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Perioperative Management of a Patient with Hereditary Hemorrhagic Telangiectasia and Deep Vein Thrombosis: A Case Report

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Abstract

Hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, is an autosomal dominant disorder characterized by telangiectasias, frequent episodes of epistaxis, and multiple arteriovenous malformations (AVMs). AVMs can develop in any organ and may lead to serious consequences such as embolic complications and high-output cardiac failure. The bleeding tendency of these patients should be considered during surgical procedures. In addition, although it is not common, HHT patients may also have a risk of venous thromboembolism.

Here, we present the perioperative management of a man with the coexistence of HHT and deep vein thrombosis, conditions predisposing him both to a bleeding tendency and to thromboembolic risk as well.

Keywords: Hereditary hemorrhagic telangiectasia; Perioperative care; Venous thrombosis

Introduction

Hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, is an autosomal dominant disorder characterized by recurrent epistaxis, mucocutaneous telangiectases and arteriovenous malformations (AVMs) in the lungs, liver or central nervous system. The approximate prevalence of the disorder is 1 in 5,000-10,000 individuals [1,2]. The majority of HHT patients may be classified into two types depending on the specific genetic mutations; HHT1 is caused by mutations in the endoglin (ENG) gene and HHT2 is caused by mutations in ACVRL1, which encodes activin receptor-like kinase 1 [3,4]. Pulmonary AVMs are known to be more common in HHT1, while hepatic AVMs are more common in HHT2. However, accurate determination of the disease subtype may not be made based solely on the clinical manifestations [3].

Due to angiodysplasia, HHT patients are susceptible to blood vessel damage and bleeding from physical stimuli. Therefore, careful attention should be paid to hemorrhagic side effects during surgery. In addition, anesthesia should be performed with care because AVM may be associated with serious complications such as embolic events and high output cardiac failure [5,6].

Interestingly, research has shown that patients with HHT have more than two-fold higher incidence rates of venous thromboembolism (VTE) compared to the general population [7]. However, the usual treatment modalities for VTE may not be applicable in the case of patients with HHT because of the enhanced risk of hemorrhage associated with HHT [8]. Although care of these patients presents a dilemma, there are few reports on the perioperative management of patients with HHT and coexisting VTE. In this case report, we present our experience with anesthetic care for a patient with HHT and deep vein thrombosis (DVT), predisposing him both to bleeding diathesis and to the risk of thromboembolism as well. Perioperative considerations for this rare disorder were reviewed along with the relevant literatures.

Case Description

A 53 kg, 160 cm, 81-year-old male patient was admitted to the hospital to undergo bilateral orchiectomy due to the metastasis of preexisting prostate cancer. The patient had a history of repeated epistaxis since his 20s. About 10 years earlier, he had been admitted to the same hospital due to epistaxis and respiratory distress with a hemoglobin level of 1.4 g/dL. Gastroscopy revealed multiple telangiectasias, which were also observed in the oral and nasal cavities, and the patient was diagnosed with suspected HHT. Afterwards, he experienced repeated admissions and episodes of blood transfusion due to recurrent epistaxis. About four years prior to the current admission, the patient had been diagnosed with suspected DVT from the left distal superficial femoral vein to the popliteal vein, based on ultrasonography. In addition, two years prior to the current admission, an abdominal pelvic computed tomography (CT) scan revealed thrombosis of the inferior vena cava (IVC) and the superior mesenteric vein (SMV) and multiple AVMs in the liver. Low molecular weight heparin and then unfractionated heparin were administered over one week. Thereafter, warfarin was started and maintained for about one year until the occurrence of severe anemia (hemoglobin level 4.3 g/dL).

At the time of the current report, the patient was admitted for bilateral orchiectomy. His initial hemoglobin level was 8.6 g/dL and it increased to 13.0 g/dL following transfusion. As part of our pre-anesthetic evaluation, we performed a chest CT to check for pulmonary AVMs, but no AVMs were found. Abdominal pelvic CT revealed thrombosis of the mesenteric vein and the right iliac vein. For prevention of pulmonary embolism, the insertion of an IVC filter was considered. However, taking into account the enhanced risk of bleeding associated with the concomitant anticoagulation with an IVC filter, it was decided that the surgery would be conducted without inserting an IVC filter.

When the patient arrived in the operating room, his blood pressure was 149/71 mmHg, his heart rate was 86 beats per minute

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(bpm), and the pulse oxygen saturation was 99%. An intra-arterial catheter was inserted to measure the blood pressure. The cardiac output was monitored using an arterial pressure-based continuous cardiac output measurement instrument (FloTrac sensor and EV1000 monitor, Edwards Lifesciences, Irvine, CA, USA). The bispectral index (BIS VISTA™, Covidien, Mansfield, MA, USA) was also monitored. Etomidate 0.2 mg/kg, rocuronium 0.8 mg/kg, and sevoflurane were used for the induction of anesthesia. Sevoflurane 1.2-4.0 vol% in air/O_2 (FIO_2: 0.5) and remifentanil 0.05-0.1 mcg/kg/min were used for the anesthetic maintenance. During the surgery, the patient's vital signs were stable and the estimated blood loss was less than 50 mL. After 35 minutes in the recovery room, the patient was transported to the ward without any particular complications. On the fourth day after the surgery, the patient's hemoglobin level was 14.2 g/dL and he was discharged without any particular problem.

Discussion

The patient in the present case was diagnosed with definite HHT according to the Curaçao Criteria [9]. The Curaçao Criteria for the diagnosis of HHT include: (1) spontaneous, recurrent epistaxis, (2) telangiectasias at the nose, lips, oral cavity, and fingertips, (3) visceral lesions such as gastrointestinal telangiectasias or AVMs in the pulmonary, hepatic, cerebral, or spinal regions, and (4) an affected first-degree relative with HHT (family history of HHT). When three or more criteria are satisfied, the condition is diagnosed as "definite" HHT. In contrast, cases which satisfy only two of the criteria are diagnosed as "possible or suspected" HHT [9,10].

In HHT patients, the bleeding tendency is due to inherent blood vessel malformation, and careful hemostasis is required during surgical procedures, along with preparation for the increased risk of intraoperative blood loss. It is important to carry out pre-anesthetic screening for AVMs, which may lead to serious complications. Pulmonary AVMs, which are found in about 50% of HHT patients [4], are mostly asymptomatic. However, they may cause pulmonary hemorrhage and dyspnea. Transient ischemic attacks and myocardial ischemia or infarction can result from paradoxical emboli associated with pulmonary AVMs. Brain abscesses may also occur as a result of thrombi; thus, a preventive antibiotic should be administered before a surgical procedure [11,12]. In addition, to prevent paradoxical air embolism, injection of even a tiny amount of air should be avoided. Because of the possibility of the above complications, a screening test for pulmonary AVMs is necessary prior to surgery. In this case, chest CT was performed as a pre- anesthetic evaluation, and no pulmonary AVMs were found. In a retrospective case series of pregnant women with HHT, the major complications occurred only in patients in whom prior screening for pulmonary and cerebral AVMs had not been conducted [13], a finding which emphasizes the importance of screening and treatment for AVMs in pregnant women. Cerebral AVMs occur in 10-20% of HHT patients and may cause hemorrhage, headache, epilepsy, and high-output cardiac failure [4]. Spinal AVMs are found in less than 1% of HHT patients [4]. El Shobary et al. reported a case of cesarean section performed under spinal anesthesia in a pregnant woman with HHT after excluding the presence of spinal AVM by head and spinal MRI; the authors stated that evaluation for spinal AVM is necessary when considering neuraxial anesthesia in HHT patients [14]. On the other hand, Gussem and colleagues successfully performed spinal or epidural anesthesia for 92 deliveries in HHT women without prior screening for spinal AVMs [13], and they suggested that screening for the presence of spinal AVMs is not routinely recommended in pregnant women with HHT. Nonetheless, to avoid the possible complications of spinal AVMs, we administered general anesthesia instead of neuraxial anesthesia in the present case, because the patient had not been screened for spinal AVMs.

In the case described here, multiple AVMs were found in the liver, but the patient did not show any abnormal symptoms related to hepatic AVM. Hepatic vascular malformations are found in about 30-70% of HHT patients and are asymptomatic in most cases [4]. The related symptoms may be high-output heart failure, portal hypertension, and biliary tract disease [15]. In the present case, the effect of the hepatic AVMs on anesthesia was considered negligible.

In our patient, thrombosis of the mesenteric vein and right iliac vein was observed, and the patient had a previous history of DVT. As in this case, there are a couple of case reports on DVT and mesenteric vein thrombosis in HHT patients [7,8]. Furthermore, Shovlin et al. suggested that the risk of VTE is increased in HHT patients [7,16]. They demonstrated that the increased risk of thrombosis in HHT patients is associated with elevated levels of factor VIII (FVIII:Ag) [16]. In another study from the same group, it was shown that low serum iron levels due to insufficient replacement of hemorrhagic iron losses are associated with elevated plasma levels of coagulation factor VIII and risk of VTE in HHT [7]. In addition to the above mechanism, VTE may occur with relation to the pulmonary AVMs or brain abscesses in HHT patients.

For the treatment of VTE and the prevention of pulmonary embolism, several modalities including anticoagulants, vitamin K antagonists, thrombolytics and IVC filters may be used [17]. In order to prevent ischemic or thromboembolic sequelae, an antithrombotic or antiplatelet agent should be considered [4]. Shovlin reported that it is possible to carefully control the dose of an antithrombotic agent for the prevention and treatment of VTE in an HHT patient [4]. However, it is difficult to manage patients with coexisting hemorrhagic diathesis and thrombosis, since antithrombotic therapy precipitates bleeding from the abnormal vascularity associated with HHT. The risks and benefits of the treatment should be carefully considered before initiation. In our case, surgery was conducted without antithrombotic measures in consideration of the patient's previous history of aggravation of bleeding following antithrombotic or anticoagulation therapy. Although insertion of a vena cava filter was considered to prevent thromboembolism, it was not performed because of concerns about the thromboembolic risk associated with the IVC filter itself and the subsequent need for anticoagulation.

When DVT occurs in a patient with hemorrhagic diathesis caused by HHT, careful management is needed to avoid both fatal thromboembolic events and aggravation of bleeding, which require treatment modalities which work in opposite directions.

In conclusion, it should be kept in mind that HHT patients may be at risk of thromboembolism as well. For the management of patients with both hemorrhagic diathesis and thromboembolic risk during the perioperative period, a careful balance between hemorrhage and thrombosis should be considered. Anticoagulation or insertion of a vena cava filter should be considered on a case-by-case basis. Pre-anesthetic screening and treatment must be performed with respect to pulmonary AVMs in a patient diagnosed with HHT, even when there are no symptoms. When choosing anesthetic techniques, the possibility of cerebral and spinal AVMs should be taken into consideration. It seems to be prudent to screen for spinal AVMs before performing spinal or epidural anesthesia even though this is not recommended routinely.

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