Hepatic echinococosis complicated with cavernous degenration of portal vein

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Abstract

Cavernous Transformation of the Portal Vein (CTPV) is a rare condition with various etiologies and diverse clinical presentations. It occurs with long-standing portal vein thrombosis (PVT) which causes portal hypertension and an 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. The main clinical presentations are gastroesophageal variceal bleeding and hematologic abnormalities due to the effects of the collateral vessels resulting in an enlarged spleen and the development of porto-systemic collaterals. However, the diagnosis of CTPV is very rarely made on an adult with signs and symptoms of obstructive jaundice. Abdominal ultrasonography, color Doppler ultrasonography, computated tomography angiography (CTA), and magnetic resonance imaging (MRI) are used to confirm its diagnosis. We report a case of a male adult in whom the diagnosis of CTPV was made during the initial workup for a hepatic mass. The symptoms that led us to this rare diagnosis were hyperbilirubinemia and an extrinsic mass compressing on the stomach and the small bowel without any signs of portal hypertension. Although the patient presented with anemia, no sources of bleeding were found, including the presence of esophageal varices

Keywords *Cavernous transformation of the portal vein; Portal vein thrombosis; Portal hypertension; Hyperbilirubinemia; Hepatic mass*

INTRODUCTION:

First proposed in 1869 by Balfour and Stewart, cavernous transformation of portal vein (CTTV) refers to the narrowing or blockage of the main portal vein and/or its branches, leading to the formation of numerous tortuous venous plexus in the hepatic portal area. These plexus, characterized by their caverous hemangiomatous changes, serve to bypass the occluded portal vein and drain blood into the liver, thus relieving

portal hypertension. CTV is a type of prehepatic portal hypertension, characterized by progressive elevation of portal pressure and clinical manifestations such as ascites, esophagogastric varices, splenomegaly with hypersplenism, and portal hypertensive biliary disease. While various factors can contribute to the development of CTTV, portal vein thrombosis is commonly considered a major cause. In rare cases, echinococcosis lesions in the first hilar region can also lead to secondary CTPV, which is associated with a difficult, high-risk, and poor-prognosis treatment. In recent years, there has been increasing focus on Echinococcus cercariae. Through a systematic review of existing literature on hepatic echinococcosis complicated by secondary CTPV, this paper examines the etiological mechanism, clinical manifestations, diagnosis, and treatment of this condition.

1 Etiology and classification of hepatic echinococcosis complicated with secondary CTPV：

The etiology and classification of hepatic echinococcosis complicated with secondary CPV is not fully understood. The condition is caused by the metacestode of echinococcus hepatis, which leads to portal hypertension. This can occur due to compression of the inferior vena cava, hepatic vena, hepatic hilum, and secondary segmental portal hypertension. Additionally, portal thrombosis and secondary CPV can also result from this condition. However, the precise mechanism by which hepatic echinococcosis causes CPV is not clear. According to Gonzalez and Khalefa AA, portal vein thrombosis and cavernous transformation of the portal vein are rare complications of hepatic cysticercosis. The presence of metacestode in the hepatic portal can cause compression of both portal and hepatic veins. This compression, especially of the portal vein, can lead to luminal stenosis or occlusion, and may also cause inflammation in the vascular wall, ultimately resulting in portal vein thrombosis and cavernous degeneration. Previous history of hepatic metacestode infection or abdominal surgery can also contribute to CPV, as changes in vascular anatomy can lead to hemodynamic changes and increased risk of portal thrombosis. Furthermore, cirrhosis of the liver can further aggravate this condition, as it is associated with increased intrahepatic blood flow and blood stasis, which can slow down the already compressed and narrow portal vein and promote the formation of portal vein thrombosis and cavernous transformation.

1.2 Typing of CT PV

There are three types of CT PV, secondary to hepatic echinococcosis, which can be classified based on the involvement of the main portal vein and its branches. Type 2 is the extrahepatic type, characterized by stenosis or disappearance of the portal vein trunk, but without involvement of the intrahepatic portal vein branch. Type 1 is the intrahepatic type, where there is involvement of the intrahepatic portal vein branch, but the extrahepatic portal vein trunk is not affected. This classification is important in determining the severity of portal vein invasion and in developing effective treatment plans.

2 Clinical manfestions.

The clinical manifestations of secondary CPV caused by metacestode of echinococcus hepatica are different from those of primary CTPV. Common symptoms include abdominal pain, ascites, splenomegaly, progressive jaundice, hypersplenism, and in some cases, gastrosophageal varices. Gastrointestinal bleeding is rare. The compression of echinococcosis can lead to obstructive jaundice, and CPV can cause portal hypertensive biliary disease, which can further aggravate jaundice in patients with Hauetibla. This occurs when the lateral branch venous plexus around the biliary tract becomes filled and expanded, causing compression and obstruction of the bile duct.

3 supplementary examination

3.1 Laboratory tests

The blood routine of patients with hepatic echinococcosis complicated with secondary CTV displayed a decrease due to hypersplenism [12]. In cases where only the number of eosinophils was elevated, it was suggested that if the cyst ruptured and led to an increase in serum and urine bilirubin, combined with yellow discoloration of the skin and sclera, the hepatic echinococcosis lesion could result in obstructive jaundice due to compression of the bile duct. Furthermore, if patients also presented with hypoalbuminemia, increased levels of alkaline phosphatase, glutamic oxaloacetic transaminase, glutamic-pyruvic transaminase, prothrombin time (PT), partial prothrombin time (APTT), and international standard ratio (INR), it indicated incomplete liver function. Additionally, ELISA and other echinococcosis immunodiagnostic tests can be utilized to aid in the diagnosis of hepatic echinococcosis. [13]

3.2 Imaging studies

3.2.1 Portography

One of the effective techniques for diagnosing CPV is portal venography, which includes direct and indirect methods. Among these, indirect portography using the superior mesenteric artery or celiac artery is a safe and reliable option. This method can reveal multiple small, twisted collateral veins in the portal vein area, as well as the severity of esophageal and gastric varices and the normal state of the superior mesenteric and splenic veins. It is useful for assessing the extent of portal vein cavernous degeneration and guiding treatment plans. In recent years, transspleen portal venography has been shown to have advantages in diagnosing and treating extrahepatic portal vein occlusion and CTPV. However, due to its invasive nature and limited usefulness in diagnosing hepatic hydatidosis, it is primarily used in the interventional treatment of CTPV complication with hepatic echinococcus metacestode.

3.2.2 Ultrasound

Ultrasonic examination offers numerous benefits in the diagnosis of hepatic echinococcosis due to its non-invasive nature, cost-effectiveness, convenience, and high success rate. In comparison to traditional B-ultrasound, color Doppler ultrasound provides even more advantages in assessing the blood supply to echinococcosis, identifying adjacent blood vessels, and measuring the diameter and blood flow of vessels. It is also capable of displaying abnormal blood flow patterns in cases of hepatic hilar disorder and dilated tortuous vessels in the diagnosis of CTPV, making it a specific and preferred option for diagnosing hepatic echinococcosis complicated by secondary CPV. Additionally, color Doppler ultrasound can be used for tracking and postoperative follow-up of hepatic echinococcosis with secondary CPV. A relatively new technique, contrast-enhanced ultrasound, has the ability to enhance Doppler signals and offers more advantages than CT and MRI in delayed imaging. Recent studies have shown that contrast-enhanced ultrasound has a higher diagnostic accuracy in identifying alveolar echinococcosis and CTPV. Despite its many benefits, factors such as patient obesity, interference from intestinal gas, and operator experience may contribute to missed diagnoses.

3.2.3 CT and MRI

The combined use of CT scan and dynamic enhancement scan not only enables the determination of location, size, and number of hepatic echinococcus lesions, but also provides a clear visualization of the relationship between the lesions and adjacent tissues. Moreover, the application of 3D reconstruction technology accurately displays the compression and displacement of hepatic echinococcus lesions, as well as the narrowing of important vessels such as the hepatic metacestode [18]. Additionally, it can also effectively demonstrate the

distribution of main branches of the portal vein system, the tortuous collateral circulation in the hepatic portal region, and the varicose esophagogastric fundus vein, which is crucial for vessel localization, disease assessment, and surgical planning. Furthermore, it offers unique advantages in cases of hepatic echinococcosis complicated by secondary CTV, MRIMRCP. The use of multi-parameter and multi-directional imaging on MRI allows for accurate characterization and localization of hepatic hydatid disease foci [19]. The addition of 2D imaging with fat suppression can reveal the formation of soft tissue masses or reticular structures around the occluded portal vein [20]. On contrast-enhanced CT scans, the late phase can display dilated, tortuous, and cavernous vessels surrounding the common bile duct, known as the 'pseudocarcinoma of the bile duct' sign [21]. MRCP plays a significant role in the diagnosis of portal hypertensive biliary disease [22]. Furthermore, the application of three-dimensional dynamic contrast-enhanced magnetic resonance angiography (3D DCE-MRA) [23] can accurately assess the severity of portal vein thrombosis and provide a clearer visualization of the entire portal system and its collateral circulation in the presence of CPV. In summary, the combination of CT scan, dynamic enhancement scan, and various MRI techniques offers valuable insights into the location, extent, and severity of hepatic echinococcosis and its complications.

4 Treatment strategies

There are multiple treatment strategies for managing complications of echinococcus multilocularis (CTPV). In the past, surgical treatment was not recommended for cases involving CTPV. However, recent advancements such as beta-blockers, endoscopic ligation or sclerotherapy for digestive tract bleeding, and ERCP or PTCD for biliary complications have expanded the options for treatment. Vascular bypass reconstruction and liver transplantation have also emerged as effective techniques for managing hepatic echinococcosis complicated with CTPV. In a study by Serdark et al in 2016, successful surgical treatment was achieved for cases of hepatic cysticercosis with secondary CTPV1 through the use of H-shaped intestinal shunt created by an allogenic illal vein bypass. This resulted in the resolution of portal hypertension and abdominal distension. The researchers emphasize that when dealing with hepatic hilar echinococcosis complicated with portal vein thrombosis and secondary CTPV, it is crucial to address both the echinococcosis cyst and portal hypertension during surgery.

Afterwards, Professor Wang Wentao and his team reported a successful case of treating alveolar echinococcosis complicated by secondary CTPV through autologous liver transplantation. To reconstruct the inflow and outflow tract during the operation, the portal and hepatic veins were repaired using autologous great saphenous vein. The method of revascularization, whether through in vivo hepatectomy or autologous or allogeneic liver transplantation, is crucial. Based on the classification and characteristics of the lesions, it was determined that type l cases only involved the main portal vein, making end-to-end anastomosis possible after resecting the affected segment. In type ll cases, the portal venous trunk and intrahepatic branches were invaded, most commonly in the right lobe of the liver extending into the caudate lobe. In these cases, the intrahepatic portal venous trunk and right branch were typically completely occluded. After removing the lesion, the left hepatic portal branch and main extrahepatic portal trunk could be anastomosed to the left hepatic portal branch. If the defect was too long, an autologous or allogeneic blood vessel graft or artificial blood vessel could be used to reconstruct it. In type lll cases, only the left and/or right branches of the intrahepatic portal vein were involved. However, due to cavernous degeneration of the portal vein, it was difficult to find suitable portal vein branches for anastomosis in the liver. Therefore, DCD liver transplantation was the preferred method. The specific treatment strategies are summarized as follows

4.1 Preoperative treatment

Echinococcosis of the liver, complicated by secondary CPV, can lead to obstructive jaundice due to compression from the echinococcosis. Additionally, CPV can result in portal hypertension and aggravate symptoms such as yellow staining of the skin and sclera. To relieve jaundice, decompression and drainage through ERCP or PTCD may be necessary to improve liver function. Surgical treatment may be pursued once the serum total bilirubin levels have decreased to below 40 g mol/L. In cases where patients with Budd-Chiari syndrome experience obstruction due to compression of the hepatic vein and inferior vena cava, balloon dilation or stent placement may improve liver function. Furthermore, patients with hepatic echinococcosis complicated by secondary CTPV may also experience ascites due to portal hypertension, resulting in abdominal distension and discomfort. In cases of significant ascites, ultrasound-guided abdominal puncture and drainage may be required. Treatment for liver dysfunction may include liver protection and other symptomatic measures. The presence of metacestode of echinococcus hepatis, combined with secondary CTPV, may result in stenosis of the portal trunk or branches due to focal compression, leading to hemodynamic changes and thrombosis. Therefore, prophylactic anticoagulation should be considered to prevent further exacerbation of portal thrombosis. Although some studies have suggested the benefits of long-term anticoagulation in the treatment of portal cavernous degeneration, specific indications and protocols have not been established and further research is warranted.

4.2 **4.2 Conventional surgery**

In the case of hepatic echinococcosis, secondary CTV involvement can complicate matters in the hilar region. CTPV is typically located on one side of the echinococcosis, and apart from portal vein invasion, other vascular structures such as the hepatic vein or inferior vena cava are not usually affected. To alleviate pressure on the hepatic hilum, conventional surgery can be used to extract the metacestode of echinococcus. However, for cases of hepatic cystic echinococcus disease, the tough outer capsule closely located to the hepatic hilum may impede decompression with simple internal capsule treatment. In such situations, resection of the outer capsule near the hilum is necessary. If the lateral branches of the dilated and tortuous portal vein in the hepatic portal make removing the external capsule challenging, alternative options such as end-to-end anastomosis of the main portal vein, enterocaval shunt, or bypass of the Rex superior mesenteric vein and intrahepatic portal vein branch can be considered based on the severity of portal hypertension and the nature of the portal vein embolism.

4.3 Excised liver resection and autologous liver transplantation

The surgical procedure of liver resection and autologous liver transplantation is a viable option for patients with hepatic alveolar echinococcosis, who also have complications of secondary CTPV and invasion of the posterior inferior vena cava and hepatic vein confluence. In such cases, the transplanted liver volume should exceed 40% of the estimated standard liver volume[13], while ensuring that the total bilirubin levels are less than twice the normal value (s 60 mmol / L) [26]. If the portal vein inside and outside the liver are normal, the peripheral branches of the portal vein can be excised and the portal vein trunk can be anastomosed end to end. Alternatively, if the defect segment is too long, bypass surgery can be performed using artificial or allogenic blood vessels.

4.4 Liver transplantation

In cases where the liver's portal vein has undergone significant cavernous transformation due to the presence of echinococcus hepatis with secondary CTPV, and there is no anastomosing portal vein trunk, routine hepatectomy or OLT procedures are not feasible.

5 Conclusion

Rarely seen in hepatic echinococcosis, cavernous degeneration of the portal vein is a serious complication. It is believed to be caused by the local compression and vascular invasion of the echinococcosis located in the liver's hilum, resulting in portal vein stenosis. In order to maintain normal liver function and ensure proper blood flow, the body compensates by forming a complex network of lateral venous plexus around the portal vein. This condition is characterized by a variety of symptoms including jaundice, abdominal pain, splenomegaly, hypersplenism, and the development of esophageal and gastric varices. If left untreated, it can lead to life-threatening complications such as portal hypertension and bleeding from the varices. Therefore, the primary treatment approach should involve the resection of the hydatid lesion to effectively alleviate portal hypertension. However, there is a lack of comprehensive research on this condition, including its surgical methods, effectiveness, and long-term prognosis. Further objective and reliable clinical studies are needed to confirm the best course of treatment for this rare complication.

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