A POSSIBLE CASE OF LEGG-CALVÉ-PERTHES' DISEASE IN AN ADULT MALE FROM MEDIEVAL/MODERN TOMAR, PORTUGAL

Curto AQ¹, Fernandes T¹

¹ Departamento de Biologia e Centro de Ciências e Tecnologias da Saúde, Universidade de Évora

Correspondencia a: a.q.curto@gmail.com

RESUMEN. La enfermedad de Legg-Calvé-Perthes (LCPD) es una necrosis avascular idiopática de la cabeza femoral, relacionada con una interrupción del suministro de sangre, probablemente iniciado con un trauma. Esta patología se pasa por cuatro etapas: la aparición de necrosis avascular, fractura subcondral, la revascularización y curación. Esta condición aparece entre los 5 y 9 años de edad y los niños son más afectados que las niñas. Además la mayoría de los casos han sido esporádicos, algunos factores, como la predisposición genética e influencias ambientales han sido implicados en la etiología de LCPD. En este estudio se reporta un posible caso de LCPD en un medieval/moderno hombre adulto de una excavación en Tomar, Portugal. En este individuo no sólo se observó las características que se encuentran normalmente en LCPD, como la cabeza del fémur en forma de hongo y una ligera rotación disto-mesial de la tibia, sino también la osteoartritis en el calcáneo y astralagus, probablemente relacionado con los cambios
en el fémur. También se observó la presencia de hueso nuevo en los últimos cuatro vértebras torácicas, en el lado derecho se ve como cera de vela derretida en la parte delantera de la T9, T10 y T11 y en la T12 se observó un llenado total del espacio del disco con hueso nuevo. También se observó un alto nivel de alteración en la entesis y formación de hueso. Esta persona muestra una aparente tendencia para osificarse los tejidos blandos y habría un movimiento muy limitado siendo fuertemente dependiente de los demás.

PALABRAS CLAVE: Enfermedad de Legg-Calvé-Perthes, formadores de hueso, Tomar, medieval

ABSTRACT. The Legg-Calvé-Perthes’ disease (LCPD) is an idiopathic avascular necrosis of the femoral head, related with a disruption of the blood supply, probably initiated with a trauma. It passes through four stages: the onset of avascular necrosis; subchondral fracture; revascularization and regeneration; and healing. This condition usually appears between 5 and 9 years old and boys are more affected than girls. Besides most cases have been sporadic, some factors, as genetic predisposition and environmental influences have been implicated in the etiology of LCPD. In this study we report a possible case of LCPD in a medieval/modern male adult from an excavation in Tomar, Portugal. In this individual not only we observed the usually features found in LCPD, like the femoral head in a form of mushroom and a slight disto-mesial rotation of the tibia, but also osteoarthritis in the calcaneus and astragalus, probably related to the changes in the femur. We also observed the presence of new bone in the last four thoracic vertebrae, at the right side it looks like melted candle wax down the front of the T9, T10 and T11 and in the T12 it was observed a total filling of the disc space with new bone. It was also noted high levels of changes in the entesis and bone forming in some bones. This person shows an apparently tendency to ossify soft tissues and would have a very limited movement being heavily dependent on others.

KEYWORDS: Legg-Calvé-Perthes’ disease, bone formers, Tomar, medieval
INTRODUCTION

The Legg-Calvé-Perthes’ disease (LCPD) is a form of idiopathic avascular necrosis of the femoral head (Waldron, 2009) and has a clear relationship with disrupted blood supply that is probably initiated by trauma (Ortner, 2003) but its etiology is unknown. This osteochondrosis affects children from 3 to 10 years of age and boys outnumber girls about 4:1 (Aufderheide e Rodríguez-Martín, 1998). In the course of this pathology the femoral head flattens due to a combination of compression fracture, lack of endochondral growth and lack of vascularization, resulting in a femoral head with a “mushroom shape” (Ortner, 2003) leaving sequels even when treated, increasing the risk of severe osteoarthritis (Onishi et al., 2011).

Although most cases of LCPD are sporadic, there seems to be some genetic components that predispose some families for this disease, specially associated with factor V Leiden and collagen mutations (Miyamoto et al., 2007; Kenet et al., 2008; Vosmaer et al., 2010). It may be due to genetic factors that some populations are more prone to develop this condition, which is more common in Caucasians, follow by Asians and Africans (Rowe et al., 2005). Besides genetic factors, environmental factors also seem to have some connection with the incidence of this pathology (Aufderheide and Rodríguez-Martín, 1998; Margetts et al., 2001; Rowe et al., 2005) and the lack of concordance in the incidence of this pathology in twins shows the environmental importance in LCPD (Perry and Hall, 2011). Tayton (2010) suggest that one of the reasons to that predispose to LCPD may be a preexisting restricted range of movements in the hip, making it more susceptible to strain injuries, once African groups and females children have a bigger range movement. Some authors (Perry and Hall, 2011) suggest that LCPD may be associated with congenital anomalies and so, have an intrauterine cause.

MATERIALS AND METHODS

The individual under study (SMOL-271) is part of a sample studied for an acquisition of a M.Sc. degree (Curto, 2011) associated with Santa Maria dos Olivais’ church from Medieval/Modern Age is
almost complete and well preserved. This skeleton was recovered from area 14, with an orientation from SO to NE, in a supine position, with his arms crossed at the thoracic area and his legs extended parallel, like the other individuals in this Christian cemetery.

This necropolis extends for 6500m$^2$, has about 4000 burials and a minimum number of individuals of 6792. So far there were studied 73 individuals from the sample recovered, 57 adults (20 females, 31 males and 6 unidentified) and 16 sub-adults (Anselmo, 2011; Curto, 2011; Gonçalves, 2011).

We used Bruzek’s method (Bruzek, 2002) for sexual diagnosis and Lovejoy’s method (Lovejoy et al., 1985) to determine the age at death. In order to compare the stature of the individual under study with the rest of the sample with the same sex we used measurements from the femur (Mendonça, 2000), tibia (Olivier et al., 1978) and second metatarsal (Santos, 2002) obtain so far (Anselmo, 2011; Curto, 2011; Gonçalves, 2011).

All the lesions in this individual were recorded and described macroscopically, in order to get the most information possible from the available material, accompanied by measurements when relevant and possible. Given the similarity of injuries among different pathologies, we used whenever possible, a differential diagnosis.

**RESULTS AND DISCUSSION**

The subject of this study is an adult male which age at death, according Lovejoy et al. (1985), is between 45 to 49 years old. Femoral head, on right femur, has a “mushroom shape” with bony growths in the joint area, porosity and eburnation in the antero-posterior face which affects almost entirely the anterior joint, yet the left femur doesn’t show such changes (Fig. 1). As there wasn’t a large shortening of the femur, once the right femur physiological length was 427cm and the left femur measure was 431cm, we can exclude the possibility that it is a displacement of the femoral head (Waldron, 2009), besides, there wasn’t observed a malalignment of the femoral head on the neck of the femur. The changes of the proximal joint of the right femur resulted in arthritis at the right
acetabulum, which was observed lipping, eburnation and porosity at the upper zone of the acetabulum.

We also observed a slight disto-mesial rotation in the right tibia, probably, related to the changes observed in the right femur and neofacets in the proximal extremity of the tibias in a distal position, being more evident in the right tibia. Still at the right side it was observed an artrose in the astragalus and calcaneus at the joint that these bones share, where there was bone destruction in the articular facets and changes in their morphology, when in the left foot there was no changes.

The individuals with this pathology usually have a delay in growth (Aufderheide and Rodríguez-Martín, 1998; Perry and Hall, 2011). In fact, we observed a smaller stature in the individual at study (SMOL-271) than in the sample mean, however, the stature calculated for SMOL-271 are in the range of the standard deviation (Table 1).
TABLE 1. Estimation of stature by measurements from second metatarsal (Santos, 2002), tibia (Olivier et al., 1978) and femur (Mendonça, 2000)

<table>
<thead>
<tr>
<th></th>
<th>Second metatarsal</th>
<th>Tibia</th>
<th>Femur</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>x</td>
<td>σ</td>
</tr>
<tr>
<td>Santa Maria dos</td>
<td>13</td>
<td>164,00</td>
<td>6,54</td>
</tr>
<tr>
<td>Olivais Collection</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SMOL-271</td>
<td>162,70</td>
<td>163,04</td>
<td></td>
</tr>
</tbody>
</table>

In the right upper limb we found artroses at the distal end of the clavicle that also shows a neofacet at the area of the costoclavicular ligament which seems to articulate with the first rib that shows a bone growth. In the right scapula we also found lipping in the glenoid cavity and porosity at the joint with the clavicle and in the right ulna we also noted a dilatation at the distal end which due to a post-mortem fracture was possible to observe the absence of the medullar cavity, characteristic of osteomyelitis when on the right radio it wasn’t observed any evidences of priostitis or osteomyelitis.

In this individual new bone deposits are present in the last 4 thoracic vertebrae. On the right side there was an ankylosis due to paravertebral ossification looking like “melted candle wax” on the proximal surface of the vertebrae T9, T10 and T11, which is not present on the left side, due to the descending aorta on this side (Waldron, 2009). The morphology of these changes are compatible with the lesions characteristics of DISH, so, we are probably dealing with an initial stage of this pathology, but we cannot exclude the possibility that this is a case of spondyloarthropathy. Between the vertebrae T11 and T12 we observed a complete filling of the intervertebral space with new bone, characteristic of an espondyloarthropathy, resulting in ankylosis connecting this 4 continuous vertebrae. There was also intra-articular fusion of the vertebral apophyses of T11 and T12, although the conservation status of the vertebrae do not allow analysis of the remaining vertebral apophysis, the absence of this lesion on the superior facets of T11 suggest that the facets above was also not fused. Also in the lumbar vertebrae of these individual were observed some
changes: when we reconstruct the curvature of the lower back it was evident a flattening in the anterior zone of L1, leading to the loss of the natural curve of this anatomical area. Although the analysis of the sacrum isn’t possible, on the auricular surface there were no evidences of ankylosis or abnormal bone growth. The vertebral column of this adult male seems to represent two distinct pathologies: DISH and an espondyloarthropathy possibly a case of ankylosing spondylitis.

CONCLUSIONS

These man movements were probably limited, especially in the lower right limb in consequence of the absence of treatment for the Legg-Calvé-Perthes disease which is highly autolimiting, especially when associated with vertebral fusion.

A better understanding of the pathobiology of LCPD will lead to the development of more effective treatments.

Acknowledgements

We are grateful to Andreia Gonçalves and Daniela Anselmo for their data, which allowed us a larger sample for the study of the stature. We are also grateful to Sónia Ferro for her information about the necropolis.

BIBLIOGRAPHY


