

CASE REPORT

Portopulmonary venous anastomosis as a rare cause of embolic stroke following endoscopic cyanoacrylate injection for gastric variceal hemorrhage: A case report and review of the literature

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Key words

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Introduction

Portal hypertension is characterized by an increase in the portal venous pressure beyond the normal pressure of 5–10 mmHg. The most common cause of portal hypertension is cirrhosis, but other recognized causes include hepatitis, portal vein thrombus, or portal arteriovenous fistulae. As the portal pressures increase beyond 5–10 mmHg collateral, variceal vessels develop that allow shunting of blood from the portal system to the systemic circulation. Portopulmonary venous anastomoses (PPVA) are portal vein collaterals that connect directly to the pulmonary venous circulation and subsequently to the left heart. These PPVA typically arise from dilated paraesophageal veins. The incidence of these collaterals in patients with portal hypertension is unclear, with a reported incidence of between 1 and 30%.^{1–8} The largest dedicated series of cirrhotic patients assessed with triple-phase computed tomography (CT) studies demonstrated PPVA in 19.6% of its cohort.⁸ One author has suggested that, in patients with enlarged gastric varices, the absence of a gastrosplenic shunt may positively predict the presence of a PPVA.⁵

The risk of cerebral emboli from variceal injection in patients with PPVA was hypothesized in 1981, but the authors did not identify any prior reports of these complications.⁹ Since 1981, there have been scattered case reports of ischemic embolic stroke after variceal injection with cyanoacrylate.^{10–14} While a

Abstract

We report the case of a non-cirrhotic 25-year-old female patient with cryptogenic portal hypertension who underwent cyanoacrylate injection for acute gastroesophageal variceal bleeding with a subsequent embolic stroke via a previously unrecognised portopulmonary venous anastomosis.

number of these cerebral emboli appeared to have occurred secondary to patent foramen ovale, others were postulated to be secondary to PPVA. We present the case of a 25-year-old female patient with idiopathic portal vein hypertension and extensive gastroesophageal varices who presented with acute ischemic stroke after cyanoacrylate injection for actively bleeding gastric varices.

Case report

A 25-year-old female patient presented to our institution with symptoms of melena and presyncope. Her medical history included a cholecystectomy and a combined splenectomy and partial pancreatectomy (carried out due to idiopathic thrombocytopenic purpura without postoperative complications of note). An endoscopy was performed, which demonstrated gastroesophageal varices with a large esophageal varix and bleeding gastric varices (Fig. 1a). Her religious denomination was that of a Jehovah's Witness, and she declined blood transfusion. Given the active bleed and limited conservative treatment options, the varices were treated via cyanoacrylate injection with successful hemostasis.

However, shortly after the procedure, she was found to be obtunded with neurological deficits, and an immediate non-contrast CT brain and CT cerebral angiogram demonstrated small, scattered bilateral hyperdensities correlating with glue

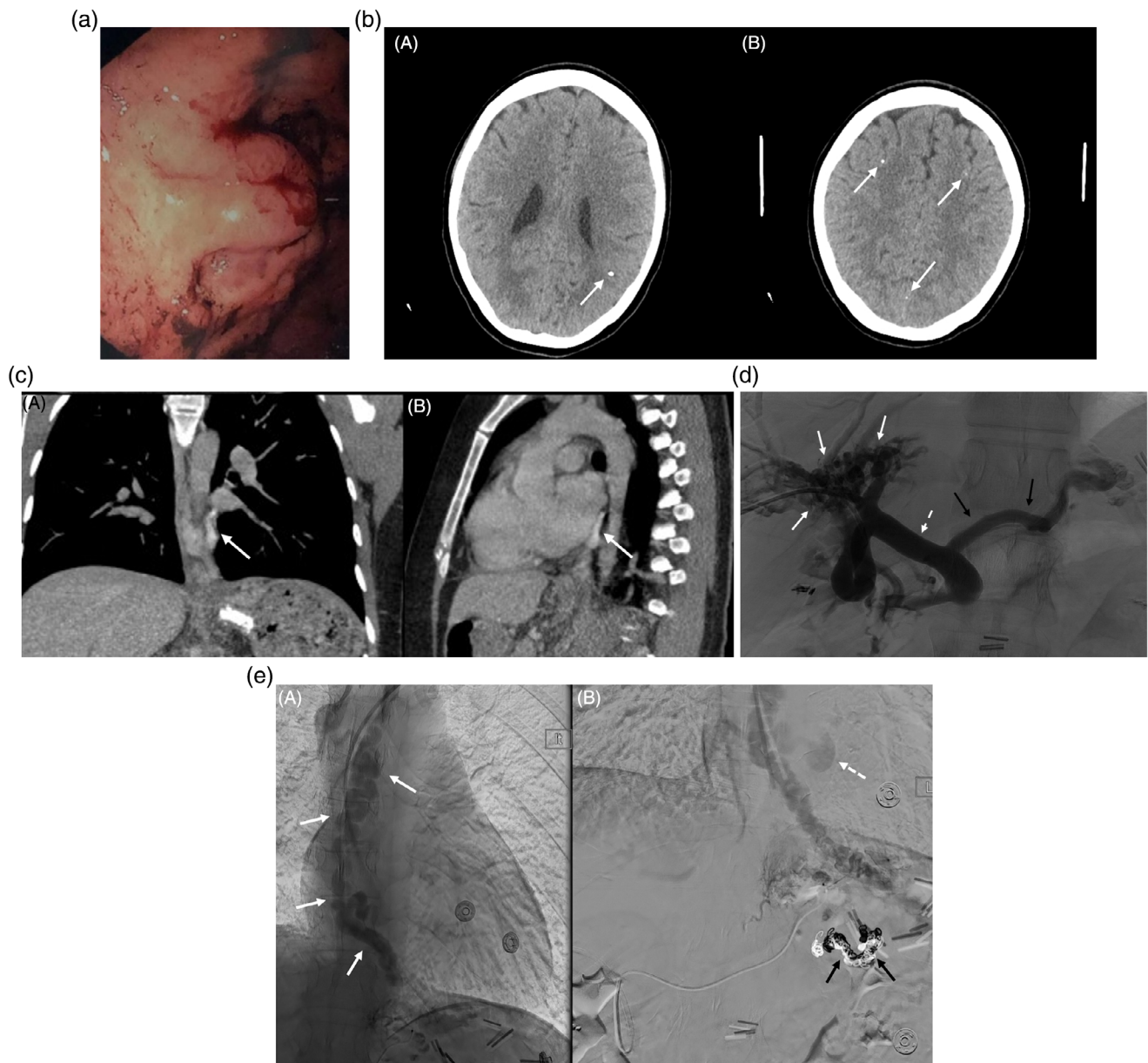


Figure 1 (a) Live image during endoscopy demonstrating large, tortuous gastric varices with active bleeding prior to cyanoacrylate embolization. (b) Images (A) and (B): Axial noncontrast computed tomography (CT) brain images demonstrating hyperdense foci at the gray–white matter interface, corresponding to glue emboli from endoscopy procedure. (c) (A) Coronal and (B) sagittal images from a contrast-enhanced CT study demonstrating a portopulmonary venous anastomosis (PPVA) from an esophageal varix connecting to the left inferior pulmonary vein (white arrows). (d) Transhepatic portal venography demonstrating cavernous transformation of the right portal vein (solid white arrows) with a normal appearance of the main portal vein (broken white arrows) and a large gastric varix arising from the left gastric vein (solid black arrows). (e) Angiographic images obtained during the gastric variceal embolization procedure (note coils [black arrows]). Transportal access was obtained, and contrast injection via a microcatheter demonstrated an extensive esophageal variceal network (A—Solid white arrows), as well as faint opacification of the PPVA (B—Broken white arrows). Surgical clips are also seen in the left upper quadrant from previous splenectomy.

emboli (Fig. 1b). The patient did recover neurologically despite a follow-up magnetic resonance imaging confirming multiple sub-acute infarcts.

This embolic ischemic stroke prompted evaluation for a right to left vascular shunt. Initially, an echocardiogram with an

agitated saline contrast bubble study was performed, which did not demonstrate a patent foramen ovale.

Endoscopic ultrasound was performed, which demonstrated an extensive gastroesophageal variceal matrix but could not identify a specific vascular shunt. Contrast-enhanced CT of

the thorax and abdomen was then performed, which demonstrated a PPVA connecting a large paraesophageal varix to the left inferior pulmonary vein (Fig. 1c). This study also demonstrated a diminutive right branch of the portal vein, although the main portal vein was normal in appearance.

The patient continued to experience symptomatic anemia requiring iron transfusions and erythropoietin. A decision was made to perform transhepatic portal venography with the aim of definitive identification of the PPVA and potential subsequent coil embolization of this vessel. The procedure was complicated by difficult access to the right portal vein, which necessitated direct main portal vein puncture. Contrast injection after portal vein access demonstrated cavernous transformation of the right portal vein (Fig. 1d) (explaining the difficulty in accessing this vessel) with extensive gastroesophageal collaterals.

A microcatheter was advanced into the left gastric vein (via the transhepatic portal access), and contrast injection confirmed a tortuous gastroesophageal variceal matrix, as well as faint opacification of the PPVA arising from a paraesophageal varix (Fig. 1e). Multiple attempts were made to access the PPVA directly; however, this was precluded by vessel tortuosity. Given the patient's persistent symptoms and anemia, a decision was made to perform coil embolization of the large gastric varices. The use of coils allowed controlled placement with minimal risk of systemic embolization. The patient recovered well after the procedure with no recurrent melena. A follow-up endoscopy demonstrated a much-improved appearance with flattening of the varices with air insufflation.

Discussion

The cause of portal hypertension in this patient is unknown. However, portal vein thrombus has been reported as a sequelae of splenectomy for hematological disorders. While symptomatic portal vein thrombosis after splenectomy is rare (2%), the total incidence of portal vein thrombosis after splenectomy is very high (53.5%).^{15–19} While we cannot definitively confirm that our patient developed portal vein thrombus after splenectomy, the presence of cavernous transformation of the right portal vein in the presence of noncirrhotic portal hypertension with gastric varices would certainly support this hypothesis.

While the exact incidence of PPVA in cirrhotic and non-cirrhotic portal hypertension remains uncertain, they have been reported in up to 30% of cirrhotic patients. These PPVA can be small and asymptomatic, but higher-flow shunts can lead to left ventricular hypertrophy and subsequent cardiac failure.^{20,21} This case demonstrates the important potential sequelae of cyanoacrylate injection in a patient with a PPVA. While these collateral pathways are uncommon and often clinically insignificant, the potential implication of systemic embolization via these vessels is substantial. In patients with portal hypertension who undergo cross-sectional imaging, dedicated evaluation of the variceal networks should be performed with the aim of identification of PPVA. However, where concern for a right to left shunt is high (e.g. in cases where oxygen saturation is significantly and unexpectedly reduced), bubble contrast echocardiography with injection via balloon-occluded retrograde transvenous venography is the most sensitive modality to identify potential PPVA.⁶ Endoscopists should be aware of the potential presence of these shunts

(particularly in the presence of large esophageal varices⁷), and where they have been previously demonstrated, injection of cyanoacrylate should be avoided. In these cases, novel endovascular techniques such as balloon-occluded retrograde transvenous obliteration of gastric varices may represent a safer technique as the PPVA itself can be identified, and controlled embolization can be performed.^{5,6}

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