C. Yılmaz MD[1]
A. Nart MD[2]
A. Uslu MD[2]
A. Aykas MD[2]

Splenic Arteriovenous Fistula: An Unusual Cause of Portal Hypertension Primarily Diagnosed by Color Duplex Sonography

Splenic arteriovenous fistulas (AVFs) are rare lesions. Herein, we report a 40-year-old man with portal hypertension secondary to splenic AVF, which was primarily diagnosed by color Duplex sonography. Sonography revealed a non-pulsatile, cyst-like mass at the splenic hilum with an unusual comma-shaped extension. In color duplex sonography, complete color fill-in of the lesion with the characteristic reversed red and blue color flow (Yin-Yang pattern) was observed. Duplex Doppler interrogation of the AVF revealed high velocity low-impedance arterial flow and moderately pulsatile venous flow within the lateral and medial part of the lesion, respectively. The comma-like extension demonstrated a low-resistance arterial waveform with very high peak systolic velocity. Based on these findings, a diagnosis of splenic AVF was made which was confirmed at surgery. Special emphasis was made on sonographic findings enabling the correct diagnosis.

Keywords: Splenic, Arteriovenous Fistula, Duplex Sonography, Portal Hypertension

Introduction

The diagnosis of splenic arteriovenous fistula (AVF) is challenging. The definite diagnosis is usually made by arteriography. Although noninvasive imaging findings such as computed tomography (CT), magnetic resonance imaging (MRI) and ultrasonography (US) may suggest the diagnosis, a high index of suspicion is required.[1-6] Approximately 100 cases have been described to date.[6] Demonstration of both high velocity arterial flow and turbulant venous flow within the splenic AVF prompted the diagnosis at color duplex sonography (CDS). To our knowledge, this finding has not been previously reported in splenic arteriovenous fistulas.

Case Presentation

A 40-year-old man with abdominal pain, diarrhea, esophageal varices and splenomegaly underwent sonographic examination to rule out cirrhosis. His past medical history was unremarkable. Laboratory tests including liver enzymes were normal. Sonography revealed a homogeneous and normal-appearing liver. The main portal vein was dilated, the largest diameter measurement was 18 mm. In color duplex sonography (CDS), the portal vein showed monophasic hepatopetal flow with normal mean flow velocity (24 cm/sec). No thrombosis was observed. The spleen was moderately enlarged with a craniocaudal length of 17 cm. In addition, a 5 cm anechoic, nonpulsatile mass, which presumed to represent a cystic lesion, was seen at splenic hilum. However, focused imaging

1. Department of Radiology, Izmir Bozyaka Training and Research Hospital, Izmir, Turkey.
2. Department of General Surgery, Izmir Bozyaka Training and Research Hospital, Izmir, Turkey.

Corresponding Author:
Cengiz Yılmaz
Address: Department of Radiology, Izmir Bozyaka Training and Research Hospital, Saim Cikrikci Street Izmir, Turkey.
Tel: +9053 2475 8094
Fax: +9023 2261 4444
Email: cengizyilmaz66@yahoo.com

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revealed a comma-like structure in contiguity with the cystic-appearing mass (Fig. 1A). This unusual appearance led us to proceed with CDS to rule out a vascular lesion. CDS revealed complete color fill-in of the lesion with reversed color flow patterns, the so-called characteristic "yin-yang" (red and blue flow) pattern (Fig. 1B). Duplex Doppler interrogation revealed high velocity low-impedance arterial flow (Fig. 1C) and moderately pulsatile venous flow (Fig. 1D) within the lateral and medial part of the lesion, respectively. The comma-like extension demonstrated a low-resistance arterial waveform with very high peak systolic flow exceeding 200cm/sec identifying it as a feeding artery (Fig. 1E). Additional findings included dilatation of the splenic vein (14 mm) and splenic artery (15 mm). Based on these findings, a diagnosis of splenic AVF was made. As we were confident with the sonographic diagnosis and the refer-

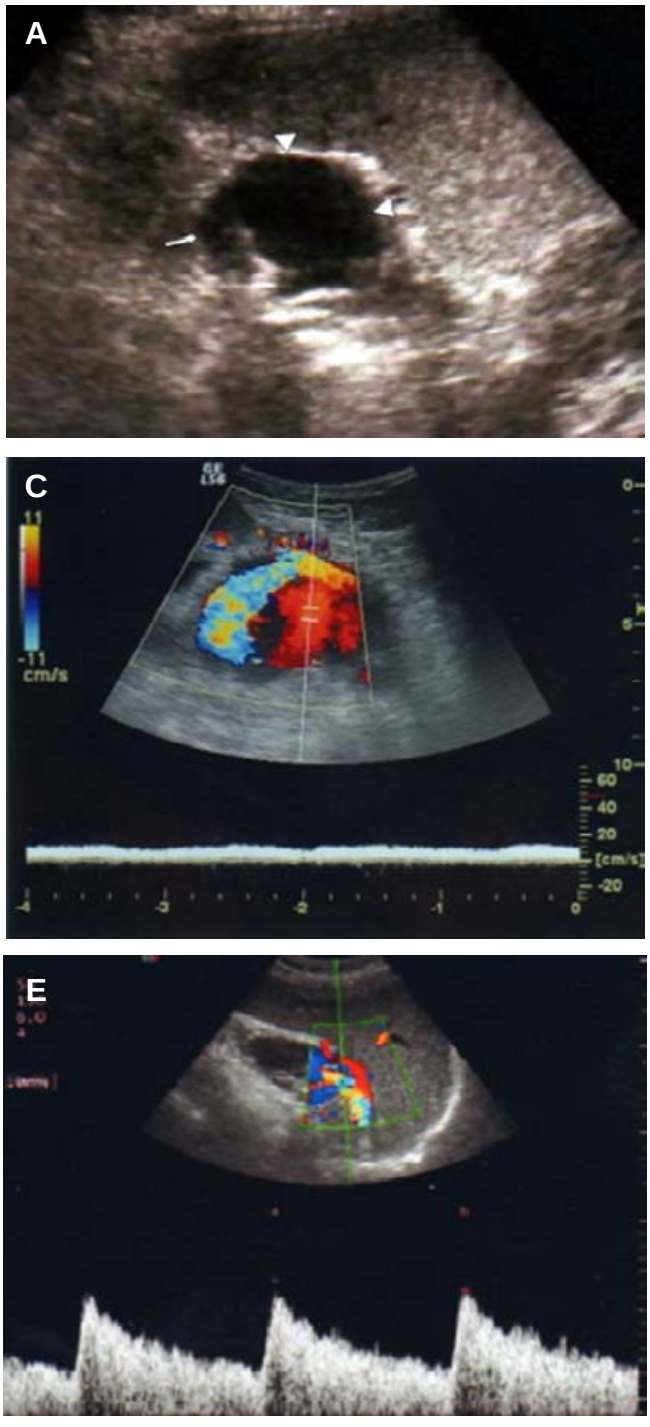

Fig. 1. A 40-year-old man with splenic AVF.

A. Gray-scale sonogram shows a 5.5 cm cyst-like structure (arrowheads) with a comma-shaped extension (arrow) at the splenic hilum. No apparent pulsations were observed at real-time imaging.

B. Color Doppler image demonstrates yin-yang (red and blue) flow pattern within the lesion.

C. Color Duplex image shows venous flow at the lateral part of the lesion.

D. Color Duplex image demonstrates arterial flow at the medial part of the lesion. Note the color-scale inversion.

E. Duplex interrogation of the comma-shaped structure demonstrates a high velocity low-resistance arterial waveform typical of a feeding artery.

ring surgeon opted to operate on the patient rather than embolisation, no further imaging with arteriography was performed. At surgery, a splenic AVF with an aneurysmatic nidus originating from the distal splenic artery and multiple, smaller draining splenic veins were found. After surgical ligation and excision of the fistula, splenectomy was performed. The postoperative period was uneventful and the patient was discharged with full recovery after 10 days of hospitalization. At 3 years clinical follow-up, the patient remains well. He did not experience any further episode of variceal bleeding.

Discussion

Splenic AVF is a rare congenital or acquired lesion, which may arise from rupture of a preexisting splenic artery aneurysm into the splenic vein due to high intraabdominal pressure during pregnancy or labor, or may develop secondary to trauma, surgery or mycotic infections.[1] This patient denied any major abdominal trauma, surgery or infection. We are not sure whether this case represents a congenital or acquired splenic AVF, since a remote trauma could not be totally excluded.

Patients with splenic AVF usually present with signs and symptoms of portal hypertension. Variceal bleeding is the most life threatening and is seen in nearly 50% of patients. Diarrhea may also be seen and is thought to be related to gastrointestinal vasculature congestion, as it was in this case.

As the signs and symptoms of splenic AVF are potentially curable, prompt diagnosis is crucial. Unfortunately, the diagnosis is challenging and delayed in most cases. The diagnosis is usually made by celiac or mesenteric angiography. Alternatively, contrast-enhanced CT or MR imaging with dynamic arterial and portal venous-phase imaging may enable the diagnosis but requires a high index of suspicion.[2-4] The diagnosis of splenic AVF may also be suggested by sonography.[2,5-7] The possibility of a nonivasive diagnosis by Doppler sonography was first suggested by Cantaro et al.[5] Piscaglio et al. also reported a case of splenic AVF in a 40-year-old multiparous woman.[6] In additon to dilatation of the portal vein, splenic artery and splenic vein, their findings at CDS included a feeding splenic artery with extremely high peak systolic flow and turbulent venous flow in the draining veins. They also noted that the gray-scale sonographic appearance of the fistula resembled a cat's head and stated that this appearance may aid in the diagnosis of splenic AVF. Splenic AVF may be mistaken for cystic lesions at gray-scale sonography. Indeed, in the presented case, the AVF looked like a cystic mass at first glance. However, at focused sonographic evaluation, a comma-shaped anechoic structure contigious with the cyst-like structure was noted. This appearance was suggestive of a vascular lesion rather than a cyst. This finding has also been reported in renal AVF.[8,9] An AVF can masquerade a noncomplicated pseudoaneurysm with no fistulous connections on CDS. Both lesions may show a characteristic pattern of reversed color flows, also referred as the yin-yang pattern. However, the demonstration of both arterial and venous flow within an aneurysmatic lesion is compatible with an AVF rather than a pure pseudoaneurysm, as it was in our case. A yin-yang color flow pattern with both arterial and venous flow components has previously been demonstrated in a patient with postnephrectomy renal AVF by Kocakoc et al. at CDS,[10] but to our knowledge, has not been reported in splenic AVF previously.

Both transcatheter embolization and surgical excision have been reported to be curative in splenic AVFs. In this case, the referring surgeon opted to operate on the patient. The patient recovered fully and did not experience any further episode of variceal bleeding during the follow-up period.

In conclusion, this case shows that both gray-scale and CDS may provide important diagnostic information in the diagnosis of splenic AVF. Familiarity with the sonographic appearance of this rare lesion may enable the correct diagnosis without further imaging work-up.

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