

## Case Reports

# SPLenic ARTERIOVENOUS FISTULA: A RARE LESION CAUSING BLEEDING ESOPHAGEAL VARICES

A.A. KESHAVARZ, M.D., M. YOUNESI, M.D., AND M. RAHBAR, M.D.

*From the Department of Gastroenterology and Pathology, Imam Khomeini Hospital,  
Kermanshah University of Medical Sciences, Kermanshah, Islamic Republic of Iran.*

### ABSTRACT

We report a case of a 40 year old man with portal hypertension caused by a splenic arteriovenous fistula that was diagnosed at laparotomy. He presented with bleeding esophageal varices and was initially treated by sclerotherapy. At laparotomy, portal pressure was 40 cmH<sub>2</sub>O but fell to 20 cm H<sub>2</sub>O after the fistula was treated with splenectomy.

All symptoms disappeared shortly after operation and the patient has remained well for the past two years.

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**Keywords:** Arteriovenous fistula, Portal hypertension.

### INTRODUCTION

Portal hypertension is a result of chronic liver disease in the majority of cases. Rare potentially curable causes of portal hypertension include vascular conditions such as hepatic or portal venous thrombosis or arteriovenous fistula. Most reported cases have been in women.

### CASE REPORT

A 40 year old man was well until 10 years ago when he developed several episodes of hematemesis necessitating hospitalization and transfusions. He was admitted in our hospital with hematemesis two years ago.

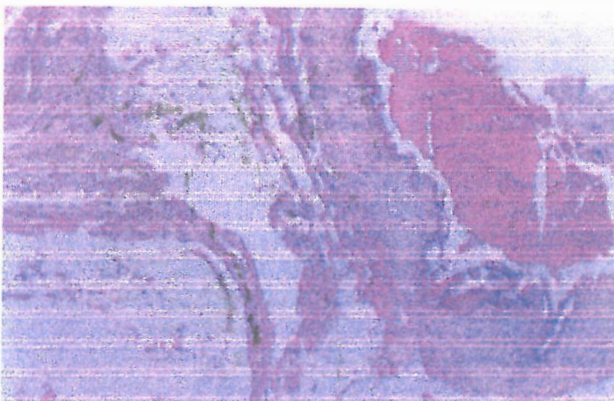
On examination he appeared ill, but there were no spider angiomas, palmar erythema, or jaundice. The abdomen was moderately distended, with no evidence of ascites. The liver was normal in size to percussion, but the spleen was enlarged 6 cm below the costal margin.

Laboratory studies revealed hemoglobin 7.2 g/dL, white blood cell count 7000/mm<sup>3</sup>, platelet count 130,000/mm<sup>3</sup>, total serum bilirubin 1mg/dL, AST 35 units/dL, ALT 40 units/dL, prothrombin time was normal, and albumin was 3.5 g/dL. HCV Ab and HB<sub>s</sub>Ag were undetectable. Ceruloplasmin was within normal limits. Sonography reported a normal liver with splenomegaly. Endoscopy revealed large esophageal and gastric varices for which repeated sessions

of sclerotherapy were done. Regardless of repeated sclerotherapy the patient had continuous bleeding. The consulting surgeon planned for a portocaval shunt.

Laparotomy revealed a few hundred milliliters of ascites. The liver was edematous and numerous collateral vessels were seen. There was a large fistula with an aneurysm at the hilum of the spleen.

The lesion was removed with a splenectomy (Figs. 1,2).



**Fig. 1.** High-power photomicrograph of arteriovenous fistula (aneurysm) showing a communication between an artery and vein without an intervening capillary bed.

## Splenic Arteriovenous Fistula



Fig. 2. High-power photomicrograph shows that the A-V fistula has vessels that are thickened and hyalinized and a fibrotic stroma with focal calcification.

A wedge liver biopsy showed dilated and congested centrilobular veins (Fig. 3).

The patient has remained well for the last two years.

### DISCUSSION

Splenic arteriovenous fistula was reported in an autopsied patient as early as 1860, but the clinical features and the cause of death were not fully described.<sup>2</sup> In a review in 1988, 15 cases were collected in which a splenic arteriovenous fistula was implicated as the cause of variceal bleeding.<sup>3</sup> At least two additional cases have since been reported.<sup>4,5</sup> 13 cases had raised portal pressure documented by direct measurement.<sup>3-5</sup> The term forward portal hypertension is often used to refer to where increased portal blood flow accompanies portal hypertension.<sup>1</sup>

In patients with splenic arteriovenous fistula, arterialization of the portal circuit accounts at least in part for increased portal blood flow and markedly raised portal pressure as seen in the present case. Although the etiology may be uncertain in some cases, splenic arteriovenous fistulae may be either congenital or acquired.<sup>6</sup>

Congenital lesions are usually intrasplenic and may resemble multiple intrasplenic hemangiomas.<sup>7</sup> Acquired lesions are more common. Up to 70% of cases are associated with atherosclerosis of the splenic artery.<sup>7</sup>

Penetrating trauma, as well as surgical and diagnostic procedures such as splenectomy may sometimes result in splenic arteriovenous fistula.

Pregnancy with vaginal bleeding has also been implicated in some reports. Most reported cases have been in

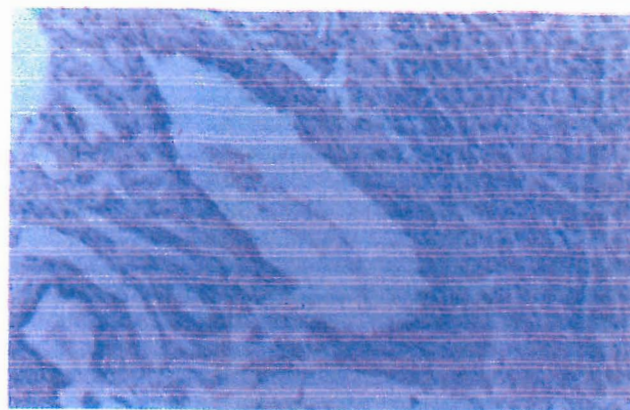


Fig. 3. Histologic picture shows liver tissue with dilated and congested centrilobular veins.

females while our case was a male patient. Therefore this treatable form of portal hypertension should be considered when the cause of portal hypertension is unclear.

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