ADOLESCENT CHONDROLYSIS OF THE HIP JOINT*

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Between the years 1958 and 1970 there was encountered at this hospital a group of cases of hip joint disease which did not conform to such tentative diagnostic labels as tuberculous, low-grade infective or monarticular rheumatoid arthritis which had been initially applied to them. Certain of these cases, however, were eventually seen to present so consistent a clinical pattern as to justify their recognition as a distinct entity, and it is proposed to report here 9 such cases. Features of the condition are mild ache in one hip, limp, progressive stiffness often becoming complete within about a year, and its occurrence is predominantly in non-White, pubertal females. The initial diagnostic confusion must be blamed for the fact that some of the earlier cases were not so fully investigated as might have been desired.

CASE REPORTS

Case 1

A 13½-year-old female was admitted on 12 August 1958 with a complaint of limp and progressive stiffness of the left hip for the past several months. On examination there was no apparent pain, but the hip was virtually fixed in 15° flexion, 20° abduction and 40° external rotation. The course was afebrile, the Mantoux test was negative and the erythrocyte sedimentation rate was between 4 and 11 mm in one hour throughout. The X-rays (Fig. 1, top) showed some osteoporosis of the left hip region, uniform reduction of the left hip joint space and a CE angle of 35° on both sides. Repeated lateral views excluded epiphyseal slip. A tentative initial diagnosis of low-grade, infective arthritis was made, and penicillin and streptomycin were prescribed for 2 weeks only following admission. After 6 weeks of skin traction a plaster spica was applied for 2 months.

On 13 August 1959 she was symptomless, had a slight limp and a painless hip without movement, fixed in 20° flexion, 10° abduction and 40° external rotation. Later X-rays showed an increase of the left CE angle to 45°, and earlier fusion of the capital and trochanteric epiphyses on the left side than on the right.

When last seen on 26 July 1968 she had no pain, could walk as far as she wanted and had recovered some movement, flexion to 45°, 5° abduction, 15° adduction, 45° external rotation and internal rotation to neutral. Radiographically there was osteo-arthritic change with slight reduction of joint space and osteophyte formation at the capital margin (Fig. 1, top right).

Case 2

A 15-year-old Bantu female was admitted on 12 June 1962 with a severe destructive arthritis of the left hip of 2 years' duration diagnosed as tuberculous. This healed by bony ankylosis following antituberculotic therapy and plaster spica immobilization including the opposite hip. On admission the opposite right hip was completely normal clinically and radiologically.

*Date received: 22 October 1970.

When on 14 March 1963 plaster immobilization was discontinued there was found to be a slight ache in the previously normal right hip and gross restriction of movement. The patient was afebrile, the erythrocyte sedimentation rate was 15 mm in one hour, and the Wassermann reaction, latex-fixation, Widal and brucellar agglutination tests were all negative. The radiographs showed osteoporosis, uniform reduction of right hip joint space, blurring and irregularity of the subchondral line and an increase of CE angle, which in earlier X-rays had been 30°, to 45°.

By 17 April 1963 right hip movement was negligible and a Batchelor-Milch excision-arthroplasty was performed on the right hip. At operation the capsule appeared normal, but the synovium, including the retinacular reflections, was oedematous, thickened and pink. The femoral articular cartilage was lustreless with fairly large areas of erosion, two of them down to bone. Oedematous synovium extended slightly from the base of the ligamentum teres onto the acetabular cartilage, which here showed underlying erosion, but was fairly normal elsewhere. No organisms were isolated from the synovial fluid. The microscopic findings will be discussed later.

When last seen on 27 August 1964 the patient had no pain, walked with one stick and had a flexion range of 40°, and an abduction-adduction range of 40°.

Case 3

A 9-year-old Coloured female, already showing pubertal changes, was admitted on 18 December 1962 with a 3 months' complaint of limp and right hip pain, finally restricting her walking range to about 500 yards.

On examination the hip was in a position of flexion, abduction and external rotation with a very slight and painful degree of movement. She was throughout afebrile and the erythrocyte sedimentation rate varied between 3 and 10 mm in one hour. The Heaf test was positive, while latex-fixation, Widal and brucellar agglutination tests were all negative. Initial X-rays showed porosis of the right hip region, uniform reduction of the joint space and accentuation of the subchondral line, the CE angle being 30° on the left side, 35° on the right. Later radiographs showed premature fusion of capital and trochanteric epiphyses on the affected side (Fig. 1, second row).

Following 3 weeks of skin traction, crutch ambulation in a plaster spica was permitted. When last seen on 29 October 1964 the patient could walk over 5 miles and had a painless but motionless hip.

Case 4

A 13-year-old Coloured female was first seen at this hospital in May 1963. A year previously she had been seen at another hospital with a complaint of limp and ache in the right groin. Tuberculous arthritis was suspected by reason of a positive Heaf test and some radiographic osteoporosis of the hip region. She was apyrexial throughout and the erythrocyte sedimentation rate varied between 4 and 10 mm in one hour. She had been treated by skin

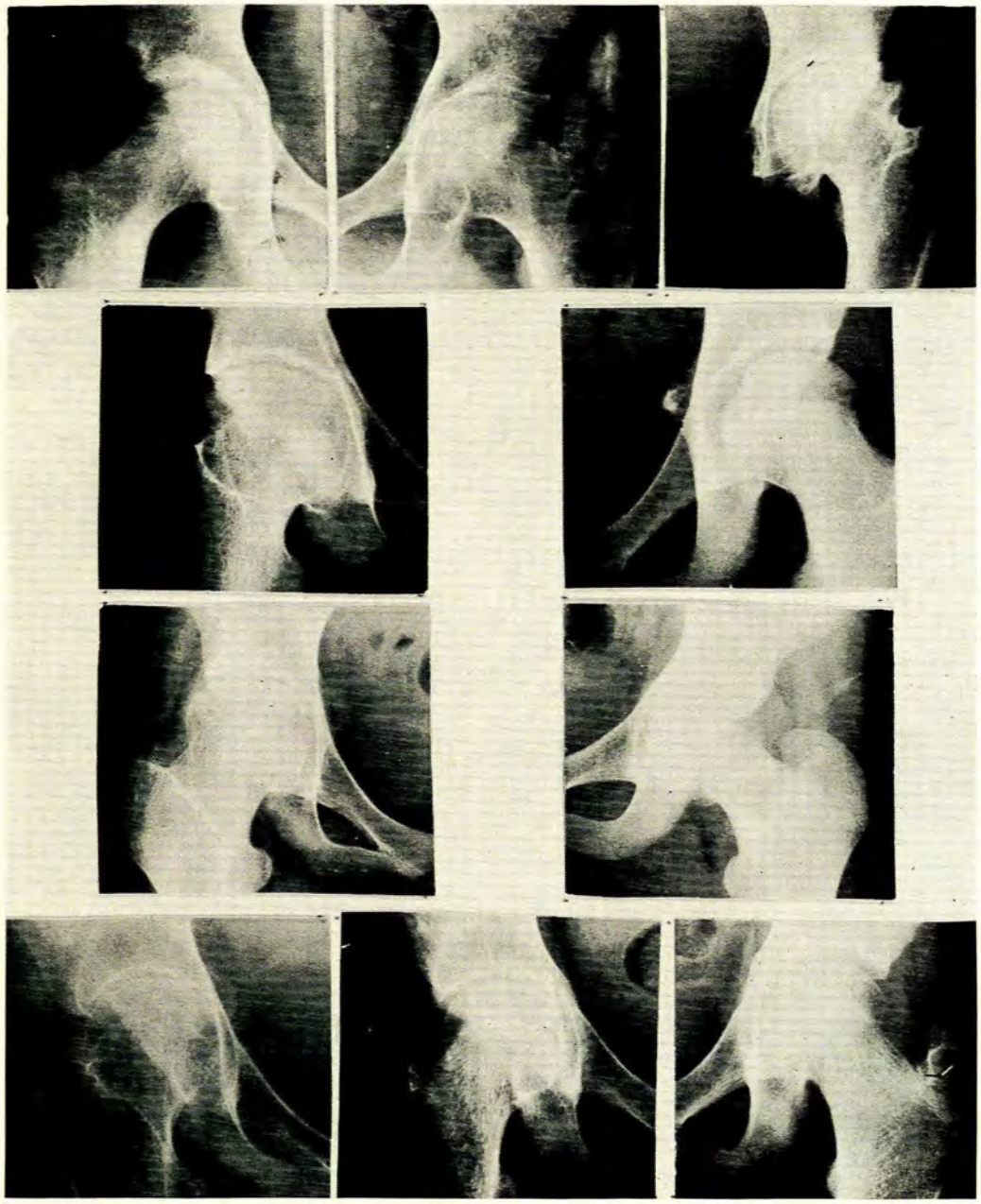

Fig. 1. Top: Case 1. Left: normal hip. Middle: involved hip in active phase. Right: involved hip 10 years after onset. *Second row*: Case 3. Left: involved hip in late active phase. Right: normal hip. *Third row*: Case 4. Left: involved hip in active phase. Right: normal hip. *Bottom*: Case 5. Left: involved hip in late active phase. Middle: involved hip in early active phase. Right: normal hip.

traction and antituberculotics for 2 months only, after which ambulation had been resumed. There was now no hip pain, but movement decreased progressively till she was referred here.

When seen here in May 1963 there was a painless limp and no movement in the hip. The erythrocyte sedimentation rate was 7 mm in one hour, the white cell count was 9 300/mm³, the blood culture negative, urine normal, serum protein electrophoretogram normal, Paul-Bunnell, Weil-Felix, Widal, latex-fixation and brucellar agglutination tests negative. The X-ray showed regional osteoporosis, uniform reduction of the right hip joint space, premature closure of the right trochanteric epiphyseal plate and a CE angle of 45° on the right side, 35° on the left (Fig. 1, third row and Fig. 2). On 18 October 1963 a corrective osteotomy of the hip was carried out, and,

as this was followed by some pain, the hip was arthrodesed on 22 December 1964. When last seen on 23 April 1970 her activities were normal and her hip was soundly fused, but there was 25 mm of shortening and occasional pain in the right knee.

Case 5

A 12-year-old Coloured female was admitted on 5 November 1965 with a complaint of ache in the right hip for which she had already been treated by skin traction for 2 months at another hospital. The hip was now painless, would flex to 105°, abduct 20°, adduct 30°, rotate externally 40°, and internally 0°. The Heaf test was negative to first strength, positive to second strength PPD, and the erythrocyte sedimentation rate was 5 mm in one hour. The white cell count was 9 550/mm³, the Wassermann, Paul-Bunnell, Weil-Felix, Widal, latex-fixation and brucellar agglutination tests were all negative. X-rays showed some regional porosis, a uniformly reduced right hip joint space and a CE angle of 45° on both sides (Fig. 2). The course had been afebrile.

She was allowed to resume ambulation, but there was occasional ache and progressive restriction of movement until, by April 1967, the hip was fixed in 30° flexion, 15° abduction and 25° external rotation. Radiographically (Fig. 1, bottom) the joint space had further decreased and the CE angle increased to 60° on the affected side.

On 26 April 1967 a cup arthroplasty was performed. Under anaesthesia a moderate movement range was obtained, suggesting that the restriction had been largely due to muscle spasm. Capsule and synovium appeared normal, but the capital and acetabular articular cartilage was thin, greyish-white and lustreless with some areas of erosion. The acetabulum was deep, uniformly hemispherical with loss of differentiation between the weight-bearing area and the acetabular fossa, and the floor was paper thin. Unfortunately histological examination was not carried out. As the hip remained symptomatic and stiff, it was arthrodesed in March 1968. When last seen in September 1970 the hip was painless and soundly fixed and her activities were unrestricted.

Case 6

A 16-year-old Bantu male was admitted in December 1964 with a painful, unstable right hip due to severe, long-standing subluxation of undetermined origin.

Following 4 months' abduction and immobilization on a frame, both hip regions were osteoporotic and the joint space of the normal left hip was reduced. Following remobilization the left hip joint space recovered to normal. On 4 November 1965 the subluxed right hip was arthrodesed and immobilized in a plaster spica including the opposite normal hip for 3 months.

On removal of the spica, very gross, painless reduction of movement of the previously normal left hip was noted, and X-rays (Fig. 3) showed osteoporosis, uniform reduction of joint space and increase of CE angle, originally 25° to 40°. The course was apyrexial and the ESR was 10 mm in one hour. The latex-fixation test was negative.

When he was last seen on 28 July 1968 the left hip was painless and virtually motionless, but he could walk a mile using crutches with a swing-through gait. Radiographs showed a much reduced joint space, increased sclerosis in the acetabular roof and some osteophytes superiorly.

Case 7

A 15-year-old Coloured female was first seen on 9 November 1967, complaining of a painless limp for several months and increasing stiffness of the right hip. The right hip was fixed in about 60° flexion. The erythrocyte sedimentation rate was 7 mm in one hour and she was apyrexial.

The Heaf, latex-fixation and brucellar agglutination tests were negative. Roentgenograms (Fig. 4, top) showed decreased joint space, some sclerosis and irregularity of the subchondral line, a CE angle of 45° on the right, 35° on the left side, and premature closure of both capital and trochanteric growth plates on the right side.

Fig. 2. Radiographic appearances in greater detail. *Left*: Case 4. *Right*: Case 5.

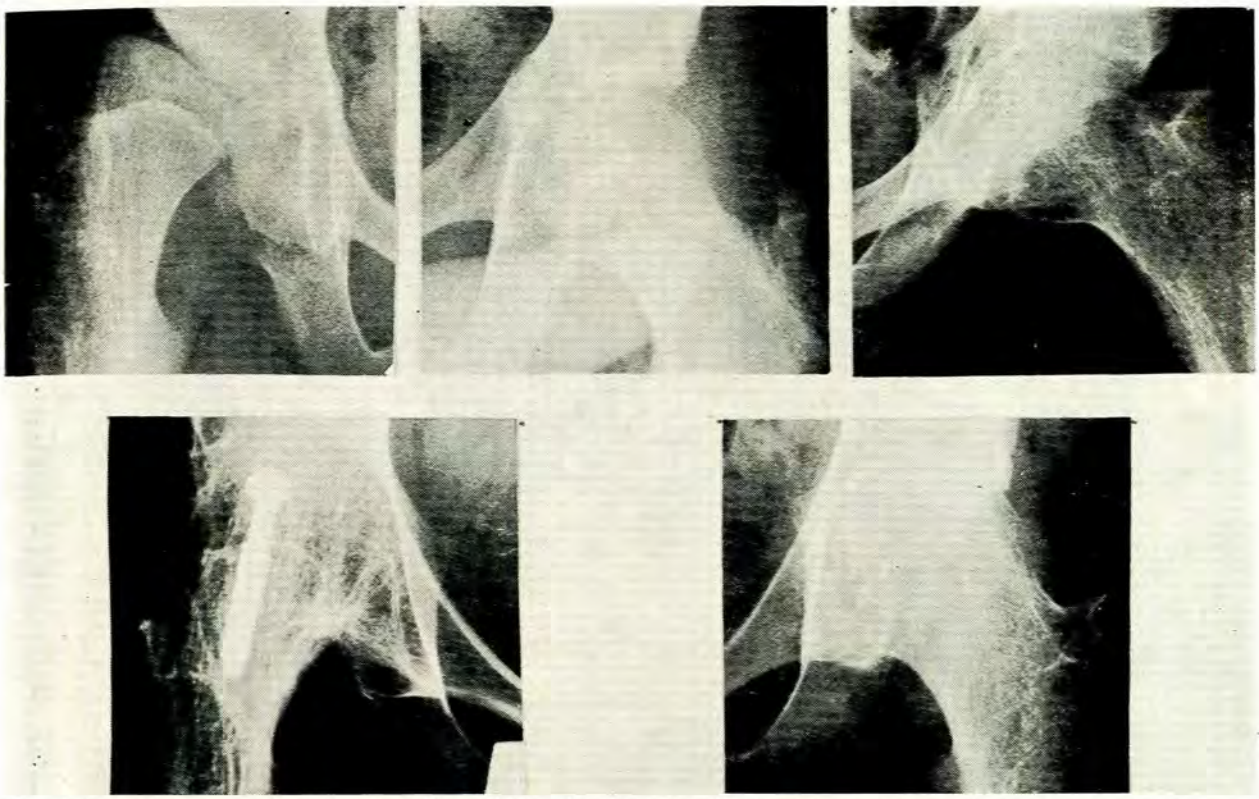

Fig. 3. Case 6. *Top*: Left: uninvolved hip. Middle: involved hip still normal before onset. Right: involved hip in early active phase. *Bottom*: Left: uninvolved hip after arthrodesis. Right: involved hip in late active phase.

On 28 February 1968 a mould arthroplasty was performed and under anaesthetic a moderate movement range was elicited. At operation both capsule and synovium appeared normal, but the capital and acetabular articular cartilage was thin and lustreless with areas of erosion, and the acetabulum was deep and its floor thin. While the gross appearances were entirely similar to those of case 5, the microscopic appearances closely approximated those of case 2 and will be discussed later.

When she was last seen on 14 November 1968 the hip was painless, would flex to 70°, abduct to neutral, adduct 20°, and rotate 10° in either direction.

Case 8

A 12½-year-old Coloured female was first seen on 18 December 1967 with a complaint of limp and pain in left hip and thigh. The hip was held in abduction and external rotation with pain on movement. She was apyrexial, the erythrocyte sedimentation rate was 10 mm in one hour and the Heaf, latex-fixation and Wassermann reactions were negative. Radiographs showed osteoporosis of the left hip region, marked reduction of the left hip joint space and a CE angle of 40° on both sides. The hip stiffened rapidly and progressively till it would flex only 30° and on 15 February 1968 a biopsy specimen was taken from the hip. Grossly there was increased vascularity of the synovium, which microscopically showed a thin surface layer of fibrin and a scanty infiltrate of plasma cells. No organisms were isolated.

When last seen on 20 August 1969 she had no pain and could walk far, but the hip had no movement. X-rays showed the left capital epiphyseal plate prematurely closed and the trochanteric plate almost closed (Fig. 4, middle).

Case 9

An 11½-year-old Coloured female with pubertal changes was admitted on 3 March 1970 with a history of pain in the left hip for 2 months. The hip was in a position of flexion, abduction and external rotation, and movement was painful though flexion to 80° was still possible.

The course was completely apyrexial, the erythrocyte sedimentation rate was 12 mm in one hour, the Heaf test was positive and the latex-fixation test was negative. X-rays (Fig. 4, bottom) showed some porosis of the left hip region, uniform reduction of the hip joint space, premature closure of the left capital and trochanteric growth plates and a CE angle of 30° on both sides. Initial treatment consisted of exclusion of weight-bearing, continued active hip movements and indomethacin. Later measures included a period of skin traction, a course of ethinyl oestradiol, and a single intra-articular corticosteroid injection. However, the abduction and external rotation deformity were not corrected and the movement range inexorably decreased. On 2 September 1970 a varus and derotation osteotomy was performed with metal fixation. Apart from limited internal rotation, all movements under general anaesthetic were little short of full. Six weeks after operation deformity was corrected and

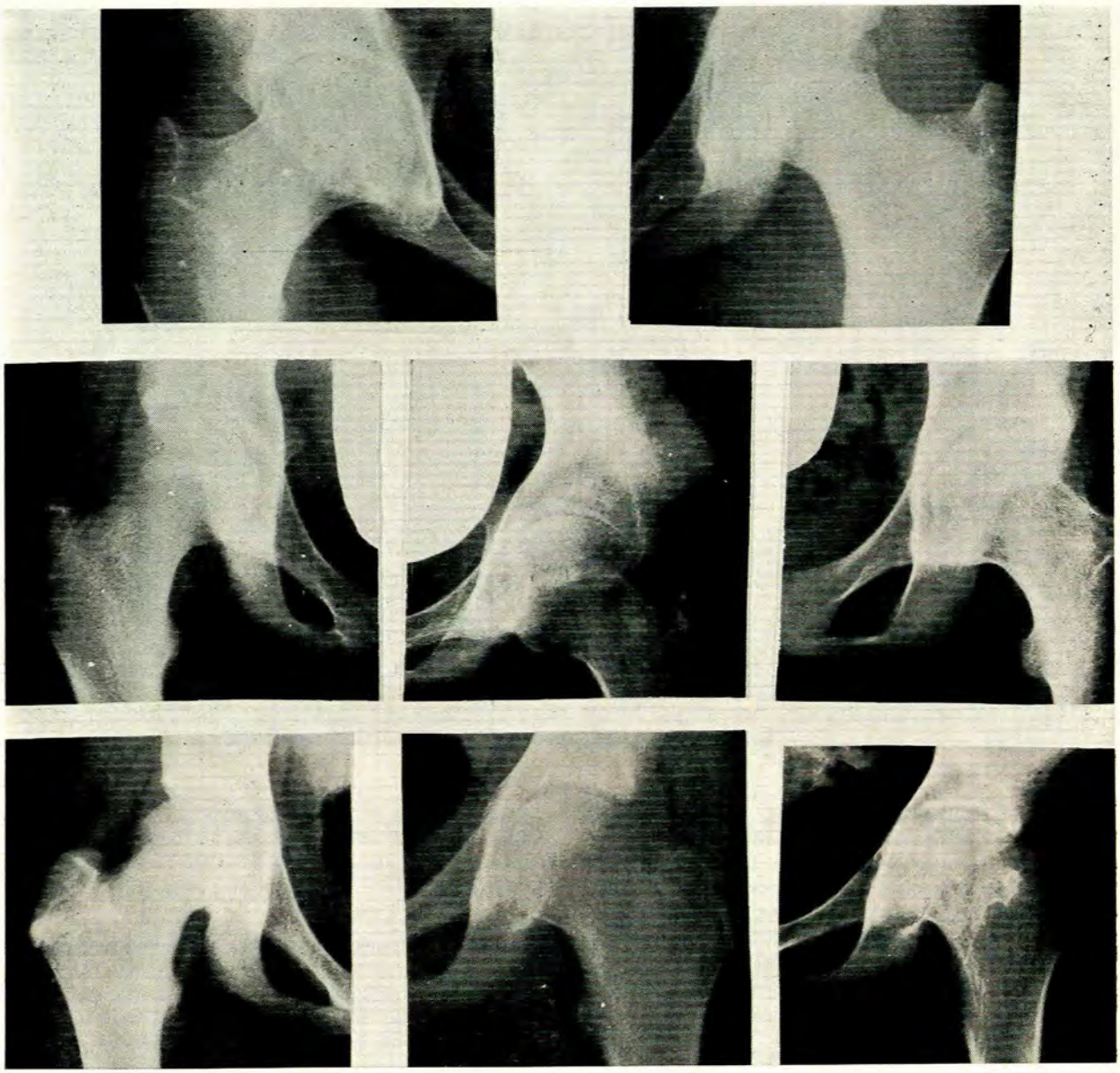

Fig. 4. Top: Case 7. Left: involved hip in active phase. Right: normal hip. *Middle*: Case 8. Left: normal hip. Middle: involved hip in early active phase. Right: involved hip in late active phase. *Bottom*: Case 9. Left: normal hip. Middle: involved hip in early active phase. Right: involved hip in late active phase.

spasm relieved, and movement range was markedly improved and progressively improving. She is still under treatment.

CLINICAL FEATURES

The over-all similarity of these 9 cases would seem to justify their recognition as a single clinical entity.

Thus all cases occurred about the time of puberty, individual ages at onset being 13½, 15, 9, 13, 12, 16, 15½, 12½ and 11½ years; all were non-White and all but one female.

Although in two cases the condition appeared to be precipitated by immobilization for a condition of the opposite hip, a tuberculous arthritis in case 2 and arthrodesis in case 6, in the majority no precipitating factor was apparent and the onset was insidious. The course was characterized by a mild ache, limp and progressive restriction of movement often proceeding within 6 - 18 months to complete clinical stiffness.

However in three such cases examined under anaesthesia in the earlier stages a moderate or good movement range was elicited, indicating that the gross restriction was largely due to muscle spasm. Nevertheless, of the 4 cases with an adequate follow-up which had not been subjected to some definitive operative procedure on the hip joint,

only one, case 1, retained or recovered a useful movement range.

Ache or discomfort was insufficient to take the patient off her feet except in the 4 cases already hospitalized elsewhere, two of them for conditions of the contralateral hip. This continued ambulation may perhaps have contributed to the fact that the characteristic position of deformity was usually one of flexion, abduction and external rotation.

All cases were apyrexial throughout and in all the erythrocyte sedimentation rate remained normal. The Mantoux or Heaf test was positive in 4 cases, negative in 4 and unrecorded in one, while the latex-fixation test was negative in all 8 cases in which it was carried out. All other accessory investigations carried out in individual cases and detailed in their records were negative.

RADIOLOGICAL FEATURES

In the earlier, active phase of the disease radiographs showed osteoporosis of the hip region, a uniform reduction of joint space and irregular blurring of the subchondral line on both femoral and acetabular surfaces.

As the disease progressed there was a further decrease in joint space which, however, never became completely obliterated even in the presence of complete clinical stiffness. In most cases there developed a degree of protrusio, with a gradual increase in CE angle as measured by Wiberg's method. This increase as compared with the normal hip was 15° in one case, 10° in 3 cases, 5° in one case, and nil in two. In two cases in which disease of the opposite hip prevented a comparison the CE angle of the involved hip increased from 30° to 45° in case 2, and from 25° to 40° in case 6. In 4 cases both capital and trochanteric epiphyses, in one the capital and in one the trochanteric alone closed sooner on the involved than on the normal side, while in cases 2 and 6 no comparison was possible. With healing the subchondral lines became denser and better defined. The later appearances in cases 1 and 6 were those of osteo-arthritis with reduced joint space and marginal, capital osteophytes. In all cases repeated lateral views excluded the possibility of associated epiphyseolysis.

PATHOLOGICAL FEATURES

Although in the active phase the synovium may be pink, thickened and oedematous as in cases 2 and 8, in the healing phases in cases 5 and 7 capsule and synovium appeared grossly normal. Articular cartilage on both capital and acetabular aspects is thin and lustreless with areas of erosion, and the acetabulum is deep and its floor very thin as in cases 2, 5 and 7. Microscopically the synovium in case 2, in the active phase, was oedematous, thickened and showed focal and diffuse areas of chronic inflammatory cell infiltration, while in case 8 there was a surface layer of fibrin and a scanty infiltrate of plasma cells. In cases 2 and 7 the articular surface had lost its covering of cartilage marginally and was invaded from the synovium by a covering of vascular pannus showing fibrocartilage formation.

Elsewhere deeper layers of the articular cartilage showed empty lacunae and/or chondrocytes with degenerate nuclei. The subchondral bony lamina was breached at numerous places with invasion of the deeper layers of the articular cartilage by vessels and pannus. The subchondral bone was normal and showed no inflammatory changes.

DISCUSSION

This is a condition affecting one hip, usually in a pubertal, non-White female, and characterized by a mild ache, insufficient to take the patient off her legs, a limp, a position of flexion, abduction and external rotation and progressive restriction of movement often terminating within about a year in complete stiffness. Radiographically osteoporosis and a uniformly reduced joint space progress to a degree of protrusio and finally typical osteo-arthritic changes. Pathologically the primary change appears to be in the articular cartilage and notably in its deeper layers.

As regards the aetiology, the negative Mantoux test in some cases and the spontaneous healing in absence of either immobilization or antituberculotic therapy excludes a tuberculous origin, and there is no evidence suggesting any other infective agent. The normal erythrocyte sedimentation rate, negative latex-fixation reaction and the relatively short, self-limiting course without subsequent involvement of other joints renders an autoimmune process unlikely.

The condition here described does, however, resemble closely the joint disorder sometimes associated with upper femoral epiphyseolysis and characterized by progressive joint stiffness and radiographic loss of joint space, normal white cell count and erythrocyte sedimentation rate. In one of three such cases, girls of 13 and 15 and a boy of 16, Waldenström[1] confirmed the cause at operation as resorption of articular cartilage. Ponseti and Barta[2] recorded three similar cases, and Lowe[3] reported an incidence of 15 cases among 100 patients with epiphyseolysis. At that time he found the prognosis poor, four hips requiring arthrodesis within 12 months of onset, but in a later article[4] he found that with prolonged, non-weight-bearing exercises a number of patients may gain reasonable recovery of joint space and movement range. Orofino *et al.*[5] emphasized the relative frequency of this condition in Negroes; in their series 52·8% of Negro patients had an epiphyseal slip. Cruess[6] found two cases of the condition among 43 Negroes with epiphyseolysis, and reported the main pathological features to be gross capsular and synovial thickening with adherence to the femoral neck, thinning or complete destruction of articular cartilage with fibrous tissue replacement and some areas of fibrocartilage formation, but complete normality of bone and no evidence of an infective process. The findings in his cases could represent late stages in the repair process. The only obvious difference between these cases and the condition presently reported is the absence of association with epiphyseolysis. It should, however, be noted that in one of Ponseti and Barta's cases and two of Lowe's it was the so-called healthy hip, free of epiphyseal slip, that was involved.

It seems that the underlying pathology in both conditions is death of articular cartilage cells, though Lowe[4]

noted in his epiphyseolysis cases cell death in the superficial two-thirds with survival of a layer of basal chondrocytes, while in the condition here described it was the latter that suffered first. The encroachment from the synovium and elaboration of fibrocartilage at the periphery of the cartilage and the breaching of the subchondral lamina, and invasion of the deeper cartilage layers by capillaries from the subchondral bone may be interpreted as secondary repair responses.

The necrosis associated with epiphyseolysis has often been attributed to impaired synovial nutrition of the articular cartilage, perhaps consequent on such aspects of treatment as manipulation or immobilization. However, the condition may occur in the absence of treatment or on the contralateral side to the slip. Certainly a synovial causation of the condition at present reported seems improbable in that it is the chondrocytes of the deeper layers of articular cartilage that appear to be primarily affected, and in that epiphyseal cartilage may also be involved with premature closure of capital and trochanteric growth plates. Nevertheless, although the onset in most cases was insidious, it appears possible that in some a local factor may play a precipitating or contributory part. Thus in two cases the involved hip had shared the immobilization of the opposite hip, in case 2 for tuberculous arthritis and in case 6 following arthrodesis.

It would seem more probable that in both epiphyseolysis and articular cartilage necrosis, whether or not associated with epiphyseal slip, a systemic factor damaging to cartilage may be responsible. Ponseti and McClintock[7] postulated as the noxious factor in epiphyseolysis nitriles consequent on a derangement of protein metabolism, though potentiation of the action of a metabolite normally concerned with the cartilage degradation that precedes endochondral ossification seems equally possible. That such metabolites gain access to cartilage by the blood stream and not by the synovial fluid is indicated by the experiments of McKibbin and Holdsworth,[8] designed to demonstrate the dual nature of the cartilage of the epiphysis. When a square of cartilage was reversed, the process of degeneration and ossification in epiphyseal cartilage was arrested as long as it was exposed only to synovial fluid, but resumed as soon as vessels from the subchondral bone had again gained access to it.

Hodge and McKibbin[9] demonstrated that the articular cartilage of immature animals derives a significant proportion of its nutrition from the subchondral bone as well as from the synovial fluid, while in adults nutrition is derived solely from the latter. If we regard this cartilage as consisting of two layers—a deeper, ossifying, epiphyseal layer and a superficial articular layer—it is possible that during growth the latter is buffered and shielded from noxious influences from the subchondral osseous circulation by the intervening epiphyseal cartilage layer. When the latter has completed its growth and is completely converted to bone the adult articular cartilage becomes sealed off from the subchondral bone by formation of a dense bony lamina. It is conceivable that delayed or defective formation of this bony lamina or breaching of it could render the deeper layers of the articular cartilage unduly vulnerable to noxious metabolic influences from the subchondral bone. The fact that both epiphyseolysis and articular cartilage necrosis occur about the time of puberty, suggests that the factor responsible for any such disordered sequence in osseo-cartilaginous maturation might be an endocrine one. This is supported by the high incidence of obesity or even a frank Fröhlich habitus in epiphyseolysis,[10] though in this condition, in which the impact is on the growth plate, there is a 2·4:1 male predominance; while in adolescent chondrolysis, which occurs almost exclusively in girls, it is the articular cartilage that is affected.

TREATMENT

Treatment remains disappointing. Much of the restriction of movement in the earlier stages, even in absence of pain, is due to muscle spasm, though complete organic stiffness may later supervene. This supports the contention of Lowe[4] that definitive measures such as arthroplasty or arthrodesis should not be resorted to prematurely. Prolonged abstention from weight-bearing certainly resulted, in his experience, in a better recovery of movement than the virtually complete restriction attending earlier resumption of ambulation in the present series. In view of the possible importance of an endocrine factor a course of oestrogens was administered to case 9 but without apparent effect. However, in this same case the early relief of muscle spasm and improvement in movement range following varus-derotation osteotomy suggests that this measure should receive further consideration.

SUMMARY

This paper describes 9 cases of a hip joint condition occurring around puberty, almost always in females and predominantly in non-Whites. The onset is insidious, the symptoms are mild, and the course, usually run within a year, is characterized by osteoporosis, decreasing joint space, a degree of protrusio acetabuli, premature fusion of capital and trochanteric growth plates and progressive loss of movement often terminating in complete stiffness.

Pathologically, chondrocyte damage in the deeper layers of the articular cartilage occasions a reparative process in the form of focal invasions from the subchondral bone and marginal synovial encroachment with deposition of fibrocartilage.

While there is no evidence of a tuberculous, other infective or auto-immune process, the condition is closely similar to, if not identical with, the articular cartilage necrosis associated with some cases of epiphyseolysis and described by Waldenström and others.

The aetiology is discussed but remains speculative, and the treatment is unsatisfactory.

I should like to thank Dr F. L. S. Visser, Medical Superintendent of the Wynberg, Orthopaedic and Convalescent Hospitals, and Prof. C. E. Lewer Allen, Professor of Orthopaedics in the University of Cape Town for permission to publish this paper; and Mr R. C. de Meneaud for the photographs.

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