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Cases Series

Simultaneous bilateral total knee prosthesis in patients with severe hemophilia with high response inhibitors.

Abstract

Two cases of simultaneous bilateral total knee arthroplasty are reported in two patients with severe hemophilic arthropathy with high response inhibitors. In the world literature there is only one case report, the first patient aged 38 years and the second aged 42 years, with severe joint destruction of both knees, both underwent a simultaneous bilateral total knee arthroplasty, where two teams with the same experience start with one knee and the next team goes one or two steps back, that optimizes the use of factor VIII, which in a country with an emerging economy, makes performing a surgical procedure in a patient with hemophilia very difficult, and especially in one with inhibitors. Recovery and rehabilitation was global, patient satisfaction was quite favorable and there was an improvement in their quality of life.

Key-words

Knee prosthesis, Hemophilia, Inhibitors, Hemophilic Arthropathy.

Introduction

Hemophilia A and B are congenital disorders linked to the X chromosome, which are caused by the absence or decrease of clotting factor in the plasma, in hemophilia A, factor VIII deficiency and factor IX in hemophilia type B. These coagulation disorders cause recurrent bleeding that commonly occurs in the musculoskeletal system in more than 90% of cases, its most common manifestation being joint bleeding, the final consequence of which is the development of hemophilic arthropathy [1], characterized by synovial hypertrophy. With the destruction of the joint surface, bone damage occurs with joint stiffness, pain and permanent severe functional disability; the knee (target organ) being the most affected joint in 44 to 50%, followed by the elbow 25%, ankle 15%, shoulder 8%, hip 5% approximately; This is a disabling disease that decreases the quality of life of the hemophilia patient. The use of the deficit factors VIII or IX concentrates means that orthopedic surgery can be performed safely and with a high expectation of success [2]. However, up to 30% of patients with hemophilia can develop inhibitors, this being the most common and serious complication of replacement therapy in patients with hemophilia A or B. Therefore, the use of recombinant factors concentrates or bypass agents is required. Thus, in the last 40 years, these patients have benefited from elective orthopedic surgery, thanks to multidisciplinary treatment and the administration of the deficient factor, recombinant or bypass agents in patients with and without inhibitors, making the surgery safe and successful. Often more than one joint is affected, so multiple surgical

procedures are indicated in the hemophilia patient. Since prosthetic knee replacement is one of the most commonly indicated major surgeries [3], the advantage of performing a simultaneous bilateral total arthroplasty is that it allows the use of the deficient coagulation factor to be optimized, thus reducing hospital costs, and a global recovery of the functionality. Eventually, this facilitates pain control, shortens the rehabilitation period and everything is performed in only one surgical event, and with a single anesthetic event of maximum 2 hours. And also, it reduces surgical stress with 2 surgical teams working simultaneously if compared to a bilateral arthroplasty in stages or a bilateral total arthroplasty performed one after the other. In Mexico there are currently more than 5,800 patients with hemophilia, and more than 50% of these are treated at the IMSS, and it is estimated that around 70% of these patients already have joint damage, and an alarming 75% do not receive adequate treatment or even no treatment at all.

Case Series

We report two cases of severe hemophilia A with high response inhibitors, both patients with Hepatitis C and referred to the UMAE service of Femur and Knee of the Traumatology Hospital "Victorio de la Fuente Narváez" to be assessed. The first patient had been suffering with disabling pain for more than two years, with limited mobility of both knees and difficulty in walking; irreversible joint damage was observed in his X-rays. Both knees were assessed as stage V on the Arnold-Hilgartner radiological scale (Figure 1). Patient two referred from the hemophilia clinic of the XXI Century Medical Center, had presented joint pain and functional limitation of both knees for more than 5 years, and x-rays revealed severe joint injury, at an estimated radiographic stage V of Arnold-Hilgartner. Both patients underwent general anesthesia without complications, during the surgical event one hour before the skin incision, a dose of rFVIIa of 120 µg/kg (Novoseven) was given [4,5,6], then the treatment was continued every two

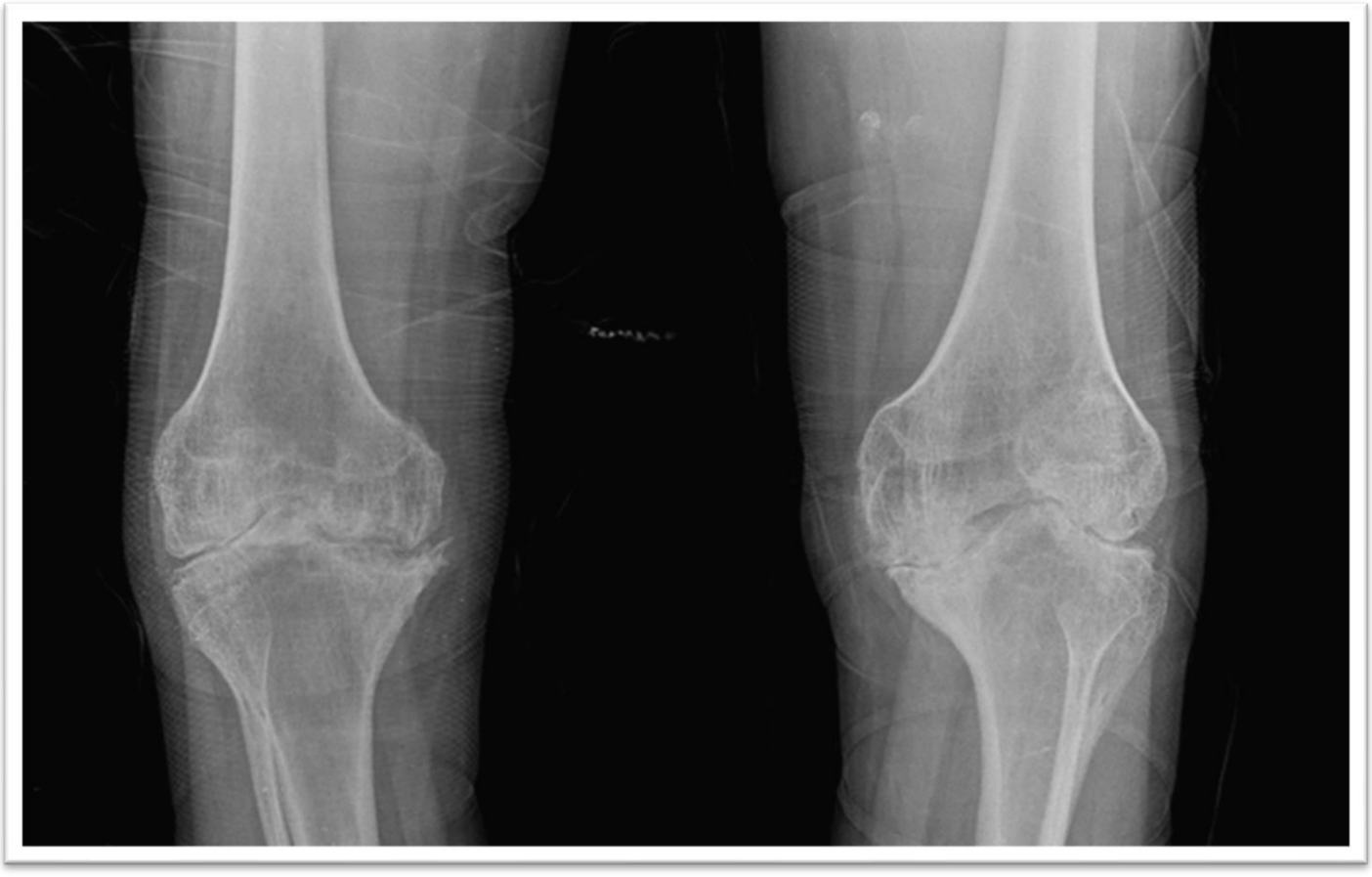

Figure 1: Patient 1 knees with Arnold-Hilgartner radiological scale stage V

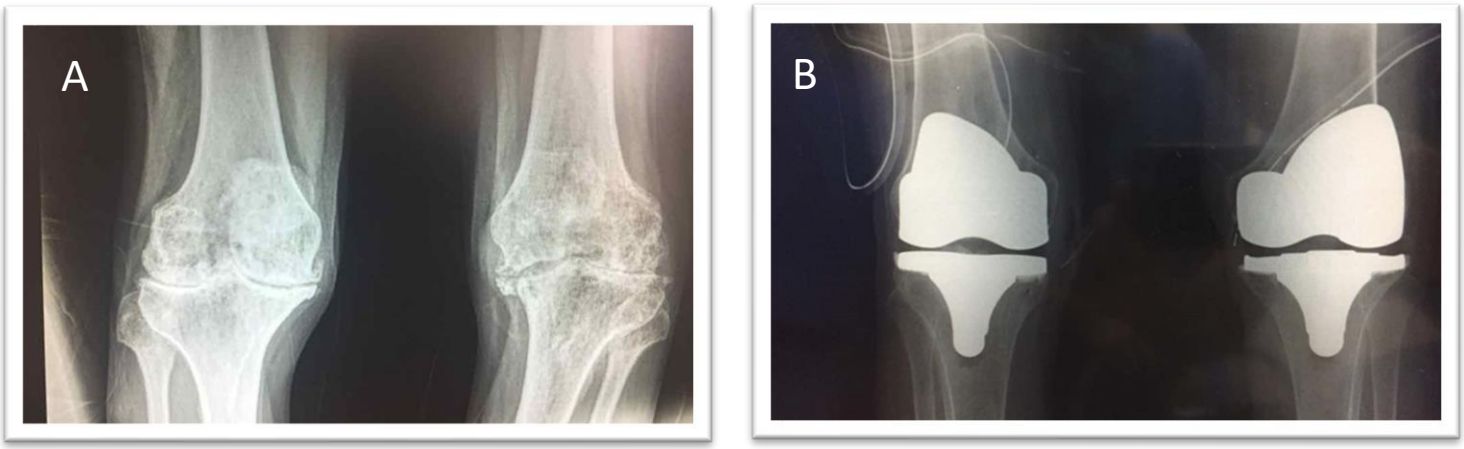

Figure 2: Patient 2 before (A) and after implant (B)

hours the first day with a dose of 90 µg/kg for 48 hrs, and subsequently a scheme of 90 µg/kg was continued every 4 hours, and then the same dose every 6 hrs for three more days. During the closure of the surgical wound, 2.5 g of Tranexamic acid was inserted intra-articularly in each knee and negative pressure drains were placed in each knee, which were opened two hours later. Simultaneous bilateral arthroplasty was performed in both patients with the aforementioned technique; the only complication, was a partial injury of the collateral ligament of the right knee, reported in patients 1 and 2, due to the deformity, resulted in patient 1 with a delay in the healing of the right knee but which resolved without other complications. Currently the patient with 1 to 5 months of evolution presents a mobilization of the right knee of 10° of extension to 70° of flexion, left knee with 15° extension and flexion up to 90°, both without pain and without instability, shown by postsurgical X-rays.

Patient two presented as a complication a partial lesion of the lateral collateral ligament which was repaired during the surgery, a triplanar splint was placed and after 4 months of evolution the right knee was observed with flexion of 10° and extension of 90° and the knee left with extension of 20° plus flexion of 70°, as shown by X-rays (Figures 8, 9 and 10). Significant improvement according to the Hemophilia joint score (HJHS), with a presurgical average of 93 and a postoperative follow-up decreasing the index to 55, which increased functionality, reduced pain and improved quality of life. Reduction in hospital costs and deficit factor concentrates for a single event, surgical time, hospital stay of 5-10 days, rapid ambulation, timely rehabilitation, and the psychological impact was favorable for the patient.

Conclusion:

Arthroplasty can relieve pain and improve the function of symptomatic hemophilic patients with advanced arthropathy, thus improving their quality of life, the knee and hip damage being the most common hemophilia complication and giving the best results [7,8]. With the advent of rVIIa recombinant concentrates and bypass agents, orthopedic and non-orthopedic surgery in patients with hemophilia and inhibitors can be performed with high expectations of success and safety [9].

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References

1) Beeton K; Rodriguez Merchan E. C. et al. Total joint arthroplasty in haemophilia. Haemophilia (2000), 6, 474-481.

2) Rodriguez Merchan E.C.; Jimenez V et al. Surgery in haemophilia patients with inhibitors, with special emphasis on orthopaedics: Madrid experience. Haemophilia. (2010), 16, 84-88.

3) Rodriguez Merchan E.C. Aspects of current management: orthopaedic surgery in haemophilia. Haemophilia (2012), 18, 8-16.

4) Jason M. Howieson A. Kingsley L. et al. Total knee arthroplasty in a patient with hemophilic arthropaty and hig-titer inhibitors. The journal of arthroplasty. Vol. 27, No. 7, 2012.

5) Mehta S, Nelson CL, Konkle BA, et al. Total knee arthroplasty using recombinant factor VII in hemophilia-A patients with inhibitors. A report of three cases. J Bone Joint Surg Am 2004; 86-A: 2519-2521.

6) Solimeno LP, Perfetto OS, Pasta G, et al. Total joint replacement in patients with inhibi- tors. Haemophilia 2006; 12 Suppl 3: 113-116.

7) Faradji A, Bonnomet F, Lecocq J, et al. Knee joint arthroplasty in a patient with haemophilia A and high inhibitor titre using recombinant factor VIIa (NovoSeven): a new case report and review of the literature. Haemophilia 2001; 7:321-326.

8) Carr ME Jr, Loughran TP, Cardea JA et al. Successful use of recombinant factor VIIa for hemostasis during total knee replacement in a severe hemophiliac with high-titer factor VIII inhibitor. Int J Hematol 2002; 75:95-99.

9) Faradji A. Bonnomet F. Lecoq J. et al. Knee joint arthroplasty in a patient with haemophilia A and hig inhibitor titre using recombinant factor VIIa (novoseven): a new case report and rewiew of the literature. Haemophilia, (2001), 7, 321-326.