A probable case of Legg-Calvé-Perthes disease in Warring States-era China

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Abstract

Reports of Legg-Calvé-Perthes disease (LCPD) in the paleopathological literature are rare. Here, the authors present a probable case of LCPD, which presents as abnormal morphology of the proximal femur. The condition was observed in an individual of the Warring States period in Shaanxi Province, China, and the morphology involves a “mushroom head” deformity of the proximal right femur and an enlarged acetabulum, along with a contralateral tibia, talus, and navicular that are enlarged and demonstrate periosteal new bone formation. The authors consider tuberculosis, septic arthritis, trauma, slipped capital femoral epiphysis, and Legg-Calvé-Perthes disease in a differential diagnosis. The authors conclude that the most likely diagnosis for the deformity is Legg-Calvé-Perthes disease. Bony changes in the hip joint and contralateral lower leg suggest that the individual had an altered gait because of the condition.

Keywords

China, paleopathology, Legg-Calvé-Perthes disease, femoral head necrosis

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1. Introduction

Osteonecrosis of the femoral head is rarely observed in the archaeological record (Smrcka et al., 2009). The condition can cause severe deformity of the hip joint in the absence of modern surgical intervention, and can involve sequelae such as infection and osteoarthritis. The case presented here involves osteonecrosis and deformity in the hip of a young adult male individual. The individual also exhibits an altered acetabulum and extensive bony changes in the lower legs, indicating that the hip deformity affected the individual's gait. The authors present a differential diagnosis of this condition and discuss its significance.

2. Case description

Individual SHSM22 is a male (Buikstra and Ubelaker, 1994; Kiales et al., 2012; Liu, 1989) aged 20-29 at death (Brooks and Suchey, 1990; Buikstra and Ubelaker, 1994; Iscan, 1991; Langley-Shirley and Jantz, 2010; Lovejoy et al., 1985), interred during the Warring States period (475-221 BCE) in the Shijiahe cemetery, in what is now northern Shaanxi Province, China. The cemetery was used, possibly simultaneously, by two groups: low status subjects of the State of Qin (N=10), who were likely moved into the area after its conquest by Qin; and a group known in historical sources as the “Rong” (N=24), who textual, archaeological, and bioarchaeological sources agree migrated from semi-arid Western China, allied themselves with Chinese states, and settled within the states' borders (Shaanxi, 2013; Sun et al., 2015). The Rong remains show a small but measurable biological distance from the Qin, though skeletal measures of their community health are nearly identical and suggest they had similar lifeways (Berger, dissertation in progress). The Qin graves have side burial chambers and are positioned around the margins of the cemetery. SHSM22 came from one of the Rong graves, in the densest area of the cemetery.

The remains were examined macroscopically at the Northwest University School of Cultural Heritage in Xi'an; radiographs were not obtainable. The right femoral head of this individual is deformed, exhibiting a "mushroom" shape. The head is flattened (Figure 1), and the fovea capitis is located at the medial inferior margin of the head. The greater trochanter is also enlarged and shows extensive periosteal new bone formation. The corresponding acetabulum is enlarged, especially on the lateral margin, with a loss of sphericity (Figure 2). No evidence of infection of the bone, or callus formation indicative of a fracture, are visible.

Figure 1 Posterior view of SHSM22 proximal right femur with "mushroom" deformity (A) and periosteal new bone on the greater trochanter (B). The opening below the femoral neck is postmortem damage.

Figure 2 Antero-inferior view of ossa coxae of SHSM22, showing enlarged lateral margin of the right acetabulum (arrow).

The tibial shafts exhibit some striated appositional bone, both woven and remodeled, resembling healing periosteal new bone. The left (contralateral) tibia is larger than the right in both length and diameter (right tibia maximum length 351 mm, maximum distal epiphyseal breadth 46.21 mm; left tibia maximum length 372 mm, maximum distal epiphyseal breadth 54.51 mm), and the distal left tibia has appositional bone on the surface (Figure 3). The left talus and navicular bone are also enlarged, with undulating solid surfaces, and lipping, faint porosity, and osteophytes (Figure 4). These changes are especially seen on the talonavicular joint surfaces and the superior articular surface of the talus. Finally, vertebra L5 shows a complete spondylolysis fracture with no reattachment.

Figure 3 Anterior view of tibiae of SHSM22. Note enlarged size, periosteal new bone (A, inset), and enthesophyte on medial proximal shaft (B) of the left tibia.

Figure 4 Medial view of tali of SHSM22. Note enlarged size of the left talus and rugosity of the superior articular surface (arrow).

A differential diagnosis suggests that this is likely a case of Legg-Calvé-Perthes disease (Table 1). The presentation also closely resembles that of slipped capital femoral epiphysis (SCFE), except that in SCFE the femoral neck is severely shortened (Ortner, 2003, p. 349),

which is not the case for SHSM22. In the case of SHSM22, the normal femoral neck length, flattened femoral head, unilateral presentation, and lack of evidence for infection are consistent with a diagnosis of Legg-Calvé-Perthes disease.

Table 1: Conditions considered in the differential diagnosis of SHSM22

Pathological condition	Diagnostic characteristics
Tuberculosis ^{ab}	Involvement of the hip is the second most common skeletal symptom after spondylitis. The disease can cause destruction of the femoral neck and head and the acetabulum. SHSM22 lacks the appearance of infection, as well as vertebral involvement.
Septic arthritis ^{ac}	Bacteria are introduced into the synovium of the joint. Hip involvement is common, and chronic sepsis results in some degree of destruction of the femoral head and neck, sometimes in addition to osteomyelitis and ankylosis of the hip joint. SHSM22 does not display destruction of the cortical bone of the femoral head and neck, only an altered shape of the femoral head that indicates collapse of the trabecular bone, and no indication of infection other than periosteal bone deposition.
Trauma (including subchondral bone injuries) ^{ad}	<p>Fracture of the femoral neck commonly leads to interruption of blood supply and avascular necrosis of the femoral head. The necrotic bone often includes visible dead trabecular bone, and the antero-lateral part of the femoral head will be most affected by structural collapse. Remodeling of the bone results in thickened trabeculae with a sclerotic rim. Advanced age is the greatest risk factor. In individual SHSM22, there is no visible evidence of a fracture in the femoral neck, or the trochanteric or subtrochanteric regions (though this cannot be assessed thoroughly without radiographs). The head of the femur is not significantly displaced, and the mushroom shape is not typical of trauma in adulthood.</p> <p>Fragmentation of articular cartilage can occur, sometimes with fracture of subchondral bone. In these cases, a sequestrum typically forms, and 90% of cases occur in the knee. There is no visible evidence of a fracture or healing callous in the subchondral portions of the femoral head of SHSM22, and the pervasive deformity of the femoral head suggests a chronic interruption of vascular supply, as in LCPD (though trauma may play a role in some cases of LCPD).</p>
Slipped capital femoral epiphysis (SCFE) ^{ae}	The unfused epiphysis is displaced, leading to displacement of the femoral head and potentially to aseptic necrosis. The epiphysis is typically displaced medially, posteriorly, and inferiorly. The condition resembles Legg-Calvé-Perthes disease (LCPD) in some ways, especially in its advanced stages, after osteoarthritis has developed. However, in LCFE,

	the femoral neck is shortened, which is not observed in SHSM22.
Legg-Calvé-Perthes disease (LCPD)/coxa plana ^{af}	The blood supply to the femoral head is interrupted for a period of several years during growth, which is followed by revascularization and often permanent deformity of the femoral head. Most cases are unilateral. The “mushroom head” results from the lack of endochondral growth and infarction of the epiphysis.

^aOrtner (2003)

^bVanhoenacker et al. (2009)

^cChoi et al. (1990)

^dAiello (2015); Khurana (2009); Ortner (2003); Taylor (2006)

^eGopinathan et al. (2012)

^fFornari et al. (2015); Khurana (2009); Wainwright and Catterall (2010); White et al. (2011).

3. Discussion

Legg-Calvé-Perthes disease (LCPD) is a developmental abnormality involving interruption of the blood supply to the femoral head for several years during growth, followed by revascularization and often permanent deformity of the femoral head. The disease's etiology is not well-understood; it may be initiated by traumatic injury, and in some cases, may be related to a systemic disorder involving a predisposition to vascular insult (Fornari et al., 2015; Khurana, 2009; Wainwright and Catterall, 2010; White et al., 2011). LCPD occurs in fewer than 20 in 100,000 people, usually presents in childhood, and occurs up to four times more often in males than in females (Fornari et al., 2015).

Femoroacetabular impingement (FAI) is a possible sequela of LCPD (Wainwright and Catterall, 2010), sometimes resulting in an enlarged acetabulum (Emary, 2010). In particular, LCPD often involves a widened acetabulum, from a combination of mechanical forces and hypertrophy of the triradiate cartilage (Huhnstock et al., 2014). However, FAI may also result from SCFE, so it does not help distinguish between LCPD and LCFE in the differential diagnosis. Enlargement of the acetabulum could also be the result of mechanical forces applied to the

acetabular margin during lateral subluxation of the femoral head, which is possible in more advanced cases of LCPD (Rab, 2005) and indicates a poor prognosis (Kotoura et al., 2015).

SHSM22's left tibia, talus, and navicular bone, contralateral to the affected hip, appear enlarged and rugose, with some remodeling periosteal new bone. The talus and navicular bone are affected by possible osteoarthritis secondary to LCPD and mechanical stress due to an antalgic gait. Osteomyelitis cannot be ruled out as a cause for the bony changes in the lower leg, but is also not evident, as there are no cloacae visible. Herrerín and Garralda (2012) report a case of LCPD with an atrophied ipsilateral tibia; the smaller diameter and shorter length of the ipsilateral tibia in the case of SHSM22 could have a similar origin. There is also a small but pronounced enthesophyte on the medial side of the proximal shaft of the left tibia. This is the location of the pes anserinus, a complex of tendons of three leg muscles, which primarily functions in flexion of the knee (Noyes and Sonstegard, 1973). An enthesophyte here, on only one side, indicates an uneven gait, which is common in LCPD (Švehlík et al., 2012; Westhoff et al., 2006; Westhoff et al., 2012).

Spondylolysis, such as that seen in the L5 vertebra, typically occurs as the result of a fatigue fracture from low-grade stress on the lower back (Ortner, 2003). This is not uncommon in adolescents, and generally results from a combination of congenital weakness in the pars interarticularis and strenuous physical activities, such as athletics or lifting heavy objects (Leone et al., 2011). This illustrates that though individual SHSM22 had an altered gait and somewhat impaired mobility, as well as chronic inflammation in the lower leg, he likely continued to participate in his community's labor activities. The dental health of this population is consistent with a diet high in starchy agricultural products (Berger, dissertation in progress), so it is possible that SHSM22 sustained the lumbar spine fracture while engaging in agricultural labor.

4. Conclusion

The effect of trauma on SHSM22's condition cannot be further assessed without radiographs, but there is no visible evidence of a fracture. Epidemiological studies have shown that incidence of LCPD in certain populations is correlated with "deprivation," but this concept is poorly defined and not linked to specific causes other than a speculative link to poor diet (Kealey et al., 2000; Margets et al., 2001). This is a possibility at Shijiahe: Berger (dissertation in progress) has found a rise in linear enamel hypoplasias (LEH) in this population over previous periods (22.6% of individuals), which may indicate growth interruptions in childhood linked to malnutrition. However, individual SHSM22 did not have LEH.

In modern populations, symptoms of mild cases of LCPD may resolve without intervention, but more severe cases can result in permanent altered gait and deformity (Wainwright and Catterall, 2010). It therefore appears that this individual suffered from a relatively severe case of LCPD. This is a significant finding, as reports of LCPD in the paleopathological literature are still rare, and the dry bone presentation is not often reported. The possible lateral subluxation of the femoral head is indicative of a more severe case of LCPD, and the enthesophyte and bony changes to the contralateral lower leg and foot indicate an altered gait. This individual therefore likely had impaired mobility. However, the spondylolysis of L5 indicates that despite his impairment, individual SHSM22 continued to engage in physical activities that put stress on the lumbar spine. Therefore, although the etiology of the condition in SHSM22 is difficult to assess, the case presents a picture of the progress of the untreated condition in an ancient individual, who still remained mobile and engaged in labor, despite his hip deformity.

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