DOI: 10.7860/JCDR/2014/8498.4821

Case Report

Internal Medicine Section

Multiple Splenic Artery Aneurysms: A Rare Cause of Extrahepatic Portal Hypertension and Massive Splenomegaly

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ABSTRACT

A 39-year-old nulliparous female was admitted with massive splenomegaly. Computed tomography of abdomen revealed multiple aneurysms in the distal half of the splenic artery. Splenic artery aneurysms are rare in nulliparous women and most cases are reported in females with a past history of pregnancy. Splenic artery aneurysms, though very rare are clinically significant as they have a high propensity for fatal rupture. Here, we report a patient with multiple splenic artery aneurysms presenting as extrahepatic portal hypertension and massive splenomegaly.

Keywords: Computed Tomography, Portal Hypertension, Pregnancy, Splenic Artery Aneurysms.

CASE REPORT

A 39-year-old female presented with one year history of dragging abdominal discomfort and fatigability of six months duration. She was non-alcoholic and had no other medical illness. General physical examination was unremarkable and abdominal palpation suggested splenomegaly of 10 cms below left coastal margin, which was firm and nontender. There was no ascites or hepatomegaly. Examination of other systems was unremarkable.

Investigations revealed normal complete blood counts, urine analysis, liver and renal function tests and peripheral smear study. Investigation reports are shown in [Table/Fig-1]. Ultrasonography (US) with color doppler of abdomen revealed splenomegaly, normal liver echotexture and cavernous transformation of portal vein with multiple spleno-renal collaterals. ECG and echocardiography was normal. Upper gastro-intestinal endoscopy showed grade 1-2 esophageal varices with portal hypertensive gastropathy. Multidetector Computed tomography (MDCT) abdomen revealed multiple aneurysms in the distal half of splenic artery, the largest one measuring (5.1cmx5.8cmx4.4cms) with dilated and tortuous course of the proximal splenic artery with multiple collaterals along the course of splenic and portal vein [Table/Fig-2,3]. Wall calcification and organized thrombus was seen in few of the aneurysms at the splenic hilum.

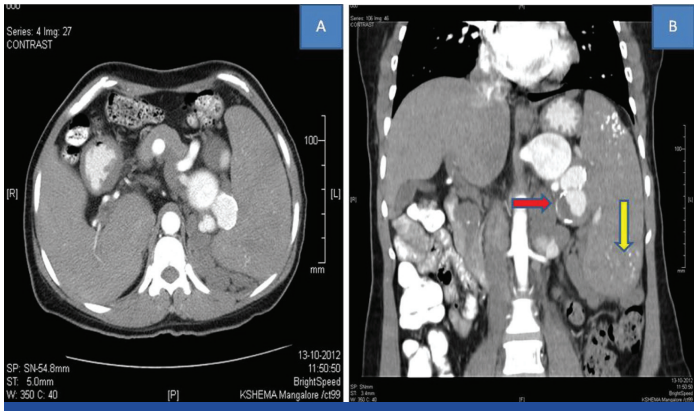
DISCUSSION

Splenic artery aneurysms (SAA) are the most common visceral aneurysms, accounting for up-to 60% of cases [1,2]. Splenic artery is the third most common site for intra-abdominal aneurysm after aorta and iliac artery [2]. It has an incidence of 0.01-0.2%, females being more commonly affected (4:1). Splenic artery aneurysm presenting as extra-hepatic portal hypertension is rare [1,3]. Here we report a patient with multiple splenic artery aneurysms presenting as extrahepatic portal hypertension and massive splenomegaly.

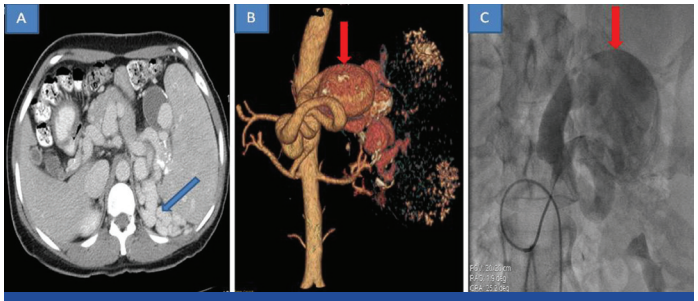
Splenic artery aneurysms are very rare with a varied prevalence of 0.01%-10.4%[4]. SAAs are associated with several conditions, including pregnancy, degenerative atherosclerosis, portal hypertension, medial fibrodysplasia, arteritis, collagen vascular disease, α1-antitrypsin deficiency, and pancreatitis [2]. Hormonal changes during pregnancy, increased blood volume and cardiac output causing portal congestion are the proposed mechanisms of SAA in pregnancy [5]. SAA are usually single, isolated and <3cms in size. It is usually located in the distal part of splenic artery. Larger aneurysms like in our patient are seldom reported [1,3,6]. The aneurysm generally develops on the main splenic artery commonly on its distal third or on intrasplenic branches.

Investigation	Patient's values	Normal range
Haemoglobin	12.3gm/dl	12.5-16 gm/dl
Total count	4,700 cells/mm3	4,000-11,000 cells/mm3
Differential count	N71L25E3M1	N40-70%L20-40%E1-6%M2-8%B0-1%
Platelet count	1.6 lakhs /mm3	1.5-4lakhs/mm3
Erythrocyte sedimentation rate	10mm 1st hour	0-25mm 1st hour
Random blood sugar	142mg/dl	70-150mg/dl
Blood urea	13mg/dl	13-45mg/dl
Serum creatinine	0.6mg/dl	<1.4mg/dl
Sodium	136.9mg/dl	130-145mmol/L
Potassium	3.8mg/dl	3.5-5.0mmol/L
Total bilirubin	0.8mg/dl	<1.0 mg/dl
Direct bilirubin	0.2mg/dl	<0.25mg/dl
Total protein	7.5gm/dl	6.6-8.3gm/dl
Albumin	4gm/dl	3.5-5gm/dl
Globulin	3.5gm/dl	2.3-3.5gm/dl
A:G ratio	1.1	1.5-2.5
SGOT	27U/L	<40U/L
SGPT	10U	<40U/L
Alkaline phosphatase	36U/L	60-170U/L
Prothrombin time	Test: 14.5 sec Control: 14.8 sec INR: 0.93	Control: 11-16 sec

[Table/Fig-1]: Investigation reports



[Table/Fig-2]: A: Multiple aneurysms in the distal half of splenic artery, three of them abutting medial surface of spleen. B- Aneurysms at the splenic hilum show wall calcification and intraluminal thrombus. Splenomegaly with multiple, coarse calcifications



[Table/Fig-3]: A- Tortuous, large calibre collateral venous channels noted along the course of splenic vein and portal vein. B- 3 D image showing dilated splenic artery and multiple aneurysms. C- Angiogram showing large fusiform aneurysm

Splenic artery aneurysms are usually asymptomatic but are increasingly diagnosed as incidental findings. However, epigastric or left hypochondrial pain can be the presenting symptom in some patients. Rarely it can manifest with features of extra-hepatic portal hypertension [1,3]. Catastrophic complication includes rupture, which is more common with pregnancy [5] and larger aneurysms. Splenic infarction may rarely complicate aneurysms due to embolisation of the clot. Rarely, the aneurysm may erode into the stomach causing upper gastrointestinal haemorrhage.

Modern imaging techniques allow early detection of asymptomatic aneurysms [7]. Direct catheter angiography, although is gold standard for diagnosing splenic artery aneurysm needs arterial puncture. With the advent of MDCT, aneurysms can be easily detected. On contrast enhanced CT, aneurysms may appear well defined and homogenous. An unruptured splenic artery aneurysm may be misinterpreted as a pancreatic mass on CT, as islet cell tumors can be hyper-attenuating in the arterial or pancreatic phase. However, 3D imaging augments the accuracy in diagnosing splenic artery aneurysm. Treatment options include aneurysmectomy, trans-catheter embolisation with coils or stent graft placement with or without splenectomy.

This case presented with abdominal pain and was found to have massive splenomegaly on evaluation. Liver function tests were normal with normal serum albumin, liver enzymes and prothrombin time. Ultrasonography of abdomen did not reveal any changes in liver echotexture. Normal hematological parameters and absence of lymphadenopathy excluded haematological cause for massive splenomegaly. Presence of esophageal varices and congestive gastropathy on endoscopy in the setting of normal liver functions pointed towards extrahepatic portal hypertension as the most probable diagnosis. Further evaluation revealed multiple splenic artery aneurysms in the distal half of the artery.

As per the literature,multiple splenic artery aneurysms are uncommon accounting for upto 20% of cases [8]. Cavernous transformation of the portal vein is likely to be due to multiple splenic artery aneurysms causing extrinsic compression of splenic vein, leading to venous stasis and thrombosis of splenic and portal vein. This is the most likely explanation for splenomegaly and portal hypertension in our patient. A case of multiple SAAs caused by fibromuscular dysplasia (FMD) has been reported by Watada S et al., [9]. FMD could be the explanation for multiple aneurysms in our patient.

CONCLUSION

Extrahepatic portal hypertension is a rare presentation of splenic artery aneurysm. SAA detection in the clinical scenario of extrahepatic portal hypertension is of utmost importance, as it is curative and treatment can prevent mortality.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Jan 15, 2014**
Date of Peer Review: **Apr 22, 2014**
Date of Acceptance: **May 06, 2014**
Month of Publishing: **September, 2014**