Case report

Idiopathic chondrolysis of the hip: a case report and review of the literature

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SUMMARY A case of idiopathic chondrolysis of the hip in an adolescent Indian girl is reported. The association between idiopathic chondrolysis of the hip and primary protrusio acetabuli (Otto's pelvis) is discussed.

Key word: protrusio acetabuli.

Chondrolysis of the hip associated with slipped upper femoral epiphyses is well recognised,¹⁻⁶ and its association with trauma and prolonged immobilisation has been described.^{4,7} Idiopathic chondrolysis of the hip was described by Jones in 1971⁸ and sporadic reports have since been made.⁹⁻¹³ The known aetiological factors include age, sex, and racial origin of the patients; the impression is that this is a disease mainly occurring in teenage female Negroes. This paper reports only the second case of idiopathic chondrolysis of the hip occurring in a female Indian girl and reviews the cases previously described in the literature.

Case report

The patient was a 14-year-old Indian girl who was admitted on 11 November 1983. She was a healthy girl with no significant previous illnesses and no history of recent injury. Her presenting symptoms were pain of insidious onset in the right hip and leg, associated with a limp which had been present approximately three months prior to admission.

On examination she appeared well with no fever or lymphadenopathy. All movements of the right hip were restricted and painful. She had a fixed flexion deformity of 20° but could flex the right hip to 100°. There was no external rotation, and internal

rotation was limited to 30°. There was also some minor discomfort in the left hip but with no restriction of movement.

Laboratory investigations showed a mild anaemia of an iron deficiency type (haemoglobin 10 g/dl; serum iron 6 µmol/l; serum total iron binding capacity (TIBC) 60.8 µmol/l; saturation 9.9% (normal value 15)) but there was no leucocytosis and the leucocyte differential count was normal; serum platelet concentration was normal. There was a slightly raised erythrocyte sedimentation rate (ESR) which was reported as 34 mm/1st h. Rheumatoid factor, antistreptolysin O (ASO) antibody titre, sickle cell test, and thalassaemia screen were normal. A biochemical screen was normal, apart from persistently raised alkaline phosphatase, which was reported as 28, 22, 24, and 20 KA units on four separate tests, with laboratory normal limits of 3-10 KA units. Serum calcium levels were normal. A skin Mantoux test was normal.

Pelvic x-rays taken on 6 December 1983 showed definite joint space narrowing affecting the right hip (Fig. 1) and possibly some slight cortical haziness affecting the right femoral head. There was regional osteoporosis. Tomographic studies confirmed the joint space narrowing and there was no definite bone destruction. A technetium bone scan showed no specific local abnormality in the region of the right hip.

The young girl was treated initially with bed rest, traction, and paracetamol analgesic. However, since

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Fig. 1 Anteroposterior of pelvis at time of admission.

her symptoms did not subside and septic arthritis could not be excluded, the right hip was explored and material taken for biopsy on 9 January 1984.

ARTHROTOMY FINDINGS

The right hip was explored by a Smith-Petersen approach. The capsule was found to be grossly thickened and oedematous. The synovium was hypertrophic, reddened, and oedematous, but there was no pannus. The articular surface appeared to be intact, though a curious indentation on the superior lateral aspect of the femoral head, i.e., the weight-bearing area, was noticed. This depression was

approximately 1 cm in diameter. There was no excessive synovial fluid in the joint. Biopsy material was harvested from the femoral neck, the superior lip of the acetabulum, and the capsule and the synovium. The capsule was not closed but the wound was otherwise closed in layers with suction drainage. Postoperatively the wound healed well and the girl's right leg was nursed on a continuous passive motion machine.

Microbiology reports on the biopsy material showed no growth and delayed cultures gave no mycobacterial growth. Histological investigation of the biopsy material of the synovium and capsule

Fig. 2 Anteroposterior of pelvis showing progression of protrusio acetabuli.

showed non-specific mild chronic inflammatory changes only. There was no evidence of rheumatoid arthritis or tuberculosis.

SUBSEQUENT MANAGEMENT

The iron deficiency was corrected with oral iron supplements. With the use of a continual passive motion machine her range of movements improved during the subsequent weeks and she was discharged from hospital on 10 February 1984. She limped less and could walk without pain. However, subsequent x-rays taken on 24 January 1984 showed further loss of joint space (Fig. 2) and the development of protrusio acetabuli. A final review six months after the time of presentation showed that she was now pain free, but the right hip was ankylosed with 30° fixed flexion and 10° internal rotation and adduction deformity.

Discussion

Jones⁸ reported the first cases of idiopathic chondrolysis of the hip in 1971, and a summary of this

paper and subsequent reports is shown in Table 1. In total 33 females (82.5%) and seven males (17.5%) have been studied, i.e., a ratio of females to males of 5:1. The average age at presentation is a little lower for females, 12.5 years (range 9–18 years) than for males, 14.8 years (range 13–20 years). There is an apparent preponderance of the right hip being more commonly affected (60% of cases), with two cases described¹³ with bilateral involvement (5% of cases).

The earlier reports suggested that this disease was limited to Negro adolescents^{8 10} until Wenger, Michelson, and Ponseti¹¹ described two white girls with the same condition. The racial incidence of the reported cases of idiopathic chondrolysis of the hip is given in Table 2, which shows that half of the cases are Negroid, and a third Caucasian.

CLINICAL FEATURES

Moule and Golding¹⁰ suggested that there could be delay in diagnosing this condition; in our review of the literature we found that 14 of the 40 cases had the time of presentation and time of diagnosis recorded. The delay in these cases ranged from 1 to 18 months. The clinical features of all 40 cases were similar. All presented with insidious onset of hip, leg, or knee pain, associated with a limp, and progressive loss of movements of the affected hip. In all cases there was no history of significant trauma, though Duncan reported that two of his nine cases had 'minor trauma'.⁹ All patients had been otherwise mobile and quite healthy before the onset of hip symptoms apart from two reported on by Jones, who had been previously immobilised because of disease in the opposite hip. All the patients had restricted hip movements at presentation, with fixed flexion deformities and fixed abduction or adduction deformities. White cell counts, blood cultures, and

Table 1 Idiopathic chondrolysis of the hip

	Female	Male
Numbers	33 (82.5%)	7 (17.5%)
Sex ratio	5	1
Age range (yr)	9–18	13–20
Average age (yr)	12.5	14.8
Side affected		
Right	24 (60%)	
Left	14 (35%)	
Right and left	2 (5%)	
Delay in diagnosis		
14/40 cases=1–18 months		

Table 2 Summary of reports of idiopathic chondrolysis of the hip

Paper	Negro	White	Hispanic	Indian	Unknown	Total
Jones (1971) ⁸	7				1	9
Moule and Golding (1974) ¹⁰	8			1		9
Sivananthan (1977) ¹²				1		1
Wenger, Michelson, and Ponseti (1975) ¹¹		2				2
Duncan <i>et al.</i> (1979) ⁹	3	5				9
Bleck (1983) ¹³	1	5	1			9
Hughes (this paper)		2	1	1		1
	20	14	2	3	1	40
	50%	35%	5%	7.5%	2.5%	100%

rheumatoid factors were normal. In only four of the 40 cases was the ESR higher than 30 mm/h. Bleck performed HLA-B27 determinations in six of his nine cases and this was negative in all six cases.¹³ Skin tests for fungal infection were all negative, but four of Jones' nine cases had a positive skin tuberculin test. No other cases were reported with similar positive skin reactions.

RADIOLOGICAL FEATURES

Bleck¹³ gave a detailed report on the radiographic changes seen in idiopathic chondrolysis of the hip. The x-ray features of the 40 reported cases are summarised in Table 3. Early changes include regional osteoporosis and blurring of the subchondral line, with reduction of the joint space. In none of the 40 cases was there evidence of epiphyseal slip. Late changes include early closure of capital, and trochanteric epiphyseal plates; more severe cases developed fibrous or bony ankylosis. In less severe cases there was a tendency to show the early changes of osteoarthritis with marginal osteophyte formation, progressive loss of joint space, and lateral buttress sclerosis. There is a tendency to develop secondary protrusio acetabuli with a progressive decrease in the CE angle of Wiberg.

PATHOLOGICAL FEATURES

Seventeen of the 40 cases had reported histological features (Table 2). Essentially these were thickening of the capsule, with oedematous changes in the capsule and synovium. There was usually no pannus formation. The synovium has been reported, in various papers, as non-specific chronic changes, or infiltration of plasma cells, or lymphocytes. There are articular surface changes with fibrillation and fragmentation and progressive loss of cartilage. Some reports suggest degeneration of chondrocyte nuclei and loss of cells in the lacunae. One case

Table 3 X-ray features of idiopathic chondrolysis of the hip

<i>Early</i>	
No epiphyseal slip	
Reduction of joint space	
Subchondral line blurring	
Protrusio acetabuli (decreased Wiberg centre edge (CE) angle) ²⁶	
Osteoporosis	
<i>Late</i>	
No epiphyseal slip	
Early epiphyseal closure	
Marginal osteophytes and lateral buttress	
Loss of joint space, ankylosis, osteoarthritis	

where some avascular necrosis was noted has been reported.¹²

DIFFERENTIAL DIAGNOSIS

Monarticular hip arthritis in adolescence presents a challenge to the diagnostician. Clinical evaluation, radiological appearances and joint tissue biopsy are the cornerstone of a firm diagnosis. A differential diagnosis includes:

- (1) trauma
- (2) slipped upper femoral epiphysis
- (3) septic arthritis
- (4) tuberculosis
- (5) Perthes' disease
- (6) pigmented villonodular synovitis
- (7) synovioma and other neoplasm
- (8) monarticular rheumatoid arthritis.

Positive features that should aid in the early diagnosis of idiopathic chondrolysis of the hip are the age, sex, and race of the patient who will present lame with hip pain and with no history of trauma or systemic illness. The patient will be aphyrexial with a stiff, contracted, painful hip joint, and laboratory data will not suggest any infections or rheumatoid aetiology. The x-rays will show osteoporosis and early joint space reduction with no effusion. Joint tissue biopsy may not be necessary and may delay any treatment¹³ but is required if there is still any doubt in the diagnosis. The histology report will be non-specific, and cultures will grow no bacterial isolates.

TREATMENT AND PROGNOSIS

In nine cases the mode of therapy was not reported. Ten cases received non-surgical treatment, including early traction followed by non-weight-bearing exercises and anti-inflammatory analgesics; five of these cases went on to develop complete loss of movement but were pain free. Twelve cases were treated with a hip spica, and eight of these subsequently developed a fusion. There have been three cases treated by soft tissue releases, and each of these later required further surgical procedures. Seven cases have received bony operations, including excision arthroplasty, mould arthroplasty. Wagner resurfacing arthroplasty, and arthrodesis.

Of the 31 cases where the treatment and progress have been reported 16 eventually lost all hip movements. However, these cases were all pain free. Nevertheless, it is evident that idiopathic chondrolysis of the hip is a progressive destructive disease for which there is no known effective therapy and which carries catastrophic end results for the children who are unfortunate enough to be affected by the disease.

ASSOCIATION WITH PRIMARY PROTRUSIO ACETABULI (OTTO PELVIS)

Otto first described protrusio acetabuli in 1824,¹⁴ and since then there have been reports of this condition occurring in childhood. The aetiology of this condition may be genetic,^{15 16} due to mechanical deformity of a soft acetabular floor,^{17 18} or due to early closure of the triradiate cartilage.¹⁸⁻²⁰ Idiopathic chondrolysis of the hip may lead to the development of protrusio acetabuli in childhood. Seven of the nine cases described by Jones⁸ developed protrusio acetabuli, as did one case of Wenger,¹¹ Moule and Golding,¹⁰ and the case reported here.

There are similarities in the clinical features of both conditions. Friedenber²¹ reports 17 cases of primary protrusio acetabuli and all were female, and six were Negro. Crichton and Curlewis²² report 67 female cases of protrusio acetabuli, and 75% of these cases were Negro, 8.5% white, and 16.5% Indian. In cases of idiopathic chondrolysis of the hip the probable cause of protrusion is the softening of the acetabular floor due to the local hyperaemia, and loss of the normal growth pattern of the triradiate cartilage.

The overlap in clinical features of these two diseases suggests that some cases diagnosed as primary protrusio acetabuli may have been cases of idiopathic chondrolysis of the hip and vice versa. Certainly the case reports of primary protrusio acetabuli described by Friedenber²³ (cases 1 and 2), Gilmour²⁰ (cases 1, 2, and 3), Brailsford,¹⁹ and again by Friedenber²¹ (cases 1, 2, and 3) could have been cases of idiopathic chondrolysis of the hip in adolescent girls.

CONCLUSION

Idiopathic chondrolysis of the hip is a rare clinical entity where, unfortunately, there is often a delay in diagnosis. There are characteristic clinical and radiological features but no effective therapy to arrest the disease. Non-steroidal anti-inflammatory analgesia has been given, but there have been no reports on the use of cortisone, which may be more effective if the aetiology is proved to have an immunological basis. This paper reports the use of continuous passive movement in treating idiopathic chondrolysis of the hip, but we cannot comment on its value. It is suggested that the disease may have immunological,²⁴ biochemical,²⁵ or viral origins.¹³ Awareness of the existence of this disease will lead to earlier diagnosis. Hopefully this in turn will remove the speculation on its aetiology and lead to effective management.

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