

Portal hypertension secondary to splenic arteriovenous fistula

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ABSTRACT

يعد فرط ضغط الدم البابي من الحالات الشائعة المرتبطة بأمراض الكبد المزمنة، ولكن نادراً ما يحدث ذلك من قبل تحويله من خلال ناسور شرياني وريدي طحالي. نستعرض في هذا المقال حالة من حالات ارتفاع ضغط الدم البابي بسبب الناسور الطحالي بين الشريان والوريد وغير متعلق بالتليف الكبدي. وتكون عادة في النساء متعددي الولادة. وهنالك أسباب أخرى مثل الحوادث أو العدوى الفطرية تحدث بشكل متساوي بين الجنسين حسب ما نشر في الإحصائيات. وغالباً ما يأتي المرضى بأعراض دوالي المريء والمعدة أو تضخم الطحال أو آلام في أعلى البطن. يعد التاريخ الطبي والفحص السريري الخطوة الأولى لتشخيص المرض، ثم تليها الموجات الصوتية والتصوير الطبقي المحوري. كما ويعد التدخل الجراحي الإشعاعي المعيار الدقيق في تشخيص هذه الحالة. يتم علاج مثل هذه الحالات بالربط الجراحي أو الانصمام الشرياني عبر القسطرة. ويفضل العلاج بالانصمام الشرياني عبر القسطرة لقلّة المضاعفات، وإمكانية استخدامه للحالات غير المستقرة. ويتساوى نجاح علاج مثل هذه الحالات بين الربط الجراحي والانصمام الشرياني عبر القسطرة.

Portal hypertension is a common condition in association with chronic liver disease; however, it is rarely caused by vascular shunting from splenic arteriovenous fistula (SAVF). We are presenting a case of non-cirrhotic portal hypertension due to SAVF. This condition is usually seen in multiparity women; however, other causes like trauma or mycotic infection are reported in the literatures with equal gender prevalence. Most of the patients present with gastric and esophageal varices, splenomegaly or upper abdominal pain. Clinical history and examination are the first step for diagnosis followed by ultrasound or contrast enhanced computer tomography. Splenic arteriovenous fistula must be then confirmed by selective celiac or splenic arteriography which is the gold standard tool. The condition is treatable either surgical ligation or intra-arterial embolization. Due to less invasive procedure, the later one is more

favorable in unstable patients. Both procedures have been reported to be equally successful in managing SAVF.

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Splenic arteriovenous fistulas (SAVF) represent a rare pathological entity that should be suspected especially in cases of acute portal hypertension not related to chronic liver disease.¹ They may be either asymptomatic or lead rapidly to the development of portal hypertension, with a clinical picture of gastrointestinal bleeding, diarrhea, ascites or unfavorable heart failure due to hyperdynamic blood flow state.¹⁻³ Rupture of a splenic artery aneurysm (SAA) into the splenic vein and formation of a fistulous tract between them consist the underlying pathology. The positive impact of transcatheter arterial embolization as an effective alternative to surgical intervention in the management of such vascular malformations is found in many literatures,^{1,3} and we are also emphasize it. This article is to emphases on the early diagnosis of the SAVF and the excellent out come of the endo-vascular embolization.

Case Report. A 54-year-old lady patient presented to our hospital with epigastric pain and abdominal distension. Her first complaint started 2 years prior to her hospitalization. It started with epigastric pain, which was localized, dull in nature, without other accompanying symptoms. On first impression, was assumed to be a peptic ulcer; thus, she was given an appointment for

workup. She lost to follow up, 2 years later she was presented to our hospital with the same epigastric pain and a new symptom of abdominal distention that lasted for 3 months. Physical examination revealed tense ascites without stigmata of chronic liver disease with grade II abdominal bruit noted in the left upper quadrant. The results of the laboratory studies were as follows: hemoglobin: 12.3 g/dl (11.5-16.5 g/dl); white blood cells: $4.0 \times 10^9/L$ ($4.0-11.0 \times 10^9/L$); platelet count: $343 \times 10^9/L$ ($151-355 \times 10^9/L$); alanine aminotransferase: 21 IU/dl (12-31 IU/dl); aspartate aminotransferase, 22 IU/dl (10-45 IU/dl); alkaline phosphatase, 82

IU/dl (45-115 IU/dl); and total bilirubin, 5 umole/L (2-22 umole/L). Iron studies, α -1 antitrypsin levels, antinuclear antibody, antimitochondrial antibody, and viral hepatitis studies were within normal limits. Endoscopy was carried out and shows grade I esophageal varicose. The treating physician started the work-up of the possibility of portal hypertension. A contrast-enhanced CT scan of the abdomen revealed enlargement of the splenic vein with several varices in the splenic hilum, calcification in the wall of splenic artery aneurysm, moderate ascites, and splenomegaly. The portal vein was severely enlarged; however, there was no liver nodularity, fibrosis, or atrophy to indicate cirrhosis. The portal vein and its main branches are markedly dilated and tortuous. Again, the splenic artery aneurysm and AVF were seen at the hilum (Figure 1).

An ultrasound revealed that splenic artery was dilated throughout its course with mixed colored signals and measured about 2 cm in diameter. There was a saccular aneurysm seen at its distal aspect measuring 5 x 3.3 cm in dimension with mosaic appearance by color Doppler (Figure 2a). There was a short fistula seen. Vigorous pulsating like venous flow waves was seen within the splenic vein (Figure 2b). Selective celiac angiography and super-selective splenic arteriography with frontal projections were performed. There is a fistula commutating the tortuous, dilated splenic artery and aneurysm-like splenic vein (Figure 3a). There was early opacification of the splenic vein with retrograde opacification of superior mesenteric and portal veins (Figure 3b). The fistula was due to atheromatous splenic artery aneurysm, which is rupturing into splenic vein. Diagnosis of splenic arteriovenous fistula and splenic venous aneurysm was made. It was deployed by 14 mm x 1 cm vascular plug amplatzer proximal to

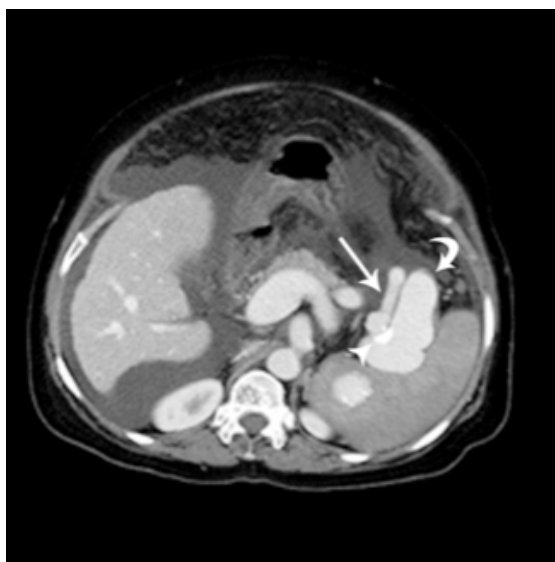


Figure 1 - A CT scan shows aneurysmal dilatation of the splenic artery with arteriovenous fistula communicating the artery with the vein. Moderate ascites and splenomegaly are seen. Straight arrow - splenic artery, arrow head - fistula, curved arrow - splenic vein

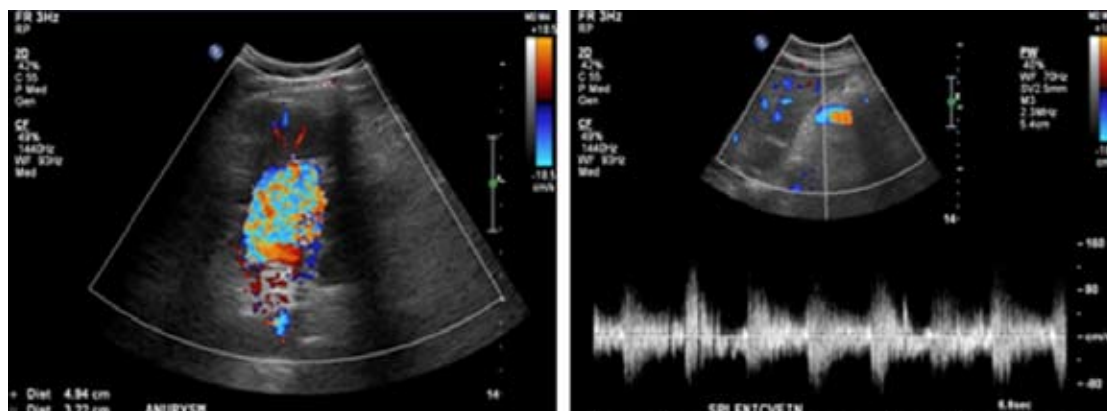


Figure 2 - Color Doppler ultrasound of the splenic arteriovenous fistulas A) A saccular aneurysm seen at its splenic artery with mosaic appearance by color Doppler. B) Splenic vein shows arterial wave by duplex analysis.

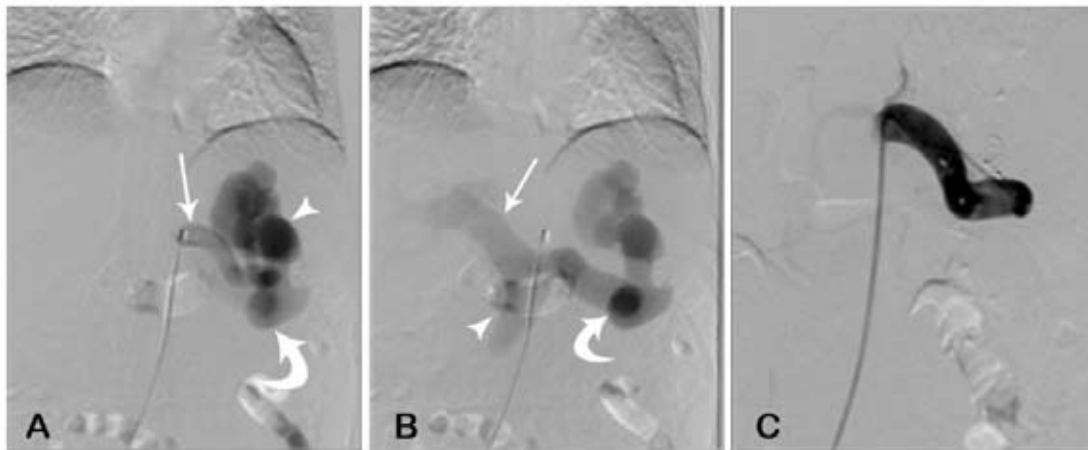


Figure 3 - Selective angiogram of the splenic arteriovenous fistulas pre- and post- embolization. **A)** Selected splenic angiogram shows splenic artery with early fill of splenic vein (straight arrow - splenic artery, arrow head - fistula, curved arrow - splenic vein). **B)** Early opacification of the splenic vein with retrograde opacification of superior mesenteric and portal veins (straight arrow - portal vein, arrow head - superior mesenteric vein, curved arrow - splenic vein). **C)** Post-embolization shows cut off of distal splenic artery.



Figure 4 - Ultrasound post-treatment shows complete thrombosis of the aneurysm.

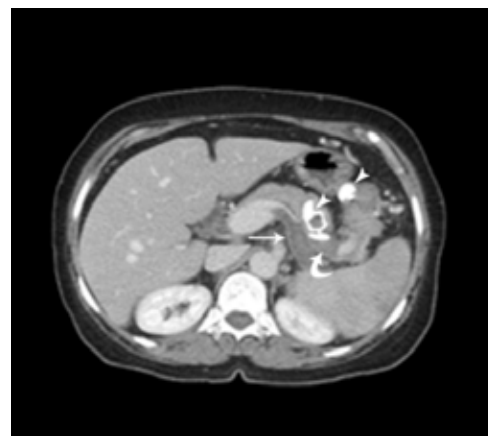


Figure 5 - A CT scan of the abdomen demonstrate 2 amplatz plugs with thrombosis of splenic artery and vein. (straight arrow - thrombosed splenic vein, arrow heads - amplatz plugs, curved arrow - thrombosed splenic artery)

the aneurysm of AV fistula site. Follow up angiogram showed slow flow (Figure 3c).

Then, another 16mm x 1cm vascular plug amplatzer was deployed proximally. Follow up angiogram showed very slow flow through the distal arterial feeding branch and AV fistula aneurysmal sac. After 2 days ultrasound was carried out and showed that splenic artery aneurysm had diminished in size and is completely thrombosed (Figure 4). The splenic vein has lost its arterialized flow. Patient was finally discharged without any symptoms. A CT scan was carried out after a month showed thrombosed splenic artery as well as the splenic vein and the aneurysmal dilatation at the site of the fistula (Figure 5). Splenic infarcts were noted in the interval;

however, most of the splenic parenchyma opacified with contrast. The ascites has resolved completely.

Discussion. Splenic arteriovenous fistulas may be due to congenital, traumatic, or infectious causes. Increased intra-abdominal pressure due to pregnancy, labor or mycotic infection will increase the risk of splenic aneurysm rupture and fistula formation. Most of SAVF occur in multiparous women (80%).¹ Fewer than 100 cases have been reported in the world medical literature.² The origin of splenic artery aneurysm is either due to medial wall degeneration caused by fibrodysplasia, portal hypertension with splenomegaly

and repeated pregnancy or due to atherosclerosis as a secondary cause. Although splenic aneurysms constitute 60% of all visceral aneurysms, SAVFs are rare and are usually associated with rupture of a splenic artery aneurysm into the corresponding vein creating a fistulous tract.^{2,3} Some splenic aneurysms are due to atherosclerotic weakening of the vessel wall, calcific changes in the aneurysm suggest atherosclerosis as a secondary, rather than a primary process.³

Symptoms and signs of SAVF are related to portal hypertension such as splenomegaly (55%), gastric and esophageal varices (52%), upper abdominal pain, ascites (35%), and secretory diarrhea.³ Other symptom is machinery-type vascular bruit or systolic murmur, which may be heard over the epigastrium, left upper abdomen, left lower chest or left flank and developed in about one third of the patients.^{2,3} Diagnosis is made by Doppler US which may demonstrate dilated splenic artery, pulsatile splenic vein, high flow through the splenic artery and turbulent flow as in our case,⁴ other modality for diagnosis include CT angiography. Splenic arteriovenous fistula must be then confirmed by selective celiac or splenic arteriography which is the gold standard tool and it will show an elongated, tortuous splenic artery with early filling of the splenic vein during the arterial phase, dense opacification of the splenoportal venous system and an aneurysm-like splenic vein, those are the characteristic findings of SAVF.³ Treating the SAVF is recommended even to asymptomatic patient to prevent the complication. The treatment of choice is surgical ligation and excision of SAVF or intra-arterial embolization. The later one is

more favorable in unstable patient. Both procedures have been reported to be equally successful in managing SAVF.^{5,6} In our presenting case, we treated the patient with intra-arterial embolization using amplatz plug, which showed excellent results in the follow up.

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