

Portal Biliopathy

Choonaik Ho*, Andrew Gunn*, Mazin Noordin**

*Surgical Department, Hospital Sultanah Aminah Johor Bahru, Johor, **Radiological Department, Hospital Sultanah Aminah, Johor Bahru, Johor

SUMMARY

Portal biliopathy is a term to describe the spectrum of abnormalities of the entire biliary tract or gallbladder associated with portal hypertension. The most common cause of portal biliopathy is extra-hepatic portal vein obstruction (EHPVO). We report a case of patient with portal biliopathy presenting with bleeding varices¹.

INTRODUCTION

Portal biliopathy is defined as spectrum of biliary abnormalities associated with portal hypertension, most commonly caused by extrahepatic portal vein obstruction (EHPVO). It is postulated that the collateral vein along the CBD was dilated and formed cavernoma which exert the pressure on the CBD leading to complications of biliary obstruction¹.

CASE PRESENTATION

A 60 years old gentleman presented with underlying myelofibrosis disorder, under haematological unit follow up more than 5 years presented with multiple episodes of upper gastrointestinal bleeding to our hospital. Multiple OGDS performed and noted bleeding large oesophageal and gastric varices. Biochemical investigation showed elevated alkaline phosphate, however bilirubin level was normal. Hepatitis B and C were non-reactive. Ultrasound and CECT abdomen noted thickening of circumferential CBD wall with portal cavernoma and splenomegaly however liver was grossly normal. MRCP revealed thrombosed portal vein with portal cavernoma, suggestive of type IIIb portal biliopathy. The patient was transfused with blood and Terlipressin was started. He was referred to Hepatobiliary unit for possible of TIPS. However he was complicated with peritonitis later and urgent CECT showed picture of pulmonary embolism with portal, splenic, IVC and renal vein thrombosis. Patient succumbed to his disease.

DISCUSSION

Portal biliopathy is a rare complication of extrahepatic portal vein obstruction (EHPVO). It is defined as abnormalities seen along the biliary tract or gallbladder associated with portal hypertension. In fact, EHPVO is the most common cause for portal hypertension and was documented up to 40% in the developing country. In adults, EHPVO is commonly associated with hypercoagulable state, chronic myeloproliferative disorder or tumour invasion.

Patient generally presents with features of obstructive jaundice or variceal bleeding as any other cause of biliary failure. In biliary tract, venous drainage passes through two venous plexus named, the epicholedochal venous plexus of Saint and the paracholedochal venous plexus of Petren. In EHPVO, the collateral veins are dilated, forming portal cavernoma and causing external compression on the biliary duct and hepatic duct, results in portal biliopathy. Besides the variceal compression, EHPVO also thought to cause scarring of the ductal wall and lead to biliary strictures due to ischemia. Shin et al documented that the mean time from the diagnosis of portal cavernoma to biliary symptoms was eight years. Majority of the patients were asymptomatic and do not require treatment.

Patient may have abnormal biochemical test due to biliary obstruction or low haemoglobin due to variceal bleeding. Investigation can be divided into invasive or non-invasive. Abdominal US is the most available imaging. It can reveal the splenomegaly, dilated portal vein and collaterals in portal cavernoma and dilated biliary duct. CT scan is good to make a diagnosis but MRCP can give a better image of the dilated vascular soft tissue. Shin et al has described the different findings based on the MRCP, such as varicoid, fibrotic or mixed type^{2,3}. Chandra et al has graded the severity of biliopathy as follows:

- Type I – involvement of extra-hepatic bile duct
- Type II – involvement of intra-hepatic bile duct
- Type IIIa – involvement of extra-hepatic bile duct and unilateral intra-hepatic bile duct
- Type IIIb – involvement of extra-hepatic bile duct and bilateral intra-hepatic bile duct.

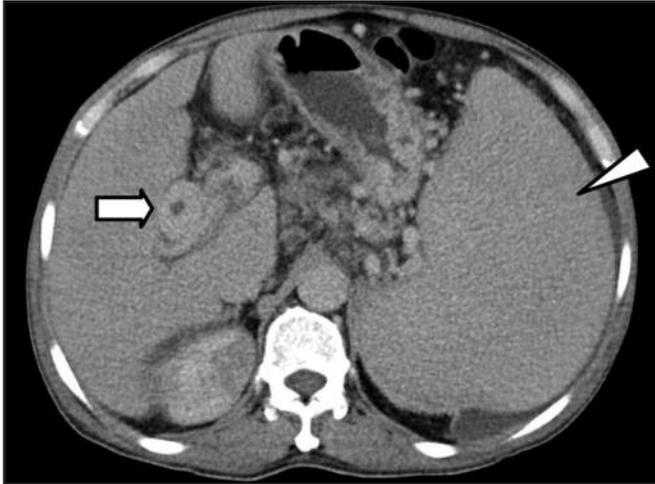
Based on the classification, our patient was classified as Grade IIIb due to involvement of bilateral intra-hepatic ducts.

For the invasive investigation, endo-ultrasound was commonly used to differentiate the cause of biliary obstruction. ERCP can be used for diagnosis and for therapeutic purpose as well. ERCP with papillotomy, dilatation of stricture and stent insertion can be used to relieve the obstruction. Sphinterotomy should be performed with caution due to coagulopathy state of the patients.

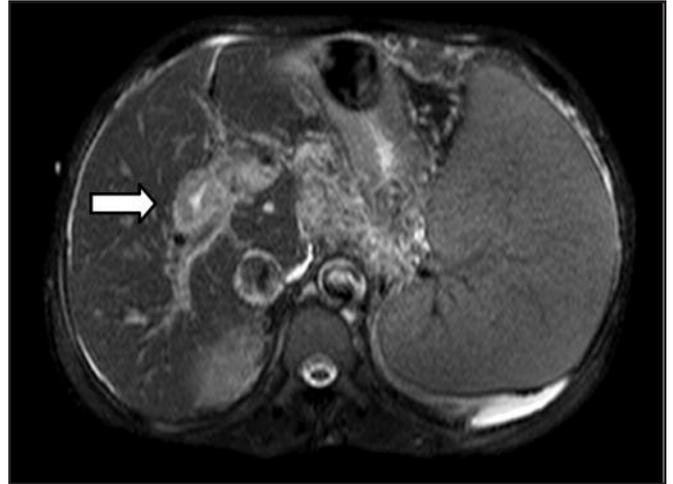
Pharmacological treatment such as somatostatin, octreotide and terlipressin are commonly used to relieve the portal hypertension, so that reducing the blood loss secondary to

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*Corresponding Author: Choonaik Ho, Hospital Sultanah Aminah, General Surgical, Jalan Abu Bakar, Johor Bahru, Johor 81000, Malaysia
Email: choonaik81@gmail.com*



Venous phase contrast enhanced CT scan showing circumferential thickening of the common hepatic duct (white arrow). There is also splenomegaly (arrowhead).



MRI (Fat suppressed T2-weighted sequence) showing smooth and circumferential thickening of the common hepatic duct.

variceal bleeding. Transjugular intrahepatic porto-systemic shunt (TIPS) can be used as last option to decompress the portal system in patient who presented with variceal bleeding. However, for those who have biliary stricture causing biliary obstruction, hepaticojejunostomy may be more beneficial.

CONCLUSION

With the help of current advanced technology, more patients are diagnosed with portal biliopathy secondary to EHPVO. Treatment should be individualized due to variations of the presentation and not all the patient having complications due to portal biliopathy.

REFERENCES

1. Reddy DN, Gokak VP. EC-01 Portal Biliopathy. *Hepatol Int.* 2011; 5(1): 3.
2. Shin SM, Kim S, Lee JW *et al.* Biliary Abnormalities Associated with Portal Biliopathy: Evaluation on MR Cholangiography. *AJR Am J Roentgenol.* 2007; 188(4): W341-7.
3. Sopirjakova P, Krajina A, Bartova J *et al.* Portal Biliopathy: Report on three cases and review of literature. *Folia Gastroenterologica et Hepatologica* 2009; 7(1): 36-41.