

## ORIGINAL ARTICLE

# Complete hepatic ischemia due to torsion of a large accessory liver lobe: first case to require transplantation

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accessory liver lobe, anatomical variation, liver transplantation.

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## Summary

Anatomical abnormalities of the liver are extremely rare. Although the majority of cases with an accessory liver are not detected, it can give rise to various clinical symptoms like recurrent abdominal pain and impaired liver function. Here we present the first case of orthotopic liver transplantation in a patient with hepatic ischemia caused by complete vascular occlusion due to a twisted accessory liver lobe. Although rare, an accessory liver lobe may cause serious and life-threatening problems and should therefore be kept in mind in patients presenting with acute abdominal pain.

## Introduction

An accessory liver is a rare anomaly in humans and in most cases an incidental finding at laparotomy, thus it is unlikely to be diagnosed before surgery. Such ectopic liver tissue, with its own mesentery, lacks the anatomical fixation by the liver ligaments and stability of the normal liver, rendering it, especially when huge in size, susceptible to rotation or even torsion. However, an accessory liver lobe might be an uncommon but serious cause of various clinical symptoms such as acute abdominal pain and impaired liver function [1,2].

Here we present the first case of orthotopic liver transplantation in a patient with hepatic ischemia caused by complete vascular occlusion due to a twisted accessory liver lobe.

## Case report

A 19-year-old woman with a history of umbilical cord hernia and left-side renal agenesis presented with acute

abdominal pain, nausea and vomiting. Physical examination revealed a painful tumor in the right mid-abdomen. On admission transaminases were elevated to more than four times normal, but bilirubin and coagulation parameters were still within normal range. Portal vein thrombosis and infarction of a large accessory lobe with inhomogeneous contrast enhancement of the main liver were discovered by CAT scan (Fig. 1). The patient's condition and liver function rapidly deteriorated.

Laparotomy performed at our center, 20 h after the onset of symptoms and initial management at an outside district hospital, revealed a large accessory liver lobe (Fig. 2) with the gallbladder embedded, weighing 1000 g. This accessory lobe, corresponding to an isolated segment I.V. with its own mesentery and hepatic vein, was twisted and completely infarcted. The mesentery pendulated from the hepatoduodenal ligament, and the vascular structures of the liver and the common hepatic duct ran through the base of the accessory lobe. Torsion of the lobe caused strangulation of the vascular supply to the main liver with consecutive portal vein thrombosis



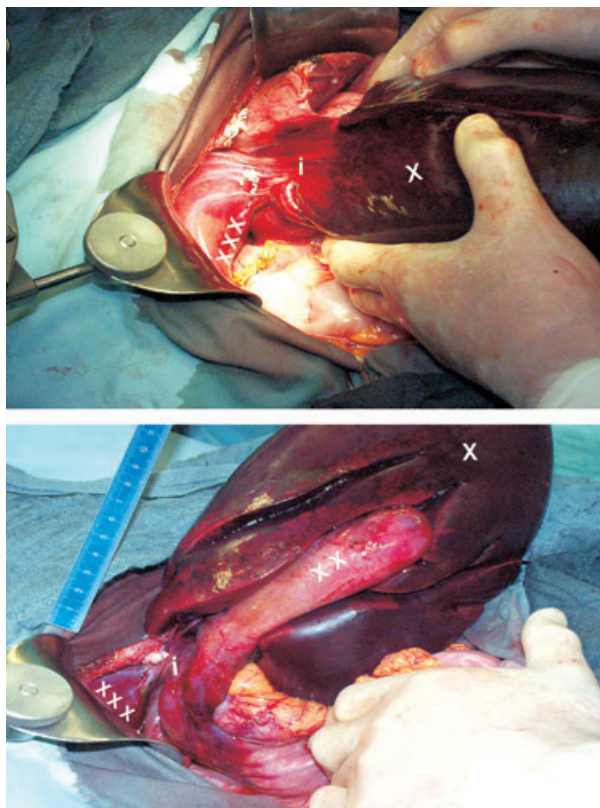
**Figure 1** (a) Preoperative CT scan (coronary reconstruction) showing infarction of a large accessory lobe (x) with inhomogeneous contrast enhancement of the main liver (xx). (b, c) Transverse scans at different indicated levels.

and complete hepatic ischemia. Resection of the necrotic accessory liver, reconstruction of the hepatic artery and bile duct and portal vein thrombectomy were performed.

After surgery transaminases and lactate decreased, whereas coagulation parameters remained at the low preoperative level. As coagulation was already impaired, the patient did not receive any further anticoagulation therapy or vasoactive drugs, following resection of the twisted liver lobe. At day 2 a sharp rise in transaminases and decrease in clotting factors were observed. The patient became encephalopathic, and the single kidney ceased to function. Doppler ultrasound revealed decreased intrahepatic portal flow, suggesting severe ischemic damage. The patient was listed for high-urgency liver transplantation and received a graft from an ABO-compatible 37-year-old donor the same day. Because of persistent anuria hemodialysis was required until day 11 after transplantation. Apart from acute renal failure, the postoperative course was uneventful and the patient discharged with normal liver and renal function on day 21 following transplantation.

## Discussion

Abnormalities of the liver are rare and generally without clinical relevance. Liver tissue adjacent to the liver with its own mesentery is defined as an accessory liver, as in our case, whereas parenchyma completely detached from the liver is called an ectopic liver [3,4]. Fairly common is the so-called Riedel's lobe, a downward tongue-like projection of the right liver lobe, which is not a true accessory lobe, but an anatomical variation [3]. Sometimes congenital anomalies of the liver, such as agenesis of the caudate lobe and lobulation of the right liver lobe, are associated with abnormalities such as umbilical hernia, omphalocele and bile duct cyst [5]. Our patient had a history of umbilical cord hernia and agenesis of the left kidney. Two similar cases, both with a hernia of the umbilical cord, which contained the gallbladder embedded in an accessory liver lobe, have been described in newborns [6]. The majority of cases with an accessory liver are not detected clinically, but noted incidentally at surgery or necropsy. In this case the accessory liver lobe had not been realized by the surgeons, when umbilical



**Figure 2** Intraoperative situs showing the large, completely infarcted and twisted accessory liver lobe (x) with the embedded gallbladder (xx) and its own mesentery (i); main liver (xxx).

hernia repair was performed, and was therefore not known to our patient. Nevertheless, depending on size and location, the accessory lobe can give rise to various symptoms such as recurrent abdominal pain [1,7] and fluctuating impaired liver function due to torsion [8] or bleeding after blunt abdominal trauma [2]. However, as

of January 2004 only 20 cases of an accessory liver lobe producing clinical symptoms had been reported [2,9]. Strangulation of the vascular supply to the liver due to the twisted mesentery of the large accessory lobe has rarely been reported in the literature. It is difficult to say whether earlier intervention would have saved the liver. Nevertheless, although rare, an accessory liver lobe may cause severe problems and should therefore be kept in mind when patients present with acute abdominal pain and impaired liver function.

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