Arterial pseudoaneurysms in hemophilia patients ; successful result of non-invasive treatment, clotting factor replacement and US-guided compression

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Introduction

Pseudoaneurysms are a potential complication following procedures involving vascular manipulation or cannulation. The dangers presented by pseudoaneurysms include arterial compromise with limb ischemia, embolization of associated thrombi, compression of adjacent venous and nervous structures and rupture with possible life-threatening hemorrhage.

Hemophilia is a sex-linked genetic bleeding disorder caused by a deficiency of plasma factor VIII or IX coagulant activity. Hemophilia patients may be at increased risk for developing pseudoaneurysms attributed to underlying hypocoagulability. Although surgery was the gold standard treatment in the past(surgical ligation), several less invasive treatment options, that include coil embolization, stent graft, and ultrasound (US)- guided compression or thrombin injection, are available.

We report two cases of pseudoaneurysms in hemophilia patients which resolved with non-invasive treatment, i.e., clotting factor replacement and ultrasound(US)-guided compression.

Case Report

A three-week-old male infant with severe hemophilia A presented with a mass in his left antecubital area. There was no significant family history about any bleeding disorder. The infant at birth had massive subgaleal hematoma and he was diagnosed to be severe hemophilia A at local clinic. During the administration at NICU, the blood coagulation test revealed activated partial thromboplastin time of 139.2 seconds, and the factor VIII(FVIII) activity was below 1%. At that time, brachial arterial puncture was performed.

The mass were firm, movable, and pulsating at the medial side of antecubital area. Size was 1.9 x 2.2cm, and distal radial pulse was intact. There was no color change of left hand.

US showed pseudoaneurysm from left brachial artery, which was 1.4 cm in size. US-guided compression was attempted. Prior to the procedure, he was infused with Advate®, FVIII clotting factor concentrates, 50 U/kg to raise FVIII activity to 100%. After 2 days, thrombus in pseudoaneurysm sac was confirmed and additional US-guided compression was performed. Follow-up US one week later showed fulfilled thrombus in brachial pseudoaneurysm, and size of thrombus was 1.4 x 0.8 x 1.2 cm (Fig. 1).

After discharge, follow-up examination showed complete resolution of pseudoaneurysm and the palpable mass 2 weeks later. Although first exposure to clotting factor was high dose and very early, inhibitor to FVIII was negative.

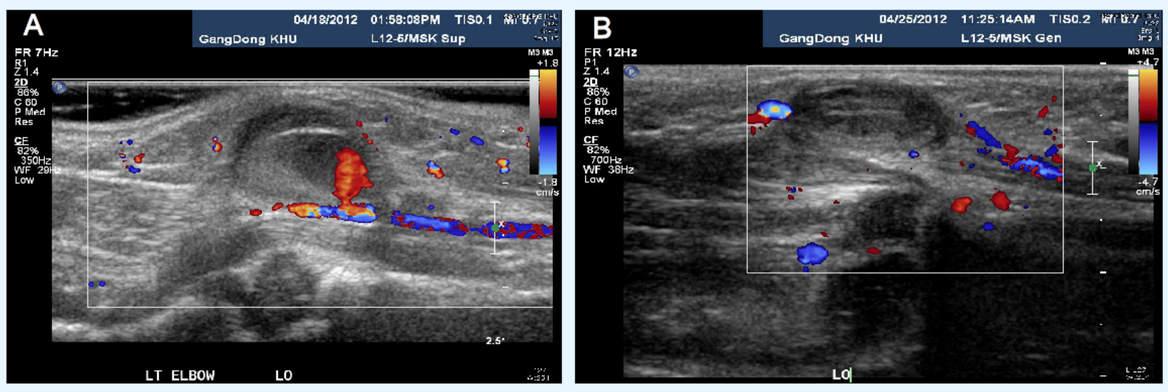

Figure 1. (A) Ultrasonography showed pseudoaneurysm from left brachial artery, that was 1.4cm in size and neck 1.0cm in size, (B) Follow-up ultrasonography was performed at 7 days after initial US-guided compression and coagulation factor administration. Findings showed thrombus in pseudoaneurysm and no intralesional blood flow into the lesion from brachail artery.

A four-week-old male infant was admitted for a pulsating mass in the radial aspects of both wrists. The infant had a history of hospital admission due to maternal gestational DM. The patient had no siblings, and his maternal grandfather had diagnosed with severe hemophilia A. Artery punctures of both radial arteries were performed in the NICU of another hospital. Coagulation assays at the time of admission to other hospital were as follows: PT 12.3 sec, PT INR 113%, and aPTT 95.8 sec. Factor assay results in our clinic showed FVIII less than 1%, and the patient was diagnosed with hemophilia A.

Wrist US showed a 0.9 x 0.5 x 0.7-cm hematoma and a completely thrombosed aneurysm in the left wrist and a 0.8 x 0.6 x 0.8-cm pseudoaneurysm arising from the right radial artery. US-guided compression after administration of Advate® on the day of admission was attempted but was not successful. US-guided compression was retried on the third and fourth days of admission after factor VIII administration, but no thrombus was visible. Due to the small size of the pseudoaneurysm and the absence of complications, the patient was discharged on the fourth hospital day to complete regular follow-up observation on an outpatient basis.

US examination two months later revealed size of pseudoaneurysm on right radial artery was decreased with formation of a partial thrombus. After one month, the pseudoaneurysm was totally thrombosed according to US (Fig. 2).

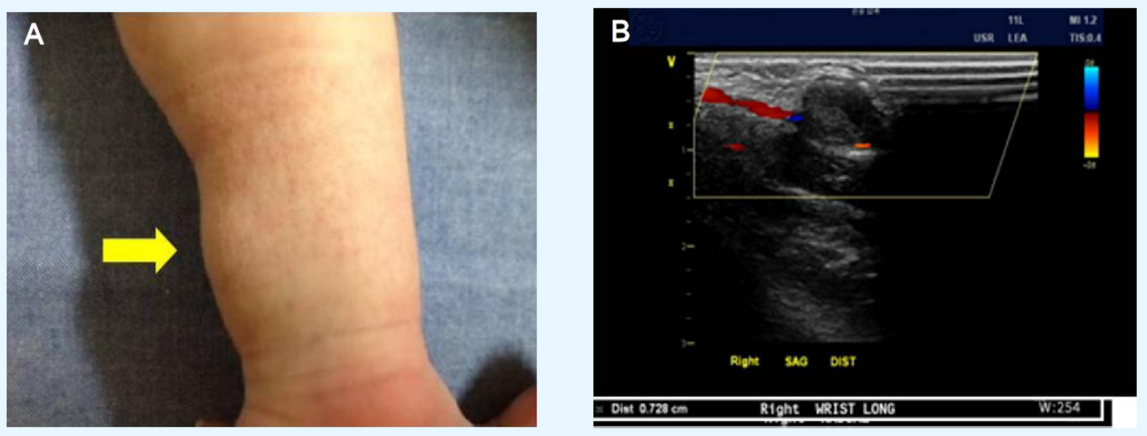

Figure 2. (A) Swelling around the right radial artery area, (B) Three-month follow-up US shows a right radial artery pseudoaneurysm with thrombus.

Summary

Arterial cannulation and procedure in hemophilia patients are very cautious because of uncontrolled bleeding and pseudoaneurysms. Our experience suggested that factor replacement and US-guided compression may be an adequate option in hemophilia patients when pseudoaneurysms are small. The primary benefit to this therapy is the avoidance of the risks associated with surgery and embolization. Procedures such as US-guided compression are safe or more cost efficient in the clinical setting.

DOI: 10.3252/pso.eu.WFH2014.2014
Poster presented at: WFH 2014 WORLD CONGRESS Melbourne, Australia, May 11-15