilage destruction lead to joint space narrowing. Marginal erosions, subchondral cyst formation, bone destruction and fractures are also seen along with this (Figure 3-7). Ultimately severe joint deformity and ankylosis develops (Figure 8-14).

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Some changes are peculiar to certain joints. Typical findings in knee include squaring of patella (Jordan's sign) or thinned and elongated patella, overgrowth of femoral condyles, pannus causing erosion and widening of intercondylar notch and widening of the groove between tibial spines (Figure 15-18). Characteristic findings in elbow are enlargement of radial head with resulting increased size of radial notch of ulna and enlargement of distal humerus with widened olecranon fossa (Figure 19, 20). Talar tilt, narrowed lateral and widened medial distal tibial epiphysis causing valgus deformity are seen in ankle. Talus may be deformed (Figure 21,22).

Hip and shoulder joints are less affected by arthropathic changes. In hip joint, the characteristic finding is avascular necrosis which can lead to femoral head resorption and eventually thinning of medial wall of acetabulum which may lead to acetabular protrusion (Figure 23). In shoulder, humeral head may enlarge initially, and later atrophy may develop (Figure 24).[2,4,5,6]

Arnold-Hilgartner and Pettersson scoring scales are the main radiographic classification systems in evaluating haemophilic arthropathy. Arnold-Hilgartner system is a progressive, simple and easy to use scale with the worst imaging finding representing the higher stage of arthropathy (Table 1). Pettersson scoring system is an additive, meticulous scale and the highest score represents a totally destroyed joint. It does not include evaluation of soft tissue changes, which is difficult to be assessed on plain radiography[7,8] (Table 2).

Other musculoskeletal manifestation is the development of pseudotumours which are non-neoplastic masses resulting from repeated bleeding in soft tissue, bone or subperiosteal areas. Intraosseous pseudotumour appears as a single or multiloculated, expanded, geographic lytic lesion with well marginated sclerotic rim. Endosteal scalloping, cortical thinning and adjacent new bone formation may also be seen. Soft tissue or sub-periosteal pseudotumour appears as a soft tissue density causing extrinsic scalloping on bone with sharp margins and periosteal reaction[2] (Figure 25-27).

Figure 1

Figure 1 shows AP radiograph of both knees of a 12yrs. old boy with haemophilia B shows soft tissue swelling and minimal irregularity in the subchondral surface. Joint spaces appear preserved.

Figure 2

Figure 2 shows AP radiograph of both knees of a 6yrs. old boy with haemophilia B shows enlarged femoral and tibial epiphyses on left side with minimal joint space narrowing, subchondral irregularity and periarticular osteopenia. Soft tissue swelling noted bilaterally (more on left).

Figure 3

Figure 3 shows minimal joint space narrowing, irregular subchondral surface and soft tissue swelling seen in AP radiograph of left knee in a 12yrs. old boy with haemophilia B.

Figure 4

Figure 4 shows AP radiograph of left knee in a 48 yrs. old haemophilic patient shows joint space narrowing, irregular subchondral surface, marginal erosions, osteophytes and periarticular osteopenia.

Figure 5

Figure 5 shows AP and lateral radiographs of right elbow in a 57 yrs. old with haemophilia shows joint space narrowing, subchondral cyst and periarticular osteopenia.

Figure 6

Figure 6 shows joint space narrowing, subchondral cyst formation and periarticular osteopenia seen in the AP and lateral radiographs of right elbow in a 21 yrs. old haemophilic patient.

Figure 7

Figure 7 shows AP radiograph of left shoulder in a 24 yrs. old with haemophilia B shows subchondral cyst formation near greater tubercle of humerus with adjacent osteopenic changes. Joint spaces appear preserved.

Figure 8

Figure 8 shows AP radiograph of right knee of a 52 yrs. old with haemophilia A. Significant joint space narrowing with ankylosis and joint deformity noted.

Figure 9

Figure 9 shows joint space narrowing with marginal erosions and ankyloses (more towards the lateral compartment) noted in the AP radiograph of right knee of a 48 yrs. old with haemophilic patient.

Figure 10

Figure 10 shows AP radiograph of both knees of a 51 yrs. old with haemophilia A shows significant joint space narrowing with ankylosis.

Figure 11

Figure 11 shows AP radiograph of right knee of a 32 yrs. old with haemophilia B. Joint space narrowing, medial slipping of femur, irregular subchondral surface with few subchondral cysts, osteophytes and soft tissue swelling noted.

Figure 12

Figure 12 shows AP radiograph of left knee of a 29 yrs. old with haemophilia B. Disorganised and subluxed joint with widened medial and narrowed lateral compartment joint spaces. Irregular subchondral surface and periarticular osteoporosis seen.

Figure 13

Figure 13 shows AP radiograph of right knee of a 44 yrs. old with haemophilia A. Depression of medial tibial plateau noted with associated osteoarthritic joint changes and varus deformity. Medial femoral condylar margins also appears irregular.

Figure 14

Figure 14 shows AP and lateral radiographs of left elbow in a 33 yrs. old haemophilic patient. Deformed and dislocated elbow with displacement of ulna upwards and posteriorly.

Figure 15

Figure 15 shows AP radiograph of left knee of a 17yrs. old boy with haemophilia B shows widened intercondylar notch, enlarged femoral and tibial epiphyses, joint space narrowing, subchondral irregularity, periarticular osteopenia and soft tissue swelling.

Figure 16

Figure 16 shows widening of intercondylar notch, widening of the groove between tibial spines, joint space narrowing, and periarticular osteopenia seen in the AP radiograph of left knee in a 32 yrs. old with haemophilia.

Figure 17

Figure 17 shows Lateral radiograph of a haemophilic teenager's knee shows squaring of patella.

Figure 18

Figure 18 shows lateral radiograph of knee in a 18 yrs. old with haemophilia shows thinning and elongation of patella. Patellofemoral and tibiofemoral joint space narrowing also noted.

Figure 19

Figure 19 shows AP and lateral radiographs of left elbow in a 32 yrs. old haemophilic patient shows enlargement of radial head, prominent radial notch of ulna, joint space narrowing and periarticular osteopenia.

Figure 20

Figure 20 shows enlarged radial head, prominent radial notch of ulna, joint space narrowing, early subchondral cyst formation and periarticular osteopenia seen in the AP and lateral radiographs of right elbow in a 32 yrs. old haemophilic patient.

Figure 21

Figure 21 shows talar tilt with narrowing of the lateral part and widening of the medial part of the tibial epiphysis causing minimal valgus deformity noted in the AP radiograph of right ankle in a 13 yrs. old with haemophilia B. Associated osteopenic changes also noted.

Figure 22

Figure 22 shows AP and lateral radiographs of left ankle in a 27 yrs. old with haemophilia B shows deformation of the talus with associated valgus deformity due to narrowed lateral part and widened medial part of the tibial epiphysis. Associated osteopenia and tibiotalar joint space narrowing also noted.

Figure 23

Figure 23 shows frontal radiograph of pelvis in a 27 yrs. old with haemophilia B also shows that right femoral head is reduced in volume and is deformed. Associated osteopenia also noted. Sclerosed acetabular margins (left>right) present.

Figure 24

Figure 24 shows AP radiograph of left shoulder in a 13 yrs. old haemophilic A patient shows resorption with atrophy and deformation of humeral head. Glenoid fossa also appears deformed.

Figure 25

Figure 25 shows AP radiograph of left knee of a 41 yrs. old with haemophilia A. Intraosseous pseudotumour appearing as a lytic lesion with cortical thinning and discontinuity noted in lateral femoral condyle.

Figure 26

Figure 26 shows AP radiograph of right knee of a 44 yrs. old with haemophilia A. Disorganised and fused joint with deformation of lower end of femur and upper end of tibia. Subperiosteal ossification noted in the lower end of femur. Intraosseous pseudotumour appearing as an expansile lytic lesion involving the tibial plateau with sclerotic margins.

Figure 27

Figure 27 shows AP and lateral radiographs of right knee in a 48 yrs. old with haemophilia A. Intraosseous pseudotumour appearing as a multiloculated expansile lytic lesion involving the tibial plateau with cortical thinning. Soft tissue pseudotumour noted as areas of soft tissue thickening causing extrinsic scalloping on upper 3rd of tibia. Lower end of femur is widened showing diffuse periosteal thickening. Tibiofemoral and patellofemoral joint spaces show significant narrowing with ankylosis.

CONCLUSION

Early diagnosis of haemophilic arthropathy requires adequate knowledge of imaging patterns in various stages of the disease. Although it is mimicked by various conditions, the clinical profile and imaging features will help in differentiating between them and reaching a definitive diagnosis.

Stage	Findings
I	No skeletal abnormalities. Soft tissue swelling
II	Osteoporosis and overgrowth of epiphysis. No erosions. No narrowing of cartilage space
III	Early subchondral bone cysts, squaring of the patella. Intercondylar notch of distal femur and humerus widened. Cartilage space remains preserved
IV	Findings of stage III more advanced. Cartilage space narrowed significantly
V	End stage. Fibrous joint contracture, loss of joint cartilage space, marked enlargement of the epiphyses and substantial disorganization of the joints

Table 1. Arnold–Hilgartner X-Ray Scale

Types of Change	Findings	Score
Osteoporosis	Absent Present	0 1
Enlarged epiphysis	Absent Present	0 1
Irregular Subchondral surface	Absent partially involved totally involved	0 1 2
Narrowing of joint space	Absent Joint space >1 mm Joint space <1 mm	0 1 2
Subchondral cyst formation	Absent 1 cyst > 1 cyst	0 1 2
Erosions at joint margins	Absent Present	0 1
Gross incongruence of articulating bone ends	Absent Slight Pronounced	0 1 2
Joint deformity (angulation and/or displacement between articulating bones)	Absent Slight Pronounced	0 1 2

Table 2. Pettersson X-Ray Scale

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