

Case Report

Hypersplenism in a young girl: managed non-surgically

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ABSTRACT

A 16-years-old female, non-alcoholic, presented with pain in left hypochondrium and distention of abdomen. Her USG Abdomen, portal vein doppler and CT abdomen revealed chronic thrombosis of intrahepatic portion of portal vein with multiple collaterals and gross splenomegaly suggestive of portal hypertension. Trans jugular liver biopsy showed no regeneration or fibrosis of liver. Endoscopy showed grade III oesophageal and gastric varices. Splenic artery embolisation was done for hypersplenism. Post procedure CT abdomen revealed large areas of splenic infarction-sequelae of splenic artery embolization.

Keywords: Hypersplenism, Portal hypertension, Splenic artery embolization, Trans jugular liver biopsy

INTRODUCTION

Hypersplenism is an overactive spleen. The spleen helps filter old and damaged cells from your bloodstream. If your spleen is overactive, it removes the blood cells too early and too quickly.¹ In 1955, Dameshek summarized that hypersplenism should be diagnosed in the presence of four conditions: i) monolineage or multilineage peripheral cytopenias; ii) compensatory hyperplasia of bone marrow; iii) splenomegaly; and iv) correction of cytopenias after splenectomy. Although these four conditions do not always apply to all cases, they have been commonly cited in the literature, and are important in the diagnosis of hypersplenism.² Here, we present a young girl who had all four manifestations.

CASE REPORT

A 16-years female, resident of Nepal, non-alcoholic, presented with pain in left hypochondrium for 6 months and distention of abdomen. There was no history of malaria, kala azar, sarcoidosis or any other infective aetiology.

On examination, she had tachycardia, pallor and bilateral pedal oedema. Per Abdomen examination revealed massive splenomegaly. Peripheral blood picture showed no howell jolly bodies. Laboratory testing showed pancytopenia (Hb- 4.3 g%, TLC-1200 mm³, Platelet count-46,000 lacs/mm³, PBS-microcytic hypochromic picture). Bone marrow aspiration showed mild erythroid hyperplasia with micronormoblastic maturation. No LD bodies seen. USG (abdomen) and portal vein doppler findings were consistent with portal hypertension. CT abdomen showed chronic thrombosis of intrahepatic portion of portal vein with multiple porto-systemic collaterals.

Endoscopy was suggestive of grade III esophageal varices and grade III gastric varices following which esophageal variceal banding was done. Trans jugular liver biopsy showed no evidence of regeneration or fibrosis. Splenic artery embolization was done.

Ct-abdomen post procedure was suggestive of marked splenomegaly with large areas of splenic infarction-sequelae of splenic artery embolization. Metallic density

lesion along course of splenic artery due to embolic material were noted. Post procedure, the patient had a rise in haemoglobin, total leukocyte count and platelet count over a period of 3 weeks.

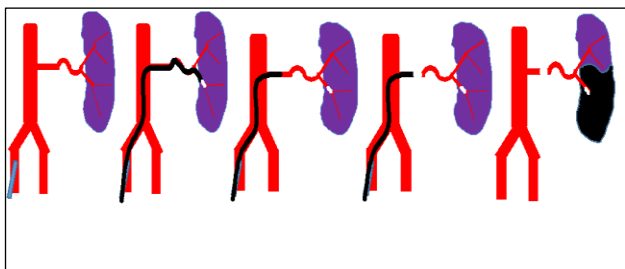


Figure 1: Diagrammatic representation of 'splenic artery embolization' procedure.

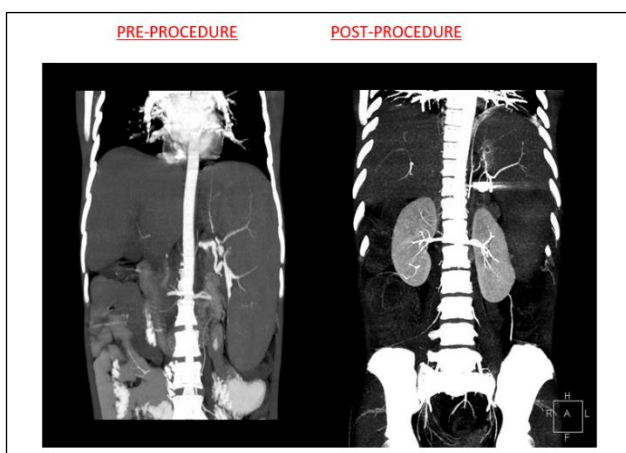


Figure 2: CT-scan showing pre-and post procedure images, with the infarction of lower pole of spleen in the latter.

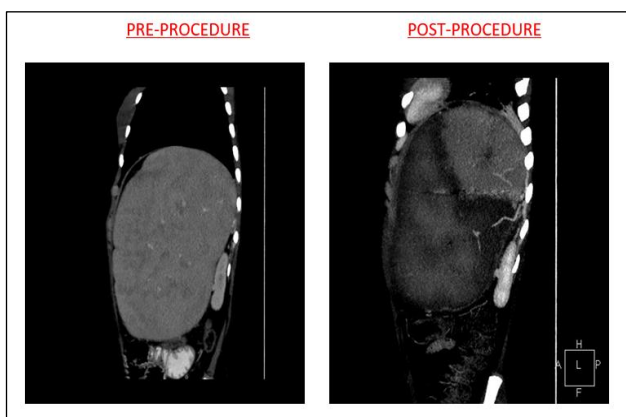


Figure 3: Marked area of splenic infarction seen in the post-procedure image.

DISCUSSION

Splenomegaly is one of the four cardinal signs of hypersplenism which include- reduction in the number of circulating blood cells affecting granulocytes,

erythrocytes or platelets in any combination, a compensatory proliferative response in the bone marrow, and the potential for correction of these abnormalities by surgical or non-surgical management.

Treatment options includes non-surgical, irradiation and ablation and surgical options. Non-surgical methods usually involve etiological treatment and treatment of concomitant diseases.³

Kenawi et al treated eight patients with liver cirrhosis, splenomegaly and hypersplenism by externally irradiating the spleens using radioactive Co-60. Laboratory data showed that the hemogram returned to completely normal in two patients and partially normal in three patients. Moreover, remission of chronic splenic pain was obtained in all patients and no significant complications were reported.⁴

In 1979, Spigos et al used partial splenic artery embolization for the first time to treat hypersplenic patients with success.⁵ This procedure not only increases platelet and leukocyte counts but also reduces splenic size, improves pancytopenia and stimulates the immune system.⁶⁻⁸

Despite some clinical success in treating splenomegaly and hypersplenism, the indications for partial splenic artery embolization are limited due to serious complications, such as splenic infarction and abscess, which could result in an elevated risk of death.⁹

Total Splenectomy in secondary hypersplenism, the underlying disease must be treated prior to splenectomy - removal of the target organ of hypersplenism.¹⁰

Splenic embolization can be used alone or in conjunction with other treatments for the mitigation of portal hypertension and associated physiological effects of portal hypertension. This technique can be used safely when total embolization volume is ~50% and the procedural and peri-procedural time periods are covered with antibiotics. In this patient population, partial splenic embolization can decrease the incidence of variceal bleeding, and protection can persist for at least a year. Additionally, liver function tests and serum cell counts can be expected to improve. Although not frequently used as primary therapy for patients with portal hypertension, splenic embolization can often be helpful as an alternative or adjunctive procedure.¹¹

Gaba RC et al studied that a total of 50 patients (male:female, 33:17; mean age, 49 years) who underwent 50 SAEs between 1998 and 2011 were retrospectively studied. All embolizations were technically successful. The procedure efficacy was 90%; five patients (10%) had a recurrent haemorrhage requiring a secondary intervention.¹²

From August 2009 to May 2012, in a study done by Wang Q et al, 14 patients underwent splenic arterial embolization. Two different embolization strategies were applied; either combined distal splenic bed particle embolization and proximal splenic artery coil embolization in the same procedure for acute haemorrhage (1-step) or interval staged distal embolization and proximal embolization in the stable patient (2-step). The one-step method was performed in three patients suffering from massive gastric bleeding, and the bleeding was relieved after embolization. The two-step method was used in 11 patients, who had chronic gastric variceal bleeding or gastric varices only. The gastric varices disappeared in the enhanced CT scan and the patients had no gastric bleeding during follow-up.¹³

Complications

Most of the patients will have post embolization syndrome consisting of fever, nausea, and left upper quadrant pain. These symptoms can usually be controlled with narcotics and antiemetics. Fever is thought to be secondary due to release of pyrogens from infarcted spleen and may be treated with steroids. Additional side effects may include anorexia, pleural effusion, ascites, and paralytic ileus, all of which can be treated symptomatically.¹²

CONCLUSION

Our patient's blood counts significantly improved post-procedure (Hb-9.7 g%, TLC-5700mm³, Platelet Count-4.80 lacs/mm³). She has been following up and is doing well currently.

Clinical implication involves hypersplenism which is an important clinical finding in many medical conditions. Among the wide list of causes of hypersplenism portal hypertension remains the commonest one. Splenic artery embolization is one of the newer techniques for the management of same with good outcome and fewer risks of having side effects. It helps in improving the liver function and hematological parameters significantly post procedure.

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Ethical approval: Not required

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