Destructive synovitis in contralateral Perthes' disease

A report of 2 cases

Lieve Vanpaemel[1], Paul J M Helders[3], Wietse Kuis[2], Hans Pruijs[1], Jonne Huber[4] and Willem Keessen[1]†

2 boys had unilateral Perthes' disease at the age of 5 years. After 6 and 7 years, respectively, both patients developed contralateral femoral head necrosis with rapid destruction leading to ankylosis of the hip. Histology of the synovium showed non-specific synovitis. Both patients fulfilled criteria for oligo-articular juvenile chronic arthritis (JCA). The association with Perthes' disease suggests a common etiology.

Departments of Pediatric [1]Orthopedics, [2]Rheumatology, [3]Physical therapy and [4]Pathology, University Hospital for Children and Youth, "Het Wilhelmina Kinderziekenhuis", P.O. Box 18009, 3501 CA Utrecht, The Netherlands
Tel +31 30-320911. Fax -334825. *Correspondence: Dr. J.E.H. Pruijs, Department of Pediatric Orthopedics.*
While finishing this case report Dr. Willem Keessen suddenly died. We lost a special friend and a highly skilled orthopedic surgeon.
Submitted 93-06-12. Accepted 94-03-21

Case 1

A 5-year-old caucasian boy was first seen in 1983 with Perthes' disease of the right hip (Figure 1). He was treated with a caliper for almost a year. In 1988 he was without complaints; the range of motion of both hips was normal and symmetric, and he was discharged from clinical control. In April 1990 he was seen for pain in his left hip. There was no preceding infection or trauma. In July 1990 he was referred to our hospital with slightly decreased flexion, internal rotation and abduction of the left hip. The radiographs showed on the left side a dense epiphysis, decreased epiphyseal index and metaphyseal reaction, and on the right side an enlarged femoral head compatible with previous Perthes' disease. Laboratory findings were all within normal range. Viral and bacterial infections were excluded by microbiological laboratory tests. There were no auto-antibodies present, including anti- nuclear antibodies and rheumatoid factor. HLA-B27 was negative. We found no signs of metabolic disease.

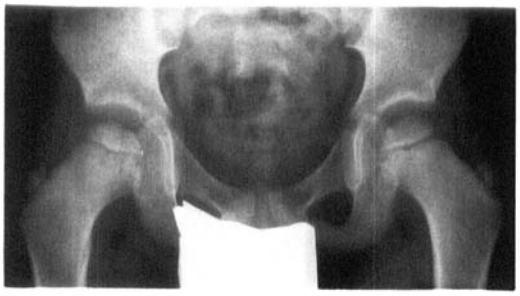
In September 1990 arthroscopy of the left hip showed a swollen, red and villous synovium with ingrowth in cartilage/bone areas. The cartilage covering the head showed small irregularities. The acetabulum and labrum were normal. Microscopic examination showed proliferation of synovial villi with minimal inflammatory changes. At the immuno-histochemical examination extensive deposits of IgG, IgM, IgA, C1q, C4, C3, properdin, fibrinogen and to a lesser degree of C5 were seen.

Initially, the patient was treated with NSAIDs (Naprosyn®) and partial weight bearing. However, the range of motion of the left hip decreased while pain increased. An open synovectomy was performed in December 1990. Histological examination showed non-specific synovitis, with extensively swollen and edematous synovial villi, proliferation of synovial lining cells and chronic perivasculitis. After the operation he had skin traction and later ambulation with non-weight bearing crutches. However, the range of motion of the left hip further deteriorated. Radiographically, destruction of the femoral epiphysis was seen. Ultrasonography and MRI showed intra-articular fluid. 3 months later the hip was nearly ankylosed in a position of flexion, external rotation and adduction.

Case 2

A 5-year-old caucasian boy developed Perthes' disease of the right hip in 1984 (Figure 2). Because of loss of containment, a varus osteotomy was performed at the end of 1985. However, the range of motion decreased and apparently because of increasing pain a valgus osteotomy was performed in 1986.

Case 1. Perthes' disease of the right hip.



1983.

1990. Flattening and increased density of the left femoral epiphysis, metaphyseal cyst formation; enlarged right femoral head and broadening of the right metaphysis, in AP view.

1991. Destruction of the left femoral epiphysis, cyst formation in the left acetabulum, joint space narrowing of the left hip.

1991. MRI. Destruction of the left hip, intra-articular mass of fluid and synovium.

Case 2. Perthes' disease of the right hip.

1984. Flattening and increased density of the right femoral epiphysis, metaphyseal reaction.

1990. Increase in density of the left femoral epiphysis, Gage's sign. Metaphyseal reaction.

1991. Destruction of the left hip with severe irregularity and lateroposition of the epiphysis, and joint space narrowing. Flattening and varus position of the right femoral epiphysis.

In April 1990 he complained about his left hip. The radiograph displayed a subchondral fracture with increased epiphyseal density and Gage's sign, comparable to images seen in cases of Perthes' disease. Technetium bone scintigraphy showed decreased perfusion of the left femoral head.

In 1991 he was referred to our hospital with considerable loss of motion and radiographical extensive destruction of the left hip. Microbiology laboratory tests revealed no viral and bacterial infections. No auto-antibodies were seen. HLA-B27 antigen was negative. Ultrasonography and MRI showed intra-articular fluid as well as thickening of the synovium. To improve the range of motion, a synovectomy with resection of the lateral part of the femoral head was performed in August 1991. Microscopic examination of the synovium showed edema of synovial villi, proliferation of lining cells and fibrin deposits. Vascular proliferation accompanied by perivascular infiltration of lymphocytes and plasma cells was seen. A regime of non-weight bearing and physical therapy was started. However, this was not successful, and the hip rapidly stiffened in flexion, internal rotation and adduction.

Discussion

Synovitis may interfere with the vascular supply of the femoral head by an increase in intra-articular pressure (Kallio and Ryöppy 1985, Royle and Galasko 1992). The most frequent type of arthritis in childhood is transient synovitis. However, the relation between this disease and Perthes' disease is controversial. Landin et al. (1987) reported development of Perthes' disease in 4 percent of patients with transient synovitis. Erken and Katz (1990) and Kallio et al. (1986) could not establish an etiological relation between Perthes' disease and transient synovitis and therefore considered them as unrelated clinical entities.

Royle and Galasko (1992) recently demonstrated scintigraphically an ischemia of the femoral head in 15 out of 92 cases of transient synovitis, 4 of whom developed Perthes' disease. Wingstrand et al. (1985) noted an increase the intracapsular pressure in transient synovitis. Gershuni et al. (1983) found that transient synovitis did not increase intra-articular pressure to a sufficient degree to cause Perthes' disease in animal experiments. However, in chronic types of synovitis Kallio and Ryöppy (1985) found a considerable increase in intra-articular pressure. This may affect the femoral head blood flow, as was demonstrated in dogs (Vegter and Klopper 1991) and in goats (Svalastoga et al. 1989).

The most frequent disease accompanied by synovitis of longer duration in childhood is juvenile chronic arthritis (JCA). The polyarticular type of JCA (4 joints or more involved) has been recognized as a cause of femoral head necrosis (Kobayakawa et al. 1989). In the two boys presented here, a chronic non-specific synovitis was present, which was compatible with the diagnosis JCA of oligo-articular type. The clinical and radiographic courses as well as the pathological findings in our cases suggest that necrosis and subsequent joint destruction may result from synovitis. The possible relation between JCA of oligo-articular type and Perthes' disease was considered by Mach and Wegener (1985). In view of the critical vascularization of the femoral epiphysis, femoral Perthes' disease may be considered as a non-specific sequel of chronic synovitis; in those with prolonged synovitis and restriction in range of motion an oligo-articular JCA might be suspected.

The finding of a chronic synovitis in Perthes' disease has consequences for diagnosis (ultrasonography and MRI) and therapy (anti-inflammatory drugs).

Acknowledgement

Dr. Hans van der Laar referred his patients.

References

Erken E H, Katz K. Irritable hip and Perthes' disease. J Pediatr Orthop 1990; 10 (3): 322–6.

Gershuni D H, Hargens A R, Lee Y F, Greenberg E N, Zapf R, Akeson W H. The questionable significance of hip joint tamponade in producing osteonecrosis in Legg-Calvé-Perthes syndrome. J Pediatr Orthop 1983; 3 (3): 280–6.

Kallio P, Ryöppy S. Hyperpressure in juvenile hip disease. Acta Orthop Scand 1985; 56 (3): 211–4.

Kallio P, Ryöppy S, Kunnamo I. Transient synovitis and Perthes' disease. Is there an aetiological connection? J Bone Joint Surg (Br) 1986; 68 (5): 808–11.

Kobayakawa M, Rydholm U, Wingstrand H, Pettersson H, Lidgren L. Femoral head necrosis in juvenile chronic arthritis. Acta Orthop Scand 1989 ; 60 (2): 164–9.

Landin L A, Danielsson L G, Wattsgård C. Transient synovitis of the hip. Its incidence, epidemiology and relation to Perthes' disease. J Bone Joint Surg (Br) 1987; 69 (2): 238–42.

Mach V J, Wegener S. HLA-examinations at Perthes disease. Beitr Orthop Traumatol 1985; 32: 590.

Royle S G, Galasko C S. The irritable hip. Scintigraphy in 192 children. Acta Orthop Scand 1992; 63 (1): 25–8.

Svalastoga E, Kiaer T, Jensen P E. The effect of intracapsular pressure and extension of the hip on oxygenation of the juvenile femoral epiphysis. A study in the goat. J Bone Joint Surg (Br) 1989; 71 (2): 222–6.

Vegter J, Klopper P J. Effect of intracapsular hyperpressure on femoral head blood flow. Laser Doppler flowmetry in dogs. Acta Orthop Scand 1991; 62 (4): 337–41.

Wingstrand H, Egund N, Carlin N O, Forsberg L, Gustafson T, Sundén G. Intracapsular pressure in transient synovitis of the hip. Acta Orthop Scand 1985; 56 (3): 204–10.