

# An unusual cause of portal hypertension in a cirrhotic patient

Deeba Ali<sup>\*1</sup>, Wilfried Christian Fambo<sup>\*</sup>, Denis Brisebois<sup>\*\*</sup>, Yilmaz Gorur<sup>\*\*\*</sup>, Lionel Bosquee<sup>△</sup> and Benoit Cardos<sup>□</sup>

<sup>\*</sup>Services des Urgences CHC Saint Joseph, rue Hesbaye 75, 4000 Liège, Belgique., <sup>\*\*</sup>Services de Radiologie CHC Saint Joseph, rue Hesbaye 75, 4000 Liège, Belgique., <sup>\*\*\*</sup>Service de Radiologie CHU Sartilmant Liège, Avenue de l'Hôpital 1, 4000 Liège, Belgique., <sup>△</sup>Service des Urgences, Clinique Andr Renard, Herstal, Belgium., <sup>□</sup>Département des Sciences de la Santé Publique, Université de Liège, Liège, Belgium.

**ABSTRACT** Gastrointestinal bleeding is a frequent and severe complication of cirrhosis. The rupture of esophageal varices is the leading cause of upper gastrointestinal bleeding in cirrhosis. It occurs in about 30 to 40% of patients with liver cirrhosis and esophageal varices. A multidisciplinary approach is needed as it is a life-threatening condition. Trans-jugular intra-hepatic portosystemic shunt (TIPS) is the elected procedure in the management of refractory bleeding to drug and endoscopic treatment.

We report the case of the casual discovery of splenic arteriovenous fistula while performing TIPS in the context of alcoholic liver cirrhosis associating oesophageal varices and portal hypertension.

**KEYWORDS** transjugular intrahepatic portosystemic shunt, arteriovenous fistula, cirrhosis, portal hypertension

## Introduction

Portal hypertension is one of the most significant complications of both acute and chronic liver diseases. It results from an increase in vascular resistance at the prehepatic, intrahepatic, or posthepatic level. Gastroesophageal varices are the most important clinical manifestation of this syndrome and are associated with a high risk of hemorrhage and mortality. However, in the refractory cases, other causes of upper gastrointestinal hemorrhage should be ruled out. [1]

## Case report

This is a 45-year-old woman admitted to our emergency department for hematemesis starting in the morning. Her medical history was relevant for alcoholic liver cirrhosis with gastroesophageal varices (CHILD PUGH stage 3), previous gastric bypass surgery complicated by a perforated gastric ulcer and acute alcoholic pancreatitis three months ago.

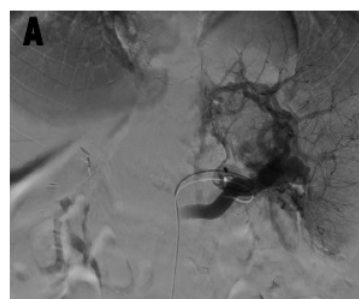
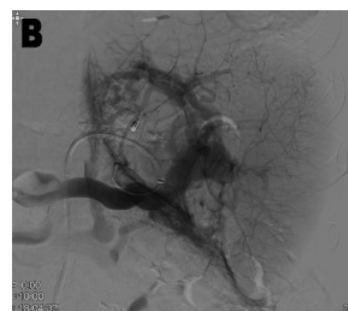


Fig A-B: A selective celiac arteriogram confirmed the arterio-venous fistula in the distal part of splenic artery. After the injection, the contrast agent is deviated into the splenic vein instead of the splenic mesenchyma.



Copyright © 2020 by the Bulgarian Association of Young Surgeons

DOI:10.5455/IJMRCR.portal-hypertension-cirrhoticpatient

First Received: October 26, 2019

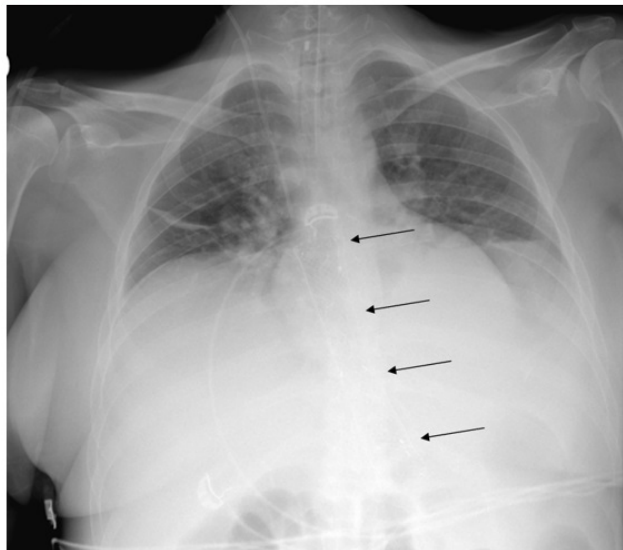
Accepted: November 07, 2019

Editor-in-chief: Ivan Inkov (BG)

<sup>1</sup>Services des Urgences CHC Saint Joseph, rue Hesbaye 75, 4000 Liège, Belgique;

Email: deeba.ali@chc.be

Figure A': Front view of a chest radiography show a expandable esophageal prosthese (arrows) from the middle part of the esophagus to the cardia.]



Upon admission to the emergency department, the physical exam revealed altered mental status, sinus tachycardia (140 bpm) and hypotension (80/60 mmHg). The cardiorespiratory exam was normal. Abdominal showed abundant ascites (grade 3). During the examination, the patient presented a new episode of hematemesis. The patient was subsequently sedated and intubated to protect the airway.

The laboratory panel showed severe anaemia (Hb 5.8 g/dl), thrombocytopenia (98000 mm<sup>3</sup>), prothrombin time(69%), hyperlactacidemia (5.4 mmol /L), slightly increase of aspartate aminotransferase and alanine aminotransferase (47 U/L each) as well as elevated gamma-glutamyl transpeptidase (156 U/l). Ethanol level was 2.25 g/dl.

An upper gastrointestinal endoscopy was describing the presence of esophageal varices stage 3 treated with five elastic ligatures. The patient received three units of blood and one of platelets before she was transferred to the intensive care unit (ICU).

At ICU, treatment with continuous proton pump inhibitor (pantoprazole) perfusion, somatostatin perfusion (3 mg per day) and anti prophylaxis with ceftriaxone 1 gr daily.Five litres of fluid were withdrawn by abdominal paracentesis and human albumin solutions administered.

During her stay at the ICU, haematemesis recurred being managed by endoscopic sclerosis, clipping and ligatures of the varices. Forty-eight hours after these procedures were performed; the patient presented a new episode of hematemesis. In this setting, a metal prosthesis (Danis Stent) was placed by the endoscopic procedure (figure A'). At three day, the patient was hemodynamically stable, she was extubated and the prosthesis removed at day five.She was transferred to internal medicine.

Unfortunately, one week after, the patient presented a new episode of haematemesis leading to hemorrhagic shock. She was readmitted to the ICU, and after a multidisciplinary approach (gastroenterologists, anesthesiologists and interventional radiologists) the performance of TIPS was retained.

The hepatic catheterization was difficult owing to the high portal pressures; the catheter is blocked at the sus-hepatic level, and there is absence of opacification of the portal vein. The sonographic Doppler study showed permeability of the sus-hepatic and portal veins. The investigations were completed with an abdominal computed tomography revealing the existence of a splenic arteriovenous communication between the hilum of the spleen leading to an opacification of the splanchnic venous system and subsequently explaining the high pressure.

Selective catheterization of the splenic artery confirmed the arteriovenous fistula in the hilum of the spleen. Considering the tortuous aspect of the splenic artery, only the proximal splenic artery is occluded (Figure A, B, C, D). The absence of hematemesis marked the clinical course and the patient was released from the ICU after three weeks of stay.

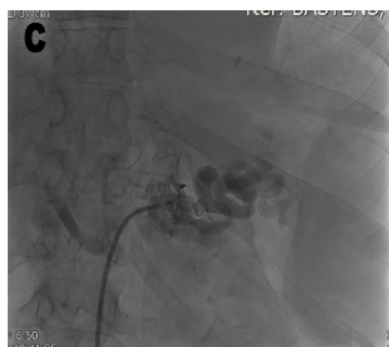


Fig. C-D: The fistula is excluded with the deployment of a vascular plug in the proximal part of the splenic artery. The device is a Amplatzer AVP4. The controle after the treatment confirme the complete exclusion of the fistula.



## Discussion

Portal hypertension (PHT) is a life-threatening condition, as in the case reported. Numerous causes of portal hypertension exist. The aetiology can be classified as prehepatic (portal vein thrombosis, splenic, arteriovenous fistula), intrahepatic (primary biliary cholangitis, sarcoidosis, liver fibrosis, cirrhosis), or posthepatic reasons (Budd-Chiari syndrome). Cirrhosis is the most common cause of portal hypertension in industrialized countries [1]. Nevertheless, many other pathologies can be at its origin, leading to a diagnostic pitfall.

Particularly in the case reported, the management was focused on the variceal disease in cirrhosis but in the presence of another entity lately diagnosed and also contributing to portal hypertension. As a result, portal hypertension was due to cirrhosis and arteriovenous fistula.

The primary complications of PHT are gastrointestinal bleeding, ascites and hepatic encephalopathy. Gastrointestinal bleeding is a frequent and severe complication of PHT being oesophageal varices ruptures the most common cause. It occurs in 30% to 40% of cirrhotic patients with oesophageal varices, and it is associated with 20% short-term mortality. Several factors of poor prognosis have been described: impaired liver function, PHT greater than 20 mmHg, active bleeding at endoscopy. Treatment of oesophageal varices rupture is well codified by the French and American consensus conferences and the recommendations of Baveno, protocols include: protection of the airways, fluid resuscitation, blood transfusion, prophylactic antibiotic therapy, vasoactive treatment (somatostatin, Terlipressine) and mechanic bleeding control by endoscopy (ligature and clip).[1, 2]

In 10 to 20% of cases, the haemorrhage is refractory to drug and endoscopic treatment, as shown in the case of our patient so the use of a self-expansive metal esophageal prosthesis could be recommended. This procedure allows an immediate bleeding control, and subsequently clinical improvement before transjugular intrahepatic portosystemic shunt (TIPS) or the liver transplantation could be performed.[3,4] In the case of refractory hemorrhage, interventional radiological TIPS could be a useful option as in this case.

This procedure was challenging to perform because of high pressure in the portal system. This fact incidentally revealed the presence of an arteriovenous fistula in this patient in whom the PHT was initially attributed exclusively to cirrhosis. Arteriovenous fistulas are uncommon, and they could be either congenital or secondarily to a surgical procedure, a biopsy puncture or aneurysmal pathology. Fistulas are commonly located in the splenic hilum or the splenic trunk. They are generally revealed by an array of portal hypertension due to excess blood supply in the portal system, depending on the size of the arteriovenous communication. Diagnosis is confirmed by computed tomography or magnetic resonance imaging with contrast injection. Interventional radiological embolization is the first choice of treatment, but the endovascular occlusion of the splenic artery could lead to complications such as massive splenic infarction progressing to an abscess, septic shock, or splenic rupture. If this procedure fails, splenectomy could be considered.[5]

In this case, the previous history of complicated gastric bypass surgery complicated probably contributed to the occurrence of the arteriovenous fistula. Its embolization has prevented the recurrence of oesophageal varices rupture by drastic diminishing portal hypertension.

## Conclusion

Upper gastrointestinal bleedings are one of the most common complications of the cirrhosis and more generally of portal hypertension. It associated with high-level mortality. In case of refractory bleeding not responding to treatment other etiologies of portal hypertension should be considered.

## Learning points

- In the case of refractory upper gastrointestinal bleeding to conventional treatment, other etiologies must be ruled out

- Portal hypertension is a complex entity
- Arteriovenous fistula could be at the origin of refractory upper gastrointestinal bleedings

## Conflict of Interest

There are no conflicts of interest to declare by any of the authors of this study.

## Funding

None

## References

1. D. Thabut, J. Massard, M. Rudler . Prise en charge des hémorragies digestives liées à l'hypertension portale. Réanimation( 2007).
2. Maeva Guillaume 1Jean-Paul Cervoni 2Carine Chagneau-Derrode. Hypertension portale: physiopathologie, cause, diagnostic et traitement. John Libbey. Volume 22, janvier 2015
3. Garcia-Pagan JC, Caca K, Bureau C et al. Early use of TIPS in patients with cirrhosis and variceal bleeding. N Engl J Med 2011;362(25):2370-9.
4. Boyer TD, Haskal ZJ. American Association for the Study of Liver Diseases Practice Guidelines: the role of transjugular intrahepatic portosystemic shunt creation in the management of portal hypertension. J Vasc Interv Radiol 2005;(5):615-29.
5. C. Henry, L Detti Off, R Mhiri, A Tournade. Fistule Artérioveineuse intrasplénique post-traumatique chez un enfant. J radiologie 1998 ;79 : 271-273