A CASE OF RIGHT HEPATIC LOBE TORSION PRESENTING WITH CHRONIC DISEASE

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ABSTRACT

A 20-year-old male presented with hepatomegaly, jaundice, and itching. Investigation revealed elevated liver enzyme levels, suggesting direct hyperbilirubinemia. Magnetic resonance imaging revealed a stable orientation of the right hemithorax, a relatively small and rotated right hepatic lobe, and a left-rotated intra-abdominal hepatic lobe, with progressive enhancement related to the foci of the hepatic fibrosis. There was an intrahepatic biliary dilatation, reaching 0.8 cm that was due to biliary compression. There was no evidence of biliary irregularity to suggest a classic primary sclerosing cholangitis pattern.

Percutaneous transhepatic biliary drainage and a cholangiogram confirmed the dilation of the left and right bile ducts with smooth, normal drainage via the common bile duct.

The symptom management strategy took the form of surgical liver transplantation. The patient demonstrated improvement following transplantation, and his total bilirubin level returned to normal within months. The patient was then discharged in a stable status, and successive follow-up appointments were scheduled every three months to monitor for disease progression or complications.

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1. INTRODUCTION

Liver torsion is a rare clinical finding that is probably underrepresented in the population owing to asymptomatic presentation in patients. This rare but significant condition presents when a liver lobe undergoes torsion. Ultrasound scanning may be the least diagnostic imaging modality for diagnosing complete torsion of a liver lobe.

Contrast-enhanced computed tomography (CT) scanning, especially CT angiography (CTA), was more frequently prioritized in the process of diagnoses. Magnetic resonance imaging (MRI) permitted the anatomy and pathology of the underlying cause to be determined to the best standards.

However, CT scan and CTA are the primary choices of imaging modality to evaluate liver lobe torsion. These tools increase the likelihood of appropriate diagnoses and increases the possibility of restoring health to the ischemic lobe or segment [1]. The clinical signs of hepatic lobe torsion may vary, with some cases being either subclinical or resulting in death. Most cases are diagnosed by laparotomy or during a later postmortem examination. Hepatic torsion is a condition that is usually incidentally discovered.

Histopathologically, the affected liver lobe exhibits an obvious loss of hepatocellular differentiation, replaced by a connective tissue which is fibrosed [2]. Even though liver torsion is rarely seen, the differential diagnosis of abdominal pain should include hepatic lobe torsion. Appropriate imaging should be performed, so the surgical teams can prepare for the next management step, which is a surgically complex procedure required for the proper correction [3]. Although it is a rare cause of acute surgical abdomen, some studies found that hepatic lobe torsion has been identified in humans. Its unusual abdominal presentation demands a high level of concern, since biliary symptoms arising from hepatic lobe torsion symptoms is an unusual occurrence [4]. This study presents a case of hepatic torsion causing biliary symptoms. We describe a rare case of primary sclerosing cholangitis (PSC) with torsion of the right liver lobe, leading to obstructive jaundice with the underlying obstructive biliary system.

Case Report

The case involved a 20-year-old male with a known case of primary sclerosing cholangitis, diagnosed by liver biopsy at the age of eleven. He had a history of esophageal atresia from birth that was corrected surgically. Written informed consent was obtained from the
patient. He presented to the hospital at the age of 12 with a case of liver mass associated with jaundice, fever, and an increase in liver enzymes, with no evidence of autoimmune sclerosing cholangitis. He was treated with ursodeoxycholic acid (250 mg) and multivitamin supplementation. The treatment was continued after radiological and laboratory investigations. The laboratory results were as follows: the liver function assays recorded included alanine aminotransferase (130 U/L), aspartate aminotransferase (89 U/L), bilirubin (22 μmol/L), gamma-glutamyl transferase (164 U/L) and international normalized ratio (1). A complete blood count was normal with a white blood count of 4.8, hemoglobin of 144, and platelet count of 135. An ultrasound showed mild right and left hepatic lobe intra- and extrahepatic biliary duct dilatation with no masses or stones (Fig. 1,2,3).

Figure 1 20-year-old male presenting with hepatomegaly. Grayscale ultrasound image of the abdomen demonstrates the visualized part of the upper midline with an unremarkable appearance.

The intravenous contrast-enhanced abdominal CT (Fig. 4) revealed an abnormally oriented dysmorphic liver; stable diffuse biliary duct dilatation, mainly in the left up to the porta hepatitis, caused by a twisted right lobe and intrathoracic herniation; patent portal veins with a small left portal vein; a solitary hepatic vein draining to the prehepatic infe-
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Figure 2 20-year-old male presenting with hepatomegaly. Grayscale ultrasound image demonstrates the right hepatic lobe with mild increased echogenicity with a smooth border. There was no suspicious focal liver lesion. A mild intrahepatic duct dilatation reaches up to 0.2 cm (arrows). The gallbladder is not visualized.

The patient underwent radiographic assessment of the biliary tract through percutaneous transhepatic cholangiography (Fig. 5). This shows dilated left and right bile duct radicles with abnormal configuration and orientation, likely due to the twisted liver. However, the contrast was drained smoothly via a normal Common Bile Duct (CBD) into the small bowel lobes. Moderate dilatation of the right and left bile duct radicles was noted, with no evidence of complete obstruction. This finding was likely due to the abnormal con-
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Figure 3 A 20-year-old male presenting with hepatomegaly. Color-flow Doppler image demonstrates that the portal vein has normal direction flow with a velocity of 26.4 cm/S. The Common Bile Duct (CBD) measures 0.2 cm.

Figure 4 Hepatomegaly with hepatic lobe torsion. Coronal, sagittal, and axial (A, B, C). Enhanced CT after contrast media injection shows partially herniated and rotated liver (A, arrowhead) with no signs of cirrhosis. Interrupted Inferior Vena Cava (IVC) along with a continuation of the left hemiazygos vein is seen (C, white arrow). The remaining hepatic vessels appear patent (B, arrows).
figuration and orientation of the liver, possibly related to the right diaphragmatic hernia, in addition to twisting of the liver.

Figure 5  A 20-year-old man with sclerosing cholangitis, dilated bile ducts (arrows), right diaphragmatic hernia (arrowhead), status post esophageal atresia repair.

MRCP (Fig. 7) found cystic intrahepatic biliary duct dilatation with alternating narrowing and dilatation, giving a beaded appearance that represented a previously diagnosed primary sclerosing cholangitis. A follow-up with abdominal MRI was recommended.

Figure 6  A 20-year-old male presented with hepatomegaly. Coronal, axial (D, E) T2- fat sat- weighted images and coronal (F) 3D MRCP demonstrate persistent smooth narrowing of the upper Common Hepatic Duct (CHD)/hepatic confluence suggestive of fibrotic process (F, arrowhead), without marked biliary irregularity to suggest primary sclerosing cholangitis (F, arrows). Persistent mild to moderate intrahepatic biliary dilatation is seen (E, arrow).

MRI scans revealed an infra-hepatic interruption of the Inferior Vena Cava (IVC) and hemiazygos continuation. An unusual hepatic anatomical position was noted. The left hepatic lobe and gallbladder were mispositioned above the diaphragm and through a right hemi-diaphragmatic defect (central diaphragmatic hernia) with a mild progressive...
enhancement, suggesting fibrosis with no stigmata of cirrhosis. As a result, the decision was made to perform an ultrasound-guided liver biopsy after 9 months. The biopsy discovered chronic cholangiopathy with ductular proliferation and early bridging fibrosis, but no significant plasma cell infiltration or cirrhosis was noted. Therefore, the decision was made to increase the patient's ursodeoxycholic acid to 500 mg b.i.d. and to start Inderal (propranolol) for his portal hypertension at a dose of 20 mg b.i.d. The patient was also given 1 tablet of multivitamins daily.

In conclusion, the patient was diagnosed with a chronic liver disease that could be related to either cholangitis or chronic cholangiopathy. However, the possibility of genetic disease, including congenital hepatic fibrosis, was also raised. At that stage, there was no indication for the need for a liver transplantation. However, the patient needed to have follow-up appointments at the clinic every 3–6 months to assess for any disease progression and manage any complications.

2. DISCUSSION

First, we concede that liver torsion is a very rare condition, and diagnosis of it based on CT imaging is challenging if the condition is not considered in the list of differential diagnoses from the start. Second, the typical population diagnosed with the condition is typically above 40 years old with female predominance. Patients usually present with right upper quadrant abdominal pain associated with nausea and vomiting. Third, most of the cases are eventually require a surgical hepatectomy, and hence the surgical teams' physicians should be prepared to perform an operative complex resection. If patients with the condition are identified and then undergo appropriate operative management, there is a decent chance for recovery [3].

Primary sclerosing cholangitis is categorized as a rare disorder which is manifested by a progressive liver disease involving strictures of the multifocal bile ducts [5]. It causes severe conditions, such as malignancies, cirrhosis, and fibrotic liver disease [6]. In the present report, we reviewed similar cases in the literature; such cases of hepatic torsion are extremely rare.

We found most of the published literature report cases of accessory hepatic lobe torsion. Most of these had occurred in situations of previous repair of omphalocoeles. A history of trauma and surgery involving the abdominal wall with the new development of acute symptoms such as abdominal pain in patients with an accessory liver lobe should highly suggest liver torsion. This should subsequently allow for a rapid transition to surgery to attempt to repair the torsion with the segments that probably become ischemic [1].
Liver torsion with infarction are extremely rare causes of surgically acute abdomen. However, it raises a high amount of suspicion when addressing unusual abdominal presentations [7]. Biliary obstruction and, in the worst scenarios, torsion of the liver can present with the liver and is enclosed in the chest [8].

An accessory liver lobe may be demonstrated on imaging as an undetermined mass. The connection of the visualized anatomical accessory lobe with the liver may be discovered by using imaging modalities such as CT or MRI scans. If the biliary drainage is independent beside the arterial, venous, and portal branches, it should raise suspicions of this rare condition.

In the case of hepatic torsion, the vascular pedicle is not visualized in such cases, making the subsequent identification of it difficult. Vascularization, loss of normal parenchymal architecture of the liver, and acute inflammatory conditions (e.g., cholecystitis and appendicitis) are usually easily detectable on enhanced CT studies with high sensitivity [9]. Regarding ultrasound scanning, we found that a twisted accessory liver lobe appears as a hypoechogenic mass because of congestion. With the Doppler ultrasound technique, it presents as an absence of the vascularity of the normal supplying blood vessels. Accessory liver lobe is a congenital anomaly which is considered rare, and it is challenging to prove the diagnosis by the clinical symptoms and radiological investigations [10].

Doppler ultrasonography is useful for assessing liver perfusion and detecting an acute outflow obstruction of the hepatic veins. CT scanning of the abdomen is proven as a definitive method for making diagnosis in similar situations [11]. Furthermore, in such patients, laboratory investigations are necessary to define the underlying, specific primary cause and it should be performed. The definitive diagnosis is made depending on the surgical exploration in most cases. Surgical intervention at the appropriate time means the patient can avoid subsequently possible morbidity and mortality [7]. Liver biopsies in patients diagnosed with PSC established by cholangiography may not be routinely necessary [8]. The combined effective therapies for primary sclerosing cholangitis are mandatory, which include anti-inflammatories, anticholestatics, and antifibrotic compounds. These halt the process of the disease's progression and increase the patient's survival rate [11]. For advanced disease, excellent outcomes may be achieved after liver transplantation [10].

Based on our knowledge, the primary sclerosing cholangitis disease with liver herniation into the chest representing a liver torsion and biliary outflow obstruction has not been reported in the available literature.

This is a rare case in which the patient was in a typical status until he later presented with jaundice and no acute symptoms. A chest x-ray revealed a small right lung volume and thoracic cage deformity, with peribronchial wall thickening of the left lung, and this
considered a rare entity of liver non traumatic right sided diaphragmatic hernia [12].

Ultrasonography and an MRI cholangiogram suggested mild to moderate intrahepatic biliary duct dilatation, consistent with sclerosing cholangitis. It was thus possible to make a diagnosis of liver torsion. After MRI scanning, we found hepatic torsion causing biliary compression, excluding other possible causes, such as extrahepatic biliary atresia and a choledochal cyst. Another difficulty was faced after management decided regarding the possibility of liver transplantation.

There are highly achievable points of learning acquired from this case. The detection of part of the liver in the chest may lead to biliary compression and, in the worst-case scenarios, torsion of the liver. Prompt identification through detailed history-taking and proper clinical physical examination is necessary, as is deciding on the appropriate imaging modality for investigation, such as CT and MRI scanning. Stable patients need follow-up laboratory and radiological examinations every 3–6 months to monitor disease progression for liver transplant assessment.

3. CONCLUSION

When the liver presents with an unusual appearance, a strong suspicion should be maintained for uncommon causes of acute abdomen. In such situations, extensive laboratory or medical imaging modalities must be conducted for the investigation. In many cases, surgical exploration requires a diagnosis. Despite its rarity, the differential diagnosis of acute abdominal pain should consider hepatic torsion as one of the provisional diagnostic susceptibilities, and accurate imaging should be performed to assist surgical teams in planning complex surgical operations.

CONFLICT OF INTEREST

The authors declare that the research was conducted without any commercial or financial relationships that could be construed as a potential conflict of interest.

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REFERENCES


