Case report

Splenic artery aneurysm with extrahepatic portal venous obstruction: A case report

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ABSTRACT

Introduction and importance: Splenic artery aneurysm is one of the most common visceral aneurysms. Patients are usually asymptomatic. Splenic artery aneurysm if untreated has the potential for rupture and is therefore life-threatening. Its association with extrahepatic portal vein obstruction is rare.

Case presentation: A 25-year female was incidentally diagnosed with a splenic artery aneurysm with extrahepatic portal vein obstruction with splenomegaly 8 years back during the 5th month of her second pregnancy. No intervention was done back then. Recently, she presented to the surgical gastroenterology outpatient department with an increasing abdominal mass. On examination, the patient was pale and splenomegaly was present. Hematological reports were suggestive of hypersplenism. The patient underwent splenectomy and aneurysmal resection with a proximal splenorenal shunt as the best course of treatment.

Discussion: Due to the rarity of the disease, the management is still challenging and needs further study. Diagnosis can be made clinically with support from imaging modalities. Surgical treatment has a good outcome in such patients. Even with the availability of less invasive procedures such as endovascular treatment, open surgery is preferred.

Conclusion: Proximal splenorenal shunt is a well-accepted surgical procedure for extrahepatic portal vein obstruction. Splenectomy and aneurysmal resection can relieve hypersplenism and treat splenic artery aneurysm in patients with isolated splenic artery aneurysm at the splenic hilum.

1. Introduction

True splenic artery aneurysms (SAA) are rare and have an estimated prevalence of 0.02%–0.1% [1]. Splenic artery aneurysm is more common in females, especially during pregnancy with a female to male ratio of 4:1. Up to 80% of the SAA are detected on routine evaluation for other problems in asymptomatic patients [1]. Giant aneurysms (>2.5 cm) are even rarer with only around 20 cases reported to date [2].

Extrahepatic portal vein obstruction (EHPVO) refers to the obstruction of the portal vein and/or its right and/or left branches and is an important cause of noncirrhotic portal hypertension, especially in third world countries [3]. SAA causing splenic vein thrombosis and subsequent portal hypertension/EHPVO is extremely rare with very few cases reported in the literature [4–7].

Here, we report one such rare presentation of splenic artery aneurysms with EHPVO. This case has been reported in line with SCARE criteria [8].

2. Case report

A 25-year G2P1L1 presenting to the antenatal clinic at her 23-week period of gestation was diagnosed incidentally with EHPVO with splenomegaly 8 years back while performing an anomaly scan. The patient was counseled for surgery but denied it due to her pregnancy. All other events during her pregnancy and post-partum period were uneventful. The patient was lost to follow-up after the delivery. However, she presented 8 years later with the complaint of increasing abdominal size and heaviness in the left upper quadrant. There was no history of hematemesis, melena, fever, bleeding from any other site, jaundice, pruritis, or ascites. In addition, her medical and surgical history were insignificant.

On examination, the patient was pale. The abdomen was soft and...
non-tender. The patient had splenomegaly (Hackett grade 3). Stigmata of chronic liver diseases were absent. The hematology report showed Hb of 10.2 g/dL, WBC count of 10,300/mm$^3$, and platelet count of 25,000/mm$^3$ with normal liver function tests (Table 1). On peripheral blood smear, dimorphic anemia, leukopenia, and thrombocytopenia were seen.

Ultrasoundography of abdomen and pelvis revealed gross splenomegaly, dilated portal vein with multiple periporal and perisplenic collateral features suggestive of portal hypertension, dilated splenic artery at splenic hilar region features suggestive of splenic artery aneurysm. The endoscopic evaluation showed fundal varices with gastrointestinal changes with erosions and antral gastritis with bile reflux. CT scan of abdomen and pelvis demonstrated portal cavernoma formation, massive splenomegaly with dilated tortuous perisplenic collateral veins, and esophageal varices along with saccular outpouching of splenic artery measuring 32 mm × 31 mm (Fig. 1a and b).

With the diagnosis of EHPVO with a splenic artery aneurysm with hypersplenism, she was planned for elective aneurysmal resection for splenic artery aneurysm, splenectomy to correct hypersplenism, and proximal splenoportal shunt for portal hypertension along with a liver biopsy by the team of surgical gastroenterologists. She was vaccinated with pneumococcal, meningococcal, and Haemophilus influenzae (Hib) vaccines two weeks prior to surgery. Intraoperatively, there was a grossly enlarged spleen, multiple tortuous collaterals at the hilum of the spleen, and a 4 cm × 4 cm calcified aneurysm in the splenic hilum. (Figs. 2 and 3). The liver biopsy was normal with no cirrhotic changes. Postoperatively, the patient was managed with analgesics, fluid, and antibiotics. She was started on aspirin (Table 75 mg once daily) on the 2nd post-operative day which was continued for three months.

Recent advances have increased the chance of a diagnosis of SAA before rupture. However, in diagnostic imaging, it is still a very unusual entity because of its low prevalence. Moreover, physicians would too easily be convinced that the bleeding in patients is due to overt portal hypertension and possible rupture of varices. Therefore, physicians usually have insufficient awareness of this rare disorder [9].

Treatment of SAA can be challenging and there is limited guidance regarding when and how to treat patients with SAA. This is most probably due to a lack of randomized trials as the disease itself is extremely rare. However, from the limited retrospective clinical series, it is evident that SAA needs to be treated if it is diagnosed in a pregnant woman or measures 2 cm or more, as these factors increase the chance of spontaneous rupture [11]. Also, in patients with portal hypertension, pregnancy, and carry the highest morbidity among all visceral artery aneurysms with maternal and fetal mortality as high as 75 % and 95 % respectively [4,14,15].

Women are affected four times more commonly by SAA, especially during pregnancy and childhood years due to hormonal changes [7]. It is common in multiparous females since the splenic artery responds to the changes of pregnancy, like high levels of estrogens and progesterone along with increased cardiac output resulting in the altered arterial wall and defective elastin synthesis, making the splenic artery more likely to dilate. Other risk factors include portal hypertension, vascular and connective tissue disorders, congenital abnormalities of the vessels, vascular trauma, and degenerative arterial disease [11]. It is worth noting that our patient was in her second pregnancy when she was diagnosed with a splenic artery aneurysm.

The pathogenesis of EHPVO is due to compression on the splenic vein by the aneurysm leading to stasis of blood and thereby predisposing to thrombosis. In our case, thrombosis was extending into the portal vein probably from the splenic vein leading to features of EHPVO. In such cases, the presentation will be similar to that of the features of EHPVO which may include gastro-esophageal varices, splenomegaly, and features of hypersplenism as in the present case [7]. Also, in portal hypertension, there exists a “splenic hyperkinetic state” promoting the development of SAAs. Among portal hypertensive patients, those with SAA have significantly increased splenic venous flow, splenic artery diameter, and portosystemic shunts which reduce the resistance thus increasing the flow through the splenic vein. Consequently, an increase in splenic artery blood flow is required to increase portal pressure and maintain liver perfusion resulting in the formation of a splenic artery aneurysm [12]. This means either splenic arterial aneurysm can cause EHPVO or portal hypertension due to EHPVO can cause a splenic arterial aneurysm. The incidental finding of SAA during pregnancy without any prior predisposing condition for the development of EHPVO can explain EHPVO secondary to SAA in our case.

Symptomatic presentation ranges from chronic abdominal pain, acute rupture with hypotension, hemorrhagic shock, and a sudden collapse which occurs in 3–10 %. However, 80 % of the patients can be asymptomatic and are detected incidentally [1,13]. In our case, the patient was asymptomatic without any features suspicious of SAA and EHPVO except for splenomegaly. SAAs are at risk for spontaneous rupture or erosion into adjacent abdominal viscera or vessels, especially in pregnancy, and carry the highest morbidity among all visceral artery aneurysms with maternal and fetal mortality as high as 75 % and 95 % respectively [9,10].

In the literature, the majority of patients with portal hypertension complicated with SAA have cirrhosis as the underlying etiology. However, limited information is available on the management of
extrahepatic portal vein obstruction with SAA [15]. The management in these patients is different from that in cirrhotics as they are good risk surgical candidates. In patients with EHPVO and SAA, the treatment option depends upon the presence of bleeding esophagogastric varices or portal biliopathy. Endovascular therapy like coil embolization, detachable balloon occlusion, and stent graft can be a good alternative in a subgroup of patients with EHPVO without bleeding or jaundice. Surgery can simultaneously treat both problems in a single setting. Excision of the aneurysm is the preferred option while ligation can be done in the case of multiple aneurysms [16,19]. In addition, the surgical treatment for patients with EHPVO (with portal hypertension and upper gastrointestinal bleed) can broadly be divided into portosystemic shunts, esophagogastric devascularization, and the more recently described Rex shunt [20]. Though endoscopic therapy (variceal ligation/sclerotherapy) has excellent results for acute control of bleeding as well as prevention of recurrent bleeding, portosystemic shunt surgery remains a single-time effective therapy in these patients [21–23]. The options for portosystemic shunts include proximal lienorenal shunt (LRS), side-to-side LRS, distal LRS, and mesocaval shunt. The long-term complications of EHPVO are shunt occlusion (5–10 %), and rebleeding.

Fig. 1. a. Computed Tomography axial and coronal view of abdomen showing massive splenomegaly (arrow). 
b. Computed Tomography, axial and coronal view showing dilated perisplenic collaterals, splenic artery aneurysm at hilar region (arrow) and features consistent with chronic portal vein thrombosis and sequelae.

Fig. 2. Pre splenectomy picture showing enlarged spleen wrapped by the omentum.

Fig. 3. post splenectomy picture showing enlarged spleen, multiple collaterals, and splenic artery aneurysm at the hilum of the spleen (arrow).
Splenic artery aneurysm associated with extrahepatic portal vein obstruction is uncommon. However, splenic artery aneurysms are clinically significant due to their potential for spontaneous and unpredictable rupture and are therefore life-threatening. SAA and EHPVO can be surgically treated simultaneously with good outcomes.

Consent

Written informed consent was obtained from the patients for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Nothing to declare.

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Author contribution

Paleswan Joshi Lakhey, Romi Dahal and Deepak Sharma = Study concept, Data collection, and surgical therapy for the patient
Niharika Pathak, Elisha Poddar, Suraj Shrestha, Khusbu Raman = Writing - original draft preparation
Ritika Ranjan and Elisha Poddar = Editing and writing
Paleswan Joshi Lakhey = senior author and manuscript reviewer
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