Various Complications after a Vascular Procedure in Patients with Hemophilia

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Hemophilia, an inherited bleeding disorder, is caused by a deficiency of coagulation factor VIII or IX. Most of patients with hemophilia need vascular procedure, which can lead to complications. Even though these complications can also occur in normal people, hemophilia and coagulopathy are particular risk factors. We reviewed medical records of patients with hemophilia who underwent vascular procedures and investigated its complications. Vessel-related complications occurred in five patients. Three patients had pseudoaneurysms after radial arterial puncture. All patients underwent coagulation factor replacement or ultrasound-guided compression and showed improvement. Neuropathy developed in one patient due to a hematoma that occurred after blood sampling. The hematoma improved, but motor and sensory deficits remained and neuropathy was confirmed. One patient died of uncontrolled bleeding after angiography. Vascular procedures require more attention in patients with hemophilia. Caution and prevention of complications is essential, even before the patient is diagnosed with hemophilia.

Key Words: Bleeding, Complication, Hemophilia, Procedure, Vascular

Introduction

Hemophilia is a rare X-linked disorder caused by a deficiency of coagulation factors VIII (FVIII) or IX (FIX). Patients with hemophilia have more chance to have bleeding, which commonly involve joint and muscle bleeding. Replacement of coagulation factors is considered the first-line treatment [1].

Vascular procedures are performed to administer intravenous coagulation factors, as well as for a variety of therapeutic purposes, including hemodialysis, embolization, and thrombolysis [2]. Angiography and interventional vascular procedures can be used to diagnose and control acute bleeding. However, this can lead to complications. Complications can also occur in normal people, but hemophilia, coagulopathy, and taking anticoagulants are particular risk factors.

Here, we describe various vessel-related complications, including pseudoaneurysms, neuropathy after vascular catheterization, and uncontrolled bleeding after angiography in patients with hemophilia.
Case Report

1) Case 1

Pseudoaneurysms occurred in three patients with hemophilia after a radial arterial puncture. Two cases of pseudoaneurysms developed after arterial sampling in hemophilia neonates, and one case was due to an arterial line procedure. All cases were diagnosed by ultrasonography. All patients received conservative management, such as coagulation factor replacement with or without ultrasound-guided compression, and improved.

(1) Case 1-1

A 29-day-old male neonate was admitted with persistent swelling of a radial artery sampling site after discharge from the neonatal intensive care unit (NICU). A right pseudoaneurysm was confirmed by wrist ultrasonography. Ultrasound-guided compression was performed and coagulation factor replacement was done. Then, a thrombosis formed, but recanalization between the pseudoaneurysm and the radial artery was observed on repeated ultrasonography (Fig. 1A). Without any additional treatment for the thrombosis, it spontaneously resolved after 3 months (Fig. 1B).

(2) Case 1-2

A male neonate was admitted to the NICU with pale skin at the time of birth and a hemoglobin level of 4.8. After an evaluation, he was diagnosed with hemophilia A. His left arm was swollen after arterial puncture for blood sampling, and a pseudoaneurysm of the left brachial artery was confirmed by ultrasonography. Elastic band compression was performed and coagulation factor replacement was done. A thrombus was observed inside the pseudoaneurysm sac 3 days later.

(3) Case 1-3

A 44-year-old male hemophilia A patient with inhibitors visited because of severe and persistent epigastric pain and was diagnosed with ischemic enteritis. He underwent a small bowel resection, and the right radial artery was catheterized for aggressive hemodynamic monitoring. A red swollen pulsatile mass occurred at the catheterization site, and Doppler ultrasonography showed a radial artery pseudoaneurysm (Fig. 2). Elastic band compression was applied and coagulation factor replacement was done in parallel with ultrasound-guided compression, and he was discharged with improvement.

Fig. 1. Ultrasound (US) images showing the progression of a pseudoaneurysm. A pseudoaneurysm from the radial artery at the wrist is shown. The recanalization between the pseudoaneurysm and the radial artery as the intra-lesional thrombus disappears. Size is about 1×0.9×0.5 cm (A). Slightly decreased size of the pseudoaneurysm at a radial aspect of the wrist with a partial thrombus; 0.8×0.9×0.5 cm (B).

Fig. 2. Ultrasound (US) image of a pseudoaneurysm from the radial artery; 0.7×0.5×1.3 cm.
2) Case 2

Neuropathy can occur as complication of hematoma or nerve damage after blood sampling. An 8-month-old boy visited another hospital due to gastroenteritis, and blood sampling was done through the antecubital vein and radial artery. Large hematomas developed at the sampling sites. He was transferred for further examination and was diagnosed with severe hemophilia A.

During the hospitalization period, coagulation factor injections, elastic bands, and ice packs were applied, and he was discharged after the hematomas improved. About 15 days after discharge, motor and sensory deficits developed on the affected arm and neuropathy was confirmed by a nerve conduction study (NCS) (Fig. 3). Median, ulnar, and radial neuropathy developed (Fig. 3). The treatment included a steroid and physical rehabilitation. The symptoms improved after 6 months, and the result was confirmed by NCS.

3) Case 3

A 43-year-old male patient with hemophilia A and inhibitors was admitted with a hemothorax due to a rib fracture and massive bleeding of the intercostal artery due to falling to the floor. A thoracentesis was performed and a large volume of blood was drained. A transfusion and bypassing agent were administered but active bleeding into the chest tube persisted. Angiography was performed, and bleeding in the left sixth and seventh intercostal artery was detected. The artery was embolized through a femoral venous puncture. Despite Doppler ultrasound-guided compression, abdominal computed tomography (CT) confirmed that a hemoperitoneum occurred due to leakage of the femoral artery embolization site (Fig. 4). Bleeding and hypotension persisted, and he eventually died of multiorgan failure and hypovolemic shock.

Discussion

Hemophilia is the most common congenital coagulation factor deficiency. Deficiency of coagulation factors disables normal hemostasis, resulting in frequent bleeding [1]. The vascular endothelial cells are essential in maintaining blood vessel homeostasis, such as expansion and contraction of the vessel wall, thrombogenesis, and thrombolysis [3]. Vascular procedures can injure vascular wall, especially the endothelial cells. Therefore, it is necessary to have regard to these features to minimize damage and excessive bleed-
Computed tomography image shows suspicious contrast extravasation from the right femoral artery with a hematoma extending along the femoral sheath.

A pseudoaneurysm is a lesion of the arterial wall that forms between the lumen of the blood vessel and the surrounding fibrous tissue. It is a pulsatile, encapsulated hematoma that communicates with the lumen of the ruptured blood vessel [4]. This can occur after blood sampling, radial arterial catheterization for patient monitoring, direct trauma, or arteriovenous shunting for dialysis [4]. Because of the lack of coagulation factors, patients with hemophilia have increased extravasation following trauma than in normal people, and a larger thrombus can form. A ruptured pseudoaneurysm is the most serious complication, resulting in massive bleeding and death [5,6]. Therefore, immediate diagnosis and treatment are important. The first choice for the diagnosis is ultrasonography, and contrast-enhanced CT should be considered as the first imaging technique in cases in which ultrasound results are ambiguous or endovascular treatment through angiography is necessary [7].

In most cases, conservative methods, such as coagulation factor replacement and ultrasound-guided compression, are successful. A thrombus that forms inside a pseudoaneurysm sac can be confirmed on follow-up ultrasonography [7]. All of our patients recovered successfully. If this management is ineffective, extended compression can be attempted, and surgical management, such as arterial ligation or embolization, should be considered [7].

In addition to a pseudoaneurysm, peripheral neuropathy has been reported as a complication. The severity of neuropathy ranges from mild and temporary sensory abnormalities to severe motor paralysis. The mechanism of neuropathy varies. The nerve can be directly damaged at the time of arterial puncture, or these injuries may occur due to excessive compression after removing the catheter [7,8]. Furthermore, as bleeding occurs, pressure inside the sheath including the nerve can increase, causing nerve ischemia followed by neuropathy [8]. In addition, a hematoma that develops at a puncture site can compress the nerve and cause neuropathy [8]. In our case, a hematoma was the cause of the neuropathy. Neuropathy that occurs after the procedure can be diagnosed by electromyography based on clinical symptoms. In addition, ultrasonography or CT can be used to confirm whether there is pressure due to a hematoma [9]. The treatment decision depends on the size of the lesion, the degree of nerve injury, and the hemodynamic status of the patient. Non-invasive conservative therapy may be performed in hemodynamically stable patients with minimal nerve damage, a small hematoma, and no active bleeding. If the hematoma is large and the nerve damage is severe, or if the disease progresses, surgical removal of the hematoma is necessary [10]. Our patient was treated with a noninvasive method, including a steroid, and both the hematoma and resulting neuropathy improved.

Invasive vascular procedures, such as arterial embolization, may be required to control bleeding, an aneurysm, or a tumor in patients with hemophilia. Complications after the procedure include bleeding at the access site or gastrointestinal bleeding. The femoral artery is used more frequently as an access site, but severe bleeding is rarely reported. However, use of the radial artery is recommended to reduce complications in high-risk patients, such as those with hemophilia, because the radial region is easier to compress, more amenable to early bleeding detection, and shows reduced bleeding compared to the femoral region [6,11,12]. In our case, angiography and therapeutic embolization procedures were effective for bleeding control, but more bleeding developed at the femoral sheath.
Vascular procedures require more attention in patients with hemophilia. Before complications arise, prevention measures such as sufficient coagulation factors and skilled execution of a procedure are needed. A thin needle should be used for vascular procedures followed by manual compression of the puncture site for sufficient time to ensure hemostasis. Early detection and an immediate approach are critical, because death can occur due to possible complications. If swelling, pain, or sensory abnormalities occur after the procedure, the patient should visit the hospital immediately for an evaluation.

In conclusion, if there is a bleeding tendency, careful management should be taken even before diagnosing hemophilia.

References