CAVERNOUS TRANSFORMATION OF PORTAL VEIN - A RARE CAUSE OF MASSIVE HEPATOMEGALY

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ABSTRACT

There are very few reported cases of Cavernous Transformation of Portal Vein (CTPV) in adults. We present a case of 79 years old male who was found to have this complication due to portal vein thrombosis (PVT). A 79 year old male with background history of JAK2 positive Myeloproliferative disorder (MPD) was referred with abnormal liver function tests. Patient was clinically well and asymptomatic. During initial workup for his abnormal LFTs, patient was noted to have enlarged caudate lobe of liver. Further abdominal imaging studies showed massively enlarged caudate lobe of liver with Cavernous Transformation of Portal Vein (CTPV), a very rare complication of portal venous thrombosis. Cavernous transformation of portal vein is a very rare cause of enlarged caudate lobe of liver. The management of CTPV is mainly symptomatic. Most of the patients are asymptomatic at presentation. Complications mostly occur due to portal hypertension which can be life threatening. There is no consensus on the management of Cavernous Transformation of portal vein itself. Patients with cavernous transformation of portal vein should be kept under regular follow up.

***Key Words**: Cavernous transformation, Portal hypertension, Hepatomegaly.*

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INTRODUCTION

Cavernous Transformation of the Portal Vein (CTPV) is a rare condition with various aetiologies and diverse clinical presentations[1]. It occurs with long-standing portal vein thrombosis (PVT) which causes portal hypertension and occlusion of the portal vein leading to the development and dilatation of multiple small vessels in and around the re-canalizing main portal vein. It has been found to occur commonly in patients with healthy livers with chronic non-cirrhotic and non-tumoral PVT. Yet, the causes of CTPV are unknown[2]. The main clinical presentations are gastroesophageal variceal bleeding and haematological abnormalities due to the effects of the collateral vessels resulting in an enlarged spleen and the development of Porto-systemic collaterals. The diagnosis of CTPV is very rarely made on an adult with signs and symptoms of obstructive jaundice. Abdominal ultrasonography, colour Doppler ultrasonography, computated tomography angiography (CTA), and magnetic resonance imaging (MRI) are used to confirm the diagnosis. We report a case of an adult male in whom the diagnosis of CTPV was made during the workup for obstructive liver function tests. The imaging led us to a rare cause of massively enlarged caudate lobe of liver compressing inferior vena cava, without any other sings of portal hypertension. This is the first reported case of such massively enlarged caudate lobe of liver secondary to portal cavernoma.

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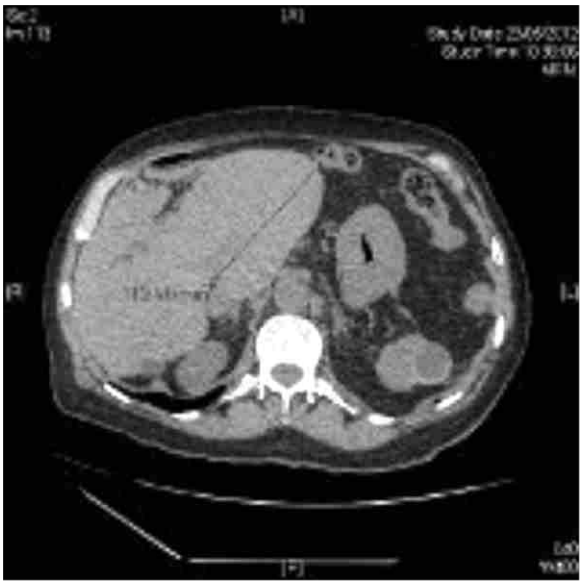
CASE REPORT

A 79 years old male, was referred to the gastroenterology services for deranged liver function tests. Patient was clinically asymptomatic, with no history of abdominal pain or jaundice. He is a non-smoker and does not drink alcohol. His background history includes JAK 2 positive Myelo-proliferative disorder (Essential thrombo-sytosis) for which he was receiving chemotherapy and was well controlled. He also has history of prostatic carcinoma which was treated with

Figure 1: CT Scan Liver Section with measurements of caudate lobe



radiotherapy in the past and the disease was in remission. Patient has been diagnosed with portal vein thrombosis in the past along with splenic infarcts. Patient did not receive any anti coagulation for the portal venous thrombosis as it was picked up late as an incidental finding and by that time patient had seen through the acute phase of the disease process.

On examination patient did not had any signs of jaundice or anaemia. Abdominal examination revealed enlarged liver which was palpable 3cms below costal margin. There was no splenomegaly and no other stigmata of portal hypertension were present. Initial laboratory workup showed normal FBC, U&Es, CRP, INR. His LFTs showed ALP of 142(70-120), gamma GT of 411(110-160) both were elevated with normal ALT and bilirubin levels. The trend of his liver function tests over the last 18months showed that his ALP has been in the range of 120-160 and his GGT was in the range of 360-450 with normal Bilirubin and ALT.

On his first consultation in the clinic, we requested a full liver screen including viral markers, tumour markers and autoimmune screen to exclude any other potential causes for portal hypertension. An ultrasound examination was also requested as an initial imaging tool and since patient was asymptomatic he was sent home with an outpatient follow up arranged. Patient was reviewed after 6 weeks. His liver screen including viral markers, autoimmune profile, ferritin, immunoglobulins, alfa fetoprotein were all found to be within normal range. His US scan showed enlarged liver, and evidence of portal venous thrombosis with partial recanalization. There was no evidence of splenomegaly and ascites.

Since patient had past history of myelo-proliferative disorder and US scan revealed an enlarged liver we organised a CT scan abdomen and pelvis liver protocol. Results of the CT scan showed progressive hypertrophy of the caudate lobe which measures 12cm in diameter. The enlarged caudate lobe was compressing inferior vena cava. Evidence of portal vein thrombosis with cavernous transformation at portahepatis with retroperitoneal collateral veins was also seen. There was no ascites and spleen was shrunken with evidence of previous splenic infarcts. No evidence of malignancy was visible. Hence diagnosis of massively enlarged caudate lobe secondary to cavernous transformation of portal vein was made.

Patient underwent gastroscopy to rule out gastrooesophageal varices, which was entirely normal. The Doppler studies confirmed inferior vena caval compression but it was not significant enough to warrant any intervention.

Patient's scans were reviewed in Radiology MDT since patient was asymptomatic, and his Liver function tests have remained stable, it was decided to keep him under follow-up with 6 monthly US scans.

DISCUSSION

Portal vein thrombosis (PVT) refers to the complete or partial obstruction of blood flow in the portal vein, due to the presence of thrombus in the vessel lumen[3].

Cavernous transformation of the portal vein (CTPV) is sequelae of portal vein thrombosis and is the replacement of the normal single channel portal vein with numerous tortuous venous channels bypassing the obstructive area. First described by Balfour and Stewart in 1869, it was Kobrich in 1928 that first used the term "Cavernoma". Once the liver blood supply decreases significantly, the compensatory mechanisms are activated and collaterals begin to form within few days after obstruction and organise into cavernous transformation in 3-5 weeks[4].

CTPV induces some morphologic changes in the liver, the most common being atrophy of the left lateral segment and hypertrophy of caudate lobe. Caudate lobe hypertrophy occurs in many other conditions, such as cirrhosis, Budd-Chiari syndrome, end stage sclerosing cholangitis and congenital hepatic fibrosis[5]. The reasons for this selective hyperplasia in the liver is unclear. However, the differences in hormones, nutritional elements and hepatotrophic factors in the portal blood flow between caudate lobe and the other segments of the liver may correlate with this hyper plastic change[6]. According to Couinaud, the caudate lobe receives a specific pedicle independent of the portal vein. The right gastric vein and the Para biliary venous system occasionally drain directly into the posterior edge of the medial segment (segment 1V) or the caudate lobe[7]. These could be also be the contributing factor in these morphologic changes seen after cavernous transformation. Among the cause of portal venous thrombosis, Myelo-proliferative disorders(MPD) remains a major latent or potential cause of extra hepatic PVT[8-10].

In non-cirrhotic and non-tumurol PVT in the West, MPD (i.e. polycy-themia-rubravera, essential thrombocythemia and myelofibrosis) with a combination of several prothrombotic factors constitute the most common identifiable cause with estimated prevalence of 30-60% in various studies[4,12,13]. Patients with CTPV can present in variety of different ways. Most patients are however, asymptomatic at presentation, like our patient. The clinical presentation of CTPV can range from normal biochemical and physical findings to cholestasis and splenomegaly and abnormal hepatic function[2]. The initial diagnosis of CTPV is seldom made in adults due to various aetiologies and clinical presentations. Our patient was referred with obstructive biochemical abnormality of his liver functions. These abnormalities could be the result of the compression of biliary tracts by the venous collaterals that run along the extra hepatic biliary tree[14]. The diagnosis of CTPV is confirmed by various imaging modalities including abdominal ultrasonography, colour Doppler US, CT angiography and MRI[15].

It has been demonstrated that the aetiology of CTPV determines the outcome, since the management differs with different causes[1]. If the patient is diagnosed with a recent phase of PVT, then anticoagulant therapy is indicated to prevent total obstruction of the portal vein and stopping the cavernous transformation. In patients with established cavernous transformation of the portal vein as in our patient, anticoagulation is not indicated due to lack of evidence and increased risk of bleeding from oesophageal varices. The major complication of PVT is upper GI bleeding originating from oesophago-gastric varices, with a mortality rate of 5% in patients without liver cirrhosis, than in patients with cirrhosis (between 30-70%)[16].

The endoscopic procedures like band ligation and sclerotherapy are the first line treatments indicated in such cases. Along with endoscopic therapy medical management with non-selective beta blockade and long acting nitrates also have a role in reducing the portal hypertension. In refractory cases of variceal bleeding TIPSS placement should also be considered. Initially considered as contraindication for liver transplantation, the complete PVT is now considered to be just a relative contraindication but only in cases in which superior mesenteric vein is permeable. Partial PVT can be managed by thrombectomy or by-pass techniques. Portal biliopathy is another potential complication that can develope in the evolution of PVT with cavernous transformation. It consists of multiple successive strictures involving common bile duct and hepatic duct as a result of extrinsic compression and/or ischemic fibrosis of the biliary tract.

In symptomatic patients (cholangitis, cholecystitis, CBD stones, and secondary biliary cirrhosis) the portal biliopathy can be addressed with endoscopic treatments like ERCP (stricture dilation, stenting). However there is no consensus to the optimal management for this complication[17]. In our patient as there was no evidence of portal biliopathy, therefore no such treatment was warranted. From the systemic conditions associated with portal cevernoma, myelo proliferative disorders is by far the most common (37% of non-cirrhotic non-malignant cases of PVT)[18].

Our patient had the diagnosis of Essential thrombocy-themia, as the cause of his PVT. To prevent thrombotic complication in these patients, platelet lowering agents are used. Hydroxyurea, a ribonucleotidereductase inhibitor with myelo-

suppressive action is the first line agent. A platelet specific lowering agent (Anagrelide) is also used for patients who are intolerant to hydroxyurea. The goal of platelet lowering agents is to maintain platelet count below 40,0000/mm3 in the high risk patients[19]. Our patient was already on hydroxyurea on presentation and his platelet counts were under control.

CONCLUSION

In the absence of liver cirrhosis and neoplasia the development of portal cevernoma is usually asymptomatic until the first variceal bleeding. Apart from variceal bleeding, the natural history of portal cavernoma can lead to intestinal ischemia (due to extension of the thrombus in the mesenteric vein) and portal biliopathy. Our patient was unique in the fact that he had developed massive hyperplasia of his caudate lobe, which was compressing his inferior vena cava, associated with deranged liver function tests, without any evidence of portal biliopathy or portal hypertension. This case was unique in the fact that this level of hypertrophy of caudate lobe has not been reported in the literature previously. Further research is needed to fully understand why this hypertrophy is limited to only one segment of the liver. This case also highlights the importance of early referral to a specialist centre and multi-disciplinary team input in cases like our patient. Since there is no curative treatment for this condition, patients like ours need close follow-up with the MDT teams so that any potential future complications are picked up early.

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