Hemodialysis Access in a Patient with Severe Hemophilia: Technical Challenges

Murali Manivannan, Amalorpavanathan Joseph, Subramaniyan S Rathinavel, Elancheralathan Kalyanaraman, PS Balakumar, Ilayakumar Paramasivam, B Velladuraichi, Devarajan Ilangoovan, Prathap Kumar Sudalaiyandi, Jayanth Vijayakumar, Krishna Muralidharan

Department of Vascular Surgery and Endovascular Sciences, TNGMSSH, *Institute of Vascular Surgery, Madras Medical College, *Department of Vascular Surgery, Saveetha Medical College, *Department of Vascular Surgery, Government Stanley Medical College, Chennai, Tamil Nadu, India

Abstract

A case report on the challenges in establishing hemodialysis access for a hemophiliac with factor VIII inhibitors. A 23-year-old male patient, a known case of congenital Hemophilia A for 6 months of age, presented with recurrent hemarthroses, uncontrolled hypertension, and azotemia; on evaluation, he was diagnosed to be suffering from chronic kidney disease. He was on factor VIII supplementation for hemophilia and was recently diagnosed with factor VIII inhibitors as he was becoming refractory to treatment. The hemodialysis access for this patient is technically challenging as the patient has blood dyscrasia. Herein discussing the choices we had in this patient and challenges faced by us in securing the hemodialysis access.

Keywords: Chronic kidney disease, factor VIII inhibitors, hemodialysis access for hemophilia

INTRODUCTION

A case report on technical challenges in securing hemodialysis access for a severe hemophilia patient is rarely reported. Hemodialysis access being mandatory for all chronic kidney disease patients makes a vascular specialist role pivotal. The importance in decisionmaking and obstacles faced in such a patient is discussed here.

CASE REPORT

A 23-year-old man was suffering from congenital hemophilia diagnosed at 6 months of age and he had been on regular care of the state run Hemophilia welfare society. He had recurrent hemarthroses and was on factor VIII transfusions. He recently presented to our nephrology department with azotemia and on evaluation, he was diagnosed to be suffering from chronic kidney disease. He was on factor VIII supplementation for hemophilia and was recently diagnosed with factor VIII inhibitors as he was becoming refractory to treatment. The hemodialysis access for this patient is technically challenging as the patient has blood dyscrasia. Herein discussing the choices we had in this patient and challenges faced by us in securing the hemodialysis access.

was pale with blood pressure of 180/100 and no pedal edema or crepitations in lungs to suggest volume overload state.

The routine investigations revealed hemoglobin of 8.6 g/dL, peripheral smear showing evidence of microcytic hypochromic anemia, blood urea of 201 mg/dL, serum creatinine was 11 mg/dL, and the abdomen ultrasonography shows shrunken scarred kidney with increased echoes as well as loss of corticomedullary differentiation suggesting chronic kidney disease. On evaluation for treatment refractory hemarthroses, he was found to have inhibitors to factor VIII by mixing studies and he was managed by administration of activated prothrombin complex concentrates (aPCC) available as Factor Eight Inhibitor Bypass Agent (FEIBA) (Baxter, Belgium). He needed a hemodialysis access which could avoid any major bleeding complication. We decided to insert a permanent tunneled cuff catheter which would not require aPCC/FEIBA during every dialysis.

Address for correspondence: Dr. Murali Manivannan, E-mail: manivannan.murali@gmail.com

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Under ultrasound guidance and monitored anesthesia, the patient had been placed a permanent tunneled cuff catheter in the right internal jugular vein (IJV). Five hundred units aPCC/FEIBA was administered intravenous 30 min before and after the procedure. The procedure was uneventful [Figure 1]. The patient had bleeding from pericatheter site at the right chest wall 6 h after intervention. No hematoma at the neck where IJV was punctured. Thousand units of aPCC/FEIBA administered with compression dressings following which bleeding was controlled.

The patient continued to have similar problem on subsequent postoperative days; extra doses of aPCC/FEIBA was administered along with steroids. We administered “FLOSEAL (BAXTER)” in the pericatheter tract to achieve hemostasis [Figure 2]. The patient had no bleeding for couple of days and again started to bleed from the same site. We took four reinforcing sutures parallel to catheter tract to create hemostasis [Figure 3]. The patient responded well and was discharged.

A month later, he presented with a swelling in the neck at puncture site which was 3 cm × 2 cm size, smooth surfaced, globular in appearance, and nonpulsatile. On palpation, the swelling was compressible with no thrill or auscultatory bruit [Figure 4]. Duplex evaluation showed an eccentric ectasia of IJV possibly due to aneurismal degeneration. We managed conservatively by ultrasonography-guided compression with tight dressing for two sessions following which the venous ectasia had thrombosed and the patient was reassured.

The patient was followed up with duplex monitoring for assessing the size of sac at discharge, 4 and 8 weeks. The sac had decreased in size with a patent hemodialysis access.

The patient had multiple hemodialysis without major complications and less need for FEIBA. Eight months later, the patient was admitted for hemarthroses of multiple joints and seizures. On evaluation, he was found to have severe systemic bleeding and the patient succumbed to the blood dyscrasia.

**DISCUSSION**

Hemophilia, a blue-blooded disease, described to be the cause of hemorrhagic disease in European royalty. This disease was initially described mainly in royal family and inherited in X-linked recessive fashion. There are three variants of hemophilia described as hemophilia A, B, and C which represent deficiency of factor VIII, IX, and XI clotting factors, respectively. Acquired form of hemophilia has been described as well, and it is associated with malignancy, autoimmune

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**Figure 1:** The permanent tunneled cuff catheter being inserted without bleeding complications in perioperative period

**Figure 2:** The FLOSEAL (BAXTER) being injected to the catheter entry site to control bleeding

**Figure 3:** The sutures taken parallel to catheter to control bleeding

**Figure 4:** The patient developed venous ectasia at entry site at internal jugular vein needing compression therapy during follow-up
disease, and pregnancy. These patients require repeated transfusion of the factors to lead a healthy life. When patients have subsequent transfusions over the years, they develop antibodies to factor VIII and therefore lead to inefficiency from future transfusions. This makes them very vulnerable for bleeding. The disease is classified based on the levels of active clotting factor in their blood: mild – 5%–40%, moderate – 1%–5% and severe – <1%. Hemophiliac patients with high titers of inhibitor who experience bleeding episodes or those who undergo surgery are typically treated with recombinant VIIIa or activated Prothrombin complex concentrate (FEIBA).[1-4] KDOQI guidelines insist on early creation of optimally functioning access for best outcomes. The access of choice is an autogenous arteriovenous (AV) fistula in an otherwise healthy individual with preference to distal access sites to proximal ones.[5] In our patient, a person suffering with severe hemophilia and factor VIII resistance, the choices are complicated. We decided to give an access with least possible complications for this patient.

Chronic renal failure is a rarely reported in hemophiliacs and/or individuals with other hereditary clotting disorders, and the dialysis modality for these patients may be a difficult choice. Hemodialysis is a good therapeutic option for patients with a mild type of hemophilia B and end-stage renal disease (ESRD) requiring dialysis. Prolonged aPTT allows for HD without anticoagulation. Hemodialysis had been successfully performed without bleeding episodes in a patient with severe hemophilia A through external arteriovenous shunt in 1977 for 1 year before renal transplantation.[6] In Wrocław, Poland, a 53-year-old man with severe hemophilia B (FIX activity, 0.1%), mild hypertension, and HCV infection who developed ESRD had been on hemodialysis for 6 months.[7] Peritoneal dialysis (PD) is considered to be safer in some patients because of minimized bleeding risk.[8] Inhibitors develop in hemophilia because transfused factor VIII can be seen as a foreign protein and elicits an immune response in much the same way as any other foreign protein might elicit an immune response. However, not all hemophiliacs generate an immune response because they do not recognize factor VIII as foreign or their major histocompatibility complex phenotype is such that a cellular immune response is not initiated. PD is relatively safe in average patients without a coagulation disorder; insertion of a peritoneal catheter is associated with bleeding complications; there is some concern for intraperitoneal or retroperitoneal hemorrhage, especially in hemophiliacs.[8-11] Considering these factors, we planned to place a tunneled cuffed catheter for hemodialysis access as it carries less risk of bleeding during each hemodialysis and less requirements for aPCC/FIBEFA during dialysis. This option also has lesser chance of infection in comparison to PD catheter or temporary catheter.[12]

The postoperative bleeding was present at tunnel site alone, and the episodes were intermittent with resolution of symptoms with FEIBA. Thrombin analogue which bypasses the role of factor VIII in clotting should have helped achieve hemostasis but it wasn’t effective. The mechanical hemostasis was achieved by taking sutures parallel to catheter. The bleeding episodes in this patient was possibly due to the avulsed small pectoral venous branches. Most inhibitors occur at young age in patients with severe hemophilia, but cumulative inhibitor risk increases with age. In severe hemophilia A, it is 30% at the age of 50 and 36% at the age of 75. In moderate and mild hemophilia A, the cumulative risk is 6%, 10%, and 12% at 5, 50, and 75 years of age, respectively. For hemophilia B, the cumulative risk is much lower at the age of 75 (8%) and at all ages compared with hemophilia A.[13] The role of immune tolerance induction for inhibitor eradication in this age group of patients is unknown.[14] Risk of thrombotic complications associated with the use of bypassing agents should be considered in the aging population with hemophilia.[14] Venous ectasia at IJV puncture site is possibly due to degeneration and as any other ectatic vein, it was managed with observation and close follow-up.

**Conclusion**

Young patients with chronic kidney disease requiring maintenance hemodialysis is a major financial burden and mental trauma for the patients. Management of these patients gets complicated when they have congenital bleeding disorder. The insertion of permanent catheter is the best possible option because it gives them a fair chance to return to their livelihood early. The risk of bleeding from PD catheter insertion site or AV fistula anastomosis or the probability of pseudoaneurysm formation at puncture sites in these patients makes the permanent catheter insertion a much better and safe option. A meticulous planning of the procedure and preoperative duplex evaluation of tunnel would help us to prevent bleeding.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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