# Difficulties of Cement-less Total Hip Arthroplasty in Osteopetrosis: A Case Report from Iran

Hasan Rahimi shoorin[1], Mahmood Bahari[1], Mohammad Gharehdaghi[1], Masoud Mirkazemi[2], Maryam Asadian[3], Mehran Azami[2] , Amirreza Fatehi[2], Abbas Eydizadeh Nokhandan[2]

[1.] Associate professor of orthopedic surgery, Mashhad orthopedic and trauma research center, Faculty of medicine, Mashhad University of medical Sciences, Mashhad Iran
[2.] Resident of orthopedic surgery, department of orthopedic surgery, Faculty of medicine, Mashhad University of medical sciences, Mashhad Iran
[3.] MD, Research assistant, Orthopedic and trauma Imam Reza Research Center, Faculty of medicine, Mashhad University of medical sciences, Mashhad Iran
gharehdaghim@mums.ac.ir; rahimih@mums.ac.ir

**Abstract:** Total joint arthroplasty is often virtually the last option for treating osteopetrosis associated with osteoarthritis or other complications. Many intraoperative challenges must be overcome when performing arthroplasty in such patients. This paper presents a 36-year-old woman with autosomal dominant (AD) osteopetrosis suffering a painful nonunion of subtrocanteric fracture. She was referred to Imam Reza University Hospital, Mashhad, Iran, in June 2010. The patient had three previously unsuccessful operations as open reduction with different fixation devices, which resulted in nonunion associated with destruction of the femoral head. Therefore, we opted to use cement-less total hip arthroplasty. During the operation, we encountered difficulties, especially when preparing the femoral canal for a prosthesis stem. The short-term post-operative clinical and radiological results were good and, today, the patient walks independently and pain-free.
[Hasan Rahimi shoorin, Mahmood Bahari, Mohammad Gharehdaghi, Masoud Mirkazemi, Maryam Asadian, Mehran Azami, Amir Reza Fatehi, Abbas Eydizade Nokhandan. **Difficulties of Cement-less Total Hip Arthroplasty in Osteopetrosis: A Case Report from Iran.** *J Am Sci* 2013;9(2):340-342]. (ISSN: 1545-1003). http://www.jofamericanscience.org. 44

**Keywords:** Osteopetrosis; Autosomal dominant; Cement-less total hip arthroplasty

## 1. Introduction

Osteopetrosis is a group of sclerosing bone dysplasia conditions characterized by diminished osteoclastic mediated skeletal resorption (1), leading to universally hard, but brittle, bones (2). It was first described in 1904 by Albert Schonberg, a German radiologist (3). This disorder is divided into three clinical forms:

- Severe infantile or malignant autosomal recessive with severe bone-marrow failure, pancytopenia, bleeding, infection, failure to thrive, blindness, deafness, hydrocephalus, and early-childhood death
- Intermediate autosomal recessive or marble bone disease, with no bone-marrow failure, renal tubular acidosis, intracranial calcification, sensor neural hearing loss, and psychomotor retardation
- Autosomal dominant (AD) form (4-7)

In all forms of osteopetrosis, there is a lack of osteoclastic function, resulting in decreased bone resorption with increased cortical bone and calcified cartilage (2). The AD form can be subdivided into type 1 and type 2 as described by Bollerslev and Anderson (8). Type 1 is characterized by increased thickness of the skull, diffused osteosclerosis of the lumbar spine and pelvis, and symmetrical long bone involvement. Type 2 shows more basal skull involvement, a “rugger jersey” spine and “endobones” within the pelvis. Although the AD form is compatible with a normal lifespan (9, 10), the patient will likely encounter frequent orthopedic problems, including fractures, coxa vara, long bone bowing, osteomyelitis, osteoarthritis, and fracture nonunion (4, 9, 11, 12, and 14). Following these problems, plating and intramedullary rods are used for fractures and bone deformities, and joint arthroplasty is performed for degenerative joints. However, these operations are sometimes extremely difficult because of the bone fragility and obliteration of the marrow cavity (4, 15, and 16). There have been several early reports of cemented or hybrid hip arthroplasty, for these conditions. But there exist few reports regarding cement-less total hip arthroplasties in such patients.

## 2. Case Presentation

A 36-year-old woman presented with painful nonunion of the left hip, subtrocanteric fracture, and hereditary osteopetrosis. She was referred to Imam Reza University Hospital. Mashad, Iran, in June 2010. The radiologic image indicated destruction of the femoral head and acetabulum due to penetration of DHS nail into the hip joint space (see Fig. 1). The patient was unable to walk without

crutches and her preoperative Harris Hip Score (HHS) was about 27 of 100. We decided to perform a total hip arthroplasty for her. In the preoperative examination, blood work and laboratory exams were normal and the patient was generally in normal, healthy condition. A preoperative X-ray of the medullary canal of the femoral bone showed it was filled with sclerotic bone and there was no space for device insertion.

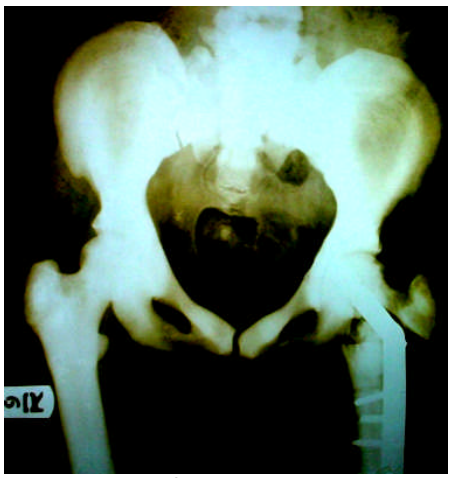

**Figure 1.** DHS failure in Osteopetrosis

Because of the difficulty of creating the wide canal and to prevent shaft fracture, we decided to use a Cone Wagner hip prosthesis that has a conical, narrow, non-cemented stem. For infection prophylaxis, we used 2 grams of Cefazolin intravenously 30 minutes before surgery and continued this for two days postoperatively. For better access to the femoral shaft and the acetabullar fossa, we used the postero lateral approach and extended it to the proximal part of femur for removing the DHS. Then, in the regular manner, we cut and removed the neck and head of the femur through the posterior capsulotomy and started to ream the acetabulum. Finally, as a press fit device, we replaced the metal and augmented it with two screws. In the next stage, as a prophylaxis of femoral shaft fracture, we used multiple levels of stainless steel wiring before reaming the femoral canal. Then, with the assistance of several times of fluoroscopy by C- arm, we tried to create a narrow canal with 3.2mm-4.5mm drill bit at the center of distal fragment and controlled it to be exactly centrally located in the femoral shaft. Then, we gradually increased the diameter of this canal, until it became so wide that the reaming with Cone Wagner reaming devices becomes possible. Finally, we used the trial stem with good position (see Fig. 2).

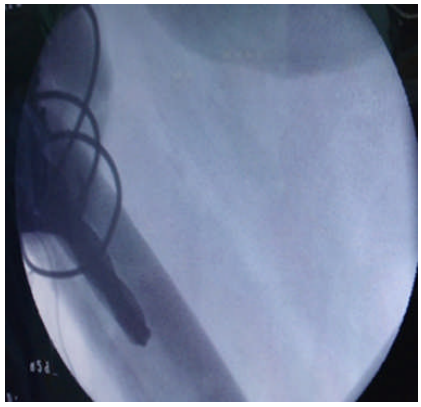

**Figure 2.** Reaming canal under c-arm guide in operation room

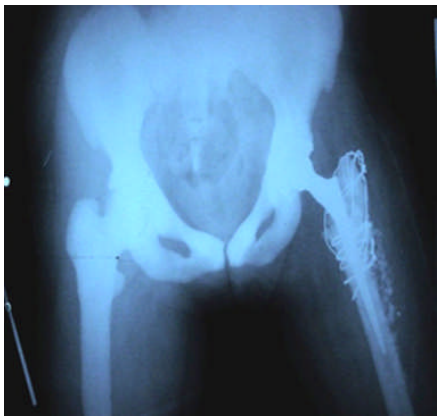

**Figure 3.** Cementless total hip arthroplasty in osteopetrosis

After trial stable reduction with standard size of neck over size 3 reamer, we inserted the original stem with size 3 Cone Wagner and the greater trochanter was fixed by multiple wire sutures and the used bone chips as the bone graft. Then, we repaired the short external rotators, muscle planes, and finally the skin over the suction drainage (see Fig. 3). The day after operation, the patient began to walk with the assistance of two crutches and toe touch. After about 36 months postoperatively, the patient has no pain and is walking well independently. Her Harris Hip Score is 70.

**3. Discussions**

Osteopetrosis as a congenital bone disease associated with failure of the osteoclast function. This malfunction with decreased resorption of bone leads to universally hard bones with thickened trabecula and decreased medullary cavity diameter, which causes inflexibility and brittleness in the bones. Fractures are often transverse and may result from minor trauma (21). Although in tarda form (AD) of osteopetrosis, it has been reported that nearly 40 percent of cases remains asymptomatic (8, 9),

orthopedic presentations – including recurrent fractures, especially in lower limbs – and degenerative arthritis are common. Fractures in osteopetrosis can occur at any age. The healing response is variable. Management of these patients is challenging and operative intervention has a high rate of intraoperative and post-operative complications (17). Patients are at risk for periarticular nonunion that resists treatment. Sometimes joint replacement remains the only viable option (18). There are few case reports of hip arthroplasty and technical difficulties arise in such patients in their follow-up, ranging from six months to 10 years (4, 11, 14, 15, 16, and 19). All published reports concern cemented or hybrid total hip arthroplasty. In 2004, Gwynne, et al., reported the first primary cement-less total hip arthroplasty in an osteopetrosis patient (20). We experienced similar problems in this procedure.

## 4. Conclusion

Since refracture may happen, or because we may need further revision in total hip prostheses in patients with underlying disease like osteopetrosis, use of cement-less prosthesis is worthy of further evaluation.

**Acknowledgements:**

The authors sincerely thank Dr. Ghayyem Hassankhani (director of orthopedics group of Mashhad University of Medical Science) and Prof. Dr. Hallaj Moghadam for performing surgery and their contribution to this study. We thank the personnel of the central operating room at Imam Reza Hospital, especially the head nurse, Mr. Sharifian.

**Corresponding Author:**
Dr. Mohammad Gharehdaghi,
Mashhad orthopedic and trauma research center,
Faculty of medicine, Mashhad University of Medical Sciences, Mashhad Iran
Email: gharehdaghim@mums.ac.ir
Tell: +098.9151152097

## References

1. Marks SC. Pathogenesis of osteopetrosis in the rat: reduced bone resorption due to reduced osteoclast function. Am J Anat 1973; 138:165–78
2. Albers-Scho¨nberg HE. Ro¨ ntgenbilder einer seltenen Knockenerkrankung. Munch Med Wochenschr, 1904; 51:365–8.
3. Shapiro F. Osteopetrosis: current clinical considerations. Clin Orthop. 1993;34–44.
4. Stein I,Stein RO,Beller ML,eds.Living bone in health and disease.4th ed. philadelphia: Lippincott, 1955:1532-4.
5. Ruffa G, Milanaccio C,Sbolgi P, Levato GL, b artocci M, Galasso V,et al.Osteopetrosis and renal acidosis;a new case of this rare syndrome.Minerva pediatr 1995;47:135-40
6. Manusov EG, Douville DR, Page LV,Trivedi DV.Osteopetrosis(‘marble bone’disease).Am Fam Physician 1993;47:175-80
7. Bollerslev J, Andersen Jr PE. Fracture patterns in two types of autosomal dominant osteopetrosis. Acta Orthop Scand 1989;60:110.
8. Armstrong DG, Newfield HJT, Gillespie R. Orthopaedic management of osteopetrosis: results of a survey and review of the literature. J Pediatr Orthop 1999;19:122-32.
9. Bollerslev J, Mosekilde L. Autosomal dominant osteopetrosis. Clin Orthop 1993;294:45-51
10. Ashby ME. Total hip arthroplasty in osteopetrosis: a report of two cases. Clin Orthop 1992;276:214.
11. Beighton P, Horan F, Hamersma H. A review of the osteopetroses. Postgrad Med J 1977;53:507.
12. Gupta R, Gupta N. Femoral fractures in osteopetrosis:case reports. J Trauma 2001;51:997.
13. Milgram JW, Jasty M. Osteopetrosis: a morphologic study of twenty-one cases. J Bone Joint Surg 1982;64A:912.
14. Cameron HU, Dewar FP.Degenerative osteoarthritis associated with osteopetrosis.Clin Orthop 1977:127:148-149
15. Casden AM ,Jaffe FF ,Kastenbaum DM ,Bonar SF.Osteoarthritis associated with osteopetrosis treated by total knee arthroplasty.Report of a case. Clin Orthop. 1989:247:202-207
16. Amit Bhargava, Manesh Vagela, C.M.E. Lennox. Challenges in the management of fractures in osteopetrosis! Review of literature and technical tips learned from long-term management of seven patients. Injury, Int. J. Care Injured. 2009:40;1167-1171
17. Justin P. Strickland, MD, and Daniel J. Berry, MD. Total Joint Arthroplasty in Patients with Osteopetrosis: A Report of 5 Cases and Review of the Literature. Journal of arthroplasty. 2005:20;6;815-820
18. Junwen Wang, Yuanxin Liang, Qingsong Z hang, Jing Jiao, Wusheng. Total joint arthroplasty in a patient with osteopetrosis:10-year follow up. orthopedics 2010
19. D. P. Gwynne Jones, B. F. Hodgson, N. A. Hung. Bilateral, uncemented total hip arthroplasty in osteopetrosis. J Bone Joint Surg [Br] 2004;86-B:276-8.
20. Dahl N, Holmgren G, Holmberg S, Ersmark H. Fracture patterns in malignant osteopetrosis (Albers-Schonberg disease). Arch Orthop Trauma Surg 1992;111(2):121–3.

12/12/2012