Pulmonary Embolism after Ankle Fusion in a Patient with Hemophilia A: A Case Report

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Abstract

Deep vein thrombosis (DVT) and subsequent pulmonary embolism (PE) are uncommon postoperative complications of operative procedures for treating injuries of the foot and ankle. Because the disorder of hemophilia A prevents blood clotting and increases bleeding, patients with this condition have been even less likely to develop DVT and PE. We present a 36-year-old man with hemophilia A in whom operative ankle fusion for treating hemophilic arthropathy of the left ankle led to DVT and PE. After decreasing dosage of antihemophilic medication and administering enoxaparin, the symptoms improved and the patient was discharged from the hospital on postoperative day 5. At 3-month follow-up with continued dosage, no complications were reported. Surgeons should be aware of possible DVT and PE in patients with hemophilia A and consider multidisciplinary efforts to successfully treat the resultant symptoms.

Introduction

Deep vein thrombosis (DVT) is an uncommon complication after operative treatment of the foot and ankle, with a documented prevalence between 0.03% and 0.27% in patients after undergoing elective hindfoot fusion.[1,2] Currently, about 43% of physicians have recommended routine use of chemical thromboprophylaxis in operatively treating the foot and ankle of patients without history of DVT.[3] However, the American Academy of Orthopaedic Surgeons has no specific recommendations for DVT prophylaxis after foot and ankle procedures.[4]

In particular, DVT and subsequent pulmonary embolism (PE) rarely occur in patients with hemophilia A, an X-linked disorder of factor VIII. Patients with this condition have difficulties with forming blood clots and are thus at risk for developing spontaneous bleeding into joints, which can severely damage cartilage and result in arthritis. Owing to infrequent blood clotting, it is counterintuitive that patients with hemophilia A could develop blood clots associated with DVT and PE. We performed left ankle fusion to treat hemophilic arthropathy in a 36-year-old man with hemophilia A, which resulted in development of DVT and PE.

Case Report

A 36-year-old man with a medical history of hemophilia A presented to our institution with pain and history of bleeding in the left ankle. Previously, the patient had undergone right ankle fusion for treating severe degenerative hemophilic arthropathy, resulting in almost complete resolution of symptoms. He returned to the clinic several years later, with progressive worsening of pain in the left ankle. Radiographs obtained at evaluation showed arthritic changes suggestive of hemophilic arthropathy about the left ankle joint. Loss of joint space and signs of osteophytes were noted (Figure 1). The patient had considerable pain despite using previous nonoperative methods for treatment.

Preoperatively, the patient underwent a routine evaluation led by hematologists and oncologists. A perioperative plan for dosage of antihemophilic factor VIII (50 units/kg) was established, which included one preoperative dose in the morning and two doses per day after the procedure, with close follow-up monitoring of factor VIII levels.

A total of 5090 units of antihemophilic factor VIII was administered to the patient. He underwent ankle fusion without observed complications (Figure 2) and was after admitted to the hospital for standard care. On postoperative day 1, he began receiving doses of antihemophilic factor VIII twice per day. Initial outcomes were promising, with limited pain and no symptoms of increased bleeding in the left ankle. On postoperative day 3, labored breathing was noted and thereby oxygen levels were increased. The patient underwent a computed tomography angiogram of the pulmonary arteries, during which PE was diagnosed in the right main pulmonary artery with extension into the right-middle and lower-lobe branches. Hematologists and oncologists were consulted. Subsequently, the dosage of antihemophilic factor VIII was decreased to once a day and

the patient was administered low-molecular weight heparin (ie, enoxaparin) at doses of 1.5 mg/kg for therapeutic purposes. Clinical signs of improvement were observed during the next 2 days, without further progression of symptoms. He was discharged on postoperative day 5.

Based on factor VIII levels at discharge, the dosage of antihemophilic factor VIII was reduced by 50% and the patient remained on enoxaparin for therapeutic treatment of PE. He was followed at regular intervals by orthopaedic, hematologic, and oncologic physicians. At 3 months postoperatively, symptoms of pain and PE had resolved, without any additional complications. Use of antihemophilic factor VIII and enoxaparin were discontinued.

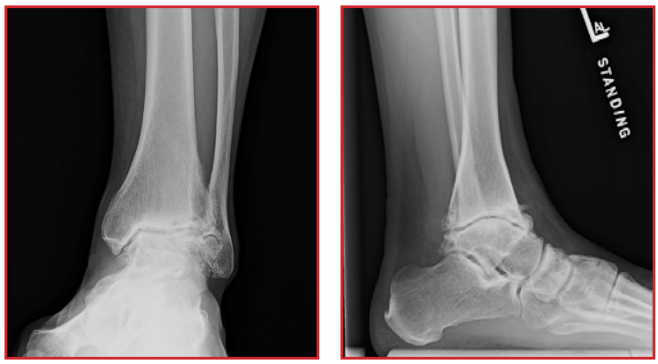

Figure 1. Preoperative radiographs of the left ankle joint, showing arthritic changes suggestive of hemophilic arthropathy, with loss of joint space and presence of osteophytes.

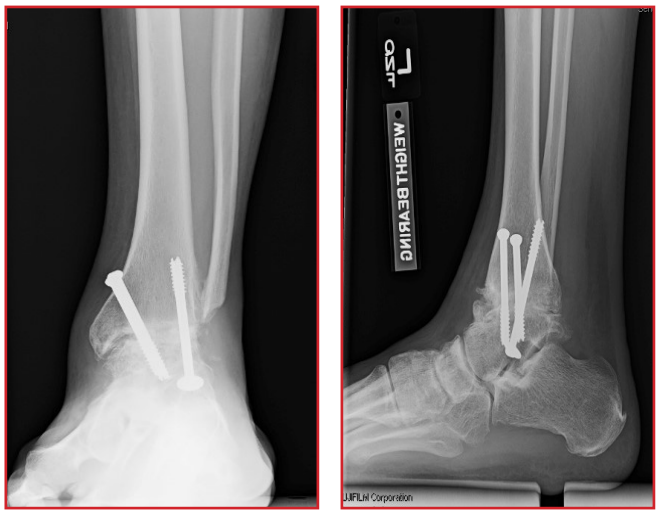

Figure 2. Postoperative radiographs of the left ankle joint obtained immediately after ankle fusion, showing initial improvement of joint space.

Discussion

Hemophilia A is characterized by a deficiency in coagulation of factor VIII, which typically results in hemorrhagic complications after operative treatment. However, the results of the current case suggest that the disorder does not completely prevent development of DVT and PE. Only 13 cases have found non–catheter-related thrombus in patients with hemophilia A, in which none involved operative treatment of the foot and ankle.[5] None of these cases involve surgery about the foot and ankle.

Patients with hemophilia A undergoing replacement therapies of factor VIII (eg, factor VIII inhibitor bypassing activity and recombinant activated factor VII) may have a closer risk factor for DVT and PE to that of the patients without the disorder. In other words, the most prevalent risk factor in patients with hemophilia A could be the presence of replacement therapies. However, the incidence of DVT and PE in patients with hemophilia A undergoing replacement therapies has been low, in which no definitive protocol exists for treating perioperative thromboprophylaxis in major hemophilia treatment centers. Previous case studies have noted that some patients may have a concomitant inheritable prothrombotic condition such as protein c deficiency or factor V Leiden.[5] In treating patients with family history of DVT and PE, Dargaud et al[6] suggested performing a complete thrombophilic evaluation of inheritable risk factors to identify potential indications for thromboprophylaxis.

No standard protocol exists for optimizing the type or dosage of anticoagulation medication using appropriate techniques for replacing clotting factors (ie, factor VIII). Additionally, consensus has not been established regarding the duration of such treatment. Reported cases suggest that low-molecular weight heparin or unfractionated heparin should be used initially for replacement of factors VIII and IX. To simultaneously achieve peak activity levels of factors VIII and IX, therapeutic dosage of heparin has been recommended in precise coordination with replacement medication. The authors recommended a treatment duration of 3 and 6 months for temporary risk factors (eg, operative treatment) and permanent risk factors (eg, inheritable traits), respectively.[6]

The occurrence of DVT and PE in patients with hemophilia A is rare. A detailed personal and family history of clotting should be relayed to hematologists to allow for appropriate workup. The plan for treatment with thromboprophylaxis, if indicated, can be completed before operative treatment. Orthopaedic surgeons should be aware of these possible complications in patients with hemophilia A, in which successful treatment can involve multidisciplinary efforts with hematologists and oncologists.

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Conflict of Interest

The authors report no conflicts of interest.

Informed Consent

The patient was informed that the data concerning the case would be submitted for publication, and he provided verbal consent.

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