CASE REPORT

Bilateral total hip arthroplasty in a young man with hereditary multiple exostoses

Raju Vaishya,¹ Sandeep Swami,² Vipul Vijay,¹ Abhishek Vaish³¹Orthopaedic & Joint Replacement Surgery, Indraprastha Apollo Hospitals, New Delhi, India²Department of Orthopaedics, Indraprastha Apollo Hospitals, New Delhi, India³Department of Orthopaedics, Sancheti Hospital, Pune, Maharashtra, India**Correspondence to**
Professor Raju Vaishya,
raju.vaishya@gmail.com

Accepted 18 December 2014

SUMMARY

We report a rare case of a young man with hereditary multiple exostoses (HME) who presented with severe pain in the hips, requiring bilateral total hip arthroplasty (THA), along with excision of the exostoses. The reported incidence of exostoses around the hip has been reported from 30% in some series to 90% in other studies of HME. These patients may remain asymptomatic, but if not, their symptoms are usually related to the presence of the exostoses itself and the possibility of hip osteoarthritis is frequently overlooked. This may account for a low incidence of THA for this condition. Surgical management often remains challenging as such lesions are not commonly encountered in the clinical practice and are often very difficult to treat.

BACKGROUND

Bilateral hip lesions in hereditary multiple exostoses (HME) are rare and complex.¹⁻⁴ They may be associated with premature osteoarthritis (OA) of the hip. The treatment of multiple exostoses with OA should not only involve removing the exostoses but must also address the OA with total hip arthroplasty (THA) in severe and symptomatic cases. We performed bilateral THA in our young patient. The surgery was challenging and complex due to disturbed anatomy.

CASE PRESENTATION

A 27-year-old man presented with bilateral hip pain and difficulty in walking for the past 1.5 years. The pain was severe enough to prevent him from walking and performing activities of daily living independently. It first developed in the left hip and subsequently also affected the right hip. It was insidious in onset and mild in intensity initially, but gradually progressed to severe and constant pain. The patient had multiple bony swellings over his body since childhood, which had grown gradually as he matured. He also had a family history of similar swellings in his younger brother and mother. There were no associated comorbid medical problems noted.

On examination, the patient was short statured (figure 1) with a thin build; he walked with a waddling gait. There were multiple hard bony swellings over his hips, proximal tibia, distal femur, scapula and proximal humerus. Both hips had limited range of motion with bilateral fixed flexion deformity of 30° and bilateral fixed external rotational deformity (20°). There was a large hard bony swelling (2"×3") present in the right inguinal and left iliac region. The right lower limb was longer than the left by 1 cm.

INVESTIGATIONS

Anteroposterior X-ray of the pelvis showed exophytic bony growth in the femur and pelvic bone with degenerative changes in the hips and coxa valga, decreased centre edge angle and increased neck head width ratio (figure 2). CT of the pelvis confirmed exophytic growth with cartilaginous cap in the femur and pelvic bones (figures 3 and 4) and significant secondary OA changes in the hips.

DIFFERENTIAL DIAGNOSIS

- ▶ Loose bodies
- ▶ Chondrosarcoma
- ▶ Heterotopic ossification
- ▶ Multiple epiphyseal dysplasia

TREATMENT

The left hip was operated on first, using an antero-lateral approach (modified Hardinges), where excision of exostoses from the femur and acetabulum

Figure 1 Multiple bony swellings (exostoses) over the scapulae, pelvis and arm.

CrossMark

To cite: Vaishya R, Swami S, Vijay V, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-207853

Figure 2 Anteroposterior X-ray of the pelvis showing exophytic growth in bilateral femur and pelvis.

was carried out; the arthritic hip was replaced with uncemented prosthesis using a 36 mm head with 58 mm cross-linked polyethylene acetabular cup (Stryker). One week later, the right hip was operated on, using an anterior approach. The osteochondromas were first excised from the pelvis through an anterior ilioinguinal approach; subsequently the THA was performed using a modified Smith Peterson approach. An uncemented THA using SROM prosthesis (Depuy) was carried out. A corrective femoral osteotomy was required in the upper third of the femur to accommodate the femoral stem of the prosthesis (figure 5). This osteotomy was fixed using one eight-hole low-contact dynamic compression plate.

The left hip was found to be dislocated after right hip arthroplasty (figure 6). After closed reduction the hip was found unstable, hence an open reduction with revision to a larger stem size 2–3 (with improved version) and longer neck length, from size +5 to +10, was made. A boot and bar cast was applied on the left leg for 3 weeks, to prevent dislocation.

OUTCOME AND FOLLOW-UP

At follow-up, after 12 months both hips were found stable (figure 7), and the osteotomy on the right side had healed. The patient was mobilising with full weight bearing. Harris hip score improved from 15 preoperatively to 78 postoperatively on the right side and 18–86 on the left side.

Figure 3 CT of the pelvis showing exostoses in bilateral femur and pelvis.

Figure 4 CT scan showing osteoarthritis changes in bilateral hips.

DISCUSSION

HME are a type of skeletal dysplasia characterised by the presence of multiple exostoses.⁵ This uncommon condition has an incidence of 1 in 50 000 population, with autosomal dominant inheritance of variable penetrance.^{6 7} The exostoses in HME are usually diagnosed in the first decade of life in affected individuals (80%). These cases are discovered relatively early due to their positive family history.¹ Exostoses may develop in any bone preformed in cartilage but are usually found on metaphysis of a long bone near the physis and are most commonly seen on the distal femur, proximal tibia and proximal humerus.⁵ Deep-seated osteochondromas in areas such as the proximal femur or pelvis are not easily diagnosed as they rarely cause symptoms.⁷

The presence and severity of symptoms in the hip are related to the location of osteochondromas.⁸ Lesions in the medial femur and periacetabular region cause more pain as they alter the hip parameters and its development resulting in acetabular dysplasia and coxa valga. Acetabular dysplasia and coxa valga occur in approximately 25% of patients with HME.^{9 10} The femoral head itself does not appear to be primarily affected, hence findings of coxa valga may require intervention as varus osteotomy, which may delay arthritis.¹ As compared to medial and periacetabular lesions, lateral femoral lesions occur less often and also cause less pain and discomfort, and do not alter the hip geometry.⁸

Figure 5 Bilateral total hip arthroplasty showing revision of the left hip femoral stem (with larger stem and longer neck length).

Figure 6 X-ray of the pelvis of bilateral hip arthroplasty showing dislocated left hip and uncemented S-ROM right hip with femoral osteotomy fixed with low-contact dynamic compression plate.

The reported view about the association of symptoms to hip pathology in HME has changed over the years. Previously, pain and limitation in hip movement were related to mechanical restriction by exostoses around the femoral neck.⁴ However, more recently, it has been recognised that other hip pathologies due to skeletal dysplasia are also responsible for causing symptoms. These include coxa valga, acetabular dysplasia and partial uncoverage of femoral head.^{10 11} Scarborough and Moreau¹² had attributed premature osteoarthritis to acetabular dysplasia and coxa valga. Since then, the concept of premature osteoarthritis of the hip as a result of abnormalities of the proximal femur and acetabulum has gained importance as a cause of hip symptoms in patients of HME.

Total hip replacement has seldom been reported as a treatment for hip involvement in HME. There are only a few reports present in which THA was performed for the condition.^{4 13 14 15} It has been recognised that THA is a technically difficult procedure in HME due to distorted hip anatomy, such as coxa valga, acetabular dysplasia, short stature and limb length discrepancy, which creates difficulty in placing properly sized implants.¹⁵ THA for severe hip involvement in HME leads to good

symptomatic relief with improvement in Harris hip scores and good functional outcome.¹³ We believe that THA (in association with removal of most accessible and symptomatic osteochondromas) can offer a one stage and definitive solution to treat osteochondroma associated with secondary OA due to HME.

Patient's perspective

I was feeling hopeless due to the complex nature of my problem and as no definitive treatment was offered to me. I was suffering with severe agonising pain in the hips. However, after bilateral hip surgery, I felt as if a new lease of life had been given to me.

Learning points

- ▶ Hereditary multiple exostosis is a rare developmental disorder which also affects the hips.
- ▶ Premature osteoarthritis of the hip can be a presenting symptom in multiple hereditary exostoses.
- ▶ Total hip arthroplasty can be a definitive procedure to treat hip arthritis in hereditary multiple exostoses.

Contributors All the authors have contributed in writing this manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Steiber JR, Dormans JP. Manifestation of hereditary multiple exostoses. *J Am Acad Orthop Surg* 2005;13:110–20.
- 2 Saglik Y, Altay M, Unal VS, *et al.* Manifestations and management of osteochondromas: a retrospective analysis of 382 patients. *Acta Orthop Belg* 2006;72:748–55.
- 3 Felix NA, Mazur JM, Loveless EA. Acetabular dysplasia associated with multiple hereditary exostoses. A case report. *J Bone Joint Surg Br* 2000;82:555–7.
- 4 Singh V, Carter S. Total hip replacement in diaphyseal aclasis: a case report. *Int J Orthop Surg* 2007;5:10.
- 5 Heck RK, Jr. *Campbell Operative Orthopaedics*. 11th edn, 2008 chap 20:860–2.
- 6 Makhdum AM, Jiang F, Hamdy RC, *et al.* Hip joint osteochondroma: systemic review of literature and report of further three cases. *Adv Orthop* 2014;2014:180254.
- 7 Porter DE, Benson MK, Hosney GA. The hip in hereditary multiple exostoses. *J Bone Joint Surgery[Br]* 2001;83-B:988–95.
- 8 El-Fiky TA, Chow W, Li YH, *et al.* Hereditary multiple exostoses of the hip. *J Orthop Surg (Hong Kong)* 2009;17:161–5.
- 9 Malagon V. Development of hip dysplasia in hereditary multiple exostoses. *J Paed Orthop* 2001;21:205–11.
- 10 Shapiro F, Simon S, Glimcher MJ. Hereditary multiple exostoses: anthropometric, roentgenographic and clinical aspects. *J Bone Joint Surg[Am]* 1979;61-A:815–24.
- 11 Weiner DS, Hoyt WA Jr. The development of the upper end of the femur in hereditary multiple exostoses. *Clin Orthop Relat Res* 1978;137:187–90.
- 12 Scarborough MT, Moreau G. Benign cartilage tumors. *Orthop Clin North Am* 1996;27:583–9.
- 13 Moran M, Kreig AH, Boyle RA, *et al.* Bilateral total hip arthroplasty in severe hereditary multiple exostoses: a report of two cases. *Hip Int* 2009;19:279–82.
- 14 Annareddy SR, Chapman TWL, Chamley GJ. Dual aetiology hip pain requiring total hip arthroplasty in hereditary multiple exostoses: a case report and review of the literature. *Hip Int* 2002;12:397–9.
- 15 Mesfin A, Goddard MS, Tuakli-Wosornu YA, *et al.* Total hip and knee arthroplasty in patients with hereditary multiple exostoses. *Orthopedics* 2012;35:e1807–10.

Figure 7 Twelve months' follow-up X-ray of both hips showing well-reduced hips and healed right femoral osteotomy.

Copyright 2015 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow