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A Rare Case of Extra Hepatic Portal Hypertension as a Consequence of Chronic Pancreatitis

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ABSTRACT

Background: Chronic pancreatitis is a rare cause of portal hypertension. As hepatic pathology mostly contributes to the same. Extrahepatic portal hypertension followed by pancreatitis is a rare condition with most patients being asymptomatic.

Objective: This paper aims to present clinical presentation, diagnosis and management of extrahepatic portal hypertension because of chronic pancreatitis.

Case Study: A 40-year-old male presented with chief complaints of pain epigastrium and nausea from past 3-4 days which was sudden in onset, progressive in nature, severe in intensity radiating to the back, aggravating on taking food, relieved on leaning forwards and associated with nausea.

Results: Clinical features, radiological and other investigations suggested rare diagnosis.

Conclusion: Extrahepatic portal hypertension is an important cause of the upper gastrointestinal bleed. Pancreatic aetiology should be considered and most of the times it asymptomatic.

Key Words: Chronic pancreatitis, Extrahepatic portal hypertension, Gastric varices, Splenomegaly

INTRODUCTION

Extrahepatic portal hypertension is a rare cause of gastrointestinal bleeding which is characterized by splenomegaly, isolated gastric varices and normal liver condition. Sixty percent of the cases of extrahepatic portal hypertension are caused by pancreatitis specifically chronic pancreatitis accompanied by splenic venous thrombosis.² Isolated gastric varices occur as a result of thrombosis or obstruction of the splenic vein leading to back pressure changes in the left portal system. The primary pathology manually arises in the pancreas, common etiologies being pancreatitis and pancreatic neoplasm.3 However not all patients with extrahepatic portal hypertension would experience bleeding complications. Agrawal et al reported that 22 percent of patients having chronic pancreatitis had imaging evidence of splenic vein thrombosis, only 15% of those had gastrointestinal bleeding.4 More evidence suggests watchful waiting as acceptable course of management in asymptomatic individuals.5

CASE PRESENTATION

A 40-year-old male presented with chief complaints of pain epigastrium and nausea from the past 3-4 days which was sudden in onset, progressive in nature, severe in intensity radiating to the back, aggravating on taking food, relieved on leaning forwards and associated with nausea. The patient was known case of diabetes mellitus for six months. The patient had a history of a similar episode eight months ago. Routine investigations were done along with serum amylase levels. The patient was a chronic alcoholic and smoker and quit alcohol after the first episode. There was no history of fever, loose stools or vomiting. On general examination, the patient's vitals were stable and he was afebrile. On Per abdomen, findings revealed tenderness in the epigastric region with guarding present. Rest of the examination was normal with no signs of peritonitis or ascites. Air entry was bilaterally equal with chest X-Ray showing no signs of pleural effusion. The patient was kept nil per oral, put on intravenous proton pump inhibitors, fluids and analgesics. The blood

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sugar level monitoring was done intensively and managed accordingly. All routine investigations were within the normal limits including LFT and serum amylase levels. Lipid profile and serum calcium levels were also normal. A contrast-enhanced CT scan of the abdomen was suggestive of chronic pancreatitis with portal hypertension with perigastric varices. USG abdomen was done to rule out liver cirrhosis and cholelithiasis. On ultrasonography, echotexture and size of liver were normal with no evidence of gall bladder and common bile duct stones.

USG colour doppler of the splenic vein was done with no evidence of splenic vein thrombosis. Hence, the diagnosis of extrahepatic portal hypertension as a complication of chronic pancreatitis was made. Upper gastrointestinal endoscopy was done for variceal status, which revealed no oesophagal varices and no actively bleeding perigastric varices. The patient was started on non-selective beta-blockers. The patient was managed conservatively. The patient improved symptomatically with no complications to date and he is on regular follow up.

RESULTS

The investigations were as follows- Hb = 12.7gm/dl, TLC = $9000/\mu l$, Platelet count= 2 lacs/ μl , serum amylase = 47 U/l, serum creatinine = 1.0 g/dl, SGPT = 45U/l, SGOT= 30U/l, serum calcium= 9.2, lipid profile was within normal limits. Arterial blood gas analysis revealed pH= 7.41, HCO₃ = 25mEq/lt, pCO₂ = 39mm of Hg, pO₂ = 90mm of Hg.

DISCUSSION

Extrahepatic portal hypertension (EPH) is defined as hypertension of the extrahepatic component of the portal venous system which does not result due to liver cirrhosis. Initially, this entity had been thought to be restricted solely to the splenic vein, so that in the past it has been referred to as segmental, left-sided, regional, localized, splenoportal, sinistral, compartmental, or lienal hypertension.

It represents a lesser common complication of chronic pancreatitis and involves either the individual superior mesenteric or splenic venous branch or may involve the whole splenomesentericoportal axis. The latter possibility appears less likely as in most cases of extrahepatic portal hypertension complicating pancreatitis the splenic vein alone is involved because of its proximity to the pancreas. As this condition was observed with increasing frequency, it became apparent that every part of the splenomesentericoportal venous axis may be involved. The most probable cause seems to be the progressive fibrosis characteristic of chronic pancreatitis, leading to progressive constriction of the splenomesentericoportal axis that passes through the pancreatic substance. 10

The pathology of EPH is characterized by two major forms; a complete occlusion of branches of the venous splanchnic system (occlusive form) ¹¹ or a subtotal obstruction of one or more of these branches (non-occlusive form). ¹²

Greenwald and Wash in 1939 were the first to present a case of left-sided portal hypertension occurring due to splenic vein obstruction (thrombosis or from outside pressure). 13 Due to the occlusion of the splenic vein, the blood from the spleen having no other outflow tract flows into the vasa brevia and other collaterals of splenic and pancreatic circulation. Gastric or oesophagal varices may then develop from the resulting increased left-sided splanchnic pressures. Extrahepatic portal hypertension is a rare clinical syndrome characterized by splenomegaly, isolated gastric varices that may bleed, and normal liver condition. 14 Pancreatic disease is the most common aetiology. Extrahepatic portal hypertension may complicate chronic pancreatitis as a result of splenic vein thrombosis/obstruction.¹⁵ Gastrointestinal bleeding with normal liver function and unexplained splenomegaly should raise suspicion for extrahepatic portal hypertension.¹⁵

Splenectomy is the preferred treatment modality for symptomatic extrahepatic portal hypertension. In symptomatic patients of chronic pancreatitis undergoing operative treatments, concomitant splenectomy should be strongly considered if extrahepatic portal hypertension and gastroesophageal varices are also present. However, just the presence of extrahepatic portal hypertension does not warrant intervention. Expectant management is justifiable in asymptomatic patients with pancreatitis. In most cases, the varices are formed in the fundus of the stomach, while sometimes severe gastrointestinal bleeding may occur due to rupture of oesophagal varices. Extrahepatic portal hypertension is commonly an incidental finding seen in diagnostic imaging for other conditions due to its asymptomatic presentation. Is

CONCLUSION

Extrahepatic portal hypertension is an important cause of life-threatening upper gastrointestinal bleeding. Primary pancreatic pathology should be considered in patients with isolated gastric varices, splenomegaly with the normal status of the liver. Most of the cases are asymptomatic. Splenectomy is the appropriate management option when it is associated with major upper gastrointestinal bleeding. Extrahepatic portal hypertension occurs when a pathological process causes occlusion of the splenic vein. Extrahepatic portal hypertension followed by chronic pancreatitis is a rare condition, which is usually asymptomatic.

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