

Hypoplasia of the Left Portal Vein Territory of the Human Liver: A Case Study

Although reports of hypoplasia or absence of the liver of left lobe are not few, descriptions of the intrahepatic vessels are rare but valuable for discussion of the pathogenesis. The present report demonstrates a case of the left surgical lobe hypoplasia that is characterized by 1) the scar-like lobe with few parenchymal tissue and dilated bile ducts, 2) no Spiegel's lobe with the portal vein stuck to the inferior vena cava, 3) unusual configurations of the right hepatic vein and the 8th segmental portal vein branch, 4) the hepatic groove on S8, and 5) the trifurcation pattern of the portal vein primary division. According to the macroscopic and histological observations, we hypothesized that the secondary abnormal peritoneal fusion occurred in utero and/or during the postnatal growth, and that it involved the left portal vein and other adjacent structures, resulting in severe atrophy of the left surgical lobe.

Key Words : Abnormalities; Liver; Portal Vein; Hepatic Veins

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INTRODUCTION

Hypoplasia or absence of the left lobe of the liver has been found in more than 20 patients in the world. To the best of our knowledge, the first report was conducted by Arnold and Ashley-Montagu (1), and it was followed by Benz et al. (2) and Belton and vanZandt (3). Later, from 1984 until now, most cases were found in Japan under radiographic and/or ultrasound examinations (4), while a few cases were reported in Korea (5, 6). However, there is little information about the associated observations. The associated absence of the gallbladder was seen only in Saigusa et al. (7). Merrill (8) and Saigusa et al. (7) described the usual morphology of S4 and Spiegel's lobe in their specimen, but other reports had no descriptions as such. Detailed description of intrahepatic vessels was limited to that by Saigusa et al. (7).

The present case study aimed to describe the extra- and intrahepatic morphology of a left anatomical lobe hypoplasia to provide better understanding of the pathogenesis, as well as heterogeneity in those anomalous morphologies.

MATERIALS AND METHODS

We obtained an anomalous liver, 165 × 90 mm in size, 355 g in weight, from a Korean female cadaver in the dissection course for undergraduate anatomical education in Chonbuk University in 2002. She died at 59 yr old with stomach cancer. In the present report, terminology for the intrahepatic vessels was based on system of Couinaud (9).

Immunohistochemical and hematoxylin-eosin (HE) staining was conducted for fragments of the specimen after macroscopic observations. We chose 3 parts of the liver corresponding to the abnormal left anatomical lobe and quadrate lobe for consideration of the pathogenesis and S6 for the control. The vascular lumen of the middle hepatic vein tributaries in the left lobe was labeled by carbon particles for easy identification on histology. The 3 fragments were dehydrated, embedded in paraffin (melting point, 58°C) and processed into 10-15 μ m-thick sections. The primary antibodies used in the immunohistochemical evaluation were Dako-monoclonal anti-human alpha-smooth muscle actin (mouse IgG2a, kappa, clone No. 1A4, code No. M151, 1:50 solution in phosphate-buffered saline [PBS]) and Dako-polyclonal anti-

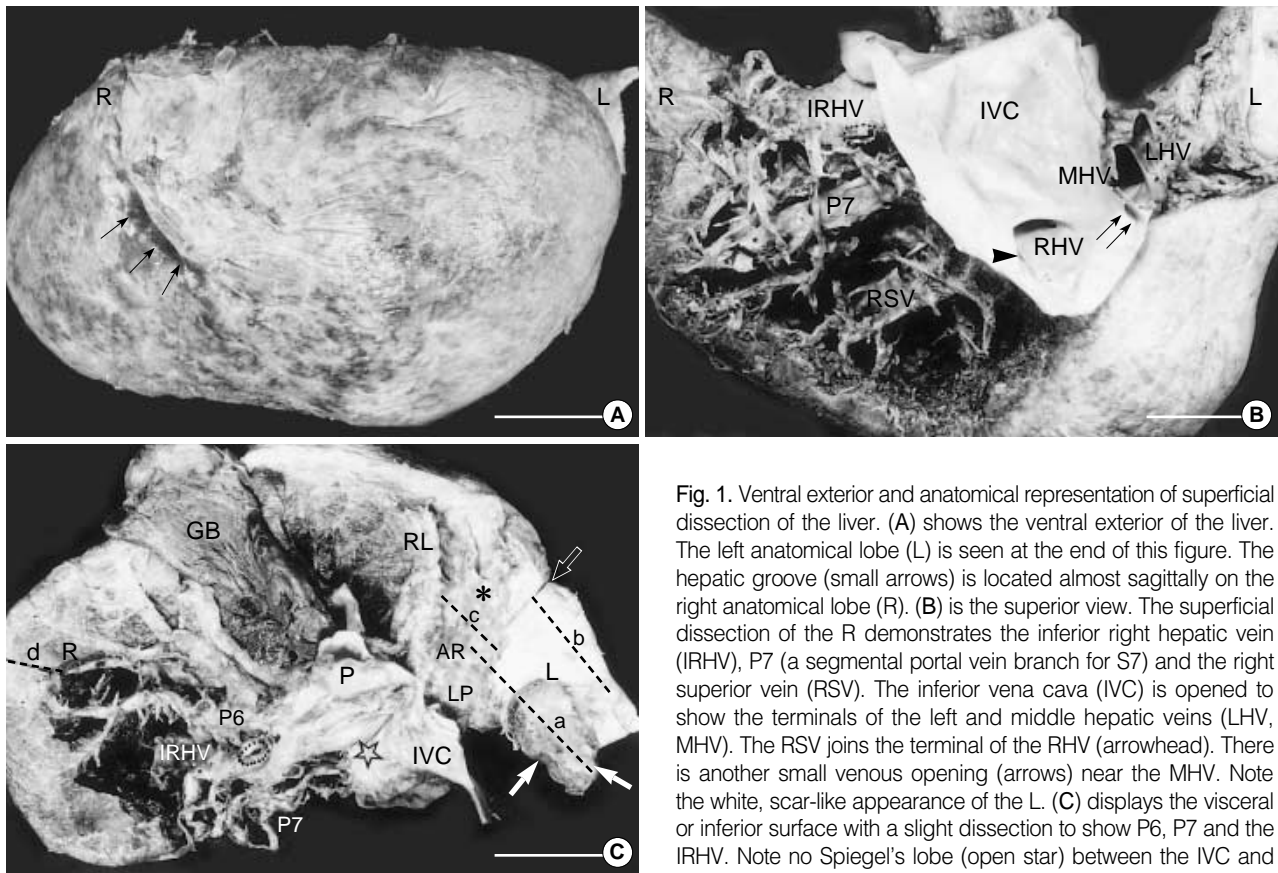


Fig. 1. Ventral exterior and anatomical representation of superficial dissection of the liver. (A) shows the ventral exterior of the liver. The left anatomical lobe (L) is seen at the end of this figure. The hepatic groove (small arrows) is located almost sagittally on the right anatomical lobe (R). (B) is the superior view. The superficial dissection of the R demonstrates the inferior right hepatic vein (IRHV), P7 (a segmental portal vein branch for S7) and the right superior vein (RSV). The inferior vena cava (IVC) is opened to show the terminals of the left and middle hepatic veins (LHV, MHV). The RSV joins the terminal of the RHV (arrowhead). There is another small venous opening (arrows) near the MHV. Note the white, scar-like appearance of the L. (C) displays the visceral or inferior surface with a slight dissection to show P6, P7 and the IRHV. Note no Spiegel's lobe (open star) between the IVC and portal vein (P) at the hilar bifurcation. The round ligament (RL) is

shifted to the right side of a fissure (black-filled-white arrow) between S4 (asterisk) and the L. Arantius duct (AR) is clearly observed. A small parenchymal mass (white arrows) is attached on the inferior surface of the scar-like L. Broken lines with a-d on the lateral, quadrate and right lobes correspond to histological sections. The a-c correspond to photographs shown in Fig. 3 and the d was used as a control (histology not shown). GB, gall bladder. White bars in the right down corner indicates 3 cm in the length.

human vonWillebrand Factor (Factor VIII Related Antigen, Code No. A0082, 1:200 in PBS). No counterstaining was performed.

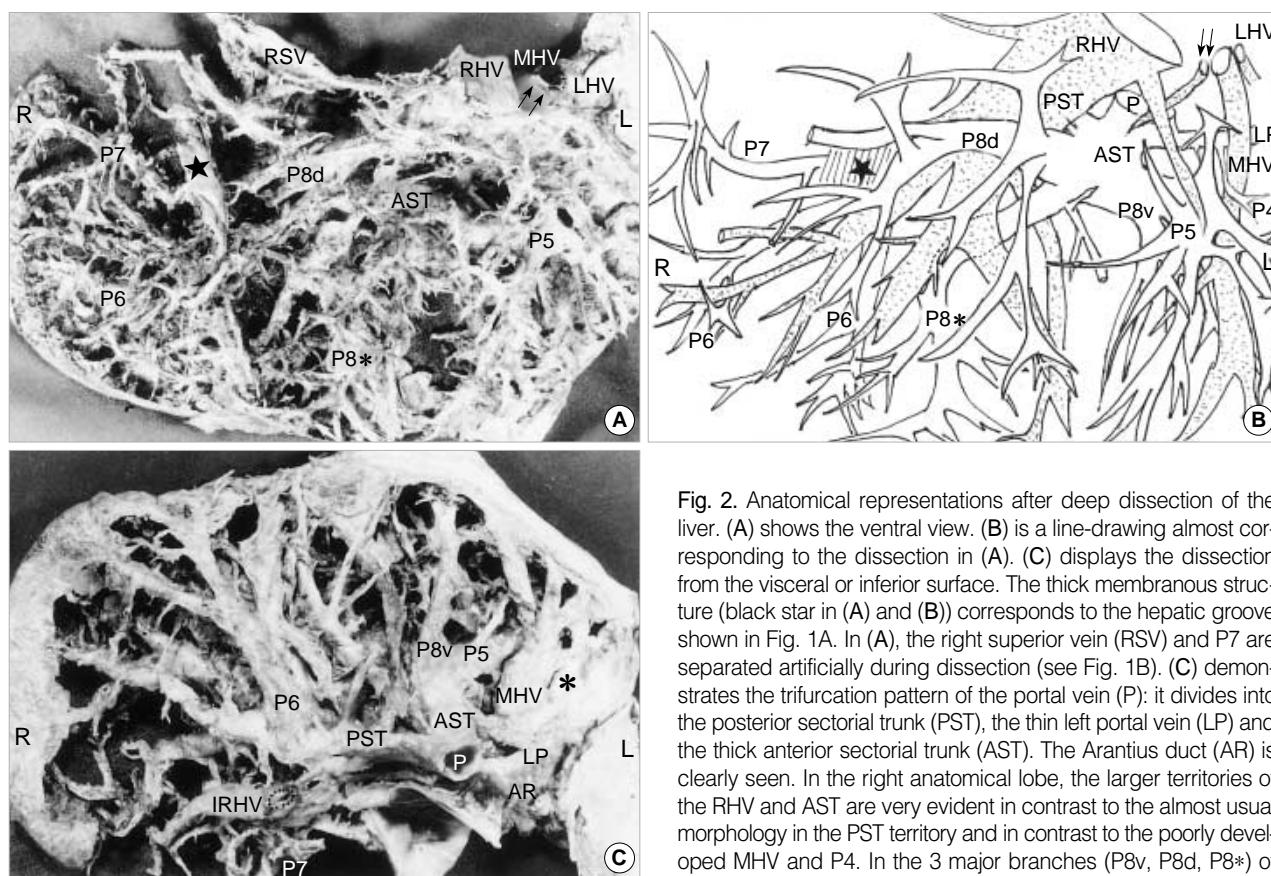
RESULTS

Macroscopic observations

The peritoneal fusion around the liver was evident when the liver was removed. The anomalous liver carried flat, white, scar-like left anatomical lobe with a small parenchymal mass (25 × 12 mm) on its visceral or inferior surface (Fig. 1A). The round ligament was slightly shifted to the right side of a deep fissure along the right margin of the scar-like lobe. This fissure was filled with the connective tissue mass continuous to the round ligament, and it looked like the main lobar fissure. The round and falciform ligaments were stuck to a connective tissue mass, occupying the porta hepatis. Any vessels and ducts at the porta were embedded in the connective tissue mass. Notably, the portal vein was stuck to the

inferior vena cava (IVC) because of no Spiegel's lobe. The gall bladder occupied near the usual position on the right anatomical lobe. On the right ventrosuperior side of the right anatomical lobe, a hepatic groove was observed (50 mm in length, and 15 mm in maximum depth). We did not investigate the extrahepatic courses of the portal vein, hepatic artery, and bile duct.

The portal vein showed the trifurcation pattern (Fig. 2). It was divided into posterior sectorial trunk, thin left portal vein, and thick anterior sectorial trunk. We could easily identify the branches of the posterior sectorial trunk as a P7 and multiple P6s, according to Hata et al. (10). The anterior sectorial trunk supplied a parenchymal area much larger than usual and than the posterior sector of the specimen. The P8-like thick branches were counted at least 3 in number and, in the present report, we temporarily termed them P8 ventral (P8v), P8 dorsal (P8d) and others (P8*). This P8d crossed the bottom of the hepatic groove and reached the right side of the groove. Notably, P8* crossed in the dorsal side of a thick tributary of the right hepatic vein. P5 was identified as the ventromedial subdivision of P8v. The left portal vein



appearances of S4 (asterisk) and the left lobe (L). The hepatic vein openings (RHV, MHV, LHV and a small opening with arrows) correspond to Fig. 1B.

issued thin branches for the scar-like S4. Along the left portal vein, we could not find a dilated portion corresponding to the umbilical portion. P2 and P3 were not identified, but several thin twigs supplied the left parenchymal tissue scattering in the scar-like left lobe. The portal vein branches showed tendency to stick not only to bile ducts and to arterial branches but strangely, also to the hepatic vein tributaries. However, the intrahepatic anastomosis was not evident between the portal vein and hepatic vein systems. Thick portal vein branches were accompanied by the dilated bile ducts, although the present figures did not provide the observations. The dilation was evident in one part of the quadrate lobe.

Topographical anatomy of terminal portions of the major three hepatic veins looked usual when the internal surface of the retrohepatic IVC was observed after opening (Fig. 1B). However, the right hepatic vein was very thick and it drained most of the right anatomical lobe, including a part of S5. The inferior right hepatic vein (11) and the right superior vein (12, 13) were present at usual positions, i.e., the former was located almost along the S6/S7 border, while the latter drained uppermost portion of S7. Thus, the drainage pattern seemed to correspond to a rare population (14). One of

the major tributaries of the right hepatic vein interdigitated with P8* and P8v, while another ran between S8 and the posterior sector. The middle hepatic vein exhibited a short upward course between S5 and the scar-like S4. Although it was obliterated, the Arantius duct was clearly identified as a bundle connecting the left portal vein and the uppermost portion of the retrohepatic IVC. A small opening near the middle hepatic vein terminal was very similar to the opening for the high dorsal drainage route of Spiegel's lobe (15). However, the corresponding vein drained a liver parenchyme ventral to the porta hepatis because of no Spiegel's lobe.

Histological observations

In the anatomical left lobe, the liver parenchyme was enclosed and fragmented by the scar-like tissue (Fig. 3A, B). The portal triad in the parenchyme appeared normal as seen in S6. However in S6, as well as in the left anatomical lobe, bile ducts were not dilated, but even thin and small in number. In both parts, liver parenchyme also appeared atrophic and the hepatic cell plate looked thin. The scar-like tissue of the left lobe, except for vessels, did not contain either of the smooth muscle tissue and factor VIII-positive structures (immuno-

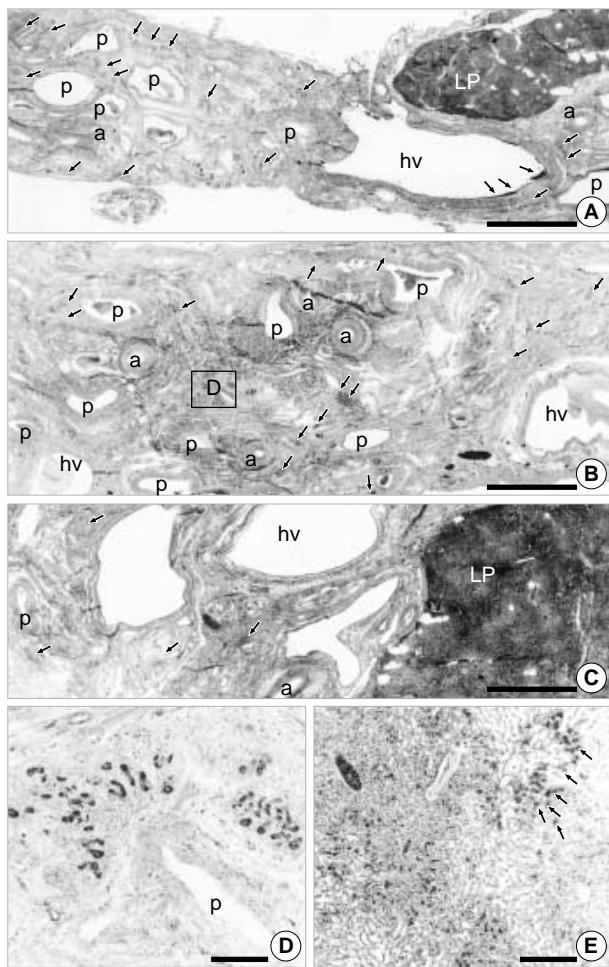


Fig. 3. Histological observations. Broken lines with a-c on the lateral and quadrate lobes in Fig. 1C indicate the location of the histological sections A-C. In A-D, p, a, and hv indicate the portal vein branches, arterial branches and hepatic vein tributaries, respectively. Black-filled-white arrows indicate clusters of thin bile ducts in A-C (higher magnification, see Fig. D). (A) shows the left anatomical lobe with a small parenchymal mass on the visceral surface of the liver (see Fig. 1C). The left hepatic vein tributary is identified by labeling with carbon particles (arrows). LP, liver parenchyme. Bar indicate 10 mm. (B) displays the scar-like, major portion of the left anatomical lobe. Note that the wall of p is as thick as that of a (but the p contained no or little smooth muscle tissue: see text). Bar indicate 3 mm. (C) Scar-like tissue in the quadrate lobe with a parenchymal mass. Bar indicate 3 mm. (D) Higher magnification view of a square containing D in Fig. 3B. Note clusters of thin bile ducts and also note thick wall of the portal vein branch (p). Bar indicate 0.1 mm. (E) Higher magnification view of the parenchymal tissue in the lateral lobe. Arrows, blood congestion in the sinusoid. Bar indicate 0.2 mm.

histochemistry not shown). In contrast to usual thick hepatic veins, the hepatic vein tributaries had poor contents of the media although dilated. The scar-like tissue was characterized by rich contents of vessels, bile ducts and nerves (Fig. 3A-C). Difference in tissue components was not evident between the quadrate lobe and other parts. In the scar-like part, the

arterial branches carried the hypertrophic media much thicker than those in the parenchyme. Moreover, the portal vein branches also carried the media-like, thick venous wall, and those were similar to the arterial wall (Fig. 3B). Notably, however, these venous wall of the portal vein system contained no or little smooth muscle tissue according to the immunohistochemistry (photograph, not shown). Clusters of very thin bile ducts were also evident in number (Fig. 3D), and those suggested budding and/or proliferative change of bile ducts during or after scar-like change of the left parts of the liver. The cluster of fine bile ducts was not usually located near the portal vein branches (Fig. 3B). Using labelling with carbon particles, we discriminated the left hepatic vein tributaries from those of the middle hepatic veins (Fig. 3A). The territorial border between the left and middle hepatic veins was located along the line corresponding to the main lobar fissure, i.e., the territory of the middle one was restricted in the scar-rich quadrate lobe part in contrast to other left lobe anomaly (16) in which the middle hepatic vein drained most of S3.

DISCUSSION

The present work reported a left lobe hypoplasia of the liver that was characterized by 1) the scar-like left surgical lobe with few parenchymal tissue, 2) no Spiegel's lobe, 3) unusual configurations of the right hepatic vein and the anterior sectorial trunk of the portal vein. The present hepatic groove was not evident because the groove crossing P8 course was common (17), although the groove associated with left hypoplasia seemed not to be reported. We believe that the thickest primary division of the portal vein was the anterior sectorial trunk (P5+P8) because the present case seemed to correspond to the trifurcation pattern, i.e., one of the major variants of the portal vein primary division (18) and because, in the present specimen, there was another definite right trunk (i.e., the posterior sectorial trunk) showing the usual morphology. Moreover, Saigusa et al. (7) also reported well-developed P5 and P8, especially P5, with extended territories in combination with poorly-developed P4 in a liver with the absence of S2 and S3. Their pattern of the ramification is similar to that in the present case, although the middle hepatic vein was usual in Saigusa et al. (7). In a variant liver with the left hepatic vein (or the left IVC) draining directly into the right atrium near the opening of the coronary sinus, Yoshinaga and Kodama (16) described an abnormal right hepatic vein crossing ventrally to the anterior sectorial trunks in combination with the thick inferior right hepatic vein. This case seems to be similar to the present right hepatic vein interdigitating with branches of the anterior sectorial trunk. Overall, the present identification of abnormal intrahepatic vessels seems to be almost adequate and logical.

Why did the left surgical lobe develop or change to be scar-like? Peritoneal fusion around the liver appeared to provide a clue. We speculated that, in the late embryonic stage or even during postnatal growth, scar formation happened at and around the porta hepatis, and it involved the round ligament and left portal vein. We can easily speculate a small injury during birth or a restricted infection around the cut terminal of the umbilical cord. Thus, the round ligament was shifted from the main lobar fissure. We speculate that clusters of very thin bile ducts in the left lobe are caused by secondary budding and/or proliferative change of bile ducts during or after scar formation in the left liver. Conversely, the basic portal triad structures seemed to be preserved even in the scar-like tissue. Therefore, we hypothesized that the secondary hypoplasia happened due to the peritoneal fusion involving the left liver structures. Dilated intrahepatic bile ducts in the quadrate lobe suggested an obstruction due to the peritoneal fusion. Moreover, in contrast to the present specimen, Spiegel's lobe is clearly seen in a left hypoplasia according to the case reported by Saigusa et al. (7). Notably, we found a hepatic vein similar to the typical superior caudate vein for the high dorsal drainage (15). Thus, the secondary fusion seemed to be likely to involve the IVC near the porta hepatis, which resulted in abnormal fusion between the portal vein and IVC and atrophy of Spiegel's lobe. Finally, we have to discuss about the topographical anatomy between P8* and a thick tributary of the right hepatic vein because a branch of P8 should be located ventral to the hepatic vein. Hata et al. (14) reported that such a thick tributary (over 5 mm in the outer diameter) was present in 9.1% of 77 usual adult livers. Thus, subsegmental branches of P8 can sandwich the hepatic vein. Nevertheless, the abnormal right hepatic vein crossing ventrally to the anterior sectorial trunks was associated with a congenital anomaly (16). We speculated that the hepatic vein course frequently shows significant variations such as those disturbing the basic topographical anatomy between the intrahepatic portal and hepatic veins.

Consequently, the present case seemed to carry secondary atrophy for the pathogenesis. It seemed to be different from those in cases reported by Saigusa et al. (7) and Merrill (8) because their specimens were associated with absence of the gallbladder and/or the usual morphology of S4 and Spiegel's lobe.

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