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CASE REPORT

Case Reports

Legg-Calvé-Perthes Disease in Czech Archaeological Material

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Abstract Legg-Calvé-Perthes disease (osteochondrosis of the femoral head) has been recognized in archaeological material for nearly a century but is extremely rare. We describe two Czech cases from archaeological findings. The first case was diagnosed in the skeleton of a man older than 50 years with the left hip affected. The skeleton was in grave Number 2 of the Langobard cemetery at Lužice (Moravia) and dated to the end of the fifth century and the beginning of the sixth century AD. The second case was described by J. Chochol in 1957 on the left femur and half of the pelvis of a skeleton from an archaeological investigation in Brandýsek (Bohemia), ninth to tenth centuries AD. Using the diagnostic criteria of Ortner and Putschar, we excluded slipped capital femoral epiphysis in both cases. We discuss the differential diagnosis of Legg-Calvé-Perthes disease versus unilateral and bilateral osteochondroses of the femoral head in archaeological and current clinical material.

Each author certifies that he or she has no commercial associations (eg, consultancies, stock ownership, equity interest, patent/licensing arrangements, etc) that might pose a conflict of interest in connection with the submitted article.

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Introduction

Osteochondrosis of the femoral head, known as Legg-Calvé-Perthes disease, generally is believed to be related to avascular (aseptic, ischemic) necrosis of subchondral bone of the femoral head. The disease initially was described by three authors independently in 1910: Arthur T. Legg in Boston [8], Jacques Calvé in France [1], and George Perthes in Germany [15]. In 1909, Hening Waldenström described the radiographic characteristics of the disease, but he presumed it was a form of bone tuberculosis [24].

Although Legg-Calvé-Perthes disease has been recognized in archaeological material for a century, the entity and slipped capital femoral epiphysis rarely have been observed. Ortner and Putschar [14] reported on a right femur from the Valley of Chicama in Peru as a possible example of Legg-Calvé-Perthes disease. The archaeological age of their case is unknown. In a review of the paleopathologic bibliography [3], we identified only one case of Legg-Calvé-Perthes disease [13].

We describe two cases of archaeological Legg-Calvé-Perthes (osteochondrosis of the femoral head) using the diagnostic criteria of Ortner and Putschar [14], and discuss the differential diagnosis of Legg-Calvé-Perthes disease against unilateral and bilateral osteochondroses of the femoral head in archaeological material and the clinical situation.

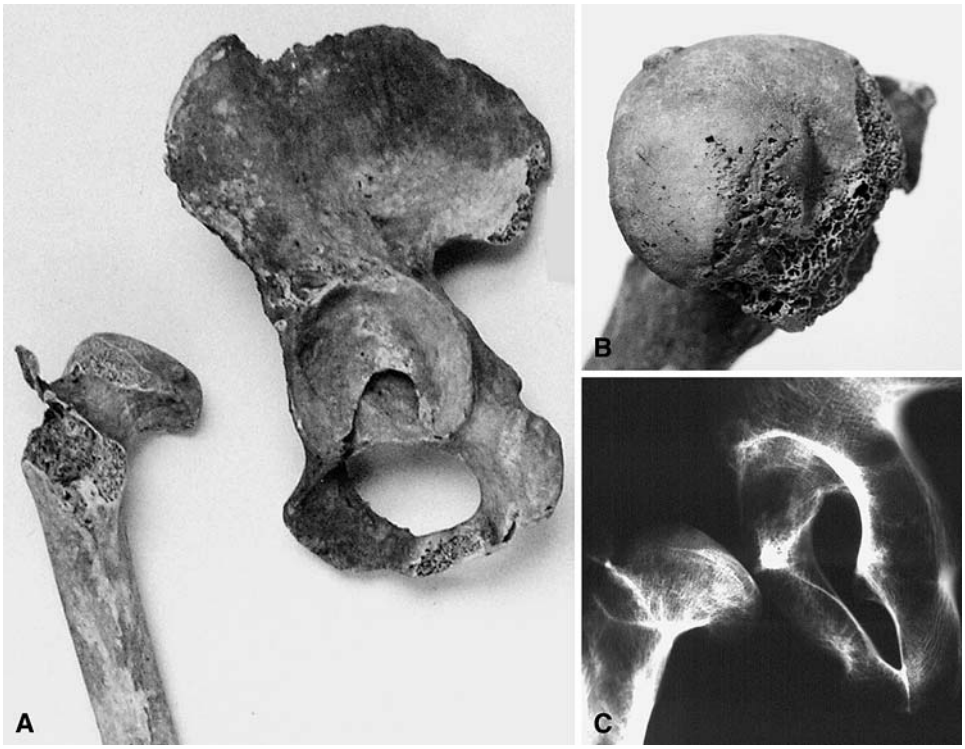

Fig. 1A–C (**A**) A photograph shows a posterior view of the left hip and femur of the archaeological skeleton from grave Number 2 of the Langobard cemetery at Lužice (South Moravia). (**B**) This photograph shows a medial view of the flattened left femoral head. (**C**) A radiograph of the left hip (anterior view) shows a steep acetabulum with sclerotic structure of the bone tissue in the front part of the facies lunata as a result of excessive functional loading. The beak-shaped head is elongated in the medial direction, flat, and big. The neck is relatively short and thick, and the greater trochanter is higher than the center of the head. The bone structure of the Adams' arch and the trabecular structure of the ventromedial quadrant of the head are sclerotic.

Archaeological Material

Case 1

This case came from the skeleton of a man older than 50 years [5, 26]. The skeleton was in grave Number 2 of the Langobard cemetery at Lužice in South Moravia and dated back to the end of the fifth century and the beginning of the sixth century AD. This cemetery is the largest discovered cemetery from the so called "Migration Period" in the region during which the migration of peoples lasted approximately from 375, when the advances by the Huns started a chain reaction of displacement of other tribes, first in the region north to the Black Sea through the lower and middle Danubian basin and further into Europe; at the same time, the Roman Empire was declining and contracting. The end of this period corresponds to the end of the sixth century in Middle Europe [19]. In the 1980s, the Archaeological Institute of the Academy of Sciences in Brno excavated 120 skeletal graves. The anthropological material was deposited in the National Museum in Prague.

The left femoral head of this case was flat with edges widening into a mushroom shape (Fig. 1A). The medial third of the head was separated by a groove (Fig. 1B). The groove was deeper on the front side and shallower in the dorsal direction. At the medial edge of the femoral head, the curvature had been formed into a beak shape and there are osteophytes (Fig. 1A). There was no substantial dislocation of the center of the femoral head from the axis of the shortened and thickened femoral neck. Only part of the greater trochanter remained and was evidently at a higher position than the center of the head.

The head of the right femur was not preserved. Diaphyses of both femurs had the same shape and width. The left acetabulum was markedly shallower in comparison to the right; in diameter and width of facies lunata, it was larger by 1 cm. The rear edge of the acetabulum was more rounded than is seen on the right side. There were no traces of neoacetabulum in either the proximal or dorsal direction (Fig. 1A).

A radiograph of the left hip (anterior view) shows a steep acetabulum with sclerotic structure of the bone tissue in the front part of the facies lunata as a result of excessive functional loading. The beak-shaped head was elongated in the medial direction, flat, and big. The neck was relatively short and thick and the greater trochanter higher than the center of the head. The bone structure of the Adams' arch and the trabecular structure of the ventromedial quadrant of the head were sclerotic (Fig. 1C).

Case 2

Deformation of the left hip was diagnosed as Legg-Calvé-Perthes disease in the skeleton of an adult man [2] at grave Number 9 of the archaeological investigation Kytlicová (1956), locality Brandýsek (Bohemia; 47 graves), ninth to

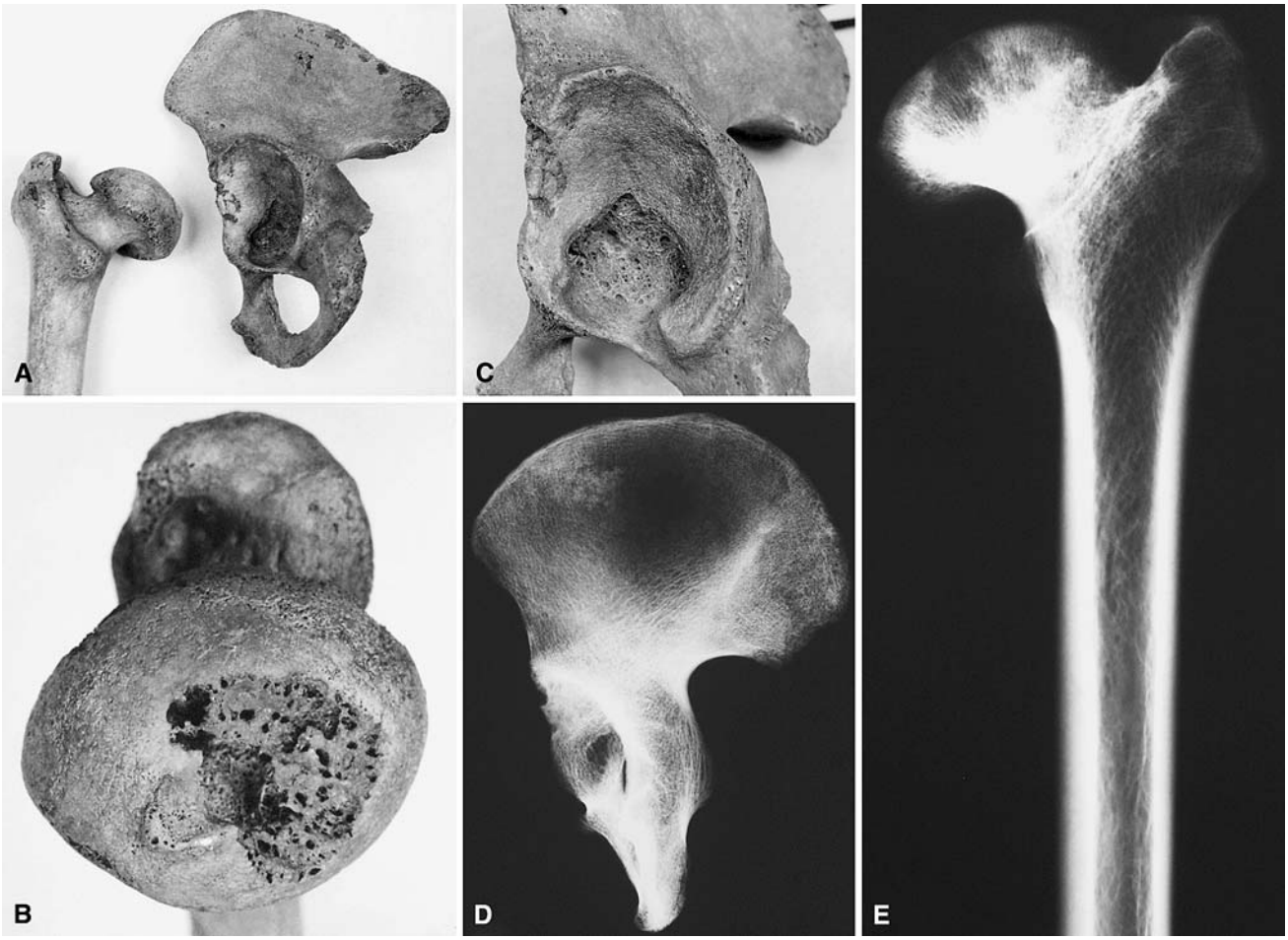

Fig. 2A–E (**A**) A photograph shows a posterior view of the left femur and the left half of the pelvis of the archaeological skeleton from grave Number 9 (Ao 1616) of archaeological investigation Kytlicová (1956), locality Brandýsek (Bohemia). Photographs show (**B**) a medial view of the postmortem-damaged and markedly flattened femoral head, and (**C**) the large and shallow acetabulum with a big acetabular notch. Radiographs of (**D**) the left half of the pelvis and (**E**) the proximal femur (anterior view) show a steep and flat acetabulum, considerable sclerosis of the neck, and extensive porous degeneration with thickening of the trabeculae of the head. The proximal epiphysis of the femur is fully fused, indicating adult age. The center of the head is below the level of the greater trochanter.

tenth century AD. The left femur and left half of the pelvis (Fig. 2A–C) of this skeleton were described previously [9].

Part of the markedly flattened femoral head in front of the fovea capitis femoris was damaged postmortem. On the proximal surface of the femoral head was a superficial groove corresponding to the margin of acetabuli (Fig. 2B). This suggests the head was subluxated. The margins of the joint surface of the head were characterized by exuberant bony overgrowth creating a mushroom-like appearance. The neck was relatively short, thick, and varus (Fig. 2A). There was no substantial dislocation of the center of the femoral head from the axis of the shortened femoral neck. The diaphysis, distal metaphysis, and surface of the femoral condyles were normal. The acetabulum was large and shallow with a large acetabular notch (Fig. 2C).

Radiographs of the left half of the pelvis and proximal femur (anterior view) showed a steep and flat acetabulum (Fig. 2D) with considerable increased density of the neck and extensive porous degeneration with thickening of the trabeculae of the head (Fig. 2E). The proximal epiphysis of the femur was fully fused, indicating adult age. The center of the head was below the level of the greater trochanter.

Discussion

The etiology of the disease is unknown, although there has been speculation regarding the potential role of traumatic, genetic, metabolic, nutrition, environmental, hormonal, and hematologic factors as causative [17]. Of the proposed theories of etiology, one seems most likely; there is some experimental proof the original occlusion of the precarious blood supply to the femoral head is caused by excessive fluid pressure of a synovial effusion in the hip, either inflammatory or traumatic. Approximately 5% of children with transient synovitis of the hip have the complication of Legg-

Calvé-Perthes disease develop. As far as the etiology is concerned, it is probable multiple factors come together in a constitutionally vulnerable child to cause the disease [13].

It is possible to differentiate between Legg-Calvé-Perthes disease and slipped femoral capital epiphysis in archaeological material [14]. Slipped femoral capital epiphysis represents a stress fracture between the metaphyseal side of the growth plate and the neck of the femur. This allows medial posterior and downward displacement of the head of the femur and usually leads to some degree of aseptic necrosis in the epiphyseal bone. Early severe degenerative arthritis modifies the appearance of osteochondrosis of the femoral head and can make the differentiation from the end stage of slipped femoral capital epiphysis difficult or impossible.

The shape of the head and acetabulum at skeletal maturity is the result of functional adaptation of bone tissue (bone remodeling) during growth and is dependent in part on proper blood supply to the femoral head. The degree of deformity of the femoral head depends on the age in which the original occlusion occurred [17]. The prognosis is good in children when onset occurs before the age of 5 years, fair in children when onset occurs from 5 to 9 years of age with more than half of the head involved, and poor in children when onset occurs after the age of 9 years. In general, the younger the child when affected, the wider and better the remodeling because of the length of time remaining until skeletal maturity for remodeling of the femoral head. It explains why girls appear to have a worse prognosis than boys because of the more advanced skeletal maturation at onset [25]. Based on these facts, both of the archaeological cases of severe residual deformities in the femoral heads we investigated appear to be those with late onset.

We excluded slipped femoral capital epiphysis in both cases based on the criteria for differentiating Legg-Calvé-Perthes disease [14] (Table 1). Archaeological Legg-Calvé-Perthes disease is typical with a big mushroom-shaped head, thickening, and usually shortening of the neck, a steep wide acetabulum, and, in adults, a premature porous lesion of the femoral head. Slipped femoral capital epiphysis is characterized by head center dislocation toward the neck axis, shortening and usually thickening of the neck, and new acetabulum at the ilium.

Table 1. The criteria for differentiating Legg-Calvé-Perthes disease (PD) and slipped femoral capital epiphysis (SFCE) in archaeological material (according to Ortner and Putschar [14])

Criteria	SFCE	PD	Case 1	Case 2
Big mushroom-shaped head	−	+	+	+
Head center dislocated toward neck axis	+	−	−	−
Thickening of neck	+/−	+	+	+
Shortening of neck	+	+/−	+/−	+/−
Steep wide acetabulum	−	+	+	+
New acetabulum at the ilium	+/−	−	−	−
Porous lesion of femoral head	−	+	+	+

There are other congenital and developmental hip afflictions with bilateral hip deformities simulating Legg-Calvé-Perthes disease. Bilateral femoral head deformities may occur with hypothyroidosis, infantile coxa vara, and various bone dysplasias and metabolic osteopathies [21] (eg, multiple epiphyseal dysplasia, spondyloepiphyseal dysplasia congenita and tarda types, spondyloepimetaphyseal dysplasia, mucolipidoses, mucopolysaccharidoses, and gangliosidoses [4, 6, 18, 20, 23]. Bone dysplasias (BD) are developmental disorders of chondroosseous tissue of the whole skeleton. Primary BD result from mutated genes that are expressed in chondroosseous tissue. Secondary BD are caused by abnormalities of extraosseous factors with secondary effects on the skeletal system such as metabolic disorders and hormonal disorders [10]. Hypothyroidosis, sickle cell disease, and infantile coxa vara are other causes of bilateral femoral head deformities [18, 22]. The causes of unilateral femoral head deformities usually are acquired (eg, avascular necrosis after reduction of developmental dysplasia of the hip, osteonecrosis after neonatal purulent coxitis and tuberculous/BCG/coxitis, after growth hormone, various drug treatments, hemophilia, and osteonecrosis of the femoral head in young adults resulting from idiopathic, posttraumatic, and postinflammatory influences [7, 11, 12, 16–18, 22]). Generally, the exact diagnosis of bone dysplasia and/or bone disorder from remnants of archaeological hip material and its radiographic examination is rarely possible. In clinical practice, the diagnosis of bone dysplasias from radiographs is possible only during the growth period in approximately 25% of cases [10].

Findings in archaeological remnants of the skeleton such as congenital and/or acquired deformities of the femoral head can be used for identification of persons living in the past, and/or in the present in forensic investigations of skeletons focused on missing persons. In these cases, the degree of severity of femoral head deformity can be helpful in the retrospective assessment of the onset of femoral head osteochondrosis.

In the cases presented, we excluded slipped femoral capital epiphysis. Although we cannot strictly exclude other causes of unilateral and/or bilateral osteochondrosis of femoral heads, we suspect these cases reflect Legg-Calvé-Perthes disease according to the criteria of Ortner and Putschar.

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