Arterioportal fistula - a case report

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Abstract: An intrahepatic arterioportal fistula is a rare occurrence in liver cirrhosis. Arterioportal fistulas have been said to result from congenital arteriovenous malformation, ruptured hepatic aneurysm, trauma, iatrogenic causes, liver cirrhosis, or liver tumor. We report a case of arterioportal fistula in the setting of cryptogenic cirrhosis.

Keyword: arterioportal fistula, cirrhosis

CASE REPORT 70 yr old female presented with abdominal distension and pain in right hypochondrium of 1 month duration. On examination patient had free fluid in the abdomen and right pleural effusion. Her routine investigations were normal. Liver biochemistry showed low albumin (3.3 gm %) only with A/G reversal. Ascitic fluid analysis revealed high SAAG >1.1 with a cell count of 150 cells/mm³. Ultrasound imaging revealed features of cirrhosis, portal hypertension and ascites. Portal doppler showed ascites with arterioportal shunting. Contrast enhanced CT of abdomen showed large arterioportal vascular shunt with early drainage into the right portal vein. Pleural fluid analysis revealed low ADA with a transudative effusion. Work up for cirrhosis which includes viral markers, iron studies, ceruloplasmin and autoimmune markers were normal. Alfa-feto protein levels were within normal limits and gastroscopy showed grade I esophageal varices. Cardiac work up including ECG and ECHO were normal. Patient was managed with diuretics,
fig. 1 and fig. 2 - shows prominent right portal vein during arterial phase occurring in arterioportal fistula.

DISCUSSION:
Patients with arterioportal fistula may be asymptomatic or present with a protean clinical syndrome including the complications of portal hypertension (gastrointestinal bleeding and ascites), heart failure, and intestinal ischemia. Arterioportal fistulas can be congenital or acquired. Congenital causes include hereditary telangiectatic diseases (Osler–Weber–Rendu syndrome, Ehlers–Danlos syndrome), arteriovenous malformations, and aneurysms (1). Most common acquired causes include trauma and liver biopsy.

Arterioportal fistula may occur in patients with liver cirrhosis, both because cirrhosis destroys the sinusoidal net and disturbs the hepatic angiostructure and because these patients more frequently undergo a diagnostic or therapeutic transhepatic procedure (2). The main symptom at presentation was gastrointestinal bleeding in 33%, ascites (26%), heart failure (4.5%), and diarrhea (4.5%) (3).

Presence of an arterioportal fistula could go unnoticed in patients with liver cirrhosis, because portal hypertension is already present in these patients and worsening of the symptoms because of the onset of an APF could be misinterpreted as being due to a decrease in hepatic function that is suggested by the natural history of the disease (2). Color Doppler US can be proposed for the screening of APFs in patients with cirrhosis. This screening can be carried out simply, as patients with cirrhosis are usually periodically examined with Doppler US (2). Early visualization of the portal vein and increased attenuation in the ipsilateral lobe bearing the fistula can be seen in contrast enhanced CT imaging (4).

We report this patient, a case of spontaneous arterioportal fistula in the setting of cryptogenic cirrhosis for its rarity.

References

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