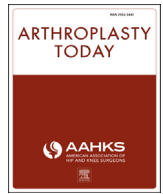




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Arthroplasty in patients with rare conditions

Total knee arthroplasty in a patient with hypofibrinogenemia

Christopher R. Nacca, MD ^{a, b, c, *}, Kalpit N. Shah, MD ^{a, b, c}, Jeremy N. Truntzer, MD ^{a, b, c},
Lee E. Rubin, MD ^{a, c, d, e}^a Warren Alpert Medical School of Brown University, Providence, RI, USA^b Department of Orthopaedic Surgery, Brown University, Providence, RI, USA^c Department of Orthopaedic Surgery, Rhode Island Hospital, Providence, RI, USA^d Department of Orthopaedic Surgery, The Miriam Hospital, Providence, RI, USA^e Division of Adult Reconstruction, Department of Orthopaedic Surgery, Providence, RI, USA

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ABSTRACT

Patients with afibrinogenemia or hypofibrinogenemia present a unique challenge to the arthroplasty surgeon as fibrinogen is a key contributor to hemostasis. Patients with these disorders are known to have a higher risk for postsurgical bleeding complications. We present the case of a patient with hypofibrinogenemia who underwent an elective total knee arthroplasty. Our colleagues in hematology-oncology guided us initially to achieve and maintain appropriate fibrinogen levels in the early perioperative period. However, the patient developed an acute joint effusion and subsequent infection 4 weeks after her initial operation. Her fibrinogen levels were noted to have fallen below the target range by that time, and it was also revealed that the patient failed to follow-up with hematology-oncology to monitor her levels. Based on our review of the available literature, we recommend that patient's fibrinogen levels be closely monitored and maintained ideally >100 mg/dL not only in the initial perioperative window but perhaps for the first 4–6 weeks postoperatively as well.

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Introduction

Afibrinogenemia and hypofibrinogenemia, defined as plasma fibrinogen levels <150 mg/dL, are rare coagulation disorders with an incidence of 1 in 1 million [1,2]. Of all coagulation disorders, fibrinogen deficiency has been reported approximately 8% of the time [3].

Fibrinogen serves as a key contributor to hemostasis by assisting in clot formation, platelet aggregation, and fibrinolysis [4,5]. Treatment for congenital fibrinogen deficiency can consist of replacement with fresh frozen plasma (FFP), cryoprecipitate (cryo), or fibrinogen concentrate (FC). Clinically, symptoms may

vary depending on the severity of the deficiency. Patients with hypofibrinogenemia are typically asymptomatic; however, they may be at risk for bleeding complications when exposed to trauma, pregnancy, or surgery. Those with afibrinogenemia tend to have a higher frequency of bleeding in comparison [3,4].

Unfortunately, there is limited literature and a lack of guidelines regarding the optimal perioperative management of these patients. The few reported cases consist of those with severe or complete deficiency of fibrinogen in the setting of trauma and pregnancy where significant bleeding is often expected [6]. In all cases, current recommendations suggest maintaining fibrinogen level above 100–200 mg/dL to prevent bleeding complications [2,7,8]. Currently, no clinical cases, guidelines, or recommendations exist for patients with congenital fibrinogen deficiency undergoing total joint arthroplasty.

We report the case of a patient with congenital hypofibrinogenemia who underwent an elective total knee arthroplasty (TKA) and unfortunately suffered a postoperative hemarthrosis and acute prosthetic joint infection. We discuss our experience with the perioperative management of this challenging patient.

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* Corresponding author. Rhode Island Hospital Department of Orthopaedics, 593 Eddy Street, Providence, RI 02903, USA. Tel.: +1 401 444 4030.

E-mail address: cnacca@lifespan.org

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Figure 1. Preoperative radiograph of the patient showing osteoarthritic changes which unfortunately not managed sufficiently with nonoperative measures.

Case history

A 67-year-old female had a medical history significant for hypofibrinogenemia and right knee pain related to moderate tricompartmental osteoarthritis. Of note, written informed consent was obtained from the patient for publication of this case report and accompanying images. She presented to our institution after a fall resulting in a closed right patella fracture with an intact extensor mechanism, which was treated conservatively. She continued to complain of right knee pain despite radiographic evidence of complete fracture healing. Physical examination and subsequent radiographs suggested that her ongoing discomfort was more likely secondary to worsening tricompartmental arthritis, particularly involving the patellofemoral joint (Fig. 1). Given her active functional status, it was determined that she would benefit from a total knee arthroplasty.

Her family history was significant for a grandmother who died because of hemorrhagic shock while delivering her father, as well as a daughter with known hypofibrinogenemia. The patient's surgical history is significant for a wisdom tooth extraction, vaginal delivery, gastric band placement, and colectomy for diverticulitis without any significant bleeding episodes. She was diagnosed with hypofibrinogenemia based on abnormal coagulation profile laboratory values obtained due to the positive family history that had prompted an in-depth hematology workup.

Before her elective right TKA, her preadmission laboratories demonstrated a fibrinogen level of 58 mg/dL (normal range: 150–480 mg/dL), international normalized ratio (INR) of 1.3 (normal range: 0.8–1.2), prothrombin time (PT) of 13.2 seconds (normal range: 9.5–12.5 seconds), and activated partial thromboplastin time (aPTT) of 31 seconds (normal range: 24–37 seconds). Hematology was consulted for perioperative management and recommended

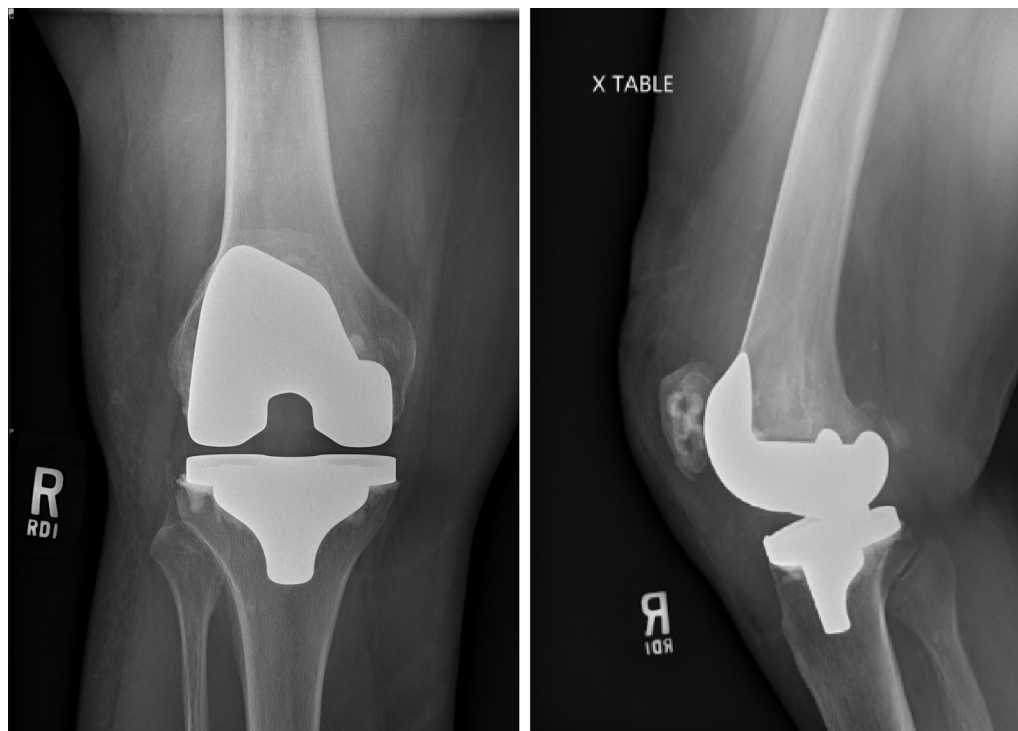


Figure 2. Postoperative radiographs after the index operation for the total knee replacement.

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