

# A Rare Quadruple Association: Fibromuscular Dysplasia, Giant Splenic Artery Aneurysm, Extrahepatic Portal Hypertension, and Hypersplenism

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## ABSTRACT

Fibromuscular dysplasia (FMD) is a non-atherosclerotic, non-inflammatory vascular lesion. It is a very rare cause of splenic artery aneurysm (SAA). An 18-year-old girl presented with hematemesis, melena, pancytopenia, and splenomegaly. Endoscopy showed esophageal varices. Computed tomography angiography showed splenic infarct and a giant splenic artery aneurysm. Portal vein showed cavernous transformation with enlarged periportal and lienorenal collaterals. The liver and pancreas were unremarkable. Microscopy of the SAA revealed intimal fibroplasia and medial dysplasia. Symptoms of extrahepatic portal hypertension were relieved by aneurysmectomy, thus proving SAA as the underlying cause. Pancytopenia was reversed post-splenectomy, thus proving hypersplenism. This is the first-ever report showing a quadruple association of FMD, splenic artery aneurysm, extrahepatic portal hypertension, and hypersplenism.

**Key messages:** Fibromuscular dysplasia can present as a giant aneurysm of the splenic artery. The resultant extrahepatic portal hypertension and splenomegaly can result in hypersplenism. Splenectomy and aneurysmectomy can reverse pancytopenia and portal hypertension.

**Keywords:** Extrahepatic portal hypertension, Fibromuscular dysplasia, Hypersplenism, Non-cirrhotic portal hypertension, Pancytopenia, Portal cavernoma, Splenic artery aneurysm.

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## INTRODUCTION

Fibromuscular dysplasia (FMD), a non-atherosclerotic, non-inflammatory vascular lesion is a very rare cause of splenic artery aneurysm (SAA) with only five reported cases.<sup>1-3</sup> Splenic aneurysm causing extrahepatic portal hypertension is also extremely rare.<sup>4-6</sup> We present the first-ever report of FMD causing giant splenic artery aneurysm with consequent extrahepatic portal hypertension and hypersplenism. Splenectomy and aneurysmectomy can reverse pancytopenia and portal hypertension.

## CASE DESCRIPTION

An 18-year-old unmarried girl presented with hematemesis, melena, and abdominal pain for 3 months. She did not have rectal bleed, pale stools, or jaundice. Endoscopy showed esophageal varices. Ultrasonography revealed massive splenomegaly while liver, pancreas, gallbladder, kidneys, and other pelvic organs were normal. Computed tomography angiography (CTA) and Doppler studies showed a fusiform aneurysm of the splenic artery just beyond its bifurcation (Fig. 1). There were portal vein cavernoma along with multiple enlarged lienorenal, mesenteric, and periportal collaterals causing compression of the suprapancreatic bile duct (Fig. 2A). The portal vein showed a peak systolic velocity of 12 cm/second, the delay suggestive of extrahepatic portal hypertension (EPHT). Serum protein was 6.2 g/dL, serum albumin 2.7 g/dL, lactic acid dehydrogenase (LDH) 392 g/dL, alkaline phosphatase 443 IU/L, serum glutamic oxaloacetic transaminase (SGOT) 89 IU/L, and serum glutamic pyruvic transaminase (SGPT) 78 IU/L. Serum amylase and renal function tests were within normal limits. Blood sugar, lipid profile, and serology for hepatitis were negative. Preoperative hematological parameters showed a hemoglobin value of 5.8 g/dL, total leukocyte count  $1.8 \times 10^3/\mu\text{L}$ , and platelet count  $58 \times 10^3/\mu\text{L}$ . Pancytopenia with massive

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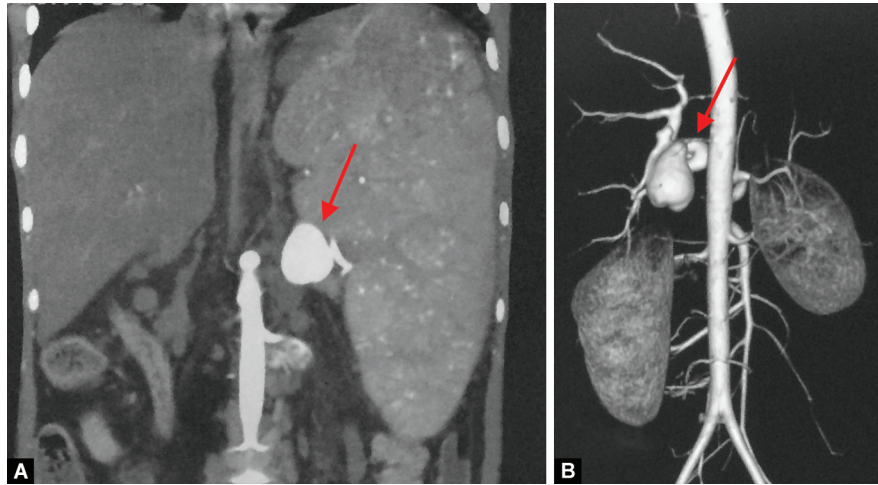
**Conflict of interest:** None

splenomegaly was suggestive of hypersplenism. Bone marrow examination was not conducted.

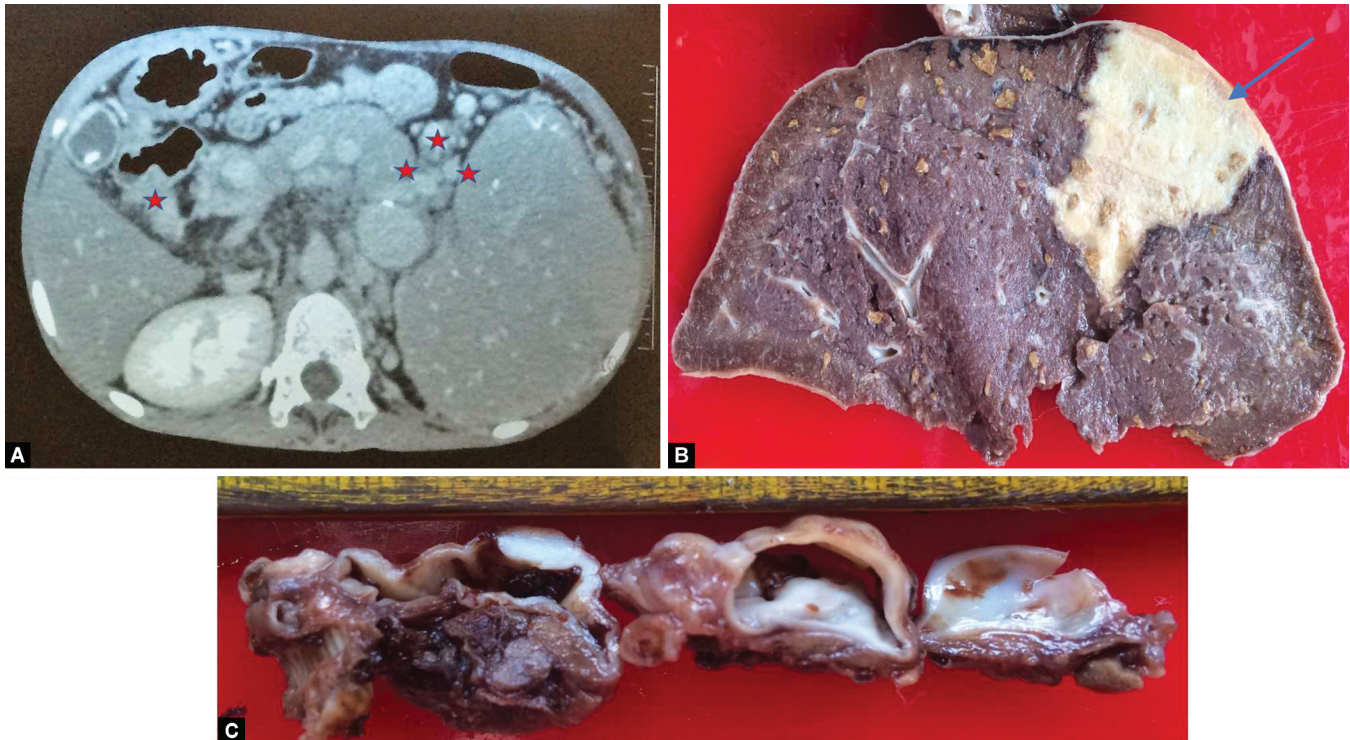
In view of hypersplenism and SAA, the patient was subjected to splenectomy with aneurysmectomy (Figs 2B and C). The resected spleen weighed 800 g and measured  $10 \times 8 \times 5$  cm. It showed fibrocongestive changes with a large infarct. Post-surgery, the hemoglobin value increased to 11.1 g/dL, total leukocyte count  $16.6 \times 10^3/\mu\text{L}$ , and platelet count  $103 \times 10^3/\mu\text{L}$ , thus confirming the hypersplenism. Histomorphology confirmed splenic infarct with multiple Gamna-Gandy bodies. Microscopy of the SAA showed fibrosis of tunica intima and tunica media along with fragmented and reduplicated internal elastic lamina, thus consistent with FMD (Fig. 3).

## DISCUSSION

Fibromuscular dysplasia is a non-atherosclerotic, non-inflammatory vascular lesion. On histology, it is classified as intimal fibroplasia, medial dysplasia, and adventitial fibroplasia.<sup>7,8</sup> Medial dysplasia is further subtyped as medial fibroplasia, perimedial



**Figs 1A and B:** (A) Computed tomography angiography in the arterial phase shows a fusiform aneurysm of the splenic artery (red arrow). The spleen shows massive enlargement; (B) Doppler study highlights the 3 × 3 × 2.3 cm splenic artery aneurysm (red arrow)



**Figs 2A to C:** (A) Computed tomography angiography in the delayed phase shows portal cavernoma along with multiple periportal, splenic, and mesenteric collaterals (red stars); (B) Spleen shows a solitary, pale infarct (blue arrow) and multiple golden-brown Gamna-Gandy bodies; (C) Serial sections of the splenic artery aneurysm show variably thickened and thinned vessel walls. No evidence of thrombus within the aneurysm

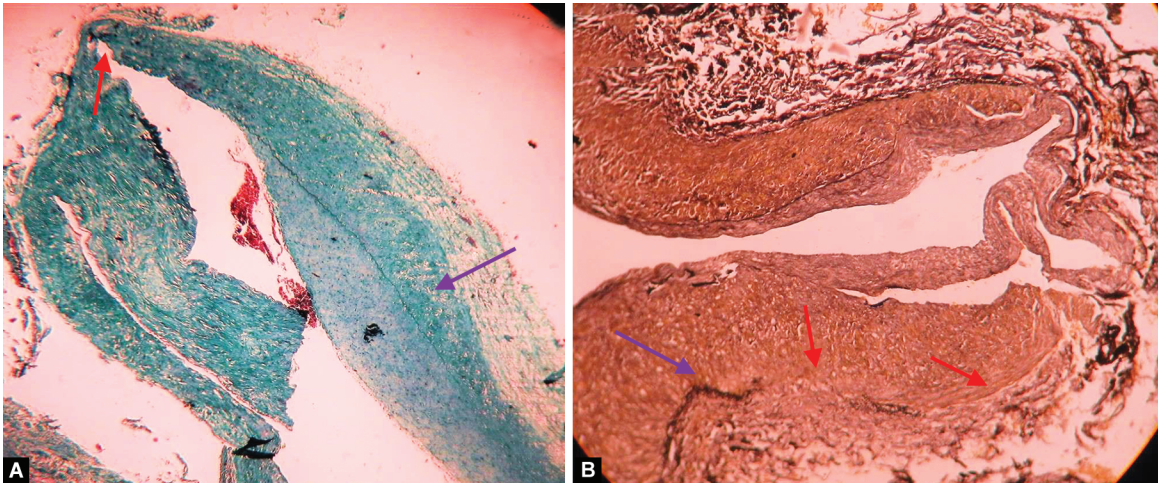
fibroplasia, and medial hyperplasia. Our case showed a mixed pattern of intimal fibroplasia and medial dysplasia. Mimics like atherosclerosis, vasculitis, and segmental arterial mediolysis were ruled out by the absence of foam cells, cholesterol clefts, transmural inflammatory infiltrate, or vacuolated smooth muscles in tunica media.<sup>7,8</sup>

Fibromuscular dysplasia shows a predilection for renal and extracranial carotid arteries.<sup>7,8</sup> Magnetic resonance angiography and CTA show 92 and 62% respective sensitivity to detect FMD. On radioimaging, intimal fibroplasia presents as single tubular stenosis while medial dysplasia shows the classic “string and bead”

appearance. Less typical images include vascular loop, vascular ectasia, arterial dissection, and aneurysm.<sup>7,8</sup> Celiac artery is rarely affected by FMD but shows aneurysm in 15.8% of cases. Our patient presented with a solitary splenic artery aneurysm without “string and bead” or stenosis on imaging.

Splenic artery aneurysm should be distinguished from splenic pseudoaneurysm.<sup>9</sup> The latter is usually associated with acute pancreatitis and pancreatic pseudocyst. Enzymatic autodigestion of the splenic artery causes extravascular hematoma and thus pseudoaneurysm formation. In our patient, pancreatitis was ruled out by normal pancreatic imaging and serum amylase levels.





**Figs 3A and B:** (A) Photomicrograph of splenic artery aneurysm shows features of fibromuscular dysplasia. There are areas of thinning (red arrow) and thickening (blue arrow) in the vessel wall. Fibrosis of tunica intima and tunica media is consistent with intimal fibroplasia and medial dysplasia. Masson Trichrome stain, 40 $\times$ ; (B) Photomicrograph shows loss of internal elastic lamina (red arrows) and reduplicated internal elastic lamina (blue arrow). Elastic von Gieson stain, 40 $\times$

The incidence of true SAA is 0.01 to 0.2%. It shows a 4:1 female preponderance, especially affecting the young reproductive age group.<sup>5,6,10</sup> The common causes of SAA include pregnancy, cirrhotic portal hypertension, trauma, and polyarteritis nodosa.<sup>6,10</sup> Fibromuscular dysplasia is a very rare cause of SAA.<sup>1-3</sup>

True SAA >2.5 cm in size is termed a giant aneurysm.<sup>6,7</sup> It remains clinically asymptomatic in 80–95% cases with a rupture risk of 2–3%. Splenic artery aneurysm can rupture into the less sac of the peritoneum, splenic vein, and pancreatic duct.<sup>9,10</sup> Initial bleed into the less sac followed by eventual peritoneal bleed and hemorrhagic shock is termed the “double rupture” phenomenon. Rupture into the splenic vein forms an arteriovenous fistula with resultant mesenteric steal syndrome. Succus pancreaticus occurs due to aneurysmal rupture into the pancreatic duct with resultant tracking of blood through the sphincter of Oddi into the duodenum.<sup>9,10</sup> Very rarely, as in our case, SAA can present as extrahepatic portal hypertension.<sup>4-6</sup>

Cirrhotic portal hypertension is associated with SAA in 7% of cases.<sup>10</sup> The portosystemic shunt with resultant increased splenic arterial flow weakens the arterial wall and predisposes to the formation of SAA. In our case, the liver showed normal size and echotexture while the hepatic vein showed normal flow, thus ruling out cirrhotic portal hypertension.

The reverse association, i.e., SAA leading to EHPT is rarely documented in the medical literature.<sup>4-6</sup> Splenic artery aneurysm can cause compression of the splenic veins, thus causing venous stasis and thrombosis.<sup>5,6</sup> This can lead to extrahepatic portal vein obstruction with consequent portosystemic shunting and enlarged collaterals. Our case showed a slowed portal flow, multiple enlarged lienorenal collaterals, and portal cavernoma, thus confirming EPHT. In addition, periportal collaterals were compressing upon the suprapancreatic common bile duct, which obstruction could explain the mildly elevated serum levels of LDH and alkaline phosphatase in our case.

Unusual features in our case include the occurrence of FMD and SAA in a non-pregnant young girl. The splenic artery FMD presented as an aneurysm on radioimaging, without the classic beaded appearance. The SAA showed a mixed pattern of intimal fibroplasia and medial dysplasia on microscopy. The symptoms

of portal hypertension were relieved by aneurysmectomy, thus confirming SAA as the cause of EPHT. Splenectomy reversed the pancytopenia, thus proving hypersplenism.

Thus, we present an unusual case of splenic artery FMD leading to solitary splenic artery aneurysm and subsequent extrahepatic portal hypertension with hypersplenism in a non-pregnant young female. To our best knowledge, this is the first-ever report of this quadruple association.

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