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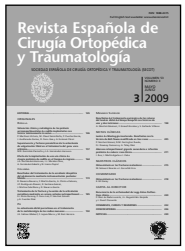

LETTERS TO THE EDITOR

Recurrence of Legg-Calvè-Perthes's disease. A case report

Recurrencia de la enfermedad de Legg-Calve-Perthes. Caso clínico

Legg-Calve-Perthes disease is a relatively common condition in the pediatric population. However, recurrence of the disease in a previously affected amd gully recovered hip is a rare occurrence.

The etiopathogenesis of this disease still remains unclear. For the condition to develop, the femoral head must experience an ischemic process leading to its destruction. There is no consensus as to what causes this ischemic process given that none of the hypotheses put forward (thrombosis of the vessels in the head because of an increase in intraepiphyseal pressure, embolism-derived interruption of blood flow) have been demonstrated.

We hereby present the case of a 4 year and 5 month-old male with a 4-month history of knee pain without symptoms of infection. Physical examination revealed a relative limitation to internal rotation of the left hip, with no other limitation affecting that limb. Radiographs showed a proximal femoral epiphyseal alteration compatible with left hip necrosis (fig. 1). The child did not present with any sign that may indicate bone dysplasia. On the basis of these clinical and radiological findings a diagnosis was made of Legg-Calve-Perthes disease. Conservative treatment was administered in the form of non-steroid anti-inflammatory drugs and an abduction Thomas splint for 3 years. The patient was followed up both clinically and radiologically. At 3 years, the child was asymptomatic and Imaging studies showed a favorable evolution: complete remodeling of the femoral head had taken place (head morphology was classified as class I according to the classification by Stulberg[1]. The patient was discharged and could lead a normal life.

When the child was 8 years old (one year after discharge) he presented to us again, this time with a limp and a 2-month history of pain in the left knee. Examination of his left hip showed that he was capable of 100° flexion, –30° extension, 10° adduction, 10° abduction, 0° internal rotation and 20° external rotation. Radiographic study showed renewed destruction of the femoral head. Complementary tests (hemogram, leukocyte formula, coagulation tests, globular sedimentation rate and C-reactive protein) were normal.

A hip arthrography was performed to determine the condition of the hip joint and femoral head coverage. On the basis of these results, an adductor tenotomy was performed and an abduction splint applied for 3 weeks, with the aim of shifting the femoral head to its anatomic center, thereby optimizing its remodeling. Ten months later, follow-up x-rays showed progression of the bone destruction and a higher degree of hip extrusion.

At age 9, the child was asymptomatic and walked normally. Physical examination showed that he was capable of 120° flexion, 0° extension, 40° abduction, 25° adduction, 15° internal rotation and 30° external rotation. X-rays showed complete remodeling of the femoral head, although an anomalous configuration was already evident.

The patient was followed up until the end of this development. At the last follow-up visit he was aged 17. His range of motion was lower than in the contralateral hip and x-rays showed an irregular femoral head (class V on Stulberg's classification)[1] (fig. 2). Further to these results, he was discharged.

Discussion

In 1910, Legg, Calve, Perthes and Waldestrom described a disease consisting in necrosis and subsequent remodeling of the femoral head. In spite of the multiple studies carried out since, the cause of this condition remains unclear. The first hypotheses blamed the disease on repeated microtrauma, transient synovitis and even congenital dislocation. In 1973, Sanchís et al[2] suggested a model of bone multi-infarction, which was later borne out by the histologic studies of McKibbin and Ralis[3]. Current wisdom holds that the cause of the pathological changes affecting the joint is ischemia.

Although the origin of the disease is unclear, its relative frequency has allowed scholars to Estbaliz its natural history: destruction of the femoral head is followed by regeneration and remodeling. Long-term prognosis of the hip joint is determined by the remodeling process; the most promising cases are those in which a congruent and spherical femoral head is obtained at the end of follow-up. When remodeling does not produce a congruent and spherical head, early degenerative changes as well as a slight leg length discrepancy are usually observed. The capacity for bone remodeling is lost with age, which means that the youngest subjects are those with a better prognosis. On the other hand, if the disease presents late in life, prognosis for the hip is usually grimmer.

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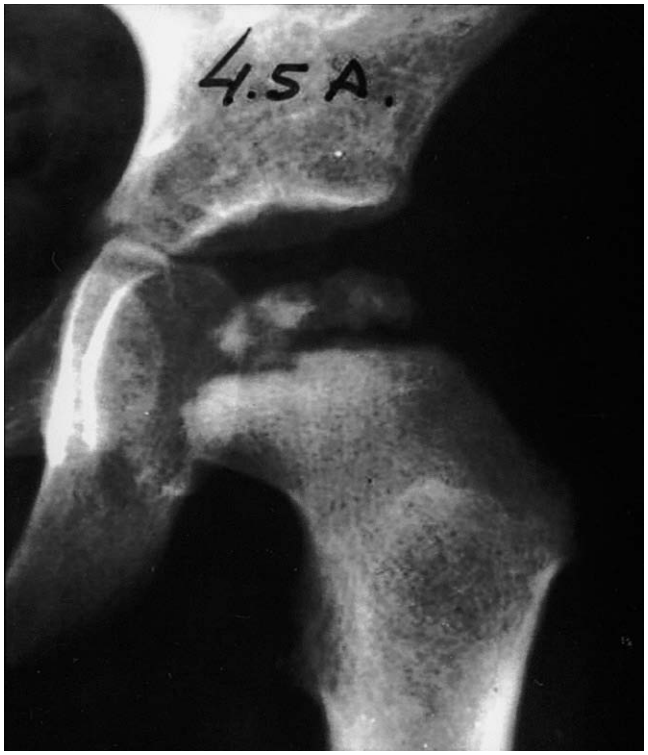

Figure 1 Anteroposterior hip radiograph of a 4-and-a-half-year-old patient showing necrosis and destruction of the lateral femoral head.

Recurrence of femoral head necrosis in Legg-Calve-Perthes disease and in Gaucher disease is exceptional: its incidence has been estimated at 0.25% of cases[4]. This phenomenon was first described in Legg-Calve-Perthes disease by Caffey[5] in 1961 and to date only 7 cases have been reported in the literature[1-8].

Functional prognosis of subjects with recurrence of Legg-Calve-Perthes disease is generally poor: hip morphology is not normal when the recurrence occurs; the second episode is normally more severe, the patient is older and therefore there is less remodeling capacity. These are probably the reasons why at the end of development these subjects show a flattened and elliptical hip. In the case described herein, after the first episode the child presented with a class I femoral head (Stulberg's classification[1]) and subsequently (at the end of development) presented with a class V femoral head, elliptical, incongruent, subluxed, hinge-shaped and with degenerative changes (fig. 2).

According to Stulberg, the morphology of the head at the end of development could be related to a greater presence of early degenerative changes[1], so it is to be expected that heads with a non-spherical and incongruent shape (class V) will present with arthritic changes before age 50.

Recurrence of the Legg-Calve-Perthes disease is a therapeutic challenge. The right indications and surgical techniques may not produce good functional results. More intensive treatment of the second episode would produce greater femoral coverage and may therefore give rise to a more congruent hip joint.

In a nutshell, although Legg-Calve-Perthes disease is fairly common in the pediatric population, the literature only contains 7 studies reporting the recurrence of the condition. The second necrosis and the destruction of the proximal femoral epiphysis result in a poorer short-term functional prognosis, both because they provoke a second reconstructive process on an already affected hip, and because they occur in older subjects with weaker remodeling capacities.

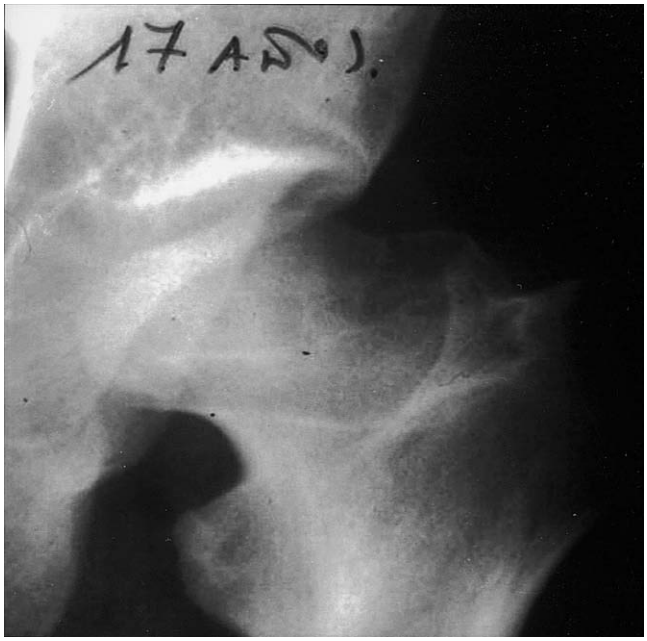

Figure 2 Final result: flattened and elliptical femoral head.

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