Turk J Gastroenterol 2003; 14(4): 266-269

Arterioportal fistula causing jejunal variceal hemorrhage

Jejunal varis kanamasına neden olan arteriyoportal fistül

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Hepatoportal arteriovenous fistulas are an uncommon cause of portal hypertension and may lead to severe gastrointestinal bleeding. Esophageal varices are the main source of hemorrhage in patients with portal hypertension. We report a 40-year-old man with a hepatic arterioportal fistula, who had gastrointestinal bleeding from jejunal varices. He had had hematemesis four years previously and was diagnosed as esophageal variceal bleeding secondary to hepatic arterioportal fistula-complicated portal hypertension. The fistula had been successfully closed with a detachable balloon but it was found to have reformed during his recent hospitalization for jejunal variceal bleeding. Alcoholism and development of portal vein thrombosis afterwards were the other contributing factors for jejunal variceal formation. Hemorrhage was controlled with medical treatment. Since he refused any surgical intervention he was discharged and advised to continue follow-up.

Key words: Hepatic aneurysm, arterioportal fistula, jejunal varices, variceal bleeding

Hepatoportal arteriovenöz fistüller, portal hipertansiyonun nadir nedenlerindendir ve ciddi gastrointestinal kanamalara yol açabilirler. Portal hipertansiyonda en önemli kanama kaynağı özofagus varisleridir. Bu olgu sunumunda 40 yaşında jejunal varis kanaması ile gelmiş olan bir hepatik arteriovenöz fistül vakası tartışılmıştır. Olgu, 4 yıl önce özofagus varis kanaması nedeniyle incelenirken, hepatoportal arteriovenöz fistül tanısı almıştı. Anjiografik balon uygulaması ile fistül kapatılmıştı. Dört yıl sonra gelişen jejunal varis kanaması medikal tedavi ile kontrol altına alındı. Cerrahi girişim istemeyen hasta izlemeye alındı.

Anahtar kelimeler: Hepatik anevrizma, arterioportal fistül, jejunal varis, varis kanaması

INTRODUCTION

Hepatic arterioportal fistulas (APF)s are rare vascular lesions and usually complicate penetrating abdominal trauma and gastrointestinal surgery (1). They can be small and asymptomatic (2, 3), but often cause symptoms as a result of portal hypertension. APFs are a reversible cause of portal hypertension; therefore, a correct diagnosis is pivotal because a definitive therapy can be proposed (4).

Esophagogastric varices are a common manifestation of portal hypertension. Bleeding from jejunal varices is a very uncommon finding. It may present as a severe, even fatal, gastrointestinal hemorrhage or as recurrent episodes of bleeding of unknown origin (5, 6).

This report describes a patient with relapsed APF presenting with jejunal variceal bleeding. Chronic alcoholism and portal vein thrombosis were the other contributing events for the development of portal hypertension.

CASE REPORT

A 40-year-old man was admitted to Türkiye Yuksek ihtisas Hospital with a complaint of hematemesis. He had reported 20 g/day alcohol consumption for 15 years; he denied any drug intake. Four years previously, he had had one episode of hematemesis, which was the first occurrence. He was diagnosed as having portal hypertension secondary to hepaticoportal fistula. Previous penetra-

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Manuscript received: 24.06.2003 **Accepted:** 02.12.2003

ting abdominal injury was thought to be the cause of fistula formation between the right hepatic artery and the right branch of the portal vein. Esophageal varices were the source of bleeding. Band ligation to the bleeding varices and detachable balloon application to the fistula were performed. Before discharge, upper gastrointestinal endoscopy was repeated and disappearance of both esophageal varices and portal hypertensive gastropathy was demonstrated.

In the ensuing years, he was completely symptom-free. He was lost to follow-up until the same complaint reappeared.

On recent admission, physical examination revealed a systolic-diastolic flow murmur over the right upper abdomen and splenomegaly. Rectal examination demonstrated the presence of black-colored, liquid stool. Hemoglobin level was 12.7 g/dl, hematocrit: 40.3%, MCV: 89.8 fL, white blood cell count: 4.6 x 109/L and platelet count: 128 x 109/L. Prothrombin time was 14.7 sec (N: 9.8-15.0 sec). Serum ALT and AST were 25 u/L and 64 u/L, respectively (N: 40 u/L for both), serum GGT: 234 u/L (N: 8-61), ALP: 138 u/L (N), total bilirubin: 3.12 mg/dl (N<1.1), direct bilirubin: 0.95 mg/dl (N<0.3), and albumin: 2.4 g/dl. Other biochemical tests were within the normal limits. Viral markers for hepatitis including hepatitis B and C viruses, autoantibodies (antinuclear, antimitochondrial, anti-smooth-muscle, anti-liver-kidney microsomal enzymes and anti-soluble liver antigen) were also negative. Serum alpha-fetoprotein was normal. Upper gastrointestinal endoscopy revealed non-bleeding esophageal and fundal varices and mild portal hypertensive gastropathy. On abdominal sonography, mild ascites was seen in addition to splenomegaly. Liver was fine-granular, normal-sized and its contours were mildly irregular. A well-demarcated mass (about 5 cm diameter) on the 4th segment of the liver was also seen. For differential diagnosis of the mass, color Doppler sonography was used, showing a turbulent arterial waveform with high peak systolic velocity, consistent with a hepatic artery aneurysm, and a fistula between the right hepatic artery and right portal vein. A partial thrombosis in the main portal vein, that was not present four years previously, was also documented. The described lesion is shown on computerized tomography in Figure 1. Selective hepatic and superior mesenteric angiography provided the definite diagnosis of an arterioportal fistula in the right lobe of the liver (Figure 2).

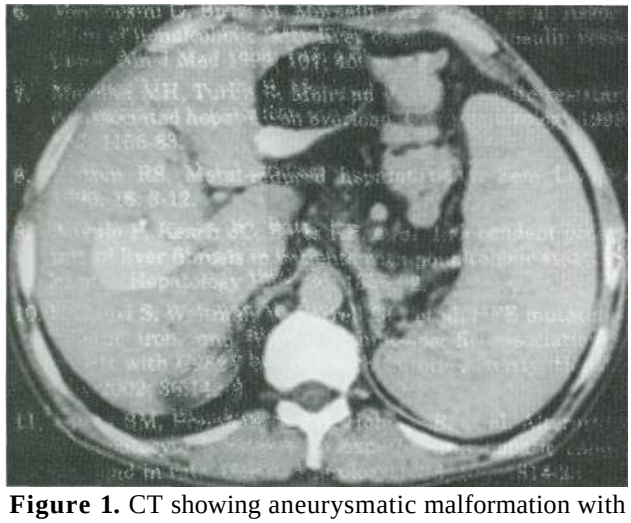

Figure 1. CT showing aneurysmatic malformation with tumor-like appearance

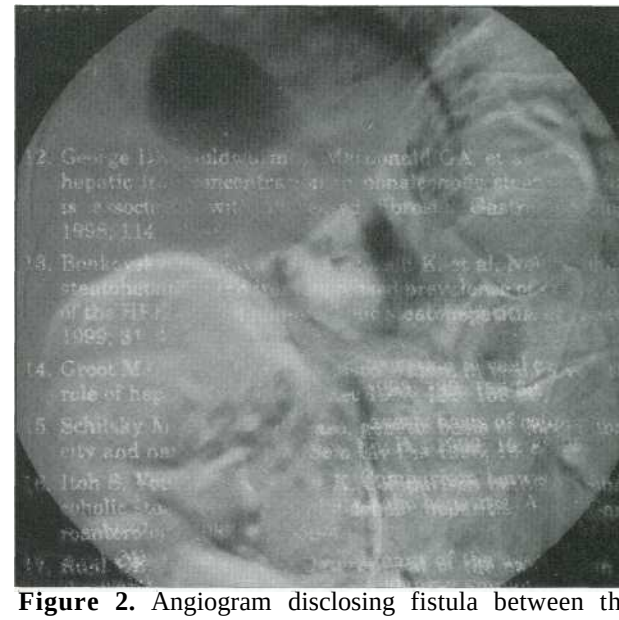

Figure 2. Angiogram disclosing fistula between the right hepatic artery and the right branch of the portal vein

During hospitalization he had melena, but no hematemesis, and blood count revealed gradually falling hemoglobin levels. Upper gastrointestinal endoscopy was repeated, showing the same findings as with the first endoscopy. Colonoscopy demonstrated black-colored blood clots throughout the colon. Push enteroscopy disclosed bleeding jejunal varices (Figure 3). Somatostatin infusion and conservative treatment were done and hemorrhage was stopped. He had no rebleeding during our follow-up.

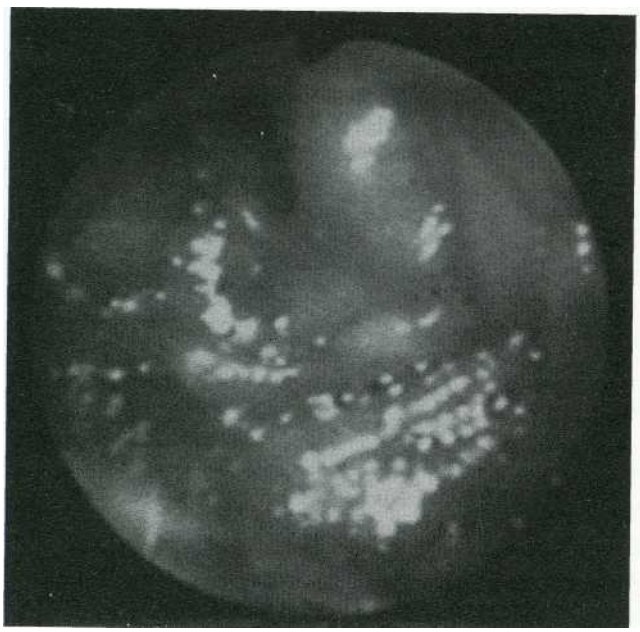

Figure 3. Enteroscopic imaging of submucosal varices in the jejunum

Because he refused any surgical intervention he was discharged and advised to continue follow-up.

DISCUSSION

Arterioportal fistulaes are a rare cause of portal hypertension (7). They may be congenital or secondary to various states including abdominal trauma, surgery, percutaneous transhepatic procedures and several liver diseases (3). Hepatic artery aneurysms, rare vascular lesions, may be the preceding pathology for fistula formation. Whether a patient will become symptomatic and, if so, after what period of time, depends mainly on the size and localization of the fistula and concomitant liver diseases (2, 8-10). The majority of symtomatic patients with APF present with symptoms like hemorrhage, hemobilia, abdominal pain, and diarrhea within weeks to months following injury. The clinical findings in some cases are related with portal hypertension or high-output heart failure, such as gastrointestinal hemorrhage, ascites, and splenomegaly (3). As seen in our patient, progression of portal hypertension and related symptoms may occur after a long-lasting asymptomatic period (11).

A correct diagnosis of APFs is important, as these are a curable cause of portal hypertension. With the advanced imaging techniques such as dynamic computerized tomography and magnetic resonance imaging, shunts are becoming more commonly encountered than in the past. Recognition of arterioportal shunt can suggest the presence of a previously unsuspected disorder and avoids false-positive diagnosis or overestimation of a hepatic disease. Although chronic alcoholism in our patient was a risk factor for portal hypertension, disappearance of esophageal varices and portal gastropathy following fistula repair demonstrates that the main contributing factor was APF formation.

The treatment options depend on the cause, size and localization of the fistula, the condition of the patient and the duration of symptoms (11). The preferred method for intrahepatic fistulas is angiographic embolization with gelfoam, steel coils, detachable balloons, or bucrylate, and the procedure has been adopted with success, providing the fistula is not too large and is accessible (8, 12, 13). Angiographic embolization with detachable balloon was successfully applied to our patient at first. Afterwards, thrombosis of the portal vein occurred, possibly due to a complication of angiographic embolization, which then aggravated the existing portal hypertension secondary to relapsed APF. Although there was no biopsy-proven cirrhosis, continuing alcohol abuse may have been another risk factor for portal hypertension.

Esophagogastric varices are the most common site of variceal bleeding due to portal hypertension. Ectopic varices may occur in the duodenum, jejunum, ileum, colon, and rectum. The prevalence of symptomatic jejunal varices is quite low (14). In the literature there are few case reports describing isolated jejunal variceal bleeding (1, 5, 6, 15). In our patient there seems to have been several factors contributing to portal hypertension and consequently to jejunal variceal formation. To our knowledge, jejunal variceal bleeding due to intrahepatic arterioportal fistula has not been reported before. Unfortunately, we cannot demonstrate the basic etiology definitively, since APF, portal vein thrombosis, and alcoholic liver disease may be the cause of portal hypertension separately or all together.

In conclusion, hepatic artery aneurysm may present as a huge mass in the liver and it should be kept in mind in differential diagnosis of space-occupying liver pathologies. As in our patient, it may resemble hepatoma in patients with chronic liver disease. Jejunal variceal bleeding must be considered for portal hypertensive patients with APFs of the liver and an unidentified source of hemorrhage.

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