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Hepatic artery pseudoaneurysm in von Willebrand's disease

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Abstract We report a case of hepatic artery pseudoaneurysm which occurred in a patient with von Willebrand's disease. The patient presented with upper abdominal pain and diagnosis was made by US and CT examinations. This case emphasizes the possible association between an established coagulation disorder and pseudoaneurysm. The patient was successfully treated by coil embolization.

Keywords Hepatic artery pseudoaneurysm · von Willebrand's disease · CT · US · Angiography

Introduction

Von Willebrand's disease (vWD) is the most frequent hemorrhagic hereditary disorder affecting 1 in 800–1000 individuals. The von Willebrand's factor (vWF) is a plasmatic vector for factor VIII in blood coagulation and promotes platelet adhesion to the vascular subendothelium.

The vWF deficiency was found to be associated with different pathologic conditions that involve the vascular system. These conditions include arterial aneurysms [1], pseudoaneurysms [2, 3, 4], intracranial arteriovenous malformations [5], mitral valve prolapse [6], gastrointestinal angiodysplasias [7] and telangiectasias [8]. We present a case of a giant hepatic artery pseudoaneurysm in a patient with vWD which has been successfully treated by coil embolization. To our knowledge, there are no previous reports of a hepatic artery pseudoaneurysm associated with von Willebrand's disease. This case highlights that a coagulation disorder may be one of the contributory factors in the pathogenesis of a pseudoaneurysm.

Case report

A 37-year-old man was admitted to our hospital with a 4-week history of upper abdominal pain. The clinical examination and blood tests (INR: 1.65; PT 48%; vWF:Ag: 0.9 mg/dl; VIII-F: 0.05 mg/dl; and positive Ristocetine test) revealed von Willenbrand syndrome type I according to the 1994 Sadler classification. The family history included haemorrhagic tendencies (mother with episodes of spontaneous gastrointestinal bleedings).

Abdominal sonography showed a large hypoechoic oval area (59×30 mm) at the level of the coeliac artery. Color Doppler sonography proved this to be a vascular lesion. Spiral CT (Fig. 1) confirmed the sonographic findings. An angiogram was performed with selective injections of the coeliac and the superior mesenteric (SMA) arteries. This showed a pseudoaneurysm at the origin of the common hepatic artery and occlusion of the distal hepatic artery (Fig. 2a). The liver was supplied by the pancreaticoduodenal arterial arcades (Fig. 2b). The splenic and the left gastric arteries were normal.

After discussion between the vascular surgeons and interventional radiologists, concerning the morphology of the lesion and the underlying coagulopathy, it was established that transcatheter embolization would be the most appropriate therapeutic option. The pseudoaneurysm was embolised via a 110-cm-long 5-F Simmons 1 catheter (Terumo, Tokyo, Japan). Six Gianturco steel coils (12–6 mm) were released into the pseudoaneurysm. The post-embolization angiogram showed the pseudoaneurysm was occlud-

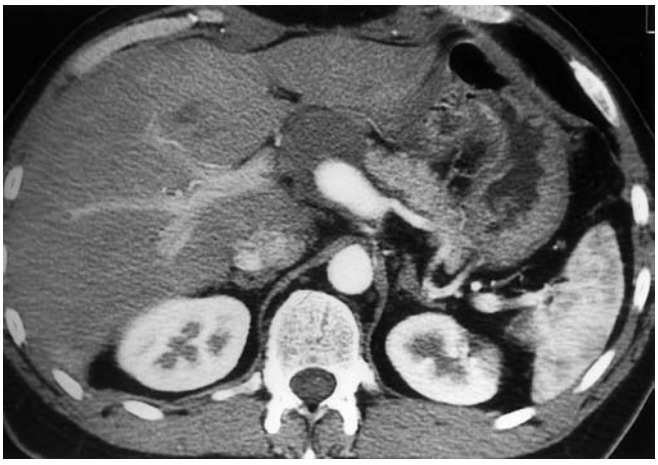

Fig. 1. Spiral CT shows giant pseudoaneurysm arising from common hepatic artery with a side partially thrombosed

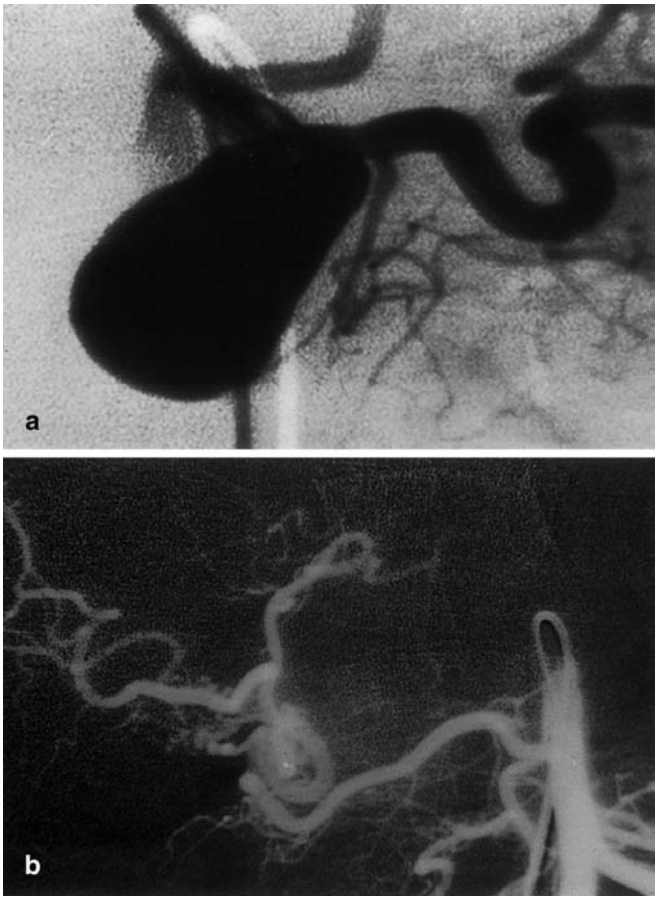

Fig. 2 a Selective angiogram showing a pseudoaneurysm of the common hepatic artery and the occlusion of the distal hepatic artery. **b** Superior mesenteric artery selective angiogram demonstrating the collateral supply to the liver by the pancreaticoduodenal arterial arcades

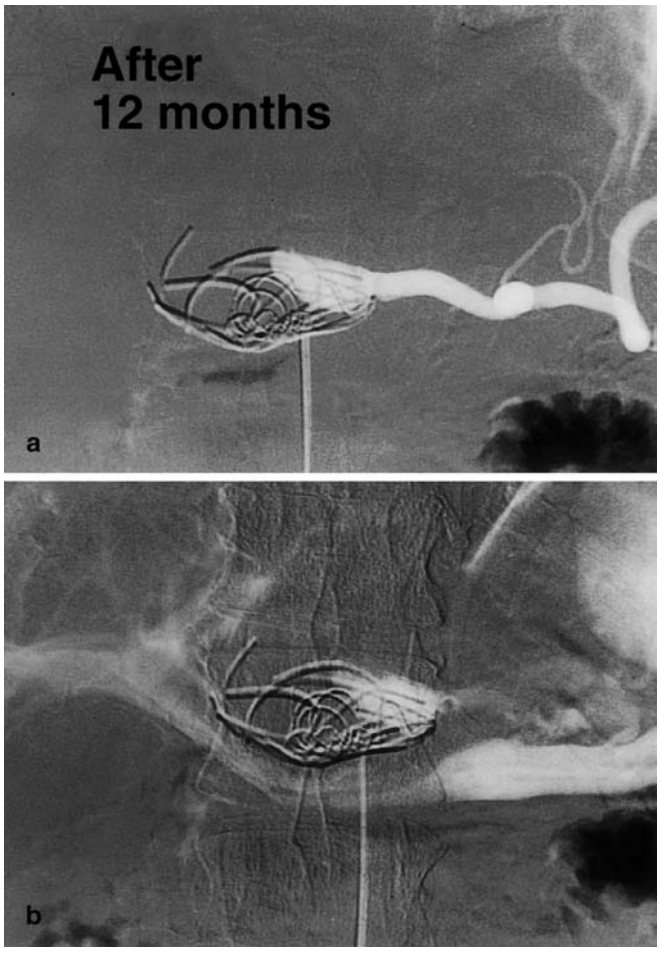

Fig. 3 a A 12-month follow-up angiogram demonstrates the pseudoaneurysm exclusion and regular splenic artery flow. **b** Partial compression of portal vein is noted

ed while the splenic and gastric arteries remained patent. The interventional procedure was concluded with closure of the percutaneous vascular approach using a Techstar-TM device (Perclose, Menlo Park, Calif.).

In the post-operative period the patient showed signs of good health and the abdominal pain gradually disappeared in the following 3 days. One week later, CT confirmed complete pseudoaneurysm thrombosis. Follow-up studies, performed at 6 and 12 months, with spiral CT and digital subtraction angiography (DSA), confirmed the exclusion of the pseudoaneurysm (Fig. 3).

Discussion

Von Willebrand's disease is one of the most common hereditary haemorrhagic disorders accompanied by a wide range of haemostatic defects. Abnormal platelet function, usually reflected in prolonged bleeding time, is the result of a quantitative or qualitative defect of the vWF. Furthermore, a secondary deficiency of factor-VIII procoagulant protein, coded for by the X chromosome, may

occur and in itself lead to an additional coagulation defect. These two glycoprotein macromolecules, vWF and factor VIII, circulate together as a combined complex. It is thought that the vWF serves as a "carrier protein" for, and has a stabilizing effect on, factor VIII.

Even if most cases are heterozygous for a mutation on chromosome 12, the genetic basis of the disease has not yet been elucidated.

Affected patients may present with spontaneous bleeding from mucous membranes, excessive bleeding from wounds or menorrhagia.

Vascular abnormalities in the form of a pseudoaneurysm associated with vWD have been previously reported. Wautier et al. [2] described a femoral artery pseudoaneurysm in a patient with familial vWD. Choung and Wei [3] described the association of vWF deficiency and traumatic pseudoaneurysm of the supraorbital artery. De Foer et al. [4] described a superior gluteal artery pseudoaneurysm associated with vWD. The vascular abnormalities in vWD are very interesting since it is known that von Willebrand's factor is synthesized by endothelial cells [8]. Evidence from literature suggests the hypothesis that vWD may be a part of a large syndrome encompassing structural and functional mesenchyma-derived structures such as bone, cardiac valves and blood vessels [6, 7, 8]. Pseudoaneurysms of the hepatic artery are rare and are usually related to abdominal surgery, intra-abdominal inflammation or penetrating abdominal trauma. Our patient did not have a clinical history of surgery, local inflammation or abdominal penetrating trauma. His haemorrhagic disorder was confirmed and the hepatic pseudoaneurysm was incidentally discovered by ultrasound examination performed for upper abdominal pain.

Presumably, the coagulation disorder could be considered in this case to be predisposing factor in the pathogenesis of the pseudoaneurysm. To our knowledge, only a few cases of hepatic pseudoaneurysm have been reported and their true incidence is still unknown. It has been suggested that the incidence may be increasing [9]. Most of the cases are unknown because of the absence or paucity of symptoms [10]. Only some patients present with the classical triad of abdominal pain, haemobilia and obstructive jaundice.

Hepatic pseudoaneurysms can rupture into the peritoneal cavity or into the common bile duct, gallbladder, duodenum or portal vein.

The usual surgical approach consists of ligation or excision of the hepatic pseudoaneurysm with arterial reconstruction and liver resection according to the anatomical location [9, 10]. Transcatheter embolization offers several advantages over surgery, such as lower mortality and morbidity risk and assessment of collateral blood flow.

The main factors that influence the radiological management of hepatic aneurysms are the site and the morphology of the lesion as well as the clinical condition of the patient [10]. In our case, interventional treatment was chosen because surgery is considered high risk in patients with vWD. Furthermore, surgical procedures in patients with vWD require complex blood products replacement therapy. Despite this, the risk of a life-threatening bleeding cannot be excluded.

In our case the hepatic artery was occluded and the blood flow to the liver was maintained by the pancreaticoduodenal arterial arcades. The presence of such a good collateral supply is likely to have reduced the risk of hepatic infarction.

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