Int J Clin Exp Med 2018;11(6):6369-6373
www.ijcem.com /ISSN:1940-5901/IJCEM0064834

Case Report

Cementless total hip arthroplasty in patient with severe haemophilia A: a case report and literature review

Jian-Guang Zhang[1], Qian Chen[1], Lu Chen[1], Wei Zhang[1], Hai-Ying Zhou[2]

[1]Department of Orthopedics, [2]Radiology, Affiliated Hospital of North Sichuan Medical College, Nanchong 637000, Sichuan, China

Received September 1, 2017; Accepted March 28, 2018; Epub June 15, 2018; Published June 30, 2018

Abstract: *Background:* Given the fact that haemophilic arthropathy of the hip is much less common than that of the knee, elbow or ankle, only limited papers with small study populations have been published to report on the surgical management and outcomes of total hip arthroplasty in haemophilic hip arthropathy. Moreover, more than half of the studies were regarding cemented implants with a relatively high rate of failure. *Case presentation:* We report a 26-year-old male with severe hemophilia A who demonstrated hemophiliac arthropathy in his bilateral hip joint, especially the right hip. Because of financial difficulty, total cementless hip arthroplasty for the right hip was performed under a modified factor VIII replacement therapy by a multidisciplinary team consisted of orthopaedic surgeons, haematologists, physiatrists and physical therapist. The patient underwent a physiotherapeutic postoperative rehabilitation program from the first day after surgery to the 3rd weekend after the patient out of the hospital. The whole process was safely managed without any excess bleeding or adverse effects. One year after the surgery, satisfactory functional outcomes were obtained. *Conclusion:* The authors experienced a cementless total hip arthroplasty in a young patient with severe haemophilic hip arthropathy and achieved satisfactory result without complications in a short follow-up period. It is supported that a successful arthroplasty and adequate replacement of clotting factor combined with a meticulous team approach are the key factors to ensure the good outcomes.

Keywords: Haemophilia, hemophiliac arthropathy, total hip arthroplasty, clotting factor

Introduction

Haemophilia is an X-linked recessive blood coagulation disorder attributed to the deficiency of coagulation factor VIII (haemophilia A) or IX (haemophilia B) with a worldwide prevalence of 1 in 10000 males and 1 in 25000 males respectively [1]. Haemophilic arthropathy is the most frequent musculoskeletal disorder in patients with haemophilia. This disorder is characterized by recurrent haemarthrosis, chronic synovitis and progressive destruction of joint cartilage, leading to considerable pain and functional deficit [2]. Haemophilic arthropathy of the hip is much less common than that of the knee, elbow or ankle, with an incidence of 4%, which may attribute to the anatomy of the hip joint [3]. Total hip arthroplasty (THA) has been considered as an effective treatment to relieve pain and improve functional status for patients with end-stage haemophilic arthropathy of the hip [4-6]. However, excessive bleeding in perioperative period, poor bone quality, and muscle contracture are the most common challenges [5]. To our knowledge, only limited papers with small study populations have been published to report on the surgical management and the outcomes of THA in haemophilic hip arthropathy [5-18] (**Table 1**). Moreover, more than half of the studies have described the results of cemented THA with a relatively high rate of failure [6-11, 13, 14] (**Table 1**). Hence, we aim to report a cementless THA performed in a 26-year old patient with haemophilic arthropathy of the hips under modified factor VIII replacement therapy.

Case report

The patient was a 26-year-old male (height 170 cm, weight 70 kg, body mass index 24.2 kg/m^2) with severe hemophilia A and HBV-positive. Since 20 years ago, he suffered unprovoked and recurrent pain on his shoulders, elbows,

Table 1. Published reports of THA in hemophilic arthropathy

Author	Year	Number of patients (cases)	Fixation type	Follow-up years (range)	Revision (%)	Prosthesis survival rate (%)
Nelson *et al.* [7]	1992	21 patients (22 THA)	Cemented	7.6 (2.7-13.1)	8 (36.4%)	63.6%
Kelly *et al.* [8]	1995	27 patients (34 THA)	26 cemented	8 (1-15)	6 (23.0%)	77.0%
			6 cementless	3	0	100%
			1 hybrid	Not mentioned	Not mentioned	Not mentioned
Lofqvist *et al.* [9]	1996	11 patients (13 THA)	Cemented	7 (1-13)	4 (30.7%)	69.3%
Heeg *et al.* [10]	1998	2 patients (3 THA)	Cemented	5	1 (33.3%)	66.7%
Habermann *et al.* [6]	2007	13 patients (15 THA)	Cemented*	11 (1-30.2)	2 (13.3%)	86.7%
			Cementless*			
			Hybrid*			
Miles *et al.* [11]	2008	24 patients (34 THA)	16 cemented	6.25 (1-17)	4 (11.7%)	88.3%
			4 cementless			
			10 hybrid			
			4 cementless (revision)			
Yoo *et al.* [5]	2009	23 patients (27 THA)	Cementless	7.7 (5-13)	2 (7.4%)	92.6%
Sikkema *et al.* [12]	2011	5 patients (6 THA)	Not mentioned	7.6 (0.6-17.9)	0	100%
Wang *et al.* [13]	2012	16 patients (18 THA)	8 cemented	8.5	2 (11.1%)	88.9%
			3 cementless			
			7 hybrid			
Ruosi *et al.* [14]	2013	2 patients (2 THA)	Cemented	4	0	100%
Lee *et al.* [15]	2015	17 patients (21 THA)	Cementless	11.2 (10-17.4)	3 (14.3%)	85.7%
Carulli *et al.* [16]	2015	23 patients (23 THA)	Cementless	8.4 (3.1-13.7)	0	100%
Wu *et al.* [17]	2017	21 patients (24 THA)	23 cementless	9.4 (5-15)	0	100%
			1 hybrid			
Strauss *et al.* [18]	2017	45 patients (49 THA)	9 cemented	11.5 (3-32)	5 (10.2%)	89.8%
			33 cementless			
			7 hybrid			

Notes: THA, total hip arthroplasty. *In this study, the cemented, cementless and hybrid implants were used for the total hip replacement, but the number of each implant was not mentioned.

knees and ankles with no swell and reduce of joint function, which can relieve itself. There was slowly progressive swelling and painful on the hips which had no apparent cause since 8 years ago. Because of financial difficulty, the patient was given no treatment about this. Since 2 months before, the swelling and pain on the right hip increased rapidly and the joint function reduced. He came to our outpatient clinic and was diagnosed as haemophilia A with the clotting factor VIII level of 1%. He had a history of bleeding from penis after slight trauma and blood transfusion at the age of 3 years. There was a history of haemophilia in his family. His elder sister's son has the same illness.

On physical examination, the right hip was tense and stiff, with muscle atrophy at the right lower limbs and a limb length discrepancy of 1 cm. Its range of motion was 85° of flexion, 5° of hyperextension, 15° of abduction, 10° of adduction, 5° of intorsion, and 25° of extorsion, and the Harris score was 46. The appearance of the left hip is no different from original, with a 115° of flexion, 10° of hyperextension, 30° of abduction, 10° of adduction, 15° of intorsion, 30° of extorsion, and a Harris score of 70. The haemoglobin level was 107 g/L, the clotting factor VIII level was 1%, no inhibitor for VIII factor was presented, and activated partial thromboplastin time was 92.8 seconds(normal reference range: 20-40 seconds). The plain X-ray images of hips which were performed with standard anteroposterior and oblique projections showed extensive joint destruction, with flattening of the femoral head, subchondral cysts and osteophytes in bilateral hips, especially the right hip (**Figure 1**). According to the radiologic changes, the right hip joint obtained a Petterson score of 11/13, while the left one gave a score of 7/13. Our

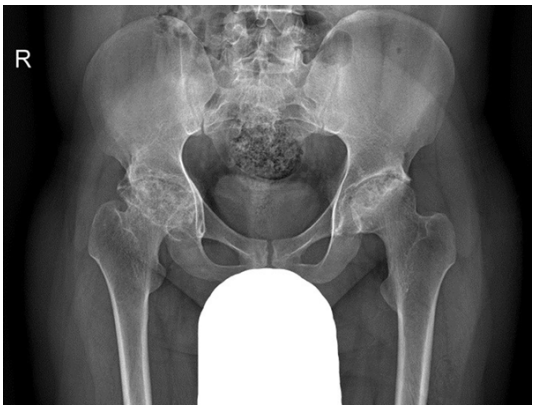

Figure 1. The plain X-ray image of hips with standard anteroposterior projection. Extensive joint destruction, with flattening of the femoral head, subchondral cysts and osteophytes in bilateral hips were showed, especially the right hip.

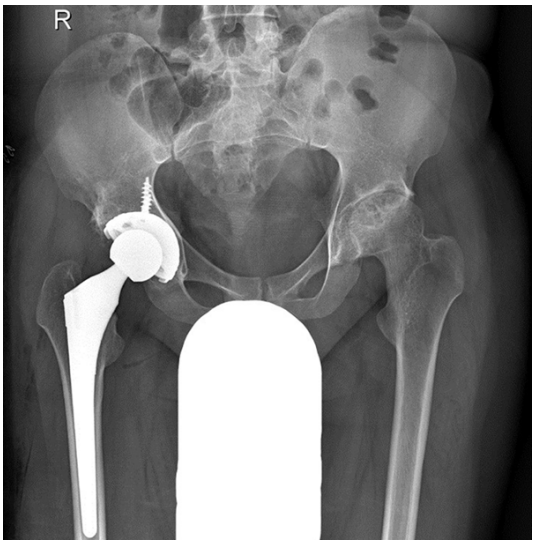

Figure 2. The plain X-ray image of hips after right total hip arthroplasty.

working diagnosis was haemarthrosis A with haemophilic arthropathy of the bilateral hips.

Because of financial difficulty, the patient only underwent THA for the right hip which was more severe under modified factor VIII replacement therapy. Factor VIII (Taibang Inc, Shangdong, China) was given intravenously 12 hours before surgery to maintain the factor level of approximately 100% on the day of surgery. Cementless THA was performed via a posterolateral approach in lateral decubitus position with the standard procedure used in other arthritis deformities under general anesthesia and antibiotic prophylaxis (**Figure 2**). Synovial hypertrophy at all part of the joint, multiple bone and cartilage defect were found intraoperatively. A Ceramic-Head acetabular shell (Trident®, Howmedica Osteonics Corp, Mahwah, NJ, US) and a Secur-Fit® femoral stem (Howmedica Osteonics Corp) were implanted. The acetabular liner was ultrahigh-molecular-weight polyethylene (Trident X3®, Howmedica Osteonics Corp), coupled with a 32 mm cobalt chrome alloy. Due to the severe abnormalities of the acetabulum and the femur head, the operation time was extended to 110 minutes instead of 70 minutes usually required for this surgery. The intraoperative bleeding amount was 600 mL and 3 units of red cell suspension were given.

A protocol was used for the management of postoperative bleeding and other complications by a multidisciplinary team consisted of orthopaedic surgeons, haematologists, physiatrists and physical therapist. Based on 2010 World Federation of Haemophilia (WFH) guideline [19], replacement treatment was administered by bolus of factor VIII concentrates to achieve trough levels of 80% in the first 72 h postoperatively and 40% for the following 10 days. The factor concentrates were dosed twice daily. Meanwhile, the patient underwent a physiotherapeutic postoperative rehabilitation program as follows: passive hip's mobilization on the first day after surgery; passive mobilization followed by sitting position in bed for one hour three times a day on the second day after surgery; walking with specific walking device on the third day after surgery. This program continued in a rehabilitation center for other 3 weeks after the patient out of the hospital.

One year after the surgery, satisfactory functional outcomes were obtained. The patient can walk well without any device, and no pain was complained for the right hip. The articular range of motion was substantially restored (flexion 0-100° from 0-85°; abduction 0-35° from 0-15°; adduction 0-15° from 0-10°; intorsion 0-15° from 0-5°; extorsion 0-30° from 0-25°), and no infection was recorded. The Harris Hip Score improved from 46 to 96 after the operation.

Discussion

With the improvement of prophylaxis and adequate replacement of clotting factors, THA is recommended as an effective treatment to

relieve pain and improve functional status for the patients with end-stage haemophilic arthropathy of the hip. According to the published studies, short- and long-term outcomes in THA for haemophiliacs have been satisfactory [3, 6, 15-18]. Nevertheless, excessive bleeding in perioperative period is a great challenge [3, 6], and the incidence of postoperative complications in patients with haemophilia was significantly higher than that in nonhemophilic patients [12, 13, 18]. Hence, adequate replacement of clotting factor combined with a meticulous team approach is the key factor to ensure the good outcomes.

According to 2010 WFH guideline, Factor VIII levels should be maintained at 120% preoperatively, at 60-80% in the first 72 hours postoperatively and at 50% on the 4-14th days postoperatively [19]. However, because of the patients' financial difficulty and limited source of coagulation factor in developing country, the coagulation factor substitution protocol is often modified and achieved good outcomes. Nelson *et al*. [7] modified and adjusted to maintain Factor VIII levels of 100% for the period of operation and 48 h postoperatively, and then at 40% for at least 12 days after operation. The amount of perioperative blood loss was approximately 300-1000 ml, and average 2-3 units of red blood cell transfusion were given. In a recent study [20], the coagulation factor levels were kept at approximately 100% on the day of operation; 80%, 60% and 40% for postoperative days 3, 6 and 9, respectively; then maintaining at 20-40% until suture removal. The intraoperative blood loss and wound drainage in this study were similar to that reported in haemophiliac patients [7] and nonhaemophilic patients [21]. In our case, factor VIII levels were only kept at 100% preoperatively, at 80% in the first 3 days postoperatively and 40% for the following 10 days. Nevertheless, the intraoperative blood loss was similar to that reported in haemophiliac patients [20] and nonhaemophilic patients [21], and no excessive bleeding occurred in the postoperative period. The modified coagulation factor substitution strategy in this study has also obtained favorable results.

In addition, THA in haemophilic arthropathy has been considered as an effective tool to relieve pain and improve functional status for patients. Historically, substantially positive outcomes have been reported, but most of these outcomes have been due to the use of old generation cemented implants with a higher revision rate [6-11, 13] than those for the general population, which is around 5%-8% at 10 years [22]. The difference has been attributed to the poor bone quality, and the microhaemorrhages at the bone-cement interface preventing adequate fixation, especially the latter [5, 11].

With the development of modern hip implants, new generation cementless implants, which are characterized by high technologic materials and coatings able to show higher bioactivity in terms of osseointegration with respect to old generation components [23], are used in severe hip haemophilic arthropathy [5, 15-18]. According to recent literature, cementless THA in haemophilic patients showed better excellent results with less complications, and a higher prosthesis survivorship than cemented THA within a short or long follow-up period [5, 16-18]. Moreover, an interesting comparison between uncemented Metal-on-Metal versus Ceramic-on-Polyethylene THAs in a series of 12 haemophilic patients with a mean follow-up of 10.4 years was reported [24]. It was demonstrated that the latter implants were superior. Based on the good outcomes of the new generation cementless THAs and the very young age of the patient, we preferred a cementless Ceramic-on-Polyethylene coupling in the case we reported and obtained a good result in a short follow-up period of 1 year after operation.

Conclusion

In conclusion, we performed a cementless THA in a young patient with severe haemophilic hip arthropathy and achieved satisfactory result without complications in a short follow-up period. It is supported that a successful arthroplasty and adequate replacement of clotting factor combined with a meticulous team approach are the key factors to ensure the good outcomes.

Disclosure of conflict of interest

None.

Address correspondence to: Lu Chen, Department of Orthopedics, Affiliated Hospital of North Sichuan Medical College, 63 Wenhua Road, Nanchong 63-7000, Sichuan, China. Tel: +86-817-2262410; Fax: +86-817-2262410; E-mail: nsmcchenlu@163.com

References

[1] Rodriguez NI, Hoots WK. Advance in haemophilia: experimental aspect and therapy. Pediatr Clin N Am 2008; 55: 357-376.

[2] Jansen NW, Roosendaal G, Lafeber FP. Understanding haemophilic arthropathy: an exploration of current open issues. Br J Haematol 2008; 143: 632-640.

[3] Carulli C, Rizzo AR, Innocenti M. Hip arthropathy in haemophilia. J Clin Med 2017; 6: E44.

[4] Wiedel J, Stabler S, Geraghty S, Funk S. Joint replacement surgery in hemophilia. Treatment Hemophil 2010; 50: 1-12.

[5] Yoo MC, Cho YJ, Kim KI, Ramteke A, Chun YS. The outcome of cementless total hip arthroplasty in haemophilic hip arthropathy. Haemophilia 2009; 15: 766-773.

[6] Habermann B, Eberhardt C, Hovy L, Zichner L, Scharrer I, Kurth AA. Total hip replacement in patients with severe bleeding disorders. A 30 years single center experience. Int Orthop 2007; 31: 17-21.

[7] Nelson IW, Sivamurugan S, Latham PD, Matthews J, Bulstrode CJ. Total hiparthroplasty for hemophilic arthropathy. Clin Orthop Relat Res 1992; 276: 210-213.

[8] Kelley SS, Lachiewicz PF, Gilbert MS, Bolander ME, Jankiewicz JJ. Hip arthroplasty in hemophilic arthropathy. J Bone Joint Surg Am 1995; 77: 828-834.

[9] Lofqvist T, Sanzen L, Petersson C, Nilsson IM. Total hip replacement in patients with hemophilia: 13 hips in 11 patients followed for 1-16 years. Acta Orthop Scand 1996; 67: 321-324.

[10] Heeg M, Meyer K, Smid WM, Van Horn JR, Van der Meer J. Total knee and hip arthroplasty in haemophilic patients. Haemophilia 1998; 4: 747-751.

[11] Miles J, Rodriguez-Merchan EC, Goddard NJ. The impact of haemophilia on the success of total hip arthroplasty. Haemophilia 2008; 14: 81-84.

[12] Sikkema T, Boerboom AL, Meijer K. A comparison between the complications and long-term outcome of hip and knee replacement therapy in patients with and without haemophilia; a controlled retrospective cohort study. Haemophilia 2011; 17: 300-303.

[13] Wang K, Street A, Dowrick A, Liew S. Clinical outcomes and patient satisfaction following total joint replacement in haemophilia - 23-year experience in knees, hips and elbows. Haemophilia 2012; 18: 86-93.

[14] Ruosi C, Rossi D, Coppola A, Liccardo S, Granata F, Barbato A, Cimino E, Di Minno MN. Our experience about the particular surgical management of total hip replacement in two patients with severe haemophilic arthropathy: case report. Haemophilia 2013; 19: e184-e186.

[15] Lee SH, Rhyu KH, Cho YJ, Yoo MC, Chun YS. Cementless total hip arthroplasty for haemophilic arthropathy: follow-up result of more than 10 years. Haemophilia 2015; 21: e54-e58.

[16] Carulli C, Felici I, Martini C, Civinini R, Linari S, Castaman G, Innocenti M. Total hip arthroplasty in haemophilic patients with modern cementless implants. J Arthroplasty 2015; 30: 1757-1760.

[17] Wu GL, Zhai JL, Feng B, Bian YY, Xu C, Weng XS. Total hip arthroplasty in hemophilia patients: a mid-term to long-term Follow-up. Orthopaedic Surgery 2017; 9: 359-364.

[18] Strauss AC, Rommelspacher Y, Nouri B, Schmolders J, Wimmer MD, Oldenburg J, Pennekamp PH, Schmolders J. Long-term outcome of total hip arthroplasty in patients with haemophilia. Haemophilia 2017; 23: 129-134.

[19] Srivastava A, Brewer AK, Mauser-Bunschoten EP, Key NS, Kitchen S, Llinas A, Ludlam CA, Mahlangu JN, Mulder K, Poon MC, Street A; Treatment Guidelines Working Group on Behalf of The World Federation Of Hemophilia. Guidelines for the management of hemophilia. Haemophilia 2013; 19: e1-e47.

[20] Zhai J, Weng X, Lin J, Qian W, Guo S. Efficacy of a modified coagulation factor substitution for total hip arthroplasty in patients with end-stage haemophilic arthropathy. Blood Coagul Fibrinolysis 2017; 28: 24-27.

[21] von Roth P, Perka C, Dirschedl K, Mayr HO, Ensthaler L, Preininger B, Hube R. Use of Redon drains in primary total hip arthroplasty has no clinically relevant benefits. Orthopedics 2012; 35: e1592-e1595.

[22] Corbett KL, Losina E, Nti AA, Prokopetz JJ, Katz JN. Population-based rates of revision of primary total hip arthroplasty: a systematic review. PLoS One 2010; 5: e13520.

[23] Matassi F, Botti A, Sirleo L, Carulli C, Innocenti M. Porous metal for orthopedics implants. Clin Cases Miner Bone Metab 2013; 10: 111-115.

[24] Panotopoulos J, Ay C, Schuh R, Domayer S, Funovics P, Trieb K, Wanivenhaus H, Windhager R. Comparison of metal on metal versus polyethylene-ceramic bearing in uncemented total hip arthroplasty in patients with haemophilic arthropathy. Int Orthop 2014; 38: 1369-1373.