SILENT AVASCULAR NECROSIS OF THE FEMORAL HEAD
IN HAEMOPHILIA

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Haemophilia is a rare cause of avascular necrosis of the femoral head. We report three cases from one centre, an incidence of 2.8%. All three cases presented “silently”, and this makes the early diagnosis difficult.

Awareness of the condition should lead to examination of the hips of haemophiliac patients at every outpatient visit and admission in the hope that hip disease can be diagnosed at an early stage. This may allow earlier treatment, less femoral head deformity, and an improvement in the long-term prognosis.

Avascular necrosis of the femoral head (AVN), has numerous causes. In adults the commonest are: alcoholism, trauma, steroid therapy, hyperuricaemia, systemic lupus erythematosus, caisson disease, haemoglobinopathies, haemophilia, and Gaucher’s disease (Boettcher et al. 1970).

In children the most common causes are: Perthes’ disease, congenital dislocation of the hip (CDH), intracapsular fracture of the femoral neck, and slipped upper femoral epiphysis. Rarer causes are associated with conditions such as Morquio-Brailsford syndrome, hypothyroidism and sickle cell disease. The association of AVN with haemophilia has been briefly mentioned in some textbooks, but it has never been discussed in depth (Sharrard 1971).

We report two cases of classical haemophilia A, and one of Christmas disease (haemophilia B), who developed AVN of the femoral head. The association of two uncommon childhood diseases in three cases from one centre suggests that AVN of the femoral head may occur more frequently in haemophiliacs than previously recognised.

PATIENTS AND METHODS

We undertook a retrospective review of all the cases of haemophilia A and Christmas disease under treatment at the Haemophilia Centre, Royal Manchester Children’s Hospital, Pendlebury. The severity of haemophilia was recorded as mild, moderate or severe according to the level of factor VIII or IX. Severe cases had factor VIII or IX levels of 0.01 μ/ml (1% or less), moderate cases had 0.01 to 0.1 μ/ml (1 to 10%), and mild cases had factor levels of over 0.1 μ/ml (10%). However, the classification of severity was modified if the clinical presentation was markedly different from that expected from the laboratory results.

RESULTS

There were three cases of AVN of the femoral head, all with advanced radiological changes. Two were in the haemophilia A group, which totalled 90 cases and one was in the Christmas disease group of 17 cases. The clinical presentation was similar in all three. They had a limp, slight restriction of hip movement, and leg length inequality of 3, 1.5 and 2 cm respectively. None had any convincing previous history of hip disease. The ages at presentation were six, 12 and nine years.

Typical radiographs at presentation are shown in Figures 1 and 2. In each of the three cases there was total epiphyseal involvement, damage to the physis, coxa vara and a “sagging rope sign” (Apley and Wientroub 1981). We consider that this latter radiological sign is produced by the anterolateral edge of the severely deformed femoral head, rather than by condensation of the spongiosa within the neck (Clarke, Harrison and Keret 1983). We concluded that all three cases had severe AVN.

The overall incidence of AVN in haemophilia A in our series was 2.2%, and in Christmas disease 5.8% (Table 1). For both diseases together the incidence was 2.8%. Our results were analysed statistically using Fisher’s Exact Test, which showed no significant
difference between the incidences in haemophilias A and B, when the mild, moderate and severe groups were compared with each other.

DISCUSSION

The incidence of haemophilias A and B is one case per 10,000 newborn male infants (Arnold and Hilgarter 1977); the diseases are of sex-linked recessive inheritance though in a third of patients no family history can be established. Classical haemophilia (Type A), shows a deficiency of factor VIII, and Christmas disease (haemophilia B), a deficiency of factor IX. Bleeding occurs in joints, most commonly in the knee, elbow and ankle, though the wrist, shoulder, foot and hip may be involved (Stoker and Murray 1974).

The commonest cause of AVN of the femoral head in children is Perthes' disease. This disease has an incidence of one in 1,200, is bilateral in 15% and is seen mainly in children from three to 12 years of age with a peak incidence at six years. Boys are affected four times more than girls (Turek 1984). “Silent” Perthes' disease has been described (Kemp 1983), but this presentation is the exception rather than the rule.

The association of AVN with haemophilia has been reported (Winston 1952; Moseley 1963), but no incidence has previously been given to compare with our finding of 2.8%. Trueta (1963) proposed that AVN in haemophilia was due to the occlusion of the epiphyseal vessels by haemarthrosis, but Moseley (1963) considered that it was secondary to intra-epiphyseal haemorrhage. Winston (1952) felt that intra-osseous bleeding before puberty could lead to the appearances of AVN, while after puberty this led to the more common changes of haemophilic arthropathy. However, Wood, Omer and Shaw (1969) was unable to verify this age relationship.

| Table 1. Incidence of AVN of the femoral head in 107 haemophiliac patients |
|-----------------------------|---------|-------|-----|
|                             | Number  | Per cent | AVN |
| Haemophilia A               |         |         |     |
| Severe                      | 39      | 43%     | 1   |
| Moderate                    | 23      | 26%     | 0   |
| Mild                        | 28      | 31%     | 1   |
| Total                       | 90      |         |     |
| Christmas disease           |         |         |     |
| Severe                      | 7       | 41%     | 0   |
| Moderate                    | 7       | 41%     | 1   |
| Mild                        | 3       | 18%     | 0   |
| Total                       | 17      |         |     |

These theories do not adequately explain the aetiology. In our three cases, AVN developed silently and the number of cases seen at our centre makes it likely that this is usual. An acute haemarthrosis of the hip with a joint pressure high enough to obstruct the epiphyseal vessels is unlikely to follow any but a major bleed, which would stretch the synovium and capsule and cause pain. The absence of pain in our cases does not suggest significant haemarthrosis. Spontaneous bleeds are not seen to occur in other epiphyses in cases of haemophilia and there are no special factors to cause this in the hip, so the theories of both Trueta and Moseley are unlikely.

Only one of our cases suffered from regular spontaneous bleeds; in the other two bleeding was usually started by trauma, though this was less severe than that required to cause haemarthrosis in a normal child. It seems possible that mild trauma caused haemorrhage at and around the epiphyseal vessels themselves, damaging the tenuous blood supply to the epiphysis without producing a significant haemarthrosis,
and thus being clinically "silent". This hypothesis is supported by Boettcher's theory (1970), which postulates that idiopathic AVN may be caused by any defect in blood coagulation resulting in bleeding, sludging or clotting of the blood supply to the femoral head.

Clinicians dealing with haemophilia should be aware of the association with AVN and also of the fact that it may occur in cases with moderate or mild disease. This may enable treatment for AVN to be instituted early, in the hope of reducing deformity of the femoral head. The value of a bone scan in such cases is difficult to assess because of the lack of clinical complaints. However, a haemophiliac presenting with a limp and no radiographic changes in the hip may benefit from this investigation. In all cases of haemophilia, the hip should be examined and leg length recorded at all outpatient visits and admissions so that an early diagnosis of AVN can be made.

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REFERENCES


