

# Neuropathic osteonecrosis of the knee in childhood

## Two cases of myelomeningocele

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Two girls, aged 11 and 7 years, with myelomeningocele had osteonecrosis in the lateral tibial epiphysis and in the medial femoral condyle, respectively. Both girls suffered from sensory impairment of the affected area and presented with warmth and swelling of the knee. No signs of infection were found. The necroses seen in radiographs were confirmed by biopsy and <sup>99m</sup>Tc-MDP scintigraphy. The lesions showed only little tendency to heal, and rather severe growth disturbances developed despite treatment in long leg calipers. One of the girls was unable to walk because of the loss of muscular power during the long period of bracing.

Four cases of neuropathic osteonecrosis of the lateral femoral condyle in childhood were reported recently (Citron et al. 1986). We have treated epiphyseal osteonecrosis of the knee in two myelomeningocele patients.

### Case reports

*Case 1.* An 11-year-old girl presented with a swollen left knee with no history of trauma. She was born with a L4 lesion from a lumbosacral myelomeningocele with anesthesia of the buttocks, feet, and lateral aspects of the lower legs and hypesthesia in the remainder of the area below the knee. She had reduced hamstring power and a total motor deficit more distally. Malpositions of the feet were treated by soft-tissue interventions and a Crice subtalar fusion on the right side. At the age of 3, she suffered from a traumatic epiphysiolysis of the left distal tibia with ensuing physeal closure and shortening of the

leg, and an epiphysiodesis of the right tibia was therefore performed at the age of 10. When 8 to 9 years old, she had several injuries of her right lower limb with fractures of the tibia. Because of dysplasia and valgus deformity of the ankle joint, she had a supramalleolar osteotomy performed.

The girl presented with no pain, but had tenderness on palpation of the medial capsular border. Radiographs showed a defect in the upper lateral part of the tibial epiphysis with surrounding sclerosis (Figure 1). Aspiration of the knee gave 50-80 ml of clear, slightly hemorrhagic joint fluid. Microscopically, only a few erythrocytes and no bacteria were found, and cultures were negative. The ESR was 17 mm/h, and the Yersinia titer, latex agglutination test, and Waaler-Rose tests were negative. Leucocytes and differential counts were in the normal range. An arthrotomy revealed a frayed and degenerated lateral meniscus and a 1×1.5 cm area of softened, loose articular cartilage overlying a subchondral bone defect, which corresponded to the radiographs. Histologic examination showed fragmented bone with necrosis, scattered new bone formation, and granulation tissue.

The girl was treated in a long leg cast followed by a long leg caliper for 2.5 years during which time the knee was continuously slightly warmer than the right knee, albeit with no joint effusion. The bone defect healed slowly forming a prominence of the lateral tibial condyle and a fusion

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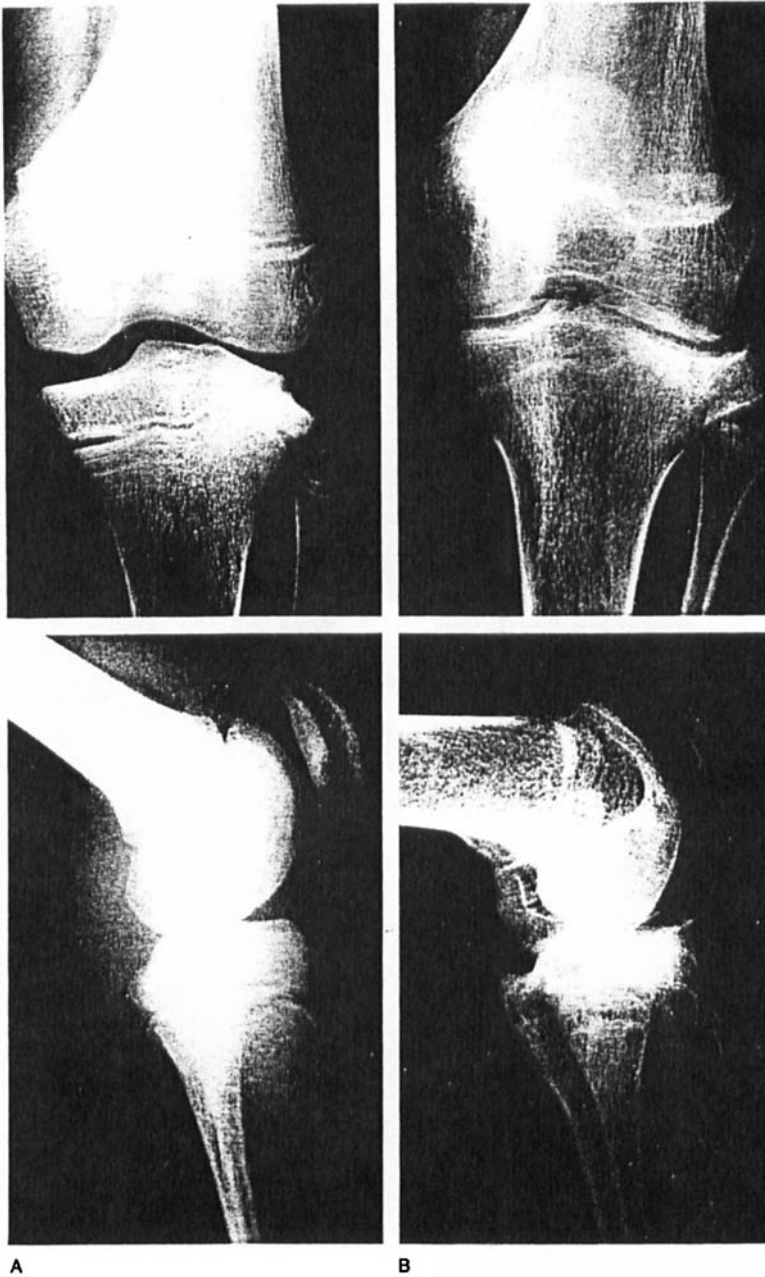
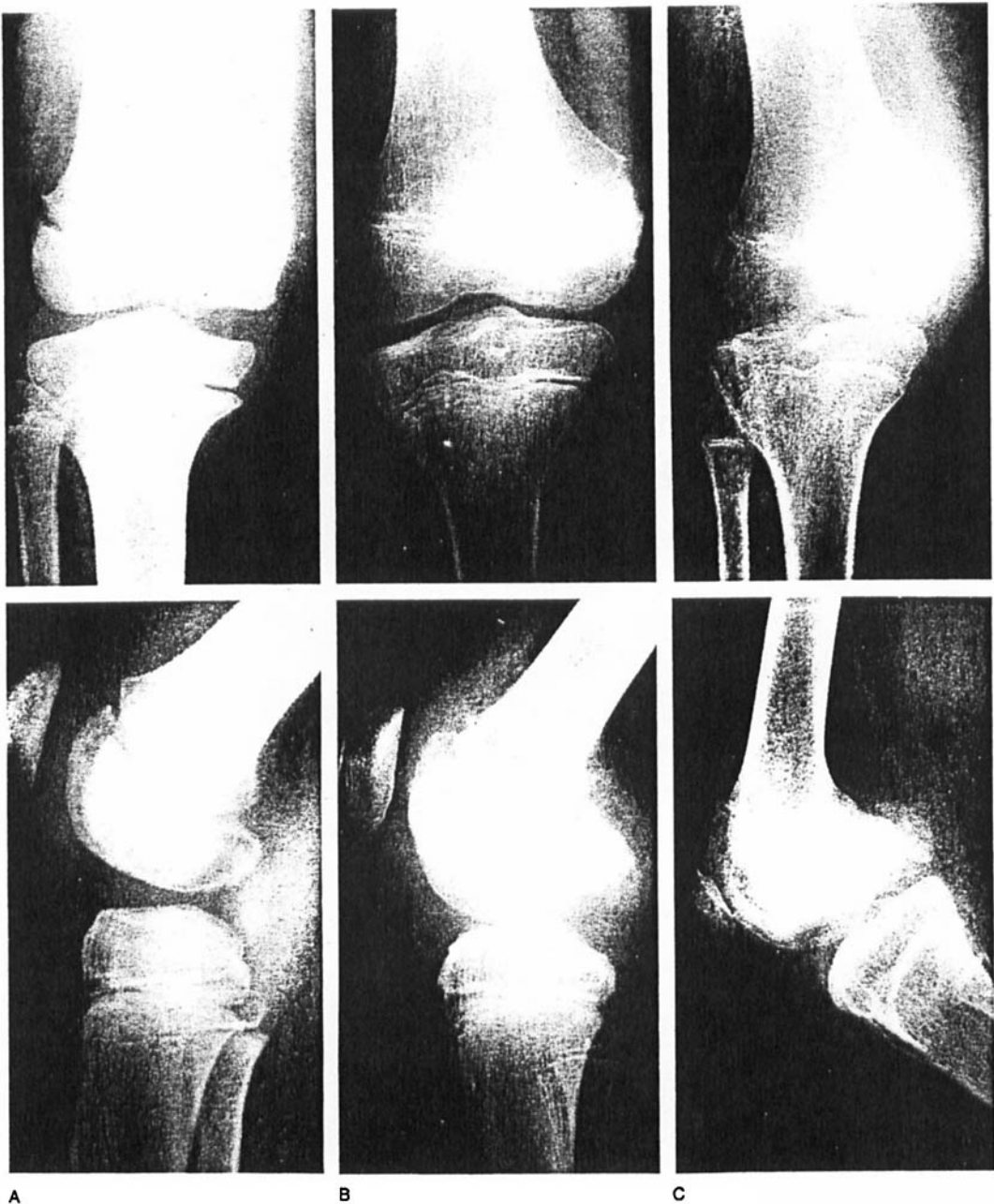


Figure 1. Case 1. 11-year-old girl with myelomeningocele.  
 A. At presentation. The lateral tibial condyle has a defect surrounded by a sclerotic halo  
 B. 5 years later.

of the lateral third of the physis creating a minor valgus deformity (Figure 1). At this time, she had no complaints and walked safely without external support. The knee was stable with no signs of inflammation and had a range of motion of 15° to 150°.

*Case 2.* A 7-year-old girl presented with a warm and swollen right knee with no history of trauma.

She was born with an L4 myelomeningocele with symmetric sensory impairment below the knee level and loss of muscle power predominantly affecting the flexors. She walked without external support at the age of 2 using low leg calipers. Muscle imbalances creating malpositions of the feet were corrected by soft-tissue operations at the age of 3 to 6 years. She had no pain and the



Case 2. 7-year-old girl with myelomeningocele.  
A. At presentation. An osteolytic process in the medial and part of the lateral femoral condyle.  
B. After 1.5 years. Healing deficiency and ossification disturbances.  
C. 5 years later.

radiographs showed no fractures; a large osteolytic process in the medial femoral condyle was found (Figure 2). Based on the former observation, it was interpreted as an osteonecrosis. The

ESR was 12 mm/h and  $^{99m}\text{Tc}$ -MDP scintigraphy showed a slightly widened physis and a high activity in the epiphysis. Despite nonweight bearing for 1 month, the condition remained un-

changed; and because radiographic progression was observed, the knee was immobilized in a long leg cast for 2 additional months followed by a long leg caliper for 4 years. During this time, intermittent effusion, warmth, and an extension lack of 10°-15° were observed. Radiographically, the lesion slowly progressed with disturbed ossification and a minimal healing tendency (Figure 2). Aspiration of the knee produced 45 ml of clear, yellow fluid in which neutrophils and synovocytes predominated, whereas lymphocytes and erythrocytes were scarce. No crystals or bacteria were found, and the culture specimens including TB were negative. The Yersinia titer was normal and the ESR was 15 mm/h. After 4 years of bracing, the patient got a wheel chair because of a plantar sore, and she did not regain her former walking ability, which was seemingly due to severe loss of quadriceps muscular power on the right. There was no knee effusion and motion was 5°-135° with no laxity.

## Discussion

Our cases had conspicuous warmth and swelling giving rise to a suspicion of infection or fracture. However, no signs of infection were found; the synovial fluid showed only a low-grade synovitis in one of the cases. In our first case a biopsy confirmed the diagnosis of osteonecrosis. In the second case the radiologic pattern of a process in the weight-bearing area of the medial femoral condyle was consistent with bone necrosis as described by Ahlbäck et al. (1968); the high uptake in the <sup>99m</sup>Tc-MDP scintigraphy further confirmed the diagnosis (Habermann & Hartzband 1984), Rozing et al. 1980, Williams et al. 1972).

The secondary bone necrosis accompanying a number of clinical conditions, such as trauma, caisson disease, hemoglobinopathies, and alcohol abuse, is well known. Bone necrosis has been described as a complication of corticosteroid

therapy either alone or combined with chemotherapy and even associated with the last group of drugs alone (Marymont et al. 1986). The cause of the "spontaneous" osteonecrosis of middle life (Ahlbäck et al. 1968) is unknown. According to Lotke et al. (1977), microtraumas create fractures and collapse of subchondral bone leading to leakage of articular fluid into the marrow space with bone necrosis as a result.

Citron et al. (1986) stressed the anatomic arrangement of the separate condylar arteries with only a few anastomoses. Grönlund et al. (1984) found hypoxia of the subchondral bone as a result of an acute increase in the intraarticular pressure in the rabbit knee. Compression of the epiphyseal vein system with its intraarticular course is suggested as an important factor. Finally, a nervous control of the hemodynamics is assumed resulting in circulatory disturbances if the spinal cord is damaged (Tøndevold 1983, Chantraine et al. 1979).

Patients with myelomeningocele and other neuropathic conditions affecting the lower limbs have all of these predisposing factors. The lack of sensitivity gives a tendency to repeated traumas and flow changes that probably also arise from the joint effluents increasing the intracapsular pressure. Further, these patients are prone to some degree of osteoporosis due to vascular changes and diminished activity. Citron et al (1986) called the condition neuropathic in order to separate it from spontaneous osteonecrosis. Even if the latter lesion is mainly located in the weight-bearing area of the medial femoral condyle, other locations have been seen, including the medial tibial plateau (Ahlbäck et al. 1968, Rozing et al. 1980, Marmor 1982).

As observed by Citron et al. (1986), treatment in a long leg caliper does not prevent progression of the lesion, and healing takes place at a slow rate producing changes of ossification. In our cases the function of the knee joint became acceptable. However, in these patients a severe loss of muscular power may result from prolonged bracing.

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