



## Case report

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

Niharika Pathak<sup>a,\*</sup>, Elisha Poddar<sup>a</sup>, Suraj Shrestha<sup>a</sup>, Khusbu Kumari<sup>a</sup>, Ritika Ranjan<sup>a</sup>, Deepak Sharma<sup>b</sup>, Romi Dahal<sup>b</sup>, Paleswan Joshi Lakhey<sup>b</sup>

<sup>a</sup> Maharajgunj Medical Campus, Institute of Medicine, Kathmandu, Nepal

<sup>b</sup> Department of Surgical Gastroenterology, Tribhuvan University Teaching Hospital, Kathmandu, Nepal

## ARTICLE INFO

## Keywords:

EHPVO  
hypersplenism  
splenic artery aneurysm  
case report

## 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 G<sub>2</sub>P<sub>1</sub>L<sub>1</sub> presenting to the antenatal clinic at her 23-week period of gestation was diagnosed incidentally with EHPVO with splenomegaly with splenic artery aneurysm 8 yrs. 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 postpartum 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

\* Corresponding author.

E-mail address: [niha.2052@gmail.com](mailto:niha.2052@gmail.com) (N. Pathak).

<https://doi.org/10.1016/j.ijscr.2022.107568>

Received 27 July 2022; Received in revised form 22 August 2022; Accepted 26 August 2022

Available online 31 August 2022

2210-2612/© 2022 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

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<sup>3</sup>, and platelet count of 25,000/mm<sup>3</sup> with normal liver function tests (Table 1). On peripheral blood smear, dimorphic anemia, leukopenia, and thrombocytopenia were seen.

Ultrasonography of abdomen and pelvis revealed gross splenomegaly, dilated portal vein with multiple periportal and perisplenic collaterals 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 gastropathic 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. 1 a 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 splenorenal 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. In addition, she has been counseled to take the pneumococcal and meningococcal vaccines every 5 years and the Hib vaccine yearly. After six months of surgery, the patient had normal blood counts and is in good condition (Table 1). She is now under regular follow-up.

**3. Discussion**

Splenic artery aneurysm is the most common visceral artery aneurysm compromising 50–75 % of all cases. After abdominal aorta and iliac artery aneurysms, they are the third most frequent intra-abdominal aneurysm, Splenic artery aneurysm is usually single and ≤2cm in size. A giant aneurysm with a diameter >2.5cm like in our patient is very rare

**Table 1**  
Laboratory parameters pre-operatively and at follow-up.

Lab parameters	Pre-operative	On follow up	Reference range
<b>Complete blood count</b>			
Hb	10.2 g/dL	13.2 g/dL	13–17 g/dL
WBC	10,300/μL	7200/μL	4000–11,000/μL
Platelets	25,000/μL	210,000/μL	150,000–450,000/ μL
RBC	3.8 mil/μL	4.7mil/μL	4.5–5.5 mil/μL
<b>Liver function test</b>			
Serum bilirubin (total)	0.8 mg/dL	0.9 mg/dL	0.2–1.2 mg/dL
Serum bilirubin (direct)	0.2 mg/dL	0.2 mg/dL	0.01–0.3 mg/dL
SGPT	24 IU/L	30 IU/L	<40 IU/L
SGOT	33 IU/L	35 IU/L	<40 IU/L
Serum alkaline phosphatase	248 IU/L	251 IU/L	80–306 IU/L
Serum total protein	6.7 g/dL	5.8 g/dL	5.5–8.0 g/dL
Serum albumin	3.8 g/dL	3.9 g/dL	3.5–5.5 g/dL
<b>Renal function test</b>			
Blood urea	21 mg/dL	32 mg/dL	15–45 mg/dL
Serum creatinine	0.6 mg/dL	0.7 mg/dL	0.6–1.4 mg/dL
Serum Na+	145 mmol/L	140 mmol/L	135–155 mmol/L
Serum K+	4.4 mmol/L	4.1 mmol/L	3.5–5.5 mmol/L

[9,10]. Women are affected four times more commonly by SAA, especially during pregnancy and childbearing 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.

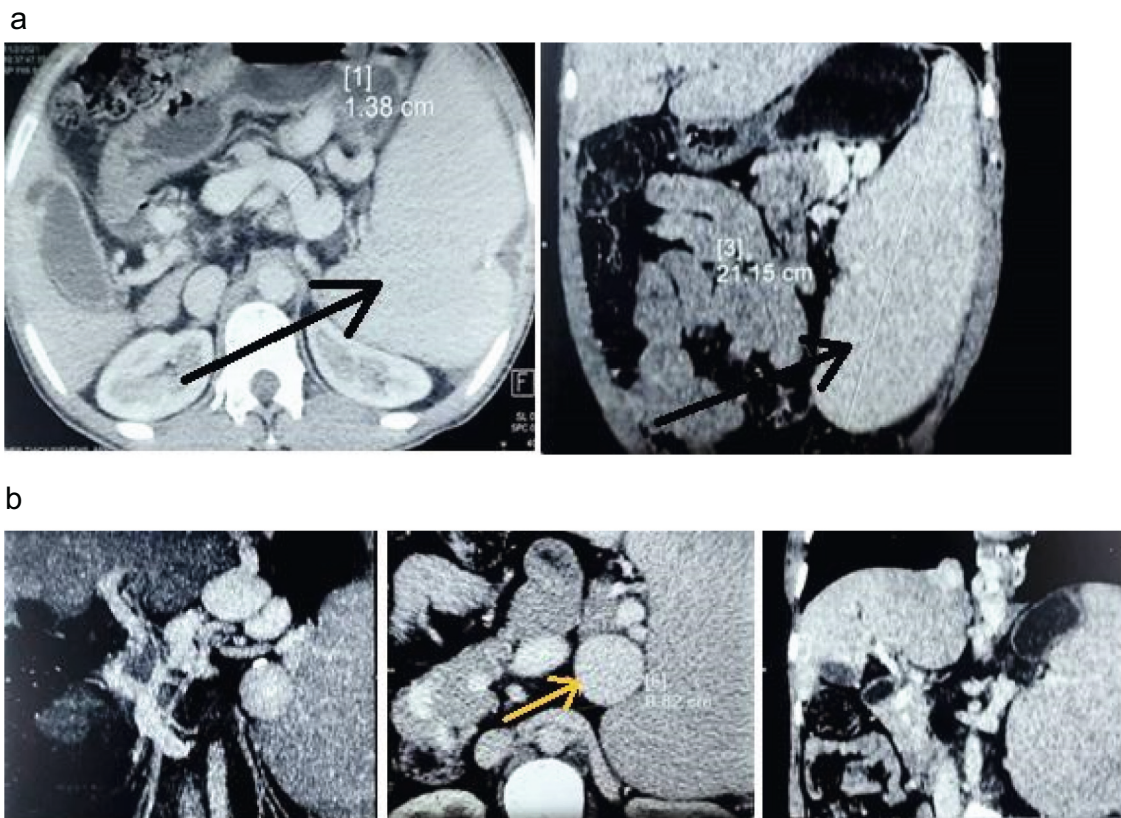
The pathogenesis of EHPVO is due to compression on the splenic vein by the aneurysm leading to stasis of blood and thereby predisposing to thrombosis. In our case thrombosis was extending into the portal vein probably from the splenic vein leading to features of EHPVO. In such cases, the presentation 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 SAA. 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 [4,14,15].

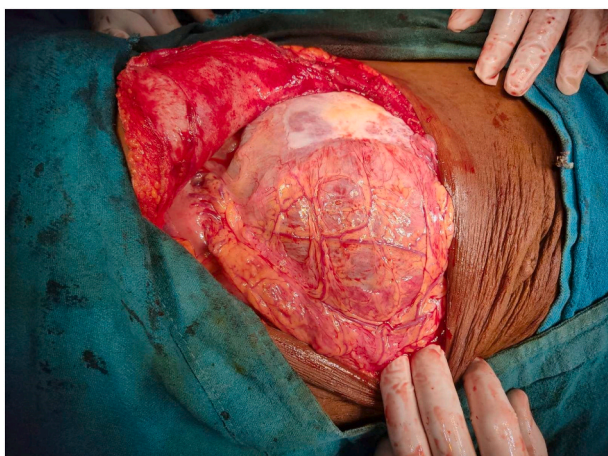
Recent advances have increased the chance of a diagnosis of SAA before rupture. However, in diagnostic imaging, it is still a very unusual entity because of its low prevalence. Moreover, physicians would too easily be convinced that the bleeding in patients is due to overt portal hypertension and possible 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, therapy is indicated irrespective of size due to the high incidence of unpredictable rupture [16]. Treatment of SAA includes aneurysmal resection with or without splenectomy and non-operative endovascular techniques. Moreover, those with non-cirrhotic portal hypertension, hypersplenism, and/or acute rupture have more benefits from splenectomy as well as aneurysmal resection [2,17,18,22]. Keeping in view the rarity of the disease and its life-threatening potential, the clinical-epidemiological picture, and the management of splenic artery aneurysm associated with EHPVO requires further study to guide more thoughtful and considerate patient care.

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



**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).

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

[20]. Among patients with normal liver function and without cirrhosis, post-shunt encephalopathy is mild to none [20,23]. In our case, the patient underwent a proximal splenorenal shunt and excision of the aneurysm with splenectomy, which was therapeutic for both the SAA and the underlying EHPVO. There were no postoperative complications due to the shunt or splenectomy in our case.

For splenic artery aneurysm presenting with atypical symptoms of EHPVO as our patient, splenectomy with excision of the aneurysm excludes the risk of aneurysmal rupture in the future and is also curative for the symptoms of EHPVO [7,10].

#### 4. Conclusion

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.

#### Source of funding

None.

#### 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  
All the authors read and approved the final manuscript.

#### Guarantor

Suraj Shrestha.

#### Registration of research studies

Not applicable.

#### Provenance and peer review

Not commissioned, externally peer-reviewed.

#### Declaration of competing interest

None.

#### Acknowledgment

None.

#### References

- [1] L.M. Messina, C.J. Shanley, Visceral artery aneurysms, *Surg. Clin. North Am.* 77 (2) (1997) 425–442.
- [2] S. Yadav, P. Sharma, P.K. Singh, et al., Giant splenic artery aneurysm: a rare but potentially catastrophic surgical challenge, *Int. J. Surg. Case Rep.* 3 (11) (2012) 533–536.
- [3] A. Abergel, M. Gasperment, F.G. Nery, et al., Extrahepatic portal vein obstruction (EHPVO) in cirrhosis, *Clin. Res. Hepatol. Gastroenterol.* 44 (4) (2020) 497–502.
- [4] J. Debnath, R.A. George, P.P. Rao, K. Ghosh, Splenic artery aneurysm—a rare cause for extrahepatic portal venous obstruction: a case report, *Int. J. Surg.* 5 (5) (2007) 351–352.
- [5] J. Arias Pérez, M.A. Aller Reyero, L. Lorente Ruigómez, L. Ortega Medina, Sacristán H. Durán, Segmental portal hypertension caused by aneurysm of the distal splenic artery, *Rev. Esp. Enferm. Apar.* 75 (5) (1989) 485–488.
- [6] Y. Hoebeke, M. Scotté, I. Leblanc, F. Michot, P. Tenière, Aneurysm of the splenic artery: an uncommon etiology of segmental portal hypertension, *Ann. Chir.* 47 (6) (1993) 557–558.
- [7] T.P. Elamurugan, S.S. Kumar, R. Muthukumarassamy, V. Kate, Splenic artery aneurysm presenting as extrahepatic portal vein obstruction: a case report, *Case Rep. Gastrointest. Med.* 2011 (2011), 908529.
- [8] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.
- [9] Y. de Miao, B. Ye, Intra-gastric rupture of splenic artery aneurysms: three case reports and literature review, *Pak. J. Med. Sci. Q.* 29 (2) (2013) 656–659.
- [10] I.H. Huang, D.A. Zuckerman, J.B. Matthews, Occlusion of a giant splenic artery pseudoaneurysm with percutaneous thrombin-collagen injection, *J. Vasc. Surg.* 40 (3) (2004) 574–577.
- [11] A. Khan, M. Ayub, I. Haider, M. Humayun, Z. Shah, F. Ajmal, Coexisting giant splenic artery and portal vein aneurysms leading to non-cirrhotic portal hypertension: a case report, *J. Med. Case Rep.* 10 (1) (2016) 270.
- [12] M. Ohta, M. Hashizume, K. Ueno, K. Tanoue, K. Sugimachi, K. Hasuo, Hemodynamic study of splenic artery aneurysm in portal hypertension, *Hepato-Gastroenterology* 41 (2) (1994) 181–184.
- [13] F.G. Grego, S. Lepidi, R. Ragazzi, V. Iurilli, R. Stramanà, G.P. Deriu, Visceral artery aneurysms: a single center experience, *Cardiovasc. Surg.* 11 (1) (2003) 19–25.
- [14] U. Sadat, O. Dar, S. Walsh, K. Varty, Splenic artery aneurysms in pregnancy—a systematic review, *Int. J. Surg.* 6 (3) (2008) 261–265.
- [15] B. Gerald, M.D. Zelenock, C. James, Splanchnic artery aneurysms, in: B. Robert, M. D. Rutherford (Eds.), *Vascular Surgery*, WB Saunders, Co, Philadelphia, 2000, pp. 1370–1373.
- [16] P.K. Mishra, S.S. Saluja, A.K. Sharma, P. Pattnaik, Management of splenic artery aneurysm associated with extrahepatic portal vein obstruction, *Hepatobiliary Pancreat. Dis. Int.* 11 (3) (2012) 330–333.
- [17] A. Robaldo, F. Gramondo, F. Beccaria, P. Colotto, Giant splenic artery aneurysm rupture, *Clin. Case Rep.* 4 (6) (2016) 620–622.
- [18] R. Pulli, W. Dorigo, N. Troisi, G. Pratesi, A.A. Innocenti, C. Pratesi, Surgical treatment of visceral artery aneurysms: a 25-year experience, *J. Vasc. Surg.* 48 (2) (2008) 334–342.
- [19] K. Karaman, L. Onat, M. Sirvanci, R. Olga, Endovascular stent graft treatment in a patient with splenic artery aneurysm, *Diagn. Interv. Radiol.* 11 (2) (2005) 119–121.
- [20] V. Mangla, S. Pal, P. Sahni, Surgery for non-cirrhotic portal hypertension: current status, *Trop. Gastroenterol.* 37 (3) (2017) 152–155.
- [21] V. Thomas, T. Jose, S. Kumar, Natural history of bleeding after esophageal variceal eradication in patients with extrahepatic portal venous obstruction; a 20-year follow-up, *Indian J. Gastroenterol.* 28 (6) (2009) 206–211.
- [22] J.G. Maksoud-Filho, M.E.P. Gonçalves, S.R. Cardoso, N.E.M. Gibelli, U. Tannuri, Long-term follow-up of children with extrahepatic portal vein obstruction: impact of an endoscopic sclerotherapy program on bleeding episodes, hepatic function, hypersplenism, and mortality, *J. Pediatr. Surg.* 44 (10) (2009) 1877–1883.
- [23] A.S. Prasad, S. Gupta, V. Kohli, G.K. Pande, P. Sahni, S. Nundy, Proximal splenorenal shunts for extrahepatic portal venous obstruction in children, *Ann. Surg.* 219 (2) (1994) 193–196.