

Total Hip Replacement in Diaphyseal Aclasis: A Case Report

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Citation

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Abstract

We describe the case of young man with multiple hereditary exostosis, who presented with degenerative disease of the hip joint. A total hip replacement was done for him. This procedure is rarely done for such cases as the hip symptoms are usually attributed to the primary condition itself and possibility of osteoarthritis of the hip is overlooked. To the best of our knowledge this is the first case report written on total hip replacement in a case of multiple hereditary exostosis.

INTRODUCTION

Diaphyseal aclasis or multiple hereditary exostosis is a common skeletal dysplasia with an incidence of one in 50 000. These patients commonly present with characteristic deformities that particularly involve paired bone in the limbs such as radius-ulna or tibia-fibula, giving rise to valgus deformity of knee, ankle and limb-length discrepancy¹. The incidence of proximal femur exostosis is around 82%¹. These are rarely symptomatic but there have been some reports of proximal femur deformity associated with secondary osteoarthritis of the hip in these cases^{1,2}. To our knowledge this is the first case report of a total hip replacement done for such cases.

CASE REPORT

A 34 years old gentleman, who was diagnosed to have diaphyseal aclasis at the age of 18 years, presented with a long history of mechanical pain in his right hip, which radiating to his right thigh and knee. In the recent months, this pain has progressively worsened. At presentation the pain was present most of the time and occasionally disturbing his sleep. He was only able to walk short distances due to this pain. He has also noticed that this right leg turned inwards when he walks. All his other joints are symptom free. He has had several surgical procedures over the years for removal of symptomatic exostosis, from various parts of his body. On examination, he was in good general health. There was some muscle wasting of the right thigh and buttock. There were well-healed scars from previous surgeries, present over the right thigh, groin and hip. Trendelenburg test was negative on both his hips. His

right leg was apparently shorter by 1 centimeter. The left hip had full range of movement but the right hip only had limited 60 degrees of flexion and was fixed in 10 degrees of adduction and 45 degrees of internal rotation. It was painful throughout its range of movement.

His pelvic radiograph showed severe degenerative changes in the right hip joint with narrowing of the joint space and subchondral sclerosis. The femoral neck and the intertrochanteric region were abnormally enlarged. There were multiple small exostosis around the intertrochanteric region with a large sessile exostosis arising from the lesser trochanter (Figure 1).

Figure 1

Figure 1: Preoperative x-ray of the pelvis including both hip joints. It shows bilateral coxa valga with broaden femoral neck and metaphysis of femur. The right hip is osteoarthritic with narrowing of joint space, subchondral sclerolosis, cyst and surrounding osteophytes. There is also a large exostosis arising from the right lesser trochanter.



This gentleman underwent a right total hip replacement. A cemented Lubinus hip with a ceramic on polyethylene articulating surface was chosen due to it's prolonged survivorship. Intra-operatively, the following was noted; the femoral head and neck was deformed with an increase neck/shaft ratio (figure 2 and 3), the acetabulum was noted to be shallow. The right hip movement was limited due to the multiple small exostosis arising from the base of the femoral neck and there was a large exostosis arising from the lesser trochanter, stretching the muscles and increasing tissue tension around the hip.

Figure 2

Figure 2: Deformed femoral head with multiple osteophytes at the base of the head, making the head more dome shaped and less spherical. There is also an increase neck/shaft angle.

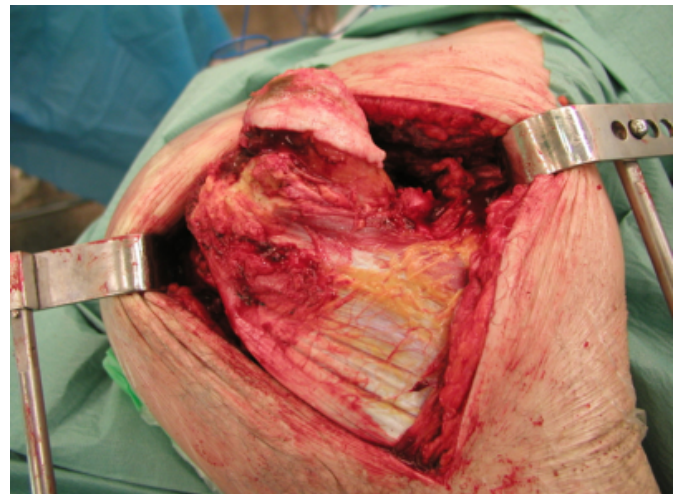
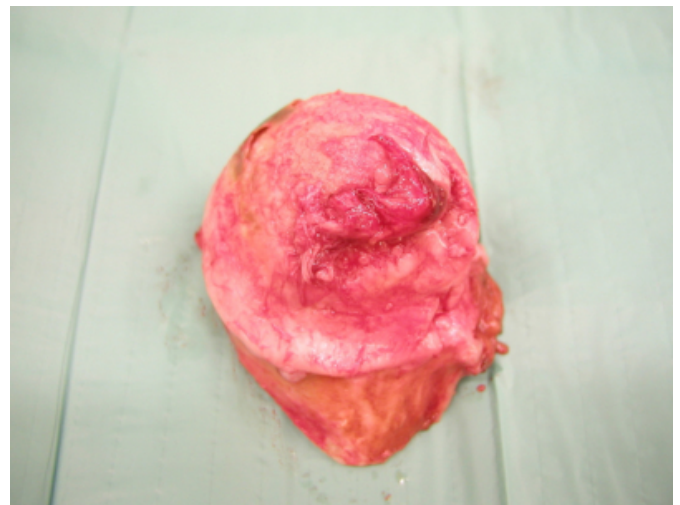


Figure 3

Figure 3: Osteotomised femoral head showing the articular surface. The articular surface is irregular with extensive chondrolysis and ebunation. The edges of the head are flared due to formation of peripheral osteophytes.



During the procedure, the excess bony protuberance was excised without structurally weakening the proximal femur and a standard total hip replacement was carried out. The acetabulum was reamed to 54mm and a 52mm cemented cup was inserted. On the femoral side, the canal was reamed to a size 2 and a Lubinus stem was cemented in with a cement restrictor. A size 28mm ceramic head was used and the hip was reduced (figure 4). It was stable in extremes of movement and the tissues were repaired in layers. In the postoperative period, the patient started mobilizing on day 2 postoperatively and was discharged after one week of

hospital stay. Postoperative x-ray is shown in figure 5.

Figure 4

Figure 4: Proximal femur with cemented lubinus SP 11 in place with a size 28mm ceramic head molded on. The cross section of the femoral neck is wider than the collar of the implant and the surrounding area around the neck is raw due to removal of the impinging exostosis.

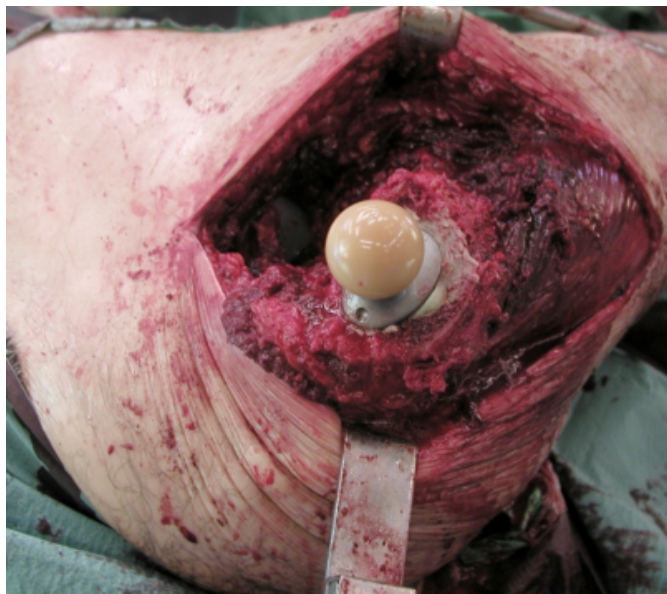
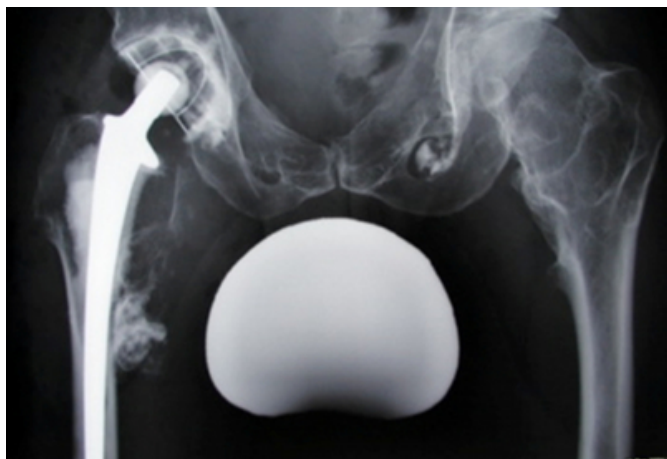


Figure 5

Figure 5: Postoperative x-ray. The hip is reduced and the implant is well cemented in.



DISCUSSION

The incidence of total hip replacement in cases of hereditary multiple exostosis, as reported by Scarborough and Moreau in their series was only 1 percent². They attributed the premature osteoarthritis to the associated coxa valga and acetabulum dysplasia, which is prevalent in these cases. Initially, it was felt that the hip in cases of Hereditary Multiple Exostosis developed along normal lines and

limitation of hip movement present was purely due to the mechanical restrictions of exostosis around the femoral neck. But this view has now changed. Shapiro in his review of such cases reported a 25 percent incidence of coxa valga and occasional incidence of acetabular dysplasia¹. Bassett and Scott also suggested the associated coxa valga was due to abnormal metaphyseal remodeling and this in return results in the femoral head being partially uncovered³. Weiner and Hoyt, in their review of 25 cases, also reported an increase valgus angle and anteversion of the femoral neck in these cases⁴. Therefore, it is obvious that these patients are prone to premature osteoarthritis as a result of abnormalities present in the proximal femur and acetabulum. Porter in his review of 12 cases found degenerative arthritis in 3 cases but only one required a total hip replacement⁵.

The choice prosthesis for total hip replacement is crucial, as these patients are young as compared to those with primary degenerative arthritis of hip. Lubinus SP11 hip was chosen for this patient because of its long survivorship as reported in the "Swedish Hip Registry". It has a longer stem (18 centimeters long), which allows it to sit further down the femoral canal, therefore providing better support. Other possible options for younger patients requiring hip replacement would be hip resurfacing and uncemented total hip replacements. The former is not possible in this case as the femoral head-neck ratio is smaller than usual due to the broad neck. Furthermore, there are still no long-term results published for this prosthesis as it's still relatively new. As for the uncemented prosthesis, their long-term survivorship historically has been poorer compared to the cemented prosthesis. However, with the development of newer forms of fixation techniques such as hydroxyapatite coating and textured titanium surfaces, the revision rates have reduced significantly. As these 3rd generation uncemented prosthesis are still fairly recent, the senior author is still not confident in its long-term reliability. As for the bearing surfaces, ceramic-on-polyethylene was chosen as it is proven to have the lowest wear rates as compared other forms of bearing surfaces. Its reported wear rate is only 0.03mm/year. On the other hand, metal-on-polyethylene articulations have a variable wear rate ranging from 0.04 to 0.14.

CONCLUSION

We feel that degenerative arthritis of the hip is more prevalent in cases of hereditary multiple exostosis. But it is often overlooked and these patient's symptoms are usually attributed to the primary condition itself. Therefore, clinicians should pay more attention to these symptoms and

institute appropriate treatment when necessary.

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