An Unusual Case of Iliofemoral Deep Venous Thrombosis in a Patient with Hemophilia B

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ABSTRACT

Hemophilia B is a disorder of coagulation due to deficiency or dysfunction of factor IX. It is characterized by provoked or spontaneous bleeding episodes, most commonly in the joints and muscles. Patients with hemophilia are at a lower risk of developing venous thromboembolism as compared to the normal population. However, when enough risk factors culminate, these patients can develop deep venous thrombosis and related sequelae. In the setting of hemophilia, the management of DVT becomes challenging due to the risk of increased bleeding tendency. Here we present a case of a young gentleman with hemophilia B who developed lower limb DVT complicated by lumbosacral plexopathy.

Keywords: Hemophilia B, Deep venous thrombosis, venous thromboembolism

INTRODUCTION

Hemophilia-B is a rare X-linked bleeding disorder that results from a deficient or dysfunctional factor IX.[1] Although hemophilia B poses a lower tendency of bleeding compared to hemophilia A,[2] prevention is essential before surgery and to avoid the complications of spontaneous bleeding such as hemarthrosis and muscle hematomas.[3] On the opposite spectrum, venous thromboembolic events (VTE) rarely occur due to the nature of the disease. Cases of deep venous thrombosis (DVT) have been reported in a minority of patients with hemophilia. The probable risk factors in such patients include orthopedic surgery with postsurgical immobilization, and administration of factor IX concentrates and activated prothrombin complex concentrates.[4] In a nonsurgical event, an expanding intramuscular hematoma can compress a vein and lead to the development of DVT.[5] Usually, it is the presence of several risk factors in a hemophiliac patient that results in VTE. Prophylaxis against DVT is routinely used in patients without bleeding disorders; however, for patients with hemophilia, the pharmacological prophylaxis and management of DVT are challenging because of the concurrent bleeding risk.[6]

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Here we present the paradoxical development and challenging management of the extensive proximal lower limb DVT in a patient with simultaneous bleeding from hemophilia-B.

CASE PRESENTATION

A 32-year-old male with a background diagnosis of hemophilia b, presented with weakness and pain in the right leg. He was initially assessed by general surgery for possible iliopsoas abscess. While optimizing him for surgical intervention, including transfusion of factor IX concentrates, he developed deep venous thrombosis (DVT) in the right leg, confirmed by a Doppler ultrasound. An abdominopelvic ultrasound was suggestive of iliopsoas hematoma. Consequently, the surgical intervention was abandoned and the patient was transferred to the medical ward for conservative management and further workup.

The patient was in obvious discomfort at the time of examination. He had a right lower limb swelling extending from the calf to the upper thigh. The power was reduced in that limb accompanied by a burning sensation. The rest of the examination findings were within normal limits. The lab workup returned within normal limits except raised prothrombin time (PT) and international normalized ratio (INR). With a background diagnosis of hemophilia B and the examination findings of reduced power and burning sensations, a compressive hematoma was suspected. An MR was requested that showed a large hematoma in the right iliacus and part of the psoas muscle compressing the right internal and external iliac veins as well as the lumbosacral nerve roots (Fig 1,2).

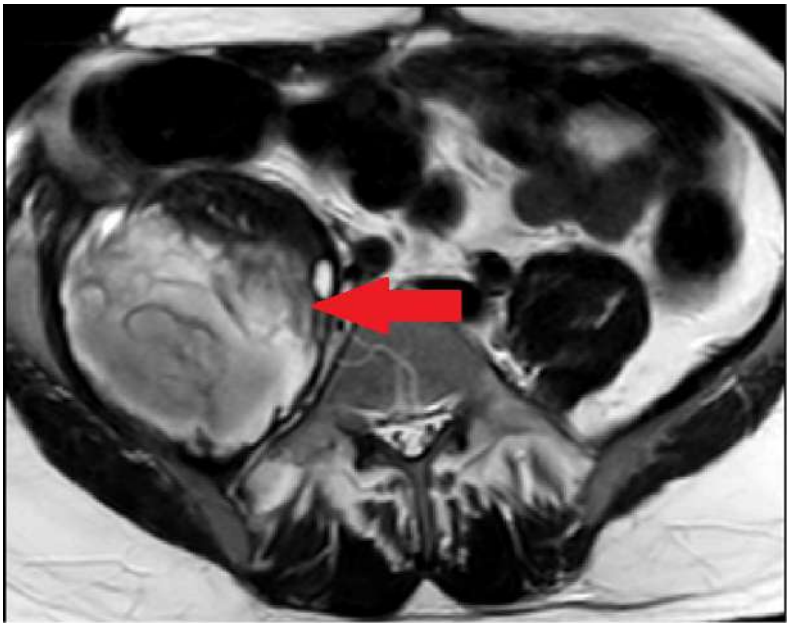

Figure 1: A large hematoma in the iliopsoas muscle (red arrow)

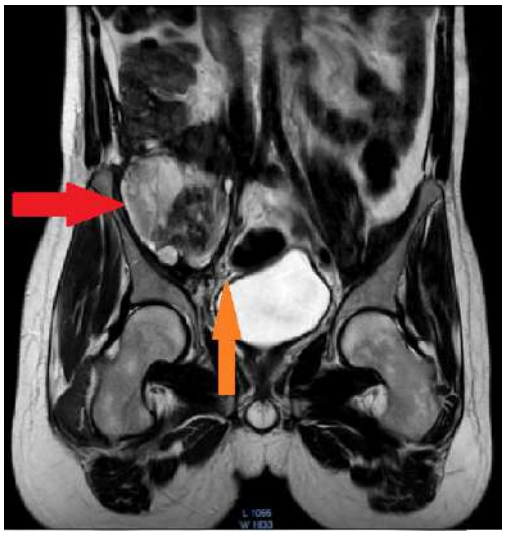

Figure 2: The same hematoma (red arrow) compressing the vessels (orange arrow)

After consultation with hematology and vascular surgery, the patient was started on an alternate-day regime of enoxaparin and factor IX concentrates. The response to treatment will be assessed at follow-up visits at regular intervals.

DISCUSSION

Hemophilia B is an X-linked bleeding disorder predominantly affecting males. The etiologies involve mutations in the F9 gene on the X chromosome (hereditary) or the production of antibodies against factor IX (acquired). The disease is inherited as X-linked recessive. Hemophilia B is characterized by a deficiency in the activity of clotting factor IX that manifests as prolonged or recurrent bleeding after injuries or surgical procedures. Patients with severe Hemophilia B (<1% factor IX) are at risk of spontaneous bleeding. The severe disease is usually diagnosed during the first two years of life.[7]

The occurrence of pre and post-op DVT in people with hemophilia is rarely reported in the literature.[8] Most of the cases seem to be related to the use of factor IX replacement and immobility following surgery.[9] Our patient had a massive iliopsoas hematoma compressing the iliac veins and also received factor IX concentrates before his anticipated surgery. Together with the immobility from compressive plexopathy, these factors were considered to be the major predisposing risks in our patient.

There are no clear guidelines regarding the optimal treatment of DVT in patients with

Hemophilia because of the rare occurrence of these events.[10] Most of the reported cases have used heparins coupled with factor VIII/IX replacement. The use of oral anticoagulants is not favored due to increased bleeding tendencies and the lack of monitoring parameters.[8]

Our patient was started on a combination of alternate-day enoxaparin and factor IX replacement after searching the literature and consultation with the hematologist. The response to treatment is satisfactory with no bleeding episodes until now. The patient is being followed up with clinical evaluation, coagulation profile, CTPA, and ultrasonographic evaluation.

LEARNING POINTS

DVT is a rare occurrence in patients with hemophilia. The risk factors vary but most of the cases seem to be related to a combination of factors including factor replacement and post-op immobility. However, in our case, the major etiology was the development of a compressing hematoma in the iliopsoas muscle. There are no clear guidelines for the treatment of DVT in hemophilia, however, the combination of heparins and factor IX has been used with success.

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