Torrential upper gastrointestinal bleeding from 'downhill' oesophageal varices complicating long term central venous access for total parenteral nutrition

Eu Jin Lim,¹ Damien L Stella,² David M Russell¹

¹Department of Gastroenterology and Clinical Nutrition Service, Royal Melbourne Hospital, Victoria, Australia

²Department of Radiology, Royal Melbourne Hospital, Victoria, Australia

Correspondence to

Dr Eu Jin Lim, Department of Gastroenterology and Clinical Nutrition Service, Royal Melbourne Hospital, Grattan Street, Parkville, Victoria 3052, Australia; ejlim@rocketmail.com

Accepted 13 January 2010
Published Online First
7 April 2010

Oesophageal varices usually develop in the setting of portal hypertension secondary to chronic liver disease. However, superior vena cava (SVC) obstruction can result in 'downhill' varices forming in the upper oesophagus. A case of torrential upper gastrointestinal bleeding from SVC obstruction due to chronic central venous access for home total parenteral nutrition is described. It is suggested that in patients presenting with gastrointestinal bleeding in the setting of SVC obstruction, 'downhill' varices should be suspected. The current literature is discussed regarding management of such varices. It is recommended that endoscopic variceal surveillance be carried out in patients with known SVC obstruction.

Clinical record

We describe the case of a 68-year-old man with no chronic liver disease who developed superior mesenteric artery occlusion which required the resection of much of his small bowel and proximal large bowel, resulting in short gut syndrome. A right subclavian vein Hickman's catheter was inserted for long term home total parenteral nutrition (TPN). After 7 years, with multiple line infections requiring intravenous antibiotics and Hickman's catheter changes utilising both left and right subclavian veins alternately, he developed facial and upper limb swelling. CT venography revealed high grade stenosis of both the left and right brachiocephalic veins, effectively causing superior vena cava (SVC) syndrome. This was thought to be thrombotic in nature but attempted thrombolysis with alteplase was unsuccessful and he was commenced on warfarin to prevent clot extension.

After 8 months of anticoagulation, he

presented to hospital with large volume melaena after recently being prescribed a course of oral antibiotics for a minor illness. On arrival, he was unwell with hypotension and tachycardia. Initial investigations revealed a haemoglobin level of 76 g/l, urea 15.4 mmol/l, creatinine 82 µmol/l and international normalised ratio 4.9. Aggressive resuscitation with blood was provided and his anticoagulation was reversed with intravenous vitamin K and fresh frozen plasma. Emergent upper gastrointestinal endoscopy showed actively bleeding oesophageal varices in the upper oesophagus, measured at 18 cm from the incisors (figure 1), with no varices noted at the gastro-oesophageal junction. Endoscopic variceal band ligation was undertaken proximal to the point of bleeding and haemostasis was achieved.

The patient's left subclavian vein Hickman's catheter was removed in an attempt to relieve the SVC obstruction. Unfortunately, no improvement was noted in his facial or upper limb swelling. A magnetic resonance venogram performed to evaluate the state of his central venous thromboses revealed no filling of internal jugular, subclavian or brachiocephalic veins bilaterally with multiple collateral veins within the chest (figure 2). It was felt that his venous thromboses were so extensive that attempted percutaneous venoplasty or surgical venous bypass would be too dangerous. On repeat endoscopy, varices were again noted in the upper oesophagus. As such, a further two variceal banding sessions, each 2 weeks apart, were done to eradicate his varices. He remains well off home TPN on an oral diet and high dose nutritional supplements. If he were to require home TPN in the future, the only central venous access available will be via the inferior vena cava.

Figure 1 Endoscopic appearance of bleeding upper oesophageal varices.

Figure 2 Magnetic resonance venogram image of loss of both brachiocephalic systems. LBCV, left brachiocephalic vein; LIJV, left internal jugular vein; LSCV, left subclavian vein; RBCV, right brachiocephalic vein; RIJV, right internal jugular vein, RSCV, right subclavian vein.

Discussion

Upper gastrointestinal bleeding from oesophageal varices is a common complication of portal hypertension secondary to chronic liver disease. Varices typically develop at the gastro-oesophageal junction and travel up the lower oesophagus in order to decompress the portal system via the formation of porto-systemic shunts. However, in our case, the use of central venous catheters for TPN resulted in stenosis of both brachiocephalic veins producing SVC syndrome. In order to decompress the SVC hypertension, varices develop in the upper oesophagus and travel down towards the portal system.¹

Upper gastrointestinal bleeding from downhill varices has been reported with SVC compression by large goitres²

and lung cancer.³ SVC thrombosis can also occur as a consequence of thrombophilia⁴ or as a complication of central venous catheters for haemodialysis.^{5,6} We have reported the first case of bilateral brachiocephalic vein stenoses effectively causing SVC syndrome due to repeated tunnelled central venous access for long term home TPN. The cause of progressive brachiocephalic venous occlusion was suspected to be due to a combination of chronic TPN use, which is known to be irritant to veins and may predispose to thrombus formation, and recurrent intravenous line related sepsis causing thrombophlebitis.

Initial therapy involves achieving haemostasis endoscopically. In this regard, variceal band ligation is preferred over sclerotherapy as the latter has a higher complication rate, including spinal cord paralysis⁷ and pulmonary embolism.⁸ After bleeding is controlled, management is then focused on relieving the SVC obstruction in order to prevent recurrent bleeding. Definitive treatment is done either by percutaneous radiologically guided venoplasty with or without stenting or the surgical creation of a venous bypass.⁹ Unfortunately, in our patient, neither the radiological nor the surgical option was viable and he embarked on a course of repeated endoscopic variceal ligations in order to eradicate his varices. He will require ongoing endoscopic surveillance to monitor for variceal recurrence.

In our patient, the presence of oesophageal varices was not detected until he presented with a life threatening upper gastrointestinal haemorrhage. We recommend that patients on long term TPN via catheters inserted into the SVC, who develop SVC syndrome, should undergo upper gastrointestinal endoscopy for variceal screening.

Conclusion

Long term central venous access for home TPN can result in SVC obstruction and one should suspect 'downhill' oesophageal varices in any patient with a history of SVC syndrome presenting with an upper gastrointestinal haemorrhage. Endoscopic variceal ligation is effective in treating bleeding 'downhill' varices. If SVC obstruction cannot be decompressed radiologically or surgically, repeated variceal banding sessions can eradicate 'downhill' varices.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; not externally peer reviewed

Contributors EJL wrote up the case report, performed a literature review and discussion, and obtained the endoscopic photographs. DLS reviewed, reported and annotated the magnetic resonance venogram. DMR consulted on the case and supervised writing up of the paper.

References

1. Felson B, Lessure AP. "Downhill" varices of the esophagus. *Dis Chest* 1964;46:740–6.
2. Fleig WE, Stange EF, Ditschuneit H. Upper gastrointestinal hemorrhage from downhill esophageal varices. *Dig Dis Sci* 1982;27:23–7.
3. Tanaka H, Nakahara K, Goto K. Two cases of downhill

- esophageal varices associated with superior vena cava syndrome due to lung cancer. *Nihon Kyobu Shikkan Gakkai Zasshi* 1991;**29**:1484–8.
4. **Vorlop E**, Zaidman J, Moss SF. Clinical challenges and images in GI. Downhill esophageal varices secondary to superior vena cava occlusion. *Gastroenterology* 2008;**135**:1863, 2158.
 5. **Lutisan JG**, Ploem S, Zijlstra JG. Bleeding non-cirrhotic fundus (“downhill”) varices in a patient on chronic intermittent hemodialysis. *Eur J Intern Med* 2006;**17**:586.
 6. **Hussein FA**, Mawla N, Befeler AS, *et al.* Formation of downhill esophageal varices as a rare but serious complication of hemodialysis access: a case report and comprehensive literature review. *Clin Exp Nephrol* 2008;**12**:407–15.
 7. **Heller SL**, Meyer JR, Russell EJ. Spinal cord venous infarction following endoscopic sclerotherapy for esophageal varices. *Neurology* 1996;**47**:1081–5.
 8. **Tsokos M**, Bartel A, Schoel R, *et al.* Fatal pulmonary embolism after endoscopic embolization of downhill esophageal varix. *Dtsch Med Wochenschr* 1998;**123**:691–5.
 9. **Froilán C**, Adán L, Suárez JM, *et al.* Therapeutic approach to “downhill” varices bleeding. *Gastrointest Endosc* 2008;**68**:1010–12.