NEUROPATHIC OSTEONECROSIS OF THE LATERAL FEMORAL CONDYLE IN CHILDHOOD

A REPORT ON FOUR CASES

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Four children are described, each with spontaneous osteonecrosis affecting nearly one-third of the lateral femoral condyle. All the children had a motor and a sensory deficit in the affected limb: two had been previously treated for neuroblastoma of the spine, one for an infected lumbar dermoid cyst and one had spina bifida. We consider that these disorders, singly or in combination, may lead to repeated excessive loading of the lateral femoral condyle, which cannot be appreciated in a knee that is not protected by normal sensation.

Osteonecrosis of the lateral femoral condyle in childhood has not previously been described. We report four children with this condition between the ages of six and 14 years seen at The Hospital for Sick Children between 1981 and 1983. All the patients had a motor and a sensory deficit in the affected limb. The presenting features and radiological appearances are described and some comments made as to the possible aetiology.

Case 1. This girl had a spinal neuroblastoma at the level of T12 successfully treated by surgery and radiotherapy at the age of one year. The residual paralysis was asymmetrical, affecting predominantly the right leg, and she ultimately developed a severe calcaneovalgus deformity of the right foot and needed to wear a caliper. Sensation to pinprick and light touch were diminished from the right knee downwards.

At the age of six she presented with a 12-month history of swelling of the right knee, and examination revealed a moderate effusion. She complained of some discomfort in the knee but pain was not a dominant feature. Radiographs showed fragmentation of the posterior third of the lateral femoral condyle, which appeared hot on a technetium scan (Figs 1 to 3). At arthroscopy the lateral meniscus was found to be shredded and whilst the lateral femoral condyle looked superficially normal, a deep fissure extending into subchondral bone was discovered when the medial aspect of the condyle was explored with a probe. An open lateral meniscectomy was performed and she was managed in an

above-knee caliper in order to prevent weight-bearing on the damaged posterior portion of the lateral femoral condyle.

Eighteen months later she still had a moderate effusion. Radiographs showed collapse of the involved portion of the lateral femoral condyle, and examination revealed some valgus laxity when the knee was examined at 30° of flexion.

Case 2. A spinal neuroblastoma at the level of T10 was diagnosed in this boy soon after birth and he was treated with radiotherapy. There was no residual motor deficit in his right leg but in the left there was weakness, mainly below the knee. A valgus deformity of the left foot was treated at the age of seven years by a Grice stabilisation procedure, but he still had a mild foot drop. There was patchy sensory loss involving the left leg from the hip downwards and also the back of the right buttock; this loss did not follow a dermatome pattern and was similar to the pattern of sensory loss seen in spinal dysraphism.

At the age of 11 he presented with a five-week history of an effusion in the left knee, which appeared after a game of rugby football. There was little pain. Careful examination disclosed patchy sensory loss in the affected limb. Radiographs showed some collapse of the posterior third of the lateral femoral condyle (Figs 4 and 5). At arthroscopy the articular cartilage overlying the lesion was intact but softened, and the lateral meniscus appeared normal. There was a low-grade synovitis. Like the first patient he was treated in a long leg caliper and remained active, but the effusion increased. Nineteen months later a loose body was palpable in the suprapatellar pouch. This was removed at a second arthroscopy during which a large irregular crater was noted at the back of the lateral condyle. Radiographs at this time showed loss of the posterior part of the lateral femoral condyle (Figs 6 to 8). There was a posterior horn tear of the lateral meniscus which was trimmed. Postoperatively the effusion diminished and his knee remains painless. It is stable in full extension, but in 30° of flexion there is

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Fig. 1

Fig. 2

Fig. 3

Case 1. Figures 1 and 2. Radiographs of the right knee showing fragmentation of the lateral femoral condyle. Figure 1—Anteroposterior view. Figure 2—Lateral view. Figure 3—Technetium scan showing increased uptake mainly in the lateral femoral condyle of the right knee.

Fig. 4

Fig. 5

Case 2. Figures 4 and 5—Anteroposterior and lateral views of the left knee showing fragmentation and collapse of the posterior part of the lateral femoral condyle. Nineteen months later (Figs 6 to 8) the posterior part of the lateral femoral condyle has been shed as a loose body and is lying in the postero-lateral compartment of the knee, above its original position. Figure 6—Anteroposterior view. Figure 7—Lateral view. Figure 8—“Tunnel” view.

Fig. 6

Fig. 7

Fig. 8

marked valgus laxity due to the loss of the articular surface.

Case 3. This boy developed an increasing paraplegia at the age of two years due to an infected dermoid cyst, associated with spina bifida at the lumbosacral level. Whilst he made some recovery he was left with saddle anaesthesia, patchy sensory loss in both legs, and drop foot with valgus deformity in both feet—worse on the right.

At the age of 13 a painless effusion developed in his right knee after a game of rugby football; radiographs showed an avulsion fracture of the upper pole of the patella and changes in the lateral condyle (Figs 9 and 10). No specific treatment was instituted. The effusion settled but recurred a year later when it was noted that he had developed a valgus deformity of 15°. Radiographs showed advanced changes of osteonecrosis of the lateral femoral condyle, best seen in the "tunnel view" (Fig. 11). Arthroscopy revealed a tear of the lateral meniscus, which was trimmed, a large crater at the back of the lateral condyle and a large loose body lying in the posterolateral compartment of the knee. At a subsequent operation the loose body was removed and a supracondylar osteotomy of the femur performed with slight overcorrection of the valgus deformity so as to unload the lateral compartment of the knee.

Case 4. This boy was born with a lumbar meningomyelocele. As a result, there was slight weakness of the quadriceps and hamstrings on both sides. With the exception of the calf muscles there was little muscle power below the knees. He retained sphincter control but there was sensory impairment involving both feet. He achieved a good functional walking pattern with no aids. Bilateral lengthening of the calcaneal tendon was performed at the age of five, and at the age of eight he had a Grice subtalar fusion on the right.

Later that year he presented with a one-month history of spontaneous onset of effusion, slight pain, and deterioration of performance in his right knee. A radiograph showed a depressed fracture of the lateral tibial

plateau, which healed leaving a residual valgus deformity. Despite this he regained his previous level of activity but, at the age of 11, he presented again, this time with mild pain in both knees after exercise and increasing difficulty when walking. Both knees had developed a fixed flexion deformity of 30° and the lateral radiograph of the right knee now showed collapse of the posterior part of the lateral femoral condyle. He underwent bilateral supracondylar extension osteotomies, with a good result on the left. The result on the right was disappointing because of continuing weight-bearing on the collapsing lateral femoral condyle, which eventually became painful. At this stage the boy was referred to us for a second opinion.

DISCUSSION

As far as we know, neuropathic osteonecrosis of the lateral femoral condyle has not previously been reported in childhood. The four children whom we describe all had pathology of the spinal cord and a pre-existing neurological deficit, with a sensory component, in the affected limb. Whilst only one knee was truly painfree, pain did not dominate the clinical presentation in any of these children, which is surprising in view of the degree of joint destruction. These are not true Charcot knees but in view of the accompanying neurological deficits we consider the term "neuropathic" to be justifiable. Despite their disability, all four children were leading an active and independent life. An effusion in the knee of such a patient, associated with little or no pain, should alert the surgeon to the possibility of this condition.

As this lesion involves the posterior third of the lateral femoral condyle, the radiological diagnosis is most readily made in the "tunnel view". The early appearance of fragmentation is followed by collapse and ultimately shedding of the posterior portion of the condyle into the joint as a large loose body. Once collapse has occurred, a step in the outline of the lateral condyle can be seen in a lateral radiograph of the knee. Whilst the early lesion bears a superficial resemblance to osteo-

Fig. 9

Fig. 10

Fig. 11

Case 3. Radiographs of the right knee showing avulsion fracture of the upper pole of the patella, and early changes in the lateral femoral condyle. Figure 9—Anteroposterior view. Figure 10—Lateral view. Figure 11—One year later the "tunnel" view shows loss of the lateral femoral condyle.

chondritis dissecans of the lateral femoral condyle (Aichroth 1971; Linden 1977) or variations in the normal pattern of ossification (Caffey *et al.* 1958), it should be distinguished by its magnitude, location, and the type of patient in whom it occurs. Technetium scans were performed in two of our patients and each demonstrated a hot lateral femoral condyle. We presume that the increased uptake occurs in a vascular junctional zone, adjacent to the plane of separation of the dead fragment.

Arthroscopy provided useful information both on the state of the condyle and on the lateral meniscus. For example, the arthroscopy in Case 1 revealed an early stage of the disease. The posterior condyle looked superficially normal and was still held in place by intact healthy articular cartilage, except on its medial aspect where a deep cleft was discovered with the probe. It should be technically possible to stabilise the fragment at this stage with a procedure similar to that which can be used for osteochondritis dissecans, but whether healing would occur in a neuropathic knee is uncertain. Furthermore, should healing be achieved would the condyle be vulnerable to further damage? A large irregular crater on the posterior femoral condyle was clearly seen in two patients when late arthroscopy was performed for removal of the shed fragment.

The cause of osteonecrosis in these patients is unknown. In the typical Charcot knee osteonecrosis is extremely rare before skeletal maturity is attained, and it is typically the medial femoral condyle that is first involved (Eichenholtz 1966). Spontaneous osteonecrosis in middle life also shows a predilection for the medial femoral condyle (Ahlbäck, Bauer and Bohne 1968). Steroids and cytotoxic agents, which are known to cause similar changes in otherwise normal knees (Aichroth 1971), had not been administered to any of our patients. A search through the Fairbank collection of radiographs at the Royal National Orthopaedic Hospital revealed only one similar patient, aged 11, in whom the diagnosis was congenital insensitivity to pain (Abell and Hayes 1964; Brunt 1967; Yoslow *et al.* 1971). We have not found any reference to this pattern of osteonecrosis in the literature on spina bifida (Dupre and Walker 1972; Sharrard 1975; Parsch and Manner 1976; Menelaus 1980).

In order to explain this lesion we have considered the vascular anatomy of the femoral condyles and the possible roles that may be played by trauma, abnormal loading and meniscal injury in a neuropathic knee. The affected portion of the lateral femoral condyle is entirely intra-articular and has no soft tissue attachment. It depends entirely on the condylar vessels for its blood supply. The intra-osseous condylar arteries fan out towards the joint surface, their vascular territories being separate, and very few anastomoses exist (Scapinelli 1968). The subchondral bone is therefore vulnerable to ischaemia if these vessels are damaged.

Despite the fact that two of our patients had sus-

tained fractures at other sites in the knee (one lateral tibial plateau, and one upper pole of the patella) with no specific history of injury, it seems unlikely that the lateral condylar lesion represents an acute osteochondral fracture. In the two patients in whom arthroscopy was performed early there was no contusion of the articular cartilage, no evidence that there had been a haemarthrosis, and the involved segment was not separated from the lateral condyle. The associated meniscal lesion in three patients appeared to be secondary to the osteonecrotic lesion, rather than a predisposing factor. In Case 2, where the lateral meniscus was normal at the first arthroscopy, a tear was seen 19 months later. In our view the most likely explanation for this lesion is abnormal loading of the knee. It may be significant that two of our patients walked with the affected leg in lateral rotation at the hip. In addition three patients had a valgus deformity of the hind foot, which had been stabilised surgically in two patients and with a caliper in one. In one patient (Case 4) valgus deformity of the knee was noted before the condylar lesion. These deformities, singly or in combination, may lead to repeated excessive loading of the lateral femoral condyle, which cannot be appreciated in a knee that is not protected by normal sensation.

Our treatment of these knees has not been satisfactory. Protection of the knee in a caliper does not prevent progression of the lesion and eventual shedding of the condylar fragment into the joint as a loose body. In the single patient who had a valgus deformity of the knee in full extension, a supracondylar osteotomy with slight overcorrection reduced the effusion, but the damage to the condyle was clearly irreparable. In the early case there may be a place for stabilisation and drilling of the necrotic fragment in the hope that healing may occur.

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