

CHINESE MEDICAL SCIENCES JOURNAL

CASE REPORT

Hepatocellular Carcinoma with Hypersplenic Thrombocytopenia and Situs Inversus Totalis: A Case Report

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Key words: situs inversus totalis; hepatocellular carcinoma; hypersplenic thrombocytopenia

Chin Med Sci J 2016; 31(2):134-136

SITUS inversus totalis (SIT) is a complete mirror image of the thoracic and abdominal viscera, occurring at an incidence of 1 in 5000-20 000 live births.^{1,2} It is supposed to originate from an abnormal rotation of the cardiac tube during embryogenesis. Although SIT is a congenital anomaly, most of patients are detected accidentally at the time of radiological investigation. Hepatocellular carcinoma (HCC) is usually associated with liver cirrhosis and portal hypertension in Chinese. Presence of hypersplenic thrombocytopenia is a common consequence of long-term portal hypertension in cirrhotic patients. We report a case of HCC with hypersplenic thrombocytopenia, which was successfully treated by splenectomy and liver resection.

CASE DESCRIPTION

A 45-year-old man with epigastric pain was admitted to our hospital in November 2009 for the treatment of suspected HCC in the left lobe of the liver. He had

presented with epigastrium unwell for nearly one month. There was no prior history of diabetes, hypertension, abdominal surgery or trauma. Physical examination showed no generalized lymphadenopathy existed. He did not present with symptoms of fever and jaundice. The apex beat was in the right fifth intercostal space, midclavicular line within 2 cm. Nothing abnormal was found in heart auscultation and the respiratory system. The abdomen was soft with little tenderness, but no rebound tenderness. The liver was enlarged with its edge 2 cm below xiphoid. The spleen was 7 cm below the right costal margin with hard texture. Nervous system did not show dysfunction.

The laboratory findings showed hemoglobin of 14.8×10^3 kg/L, white blood cells of $2.84 \times 10^3/\text{mm}^3$, and blood platelets of $35 \times 10^3/\text{mm}^3$. Serum alanine aminotransferase, serum aspartate aminotransferase, serum bilirubin, and total protein were all within normal limits. Hepatitis B surface antigen (HBsAg), hepatitis B e antigen (HBeAg), and hepatitis B core antibody (HBcAb) tests were positive and serum α -fetoprotein (AFP) was elevated to 400 ng/ml. Magnetic resonance imaging presented a distinct liver tumor with about 7 cm in diameter as well as an enlarged spleen (Fig. 1).

The patient was operated under general anesthesia with inverted L subcostal incision and the diagnosis of SIT was confirmed at laparotomy. The liver showed micronodular cirrhosis and a mass in segment 2 and 3. The size of spleen was about 17 cm×13 cm×6 cm. So we conducted the liver resection and a concomitant splenectomy (Fig. 2). The surgery was uncomplicated with a blood loss of 300 ml and an operation time of 3 hours and 20 minutes. No blood product transfusion was required. Histologic examination of the specimen showed HCC and chronic congestive splenomegaly (Fig. 3). The postoperative course was uneventful. The patient was doing well without recurrence for 3 years after the operation and symptoms of splenomegaly and hypersplenism were obviously relieved.

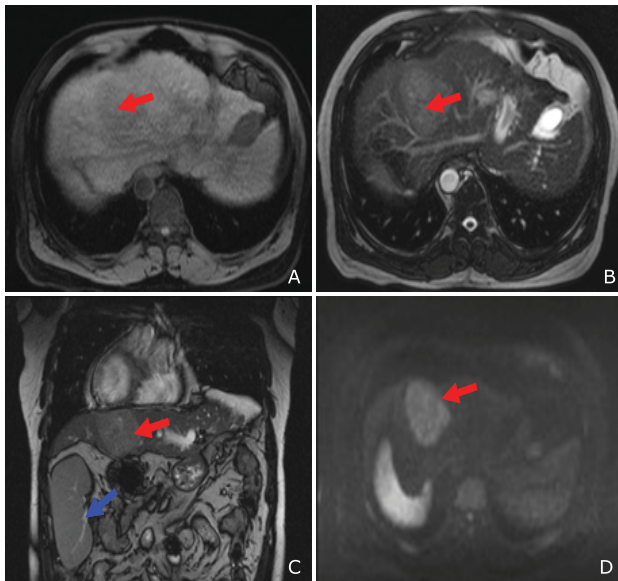


Figure 1. Magnetic resonance imaging of abdomen: a liver tumor (red arrow) and an enlarged (blue arrow) spleen with situs inversus totalis.

A. T1-weighted image; B. T2-weighted image; C. cross-sectional coronal image; D. diffusion weighted image.

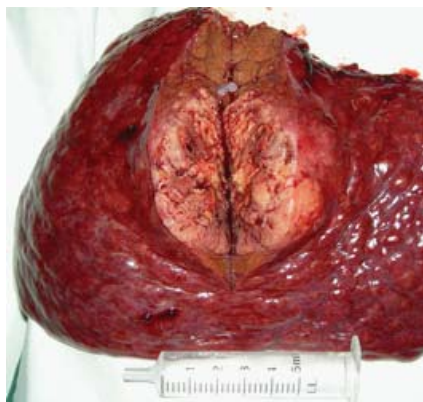


Figure 2. The open resected specimen with liver tumor.

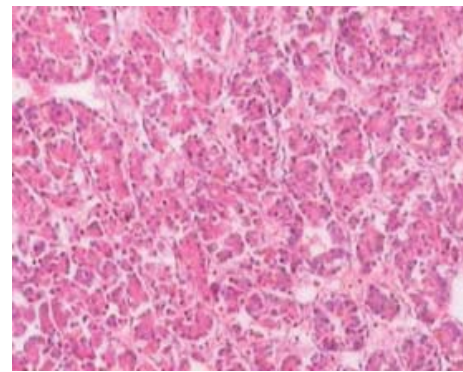


Figure 3. Postoperative pathology confirms hepatocellular carcinoma. HE staining, ×100

DISCUSSION

SIT is a rare congenital condition, in which there is a mirror-image transposition of both the abdominal and thoracic viscera. In 1600, the first case of situs inversus in humans was reported.³ Most of patients with SIT are detected accidentally at the time of radiological investigation, however, about 20% to 25% of patients present with Kartagener's syndrome⁴ and 3% to 5% of patients have congenital heart disease.⁵ Currently, no evidence shows that situs inversus patients have an increased risk of malignancy.⁶

Chronic hepatitis B virus and hepatitis C virus infection are the most common risk factors of HCC. More than one hundred million chronic hepatitis B patients live in China.^{7, 8} HCC is usually associated with liver cirrhosis and portal hypertension. The platelet count of this patient was less than $50 \times 10^3/\text{mm}^3$ caused by secondary hypersplenism. Splenectomy is suggested for the treatment of secondary hypersplenism and thrombocytopenia.⁹ Although splenectomy may induce thrombosis, liver failure, subsequent mortality, and high operative risk,¹⁰ the results of most reports¹¹⁻¹³ conducted that concomitant splenectomy in liver resection for HCC patients was encouraging. According to the Glasgow prognostic score for the operability of hepatectomy,^{14, 15} we considered a concomitant splenectomy was possible in our case.

In 1983, Kanematsu *et al*¹⁶ described the first case of a primary HCC associated with situs inversus and only 4 papers of HCC patients with SIT have been reported in Pubmed from 2000.¹⁷⁻²⁰ Transcatheter arterial embolization was performed in a HCC patient with SIT by Niki *et al*¹⁷ in 2004. Kakinuma *et al*¹⁸ reported a case of HCC with SIT treated by hepatectomy. Li *et al*¹⁹ carried out transhepatic arterial chemotherapy and embolization in a HCC patient with SIT and polysplenia syndrome. Patel *et al*²⁰ performed a right hepatectomy with the help of electrocautery in a

HCC patient with SIT. Our patient is the first reported case of HCC with SIT who underwent liver resection and splenectomy.

In our opinion, precise preoperative anatomical evaluation and good adaptation to the mirror-image anatomy are essential for successful treatment. Reversing the positions of the operator and the assistant during surgery may be a good way, however, careful and cautious assessment of abnormalities by preoperative examinations play a key way to the surgery. Hepatic resection and concomitant splenectomy in patients with SIT are recommended as long as liver function within the safe range for surgery.

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