

A Rare Clinical Problem: Intestinal Pseudolymphoma in a Patient with Haemophilia A

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To the Editor,

We had reported a 10-year-old boy with disseminated intravascular coagulation (DIC) who had undergone nasogastric topical Ankaferd Blood Stopper (ABS) administration since the endoscopic procedure had been contraindicated due to DIC and associated co-morbidities for control serious bleeding due to primary esophageal adenocarcinoma (PEA) after written informed consent regarding the off-label use of topical ABS as a means of attaining hemostasis had been obtained from the parents. To our knowledge, that was the first clinical application for topical esophageal ABS administration in a pediatric patient. The vials of ABS (Ankaferd Blood Stopper®; patent number 2007-0-114485) were donated by Ankaferd Drug Inc., Istanbul, Turkey. Six milliliters of ABS had been administered through the nasogastric tube over the bleeding esophageal site. The bleeding had stopped within a very short period of time following the topical ABS application. Re-bleeding had been not observed during the post-procedural period for several days. Due to the risk of recurrent extensive esophageal mucosal tumor bleeding, topical ABS had been re-administered three more times at the same dose and schedule. We had not observed any side effects potentially attributable to the ABS treatment. However the patient had died after three months due to disease progression. In our conclusion, topical ABS application could have been an alternative method for

controlling neoplastic GI bleeding. Further controlled clinical trials were required to validate the effectiveness and safety of topical ABS in GI bleedings in adults and children.¹

We read with great interest the paper of Turgut et al.² regarding the topical ABS in the management of critical bleedings due to hemorrhagic diathesis. In their study, Turgut et al. aimed to depict hemostatic effects of ABS in critical bleedings due to hemorrhagic diathesis, refractory to conventional measures in distinct clinical settings. In their conclusion, Turgut et al.² topical ABS application was effective for the bleeding dental surgery in Hemophilia A. However, the most serious problem in hemophilia is the bleedings inside of the body which cannot be achieved as a topical.

Intramural hematoma of the intestine is a very rare complication of this disease.³⁻⁵ We report an unusual pseudotumor in a patient with hemophilia A in whom intramural hematoma of the ileum mimicking intestinal lymphoma on ultrasonography (US). A 28-year-old man diagnosed with hemophilia when he was 6-month of age was referred to our inpatient clinics of pediatric hematology and oncology department because of having a radiological appearance mimicking intestinal lymphoma on US. His history was remarkable for many episodes of hemarthrosis.

He is a patient with severe hemophilia who is not on factor prophylaxis regularly. He presented with a history of nonspecific abdominal discomfort for a week. Palpation of his abdomen revealed tenderness. Abdominal US findings were diffuse wall thickening of the ileum and minimally enlarged mesenteric lymph nodes resembling to intestinal lymphoma-like appearances on imaging. The length of the affected small bowel segment was 12 cm. Erythrocyte sedimentation rate, urinalysis, serum glucose, urea, creatinine, electrolytes, liver enzymes, total protein, albumin, uric acid, and lactate dehydrogenase levels were resulted in normal ranges. No bleeding occurred and his platelet count, hemoglobin, and fibrinogen levels remained within the normal reference ranges. Occult blood stool test was positive. Because his activated partial thromboplastin time (aPTT) was prolonged, intermittent infusion of factor VIII (20 IU kg⁻¹ twice daily) was started immediately. Factor VIII level was found at 54%. After the successful therapy with factor VIII, US of the abdomen revealed completely resolution of the thickening and verified that this was a pseudotumor caused by intramural hematoma mimicking intestinal lymphoma. He was discharged home 10 days on a regimen of factor VIII prophylaxis (20 IU kg⁻¹ twice weekly) after the abdominal US had been resulted normal.

It should not be forgotten that the patient with hemophilia may be also a malignant disease such as lymphoma, leukemia or soft tissue tumors.⁶⁻⁸ The possible diagnosis of intestinal lymphoma should not be overlooked in a patient with hemophilia A, if the pseudotumor caused by intramural hematoma has been excluded. Administration of factor VIII concentrates should start immediately, but not be delayed until the laboratory examinations will be concluded.

Radiologists need to be aware of this pseudotumor on US or computed tomography in patients with hemophilia. If confirmation of its non-neoplastic nature is required, this can be done with US for reevaluation after the factor VIII therapy.

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