Transjugular intrahepatic portosystemic shunt placement for symptomatic non-bleeding anorectal varices in nodular regenerative hyperplasia

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Abstract

In this paper we present the case of a male 55-year old patient with known ulcerative colitis and nodular regenerative hyperplasia, a rare form of noncirrhotic portal hypertension. He presented four visits to the emergency department with rapidly progressive anal discomfort. After two weeks a transjugular intrahepatic portosystemic shunt was placed using the gun-sight technique with immediate relief of the unbearable anal pain and pressure.

To our knowledge, this is the first case where transjugular intrahepatic portosystemic shunt placement is applied for non-bleeding, congestive anorectal varices. (Acta gastroenterol. belg., **2020**, 83, **331-333**).

Key words : noncirrhotic portal hypertension, perirectal congestion, interventional radiology.

Case

A 55-year-old man presented to the emergency department (ED) with complaints of pain in the anal region since 3 days. Relevant medical history included ulcerative colitis, nodular regenerative hyperplasia (NRH) with portal hypertension and an esophageal variceal bleeding. The NRH was diagnosed by pathological examination 7 years earlier and was presumed to be secondary to the use of azathioprine, also in the presence of a heterozygous genotype of thiopurine S-methyltransferase (TPMT). The ulcerative colitis had been well controlled for years with oral mesalazine maintenance therapy and last gastroscopy 18 months earlier showed only small esophageal varices. One week before onset of the anal discomfort, his general practitioner had prescribed a course of ciprofloxacin because of aspecific lower abdominal pain. After start of the antibiotic therapy, he developed diarrhea, which was still present at the time of presentation. He described the anal pain as a continuous burning sensation inside the anus. There were no other complaints. Anal inspection and palpation did not reveal any abnormalities. Laboratory analysis yielded an elevated C-reactive protein (CRP) of 28 mg/L with a stable chronic thrombocytopenia grade 2. As an exacerbation of ulcerative colitis was suspected, rectal mesalazine was added to the maintenance therapy. The next day our patient represented to the ED because of intensified anal discomfort, prohibiting him from even sitting down. Faecal calprotectin was elevated (460 µg/g faeces) and the CRP had risen to 57 mg/L. Sigmoidoscopy revealed mild rectosigmoiditis (endoscopic Mayo score 1). Faeces culture seemed positive for Campylobacter Coli, but the diarrhea had already disappeared. Mesalazine suppositories were continued and the patient was discharged after one night hospitalization.

Three days later he reconsulted the ED because of reoccurrence of the stabbing anal sensation. There were no other symptoms. CT scan of the abdomen showed unchanged splenomegaly (22 cm) and esophageal varices, with a new important collateral circulation and extensive rectal varices. Proctologic examination confirmed the presence of diffuse anorectal varices (ARV) without thrombus or pain on palpation. Retrospectively, these varices were already present on the sigmoidoscopy images 3 days before. The patient was given an appointment for pelvic MRI scan and ileocolonoscopy, and was sent home with appropriate analgesia.

After five days our patient readmitted to the ED for the fourth time in twelve days with unbearable anal pain and pressure, preventing him from sleeping. Ileocolonoscopy the day before showed prominent ARV and no more signs of colitis (Figure 1). Laboratory analysis showed a normalized CRP. MR imaging revealed important rectal and perirectal congestion (Figure 1). The decision was made to place a transjugular intrahepatic portosystemic

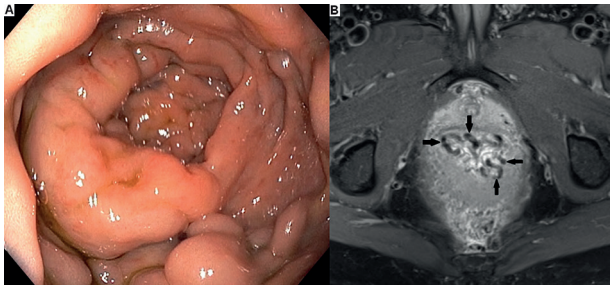

Figure 1. — Congestive anorectal varices. (A) Colonoscopy showing enlarged tortuous rectal varices as submucosal elevations. (B) An axial T2-weighted fat-saturated MR image of the rectum showing a vast submucosal varicous network (arrows).

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Figure 2. — A direct portography image showing the creation of a portosystemic shunt, before stent deployment, using ultrasound-guided transhepatic puncture (arrowheads). Note the prominent intrahepatic portal varices (arrows) that prevented a classic transjugular approach.

shunt (TIPS). Under ultrasound guidance the right jugular vein was punctured and a long 10Fr sheath was placed in the caval vein. A 4Fr Cobra catheter was used to catheterize the right hepatic vein and a 14-gauge stiffening cannula was advanced. With a 0.038 trocar stylet the veins on the portal side of the liver were punctured. However, because of strongly pronounced intrahepatic portal varices, typical of NRH with noncirrhotic intrahepatic presinusoidal portal hypertension, the portal vein could not be catheterized through a classic transjugular approach. Therefore the 'gun-sight' technique was used (Figure 2), a more challenging technique with transjugular as well as percutaneous transhepatic approach, described in 1996 by Haskal *et al.* and used in cases of unfavourable venous anatomy (1). Under continuous sonographic guidance a 22-gauge Chiba needle was transhepatically advanced through the right portal vein and further on through the right hepatic vein close to the junction with the inferior caval vein. A 0.018 nitinol guidewire was used to place a 6Fr sheath through the punctured route. A 0.035 hydrophilic guidewire was passed through the sheath into the right atrium. The long transjugular sheath was placed in the right atrium and the transhepatic wire was retrieved using a gooseneck snare. At the entrance side (right abdominal wall) the distal tip of the guidewire was fixed with a mosquito forceps, preventing it to be completely pulled through from the jugular side. Now a 4Fr C2 catheter was advanced over the wire from the transjugular side and the transhepatic 6Fr sheath was pulled (by hand) and pushed back (by the C2 catheter) under sonographic guidance till in the right portal vein. The transhepatic-transjugular wire was removed and a new hydrophilic wire was introduced through the C2 catheter and manipulated into the main portal vein and further on into the superior mesenteric vein. From now on the TIPS procedure could be continued in a standard way with deployment of an 8+2 cm long endoprosthesis (Viatorr, Gore), dilated to a diameter of 8 mm. Pressure measurements before TIPS were 5 mm Hg systemic and 20 mm Hg portal and immediately after TIPSS 6 mm Hg systemic and 17 mm Hg portal. The endoprosthesis reduced the portosystemic gradient from 15 to 11 mm Hg. The transhepatic needle tract was embolized with glue.

Immediately following the procedure, an important improvement in the anal discomfort was noticeable. Duplex ultrasonography after 3 days showed a patent TIPS. Three weeks later our patient declared to be asymptomatic since the day of TIPS placement. CT scan showed no more significant congestion in the rectal region. After 10 months he remained asymptomatic. The ARV had disappeared on sigmoidoscopy whilst MRI scan showed regression of the edematous thickening in the anorectal mucosa and decrease of the submucosal venous varices.

Discussion

Anorectal varices (ARV) are dilated submucosal veins extending proximal to the dentate line and into the rectum, being clearly distinct from hemorrhoids. In the case of portal hypertension, they represent porto-systemic collaterals connecting the superior hemorrhoidal veins of the portal circulation and the middle and inferior hemorrhoidal veins of the systemic circulation. The commonest cause of ARV in the context of portal hypertension is liver cirrhosis, with an estimated prevalence between 38% and 56% (2). In noncirrhotic portal hypertension, these rates are deemed to be higher (3). In contrary to esophageal varices, no epidemiological data are available of ARV in NRH. The diagnosis of ARV is classically based on anoscopy and flexible sigmoidoscopy, but may be missed in case of insufficient insufflation. Clinically significant bleeding from rectal varices is rare, occurring in 0,5-5% of the cases, but can be life threatening (2). As there are no prospective studies on the management of bleeding ARV, a generally accepted treatment algorithm is lacking. The first report on the use of TIPS for bleeding ARV dates from 1993 (4). Since then, successful use has been reported in several case reports (5-6). To our knowledge, no other symptomatology of complications of ARV have been described in the literature to date.

NRH is a rare liver disease characterised by diffuse transformation of hepatic parenchyma into small regenerative nodules. Approximately 27% of noncirrhotic portal hypertension cases are due to NRH, for example after prolonged exposure to thiopurines (7). TIPS procedures have been described in the setting of NRH

to alleviate bleeding complications.(8,9). Noteworthy, in presinusoidal causes of portal hypertension, the catheter is not in continuity with the actual area of increased resistance, so the recorded pressure will be that of the normal sinusoids (with normal intersinusoidal anastomosis), and the wedged hepatic venous pressure will often be underestimated as is the case in our patient (10). As the hepatic synthetic function is preserved in NRH, hepatic encephalopathy due to shunt procedures is less common.

We presumed that the rectal complaints in our patient were related to congestion of the rectal varices, analogous to the pelvic congestion syndrome in which congestion of the pelvic veins is assumed to caused chronic pelvic pain (11). Beta-blockers, which have a preventive role, were not started in this subacute onset of anal complaints. Nor did we apply obturation with histoacryl injection because it only has a mucosal effect in treatment of bleeding, whereas the unsupportable anal pain in our patient was due to submucosal swelling. Endoscopic and radiographic imaging had excluded other differentials. The spectacular resolution of the rectal complaints immediately following the TIPS procedure is a strong argument for congestion as causative mechanism. To our knowledge, this is the first case where TIPS placement is applied for non-bleeding, congestive ARV with remarkable relief of subacute severe anal complaints.

Conflicts of interest

No conflicts of interest.

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