Huge Varicose Inferior Mesenteric Vein: an Unanticipated ^{99m}Tc -labeled Red Blood Cell Scintigraphy Finding

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Abstract Ectopic varices (EcV) are enlarged portosystemic venous collaterals, which usually develop secondary to portal hypertension (PHT). Mesocaval collateral vessels are unusual pathways to decompress the portal system. Here we report the case of a huge varicose inferior mesenteric vein (IMV) that drained into perirectal collateral veins, demonstrated by ^{99m}Tc -labeled red blood cell (RBC) scintigraphy performed for lower gastrointestinal (GI) bleeding in a 14-year-old girl. This case illustrates the crucial role of ^{99m}Tc -labeled RBC scintigraphy for the diagnosis of rare ectopic lower GI varices.

Keywords Portal hypertension · Ectopic varices · Inferior mesenteric vein · Gastrointestinal bleeding · ^{99m}Tc -labeled red blood cell scintigraphy

Introduction

Varices of the lower gastrointestinal (GI) tract are an uncommon cause of hematochezia [1]. These ectopic varices (EcV) are large and convoluted portosystemic venous collaterals, which might be encountered in various sites, such as the duodenum, jejunum, ileum, colon, rectum, biliary tree, abdominal stomas, uterus, and vagina [2],

having the potential for causing 5% of all GI hemorrhages [3]. The most common cause of portosystemic collateral vessels is portal hypertension (PHT) [4], and extrahepatic portal vein obstruction (EHPVO) is the most prevalent cause of PHT in children [5]. Splenic or splenomesenteric venous stenosis and obstruction due to neoplasm, pancreatitis, or surgery are the other responsible etiologies for the development of EcV [4].

Gastroesophageal, paraumbilical, splenorenal, and inferior mesenteric collateral vessels are major pathways to decompress the portal system. On the other hand, pleuropericardial-peritoneal, pancreaticoduodenal, splenoazygos, and mesocaval collateral vessels are unusual pathways [4].

This report presents the case of an unusual varicose collateral inferior mesenteric vein (IMV) which was diagnosed first on ^{99m}Tc -labeled red blood cell (RBC) scintigraphy and then confirmed by means of ultrasonography (US) and multidetector computed tomography (MDCT) angiography.

Case report

A 14-year-old girl, presented to our hospital with the chief complaint of fresh blood in feces and mild periumbilical pain. Her mother recalled the first episode of hematochezia at the age of 3 years. Her complaints had worsened recently. On admission, physical examination yielded no positive finding except pallor and tachycardia. In laboratory studies, the white-cell count was 3,600 per cubic millimeter. The platelet count was 242,000 per cubic millimeter. The hematocrit was 21%, and the hemoglobin level was 4.8 g per deciliter. Liver function tests, viral hepatitis serology, as well as upper and lower GI series, were normal.

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Fig. 1 **a** Meckel's diverticulum scintigraphy soon after the injection of ^{99m}Tc -pertechnetate reveals a tortuous vascular structure in angiographic phase (*arrows*). **b** Sequential dynamic images of the abdomen over a period of 60 min (2 min/frame), show no abnormal radiotracer uptake to indicate the presence of ectopic gastric mucosa

The patient underwent Meckel's diverticulum scintigraphy with ^{99m}Tc -pertechnetate. Dynamic images acquired over 60 min showed a tortuous vascular structure in angiographic phase; however, the study was negative for Meckel's diverticulum (Fig. 1). ^{99m}Tc -labeled RBC scintigraphy was performed to detect the source of GI bleeding. Very shortly after the administration of radiotracer, an elongated tortuous IMV with a reverse flow extending from the epigastric region to the pelvic cavity, and anastomosing with dilated perirectal collateral veins (Fig. 2a, b). It remained unchanged in shape or intensity on subsequent delay images (Fig. 2c).

Abdominal gray-scale and color Doppler ultrasound (US) demonstrated a normal-sized liver with an acceptable diameter of intrahepatic portal vein with no detectable blood flow. These studies also confirmed IMV as a giant, tubular venous hepatofugal collateral vessel, extending from the portosplenic junction to the perirectal area with a diameter of 24 mm. The spleen was at the upper limit of normal size, showing minimal varicosity around the splenic hilum. On abdominal MDCT angiography, liver parenchyma and hepatic veins, as well as calibration of the portal vein, were normal. There was a tuft of dilated veins in the presacral region and around the rectum. An extremely

Fig. 2 **a–c** Anterior abdominal images after the intravenous administration of ^{99m}Tc -labeled erythrocytes were obtained. A 3-s sequential flow study (**a**) and selected images of the abdominal blood pool at 2, 20, 40 and 60 min (**b**) demonstrate abnormal vascular structure extending from the epigastric region to the pelvic cavity. **c** Delayed 4-h abdominal

blood pool scintigram in anterior and LAO projections of the abdomen represent huge varicose IMV (*arrows*) that drained into perirectal collateral veins (*arrowheads*), which remained unchanged in shape throughout the study

Fig. 3 Coronal contrast-enhanced maximum-intensity-projection MDCT image (a) and frontal three-dimensional (3D) volume-rendered contrast-enhanced MDCT angiography image (b) show a

dilated and tortuous IMV (*arrows*) descended into markedly dilated perirectal collaterals in pelvic region (*arrowhead* in b)

ectatic and tortuous IMV, 25 mm in width was detected, extending caudally, and communicating dilated pelvic hemorrhoidal veins with portosplenic junction (Fig. 3).

The patient underwent upper and lower endoscopies. Esophagogastroduodenoscopy did not visualize any gastroesophageal varices. At colonoscopy, there was marked venous dilation from the rectum to the sigmoid colon (Fig. 4). Additional diagnostic studies were performed to disclose any probable coagulopathies. The results for protein C deficiency, protein S deficiency, lupus anticoagulant, antinuclear antigen, anti-double-stranded DNA, anti-thrombin III, antiphospholipid antibody, homocysteine, factor VIII, factor XIII, factor V Leiden, prothrombin 20210 A, and MTFHR mutations were all negative.

The patient required 4 units of packed RBCs during hospitalization. Hence, the patient was considered to be at extremely high risk for unmanageable bleeding, she was referred to the vascular surgery service. Laparotomy was performed through a midline incision. On exploration, an extremely dilated inferior mesenteric vein was found (Fig. 5). Meanwhile, portal pressure was measured through the IMV and was found to be 14 cm H₂O. IMV-left common iliac vein side-to-side shunt surgery and liver biopsy were performed. Liver biopsy revealed no evidence of cirrhosis or fibrosis. Anticoagulation therapy was started for the patient with low-molecular-weight heparin as an inpatient treatment. Preceding discharge, it was converted to oral anticoagulation therapy using warfarin with a target

Fig. 4 Colonoscopy views, showing colonic varices at the rectum and sigmoid regions

Fig. 5 An extremely tortuous, ectatic IMV was found intraoperatively

international normalized ratio (INR) of 2–3. The goal was preventing further clot formation in the portal and mesenteric venous systems.

Discussion

Bleeding is one of the most frequent manifestations in lower GI pathologies. In this common clinical setting, accurate diagnosis and proper treatment largely depends on localization of the site of the hemorrhage. Anorectal sources contribute to lower GI bleeding, with etiologies including hemorrhoids, anal fissures, rectal varices, fistulas, and radiation proctitis [6]. However, in certain cases, it might be difficult to elucidate the exact underlying etiology of the anorectal sources of bleeding. This report focuses on a varicose IMV with significant hemorrhage from anorectal varices, as a rare condition.

Anorectal varices are enlarged portosystemic collateral veins, which develop typically in patients with PHT as a pathway for portal venous blood flow from the superior hemorrhoidal veins (portal) to and through the middle and inferior hemorrhoidal veins (systemic). In a large series of bleeding from EcV reviewed by Norton et al. [2], stomal varices were found to be the most common location accounting for 26% of cases, followed by 17% in the duodenum, 17% in the jejunum or ileum, 14% in the colon, 9% in the peritoneum and 8% in the rectum. Diagnosis of EcV in a setting of GI bleeding can be established during routine endoscopy, diagnostic transfemoral or transhepatic angiography, video capsule endoscopy, endoscopic ultrasound (EUS), ^{99m}Tc -RBC scintigraphy, computed tomography (CT) angiography, multislice helical CT, CT-enteroclysis, contrast-enhanced 3D magnetic resonance angiography, color Doppler US, laparoscopy or laparotomy, and occasionally during autopsy [7].

^{99m}Tc -labeled RBC scintigraphy is a useful method to evaluate GI bleeding. This test is more sensitive than angiography for the detection of active bleeding and allows

either continuous or intermittent imaging to be undertaken where bleeding is not continuous, thus increasing the likelihood of detecting intermittent bleeding [8]. Major criteria for identifying the site of GI bleeding include intraluminal accumulation of radiotracer that conform to bowel anatomy and usually increased in activity with time. This should move either antegrade or retrograde in the bowel. On radionuclide GI bleeding studies, a variety of abnormalities such as hemangiomas, varices, aneurysms, and angiodysplasia may be detected incidentally [9, 10]. In the present case, abnormal radioactivity extending from the epigastric region to the true pelvis was shown immediately after the administration of the radiotracer, which did not migrate or change in shape or intensity on subsequent delay images. Pooling of tagged RBCs within the varicose veins explains the scintigraphic abnormality. For the first time, Yoshikai et al. [11] reported scintigraphic recognition of an inferior mesenteric varix that was shaped like a question mark. In previous studies, intraabdominal vascular blood-pool single photon emission CT (SPECT), a safe and noninvasive method, was useful for identifying portosystemic collaterals in patients with PHT. This technique is also used for evaluating the therapeutic effectiveness of sclerotherapy, predicting the recurrence of varices, and selecting appropriate therapy [12, 13].

Noncirrhotic PHT is the major cause of symptomatic PHT in Third World countries. Common causes of non-cirrhotic PHT include schistosomiasis, noncirrhotic portal fibrosis, idiopathic PHT, congenital hepatic fibrosis, EHPVO and veno-occlusive disease. EHPVO accounts for 80–90% of cases of PHT in children [5]. The patient's complaints, huge IMV and normal result of liver biopsy were all in accordance with the diagnosis of EHPVO. Three main etiologies are involved in obstruction of the portal vein: congenital anomalies (congenital valves, webs or stenoses of the portal vein), local factors causing injury and diminished flow to the portal vein (umbilical vein cannulation for exchange transfusion, abdominal surgery, trauma and dehydration), and inborn or acquired systemic disorders leading to a thrombophilic state. In children, omphalitis and intra-abdominal infections are the most common causes of portal vein thrombosis (PVT). PVT, as an important etiology in portal vein obstruction, can present as early as 6 weeks after birth or may not become manifest until adulthood, depending on the cause [14, 15]. Considering the pathogenesis of PVT, tests for confirming the overt or occult myeloproliferative disorders, as well as risk factors for venous thromboembolism, such as factor V Leiden mutation, G20210A prothrombin gene mutation, levels of protein C and protein S, and antithrombin III can be helpful. Liver biopsy is not indicated in all patients who have EHPVO; however, other possible causes of PHT, i.e., congenital hepatic fibrosis could be ruled out. Despite

available diagnostic techniques, the precise cause of PVT remains unclear in 50–90% of children [14]; as in our case, no underlying predisposing condition could be identified. Imaging remains the mainstay for the diagnosis of EHPVO, and a variety of radiologic techniques can be used to confirm a suspected thrombosis. Color Doppler US, which might reveal absent or decreased flow across the portal vein, could be solely applied for detection of PVT. Tessler et al. [16] reported color Doppler imaging as a valuable screening procedure for the detection of PVT with a sensitivity and specificity of 89% and 92%. In our case, Doppler US revealed no flow within the intrahepatic portal vein, which is suggestive of PVT. Evidence for acute or subacute venous thrombosis in CT include enlargement of the affected vein with a non-enhancing low-attenuation center, and ring enhancement due to opacification of the vasa vasorum [17, 18]. In chronic thrombosis, the vein may be normal sized or small, with sometimes non-visualization, and opacification of collateral vessels [18].

The most common portocaval collateral to decompress PHT is the coronary–gastroesophageal route [4]. Conversely, dilatation of the IMV is an infrequent finding (2.5–5% of patients with PHT) with great importance because of its association with rectal bleeding [19]. The presenting symptom and imaging findings proposed the latter route as the most justifiable explanation for GI bleeding in our case. The most noticeable portosystemic connection in the case was mesocaval anastomosis, which connects the IMV and iliac veins mainly via the hemorrhoidal plexus. These anastomoses may rarely reach huge dimensions but this had happened to our case. Akgul et al. [20] reported a case of a giant varicose IMV with the findings on US, CT and splenoportography in a case of PHT. Two cases of giant paraesophageal and paraumbilical varices were also reported by Shapiro et al. [21].

Optimal treatment remains controversial because EcV are uncommon and none of the treatment modalities has been investigated in a large series of patients. The therapeutic alternatives for bleeding varices include conservative medical therapy, surgical options such as portosystemic shunt, or percutaneous treatment with either embolization or transjugular intrahepatic portosystemic shunt (TIPS) [2]. Portosystemic shunt has been utilized in children with extrahepatic portal vein thrombosis when variceal bleeding is refractory to medical or endoscopic therapy [22]. Accordingly, this method has been chosen for our patient. It has also been demonstrated that recent thrombosis can be recanalized in 80% of patients by systemic anticoagulants. Anticoagulant therapy increased neither the risk of GI bleeding nor the severity of bleeding [23]. However, in chronic EHPVO, the role of anticoagulant therapy is not clear and only can be considered if there

is a specific clinical history of recurrent thrombotic episodes and after shunt surgery [14].

In this case, we reviewed a huge varicose IMV as an unusual route that the portosystemic collateral vessel can develop in PHT. We conclude that ^{99m}Tc -labeled RBC scintigraphy could be an acceptable choice in the diagnosis of GI bleeding, with the added benefit of demonstrating underlying pathology in the abdomen or pelvis.

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