

A rare case of combined hypoplasia of the right lobe of the liver and a persistent right umbilical vein

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SUMMARY

Developmental anomalies in the biliary tract occur mainly in the extra hepatic part and its vascular tree. The liver itself is usually free from major structural anomalies. Anomalies of the hepatic lobes are rare. Alterations in umbilical venous anatomy, especially the persistence of a right umbilical vein is considered rare. However, with the advent of ultrasonography and its use in prenatal diagnosis, its incidence has been increased. Here we report a case of hypoplasia of the right lobe of liver associated with a persistent right umbilical vein and discuss its clinical significance.

Key words: Liver – Hypoplasia of right liver lobe – Right umbilical vein

INTRODUCTION

The liver is anatomically subdivided into a large right lobe and a smaller left lobe by the attachment of the falciform ligament and the fissures for the ligamentum teres and ligamentum venosum. The right lobe presents a caudate lobe and a quadrate lobe, separated by the porta hepatis. The inferior vena cava is to the right of the caudate lobe and the gall bladder is to the right of the quadrate lobe. The oesophagus and stomach are related to the left lobe. The right kidney,

right suprarenal gland, right colic flexure, the second part of the duodenum and the diaphragm are related to the right lobe.

During early development, the right and left umbilical veins are formed. Soon, the right umbilical vein disappears but the left vein functions and, after birth, is converted into the ligamentum teres.

Cases of agenesis and hypoplasia of the right lobe of the liver and cases of persistent right umbilical vein had been reported. Here we report a case of hypoplasia of the right lobe and persistence of a right umbilical vein in the same liver.

MATERIALS AND METHODS

During routine dissection of the abdomen of a 65 year old female cadaver by the medical students in our anatomy department, a liver with a large left lobe was found. Its relations were noted. The liver was removed and its surface features were examined.

OBSERVATIONS

The right lobe was hypoplastic and small while the left lobe of the liver was hyperplastic and large. The gall bladder was shifted to the right. The right kidney was higher than the left and the left kidney was lobulated (Fig. 1).

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Figure 1.- Posterior abdominal wall, showing the left kidney, at a lower level compared to the right kidney. RK: Right kidney; LK: Left kidney; C: Coeliac trunk.

The right lobe of the liver was extremely small. Costal impressions were present on the right lateral surface. The ligamentum teres ascended in a groove on the anterior surface, coursed to the right, and then ran in the fissure for the ligamentum teres on the inferior surface of the liver. It thus surrounded a small lobule (Fig. 2). The left lobe had prominent impressions caused by the oesophagus, stomach, transverse colon, and the first part of duodenum. The quadrate lobe was narrow, with a width of 0.5-1.0 cm (Fig. 3). The gall bladder was 8.5 cm long and 5 cm wide. The right lobe had impressions caused by the gall bladder and right suprarenal gland but not by the right kidney.

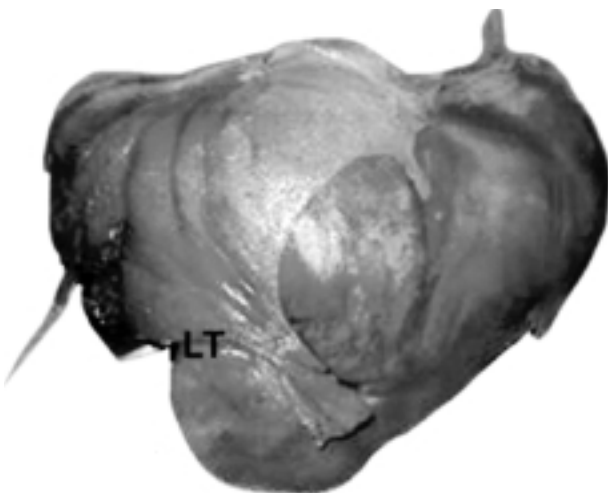


Figure 2.- Diaphragmatic surface of the liver. LT: Ligamentum teres.

To the right of the fossa for the gall bladder was a fissure, which housed the obliterated right umbilical vein (a ligament), extending from the um-

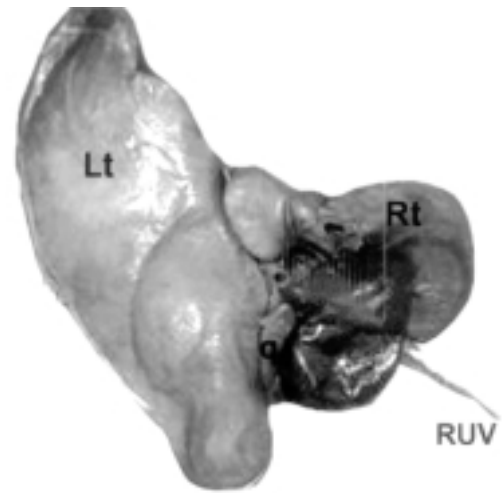


Figure 3.- Visceral surface of the liver. Lt. Left lobe of liver; Rt: Right lobe of liver; q: Quadrate lobe; RUV: Right umbilical vein.

bilicus towards the right branch of the portal vein, deep to the gall bladder (Fig. 3). Intrahepatically, the obliterated right umbilical vein was accompanied by a branch of the right hepatic artery. The portal vein was dilated and was 1.5 cm in diameter.

DISCUSSION

Congenital anomalies of the lobes of the liver are rare. The left lobe is distinctly larger in the infant than in the adult (Symington, 1914). Hypoplasia and agenesis occur more often in the left lobe (Gathwala and Sen, 2003). Agenesis of the right lobe is a rare anomaly; only 42 cases had been reported before (Gathwala and Sen 2003). Persistence of right umbilical vein had traditionally been considered to be extremely rare. Later evidence casted doubt on the veracity of this contention. Earlier these anomalies were discovered during laparotomy, autopsy, or dissection, but recently, they have been noticed frequently during routine ultrasound, computerized tomography, angiography etc. The imaging findings described included the absence of the right hepatic lobe, several degrees of enlargement of the medial and lateral segments of the left lobe and caudate lobe of the liver, the presence of a retro- and suprahepatic gall bladder, and a persistent right umbilical vein.

To add to the 25 cases of agenesis of the right lobe of the liver, Radin et al. (1987) reported 5 more cases and described the alteration in hepatic lobar morphologic characteristics. Agenesis and atrophy of the right hepatic lobe has been reported (Radin et al., 1987; Demurci et al., 1990; Kakistubata, 1991; Kanematsu, 1991; Morphett, 1992; Chou et al., 1998; Gathwala and Sen, 2003).

Agenesis of the right lobe of the liver can be diagnosed in computerized tomography by a retro- or suprahepatic gall bladder and the complete absence of the right hepatic vein, the right portal vein and its branches, and dilated right intrahepatic ducts. In severe lobar atrophy, at least one of these structures is recognizable. The absence of the right lobe occurs slightly more often in males and may be accompanied by other associated problems, such as portal hypertension, a suprahepatic gall bladder and hypertrophy of the left lobe (Radin et al., 1987; Gathwala and Sen, 2003).

A persistent right umbilical vein was detected sonographically in 17 out of 8950 low-risk fetuses (1:526) (Wolman et al., 2002), and other reports have also been published (Blazer et al., 2000; Miannay et al., 1999; Amy et al., 1999; Lai, 1998; Shen et al., 1996; Kirsch et al., 1996; Ariyuki et al., 1995; Hill et al., 1994; Jeanty, 1990; Ricklan et al., 1988; Bell et al., 1986; Theander and Karisson, 1978; Weinstein and Goldstein, 2002; Wolman et al., 2002).

Although in a few cases of persistent right umbilical vein associated anomalies of the cardiovascular, gastrointestinal, genitourinary, and musculoskeletal systems were present, in general the cases had favourable outcomes.

Our report is the first case of combined hypoplasia of the right lobe of the liver and a persistent right umbilical vein. Thus, radiologists need to bear this possibility in mind. The fact that an individual with these combined anomalies survived for 65 years shows that the outcome was favourable. However, when these anomalies are noted, a detailed evaluation must be carried out for possible associated cardiac, venous, aortic, gastrointestinal, genitourinary congenital malformations and a single umbilical artery, abdominal visceral situs inversus, hypospadias, phocomelia, portal hypertension and diaphragmatic hernia.

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