Coexistence of multiple anomalies in the hepatobiliary system

Seyed Hadi Anjamrooz¹, Hassan Azari²
¹Department of Anatomical Sciences, School of Medicine, Kurdistan University of Medical Sciences, Sanandaj, ²Department of Anatomical Sciences, School of Medicine, Shiraz University of Medical Sciences, Shiraz, Iran

Abstract: The co-occurrence of several anomalies in the hepatobiliary system is uncommon. In the present study, hepatic lobe anomalies occurring in combination with hepatic artery and biliary variations were observed in an adult male cadaver. There are no previous reports in the literature on the coexistence of such anatomical variations. Preoperative diagnosis of such coexisting anomalies is very difficult. Hence, a thorough knowledge of these variations will enable surgeons to select the most appropriate hepatobiliary surgical procedure and postoperative management.

Key words: Liver, Hepatic artery, Gallbladder, Cystic duct

Received January 2, 2012; Revised February 19, 2012; Accepted March 9, 2012

Introduction

The liver and extrahepatic bile ducts comprise the hepatobiliary system. This system is involved in the production, storage, transport, and release of bile, which plays an important role in the metabolism of carbohydrates, proteins, and fats. The liver is located in the right hypochondriac and epigastric regions, behind the lower ribs. Attachment of the falciform ligament onto the anterosuperior surface splits the liver into 2 anatomic lobes (the right and left lobes). Two additional smaller lobes (the quadrate and caudate lobes) are visible on the visceral surface of the liver. The liver is supplied with blood from 2 sources, namely, the hepatic artery proper and the portal vein [1].

The hepatic artery proper originates from the common hepatic artery and extends to the porta hepatitis alongside the portal vein and the common bile duct, where it bifurcates into the right and left hepatic arteries. The extrahepatic bile system is divided into 4 topographic portions: the gallbladder and cystic duct, the right and left hepatic ducts, the common hepatic duct, and the bile ducts [2].

For successfully performing modern surgery on the hepatobiliary system, thorough knowledge of its components is required. Hence, information on the possible anatomical variations that may occur in the liver, gallbladder, and bile ducts as well as in their associated arteries is important to surgeons.

The present case included several liver lobe anomalies along with hepatic artery and bile duct variations that have not been reported previously in the literature. Comprehensive knowledge of these abnormalities is vital for determining the technical feasibility of surgical interventions and for performing complication-free surgery in this region.

Case Report

During routine dissection in an anatomy laboratory, cadaver of an adult Iranian man showed visceral and vascular anomalies (Table 1) with no indications of previous abdominal operations. A midline abdominal incision was
made for adequate access, and the liver lobes, gallbladder, cystic duct, common bile duct, portal vein, and hepatic artery were assessed.

The vertical size of the liver was significantly greater than the horizontal transverse size. The lower end of the liver, which was at the level of the inferior pole of the right kidney, was located at the level of the L2-L3 intervertebral discs. The lower pole of the right kidney was located approximately 33 mm higher than that of the left kidney. In addition, the right kidney and suprarenal gland were positioned posterior to the right hepatic lobe. The right hepatic lobe was elongated downward and created a very deep right hepatorenal pouch (Fig. 1). Hypoplasia was observed in the right, quadrate, and caudate lobes of the liver; in particular, the posterior region of the right lobe was abnormally small. A short fissure was observed on the visceral surface of the right hepatic lobe near the neck of the gallbladder. The caudate process of the caudate lobe was posterior to the porta hepatis, and the quadrate lobe was deformed, with a papillary protrusion extending toward the porta hepatitis (Fig. 2).

Macroscopically, the gallbladder appeared to be normal. However, the fundus of the gallbladder was embedded in the hepatic visceral surface and did not reach the anterior abdominal wall. In addition, Hartmann’s pouch was absent from the neck of the gallbladder. The common hepatic duct was unusually short, and the cystic duct joined the common hepatic duct at a point immediately after the junction of the left and right hepatic ducts (Fig. 3).

Table 1. The observed abnormalities in the gross morphology of the hepatobiliary system

| Anatomical structure | Presence of anomaly | Type of anomaly                           |
|----------------------|---------------------|-------------------------------------------|
| Liver (overall shape) | +                   | Deformation                               |
|                      |                     | Increased vertical size                   |
|                      |                     | Decreased horizontal transverse size      |
| Right lobe           | +                   | Hypoplasia                                |
|                      |                     | Presence of Riedel’s lobe                  |
|                      |                     | Presence of a short fissure               |
| Left lobe            | +                   | Hypoplasia                                |
| Quadrate lobe        | +                   | Hypoplasia                                |
|                      |                     | Presence of a papillary protrusion        |
|                      |                     | Deformation                               |
| Caudate lobe         | +                   | Hypoplasia                                |
|                      |                     | Deformation                               |
|                      |                     | Abnormal location of the caudate process  |
| Gallbladder          | +                   | Abnormal location                         |
|                      |                     | Absence of Hartmann’s pouch               |
| Cystic duct          | -                   | None                                      |
| Common hepatic duct  | +                   | Short                                     |
| Right hepatic duct   | -                   | None                                      |
| Left hepatic duct    | -                   | None                                      |
| Portal vein          | -                   | None                                      |
| Hepatic artery proper| +                   | Presence of a supernumerary artery        |

Fig. 1. Photograph obtained during the dissection of the abdomen, showing the liver and right kidney. The liver lies anterior to the right kidney and suprarenal gland. The downward elongation of the right hepatic lobe created a deep hepatorenal pouch. CL, caudate lobe; GB, gallbladder; HRP, hepatorenal pouch; LHL, left hepatic lobe; QL, quadrate lobe; RHL, right hepatic lobe; RK, right kidney; RL, Riedel’s lobe.

Fig. 2. Photograph of the visceral surface of the liver showing the unusual location of the fundus of the gallbladder, hypoplasia and deformation of the hepatic lobes. CL, caudate lobe; CP, caudate process; FL, falciform ligament; GB, gallbladder; LHL, left hepatic lobe; PP, papillary process; PV, portal vein; QL, quadrate lobe; RHL, right hepatic lobe; RI, renal impression; RL, round ligament; SF, short fissure; SI, suprarenal impression.
A double hepatic artery proper entered the liver at the porta hepatis. One hepatic artery was positioned to the left of the bile duct, and the other was posterior to the bile duct (Fig. 3). These arteries had completely separate origins.

Discussion

There are no reports in the literature on the complex anatomical variants observed in the present study; however, a few similar variants have been reported.

For example, Demirci et al. [3] observed agenesis of the right liver lobe on computed tomography (CT) images. Yano et al. [4] observed downward elongation of the hepatic lobe or Riedel’s lobe in a Japanese woman. Pamidi et al. [5] reported a case in which the quadrate lobe of the liver was small and had almost separated from the liver, except at its posterior end. Han and Soylu [6] described an unusual case of a 26-year-old woman with an accessory liver lobe in the thoracic cavity. However, none of these studies reported on the coexistence of several hepatic lobe anomalies like those observed in the present case. The term “deformed liver” could be used to describe the present case, and it may be postulated that its occurrence resulted from uninhibited further ascension of the right kidney during embryonic development.

Congenital agenesis affects the left hepatic lobe to a greater extent than the right hepatic lobe [3]. However, in the present case, hypoplasia of the right hepatic lobe was more pronounced.

Lobar hypoplasia or aplasia is defined as incomplete congenital development or the absence of a hepatic lobe; however, it is not always of congenital origin.

Postnecrotic cirrhosis, chronic hepatic infection, choledochoolithiasis, idiopathic portal hypertension, malnutrition, biliary obstruction, veno-occlusive diseases, Caroli’s disease, cholangiocarcinoma, and hepatotoxic agents have been associated with atrophy or hypoplasia of a hepatic lobe or segment; however, these abnormalities can also occur before fulminant hepatitis and surgical resection [7, 8].

Liver lobe anomalies may be accompanied by abnormal conditions of the gallbladder. The section of the gallbladder that extends beyond the inferior border of the liver is known as the fundus; however, in the present case, the fundus was located on the undersurface of the liver, which has not been reported previously.

The region of maximum tenderness in acute cholecystitis is located where the fundus of the gallbladder is in contact with the anterior abdominal wall, i.e., at the intersection of the outer edge of the right rectus muscle and the ninth costal cartilage. Hence, the observed gallbladder anomaly in this case may have weakened the symptom severity of this condition. Clinicians must be aware that congenital anomalies of the gallbladder and biliary tree are difficult to diagnose during routine preoperative studies.

The presence of several rare variations of the bile ducts and arterial supply of the hepatobiliary system adds another interesting aspect to the present report.

Talpur et al. [9] performed a laparoscopic study of the extrahepatic biliary system and identified short cystic ducts in 2.67% of their 300 patients. However, no cases of short common hepatic ducts were reported.

Jones and Hardy [10] encountered hepatic artery anomalies in 43% of 180 cadavers. However, this extensive study did not report the presence of 2 hepatic arteries of equal diameter like those observed in the presented case.

According to the hepatic artery classification proposed by Michels, there are 2 types of variants: type A, a replaced hepatic artery that acts as a substitute for the absence of the normal artery and type B, an accessory hepatic artery that serves as a smaller-diameter artery, which occurs in addition to the normal artery [11]. The variant hepatic artery observed in the present case is a type C artery according to this classification because the 2 arteries are of equal diameter and are
Hepatobiliary anomalies located at the porta hepatis.

At present, surgeons have no method for distinguishing between a replaced artery and an accessory artery; however, it is important to know that an aberrant artery may supply a whole liver lobe (replaced artery) or part of it (accessory artery) because every ligation could lead to liver lobe or segment necrosis.

This review indicates that although several studies have previously reported the anomalies in the hepatobiliary system, the complex variant pattern observed in the present case appears to be unique in the literature.

References

1. Skandalakis JE, Skandalakis LJ, Skandalakis PN, Mirilas P. Hepatic surgical anatomy. Surg Clin North Am 2004;84:413-35.
2. Chen WJ, Ying DJ, Liu ZJ, He ZP. Analysis of the arterial supply of the extrahepatic bile ducts and its clinical significance. Clin Anat 1999;12:245-9.
3. Demirci A, Diren HB, Selcuk MB. Computed tomography in agenesis of the right lobe of the liver. Acta Radiol 1990;31:105-6.
4. Yano K, Ohtsubo M, Mizota T, Kato H, Hayashida Y, Morita S, Furukawa R, Hayakawa A. Riedel’s lobe of the liver evaluated by multiple imaging modalities. Intern Med 2000;39:136-8.
5. Pamidi N, Nayak S, Vollala VR. Cystogastrocolic fold and associated atrophy of the gallbladder. Singapore Med J 2008;49: e250-1.
6. Han S, Soylu L. Accessory liver lobe in the left thoracic cavity. Ann Thorac Surg 2009;87:1933-4.
7. Suh HJ, Kim WT, Kim MY, Cho YK. Combined anomaly of the right hepatic lobe agenesis and absence of the inferior vena cava: a case report. Korean J Radiol 2008;9 Suppl:S61-4.
8. Aktan ZA, Savas R, Pinar Y, Arslan O. Lobe and segment anomalies of the liver. J Anat Soc India 2001;50:15-6.
9. Talpur KA, Laghari AA, Yousfani SA, Malik AM, Memon AI, Khan SA. Anatomical variations and congenital anomalies of extra hepatic biliary system encountered during laparoscopic cholecystectomy. J Pak Med Assoc 2010;60:15-6.
10. Jones RM, Hardy KJ. The hepatic artery: a reminder of surgical anatomy. J R Coll Surg Edinb 2001;46:168-70.
11. Michels NA. Blood supply and anatomy of the upper abdominal organs with a descriptive atlas. Philadelphia: J. B. Lippincott; 1955.