An Autopsy Case of Budd-Chiari Syndrome

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INTRODUCTION

The Budd-Chiari syndrome is an uncommon disease caused by the occlusion of major hepatic veins or obstruction of the inferior vena cava (IVC) near the ostium of the hepatic veins. The syndrome was first reported by Budd[1] in 1845 and subsequently as the symptomatic hepatic vein occlusion by Chiari[2] in 1899. Although some reports described the underlying disease leading to the disease, for example polycythemia vera [3], phlebitis of the hepatic vein[4,5], tumors which caused the hepatic vein occlusion[6], etiology of many cases are still remains unexplained[7,8]. The authors have recently had an opportunity to autopsy a patient who developed a classical form of the disease which was presumed to originate from developmental anomaries of both hepatic veins in each osteal portions.

REPORT OF A CASE

A 26-year-old woman was admitted to the Yamaguchi University Hospital on July 22nd in 1975, because of persistent ascites. About 3 years before this admission she had an episode of right hypochondralgia with nausea and admitted for 11 days with a diagnosis of hepatitis. After then she had been in good health until 2 and a half years later when she noticed abdominal full sensation and edema of the lower extremities. At that time she was pointed out ascites and was treated with diuretics and occasional paracentesis, however, ascites did not subside and her general condition was gradually disturbed, then she was referred to readmit to the hospital. On the day of admission, marked abdominal distension with dilated veins on its surface and icteric discoloration of general skin were noted. A paracentesis yielded 3 liters of yellowish clear ascitic fluid, and then the hard liver edge 3.5 finger breadth beneath the right costal margin was palpated. No splenic enlargement

Table 1. Labolatory Data on Admission

Hemogram				
RBC	4.94million/cu mm			
Ht	49.0%			
Hb	15.3g/dl			
WBC	9700/cu mm			
Blood Chemistry				
Total protein	5.2g/dl	Cholinesterase	0.20	ph
Alb.	2.6g/dl	Serum ammonia	233	μg/dl
Glob.	2.6g/dl	Alk. phos.	44	U(King Armstrong U)
α_1-glob.	4.8%	Cholesterol	100	mg/dl
α_2-glob.	6.4%	SGOT	23	U/ml(Henry U)
β-glob.	10.7%	SGPT	18	U/ml(Henry U)
γ-glob.	26.0%	α-feto protein	(−)	
Total Bil.	1.3g/dl	Au. antigen	(−)	
Direct Bil.	61.5%	Wasserman reaction	(−)	
ICG test (15min.)	86.6%	RA test	(−)	

was noted. On the following day the liver was hard to palpate due to ascites, which did not lessen in spite of taking diuretics. Laboratory data indicated moderately impaired hepatic function (Table 1). Ascitic fluid revealed yellowish color, and Rivalta reaction was negative. Its specific gravity was 1.013. Cytological examination of the fluid revealed no atypical cells. Gastrofluoroscopy showed esophageal varices and gastric erosion. Portal venogram revealed dilated portal vein showing marked tapering in its proximal portion and dilated paraumbilical veins. On scintigram of the liver, there was no space occupying lesion. Subsequently, the patient complained of occasional hemorrhoidal bleeding, paroxysmal tachycardia, dyspnea, palmar erythema, edema of the face and extremities, and oliguria. During the three months after admission, her general condition progressively got worse and took a sudden turn for hepatic coma with mineral unbalance. Finally she died from apnea attack with tremor and cardiac arrest. Autopsy was performed at 4 hours post mortem.

AUTOPSY FINDINDS

Macroscopic Observation

Moderately nourished young woman (Body length 152 cm, Body weight 62 kg) showed marked abdominal distension with striae and venous dilatation, jaundice, petechiae on upper chest. Totally 10 liters of yellowish ascites was noted. Diaphragm was elevated up to the level of the 3rd

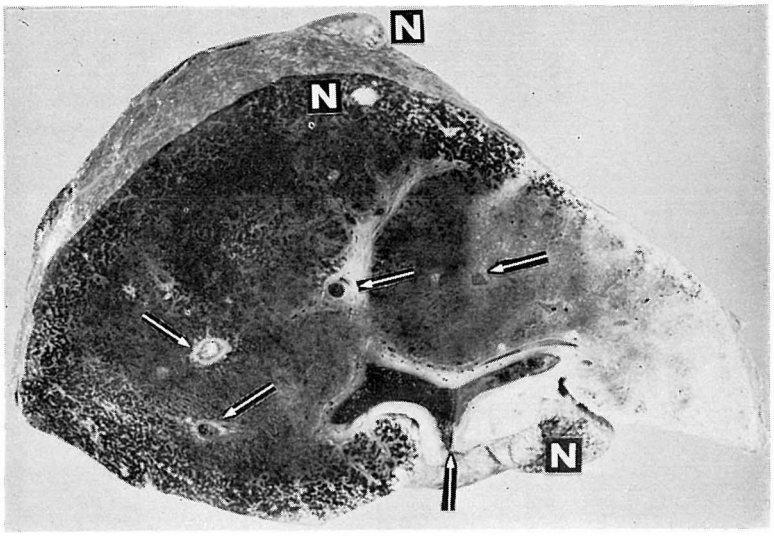
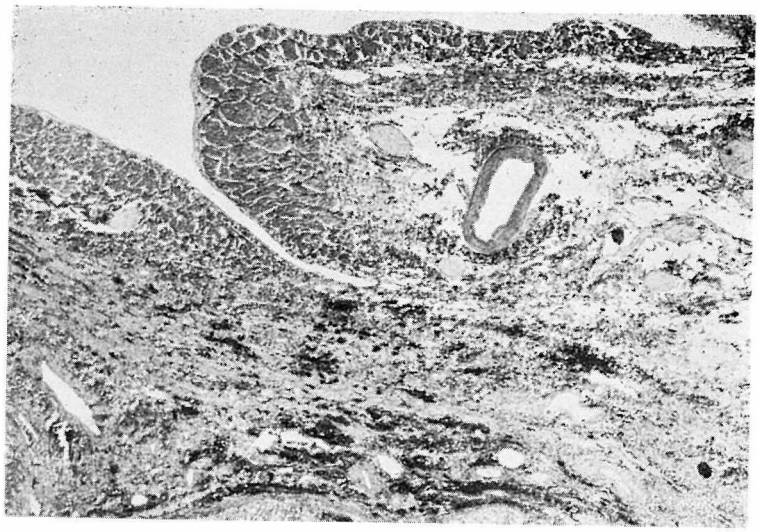


Fig. 1. Coronal section of the liver. Several large hepatic veins are occluded by thrombi (arrows) of varing stages. Intrahepatic portal veins are occluded by red thrombi (arrows) from the hilus of the liver. The right lobe shows marked congestion with hemorrhage and the left lobe is reduced in size with cirrhosis. In addition there are several nodules (N) scattered in both lobes. ×0.5

Fig. 2. Intrahepatic portion of the inferior vena cava (IVC). Note the small openings of hepatic veins (arrows). ×4



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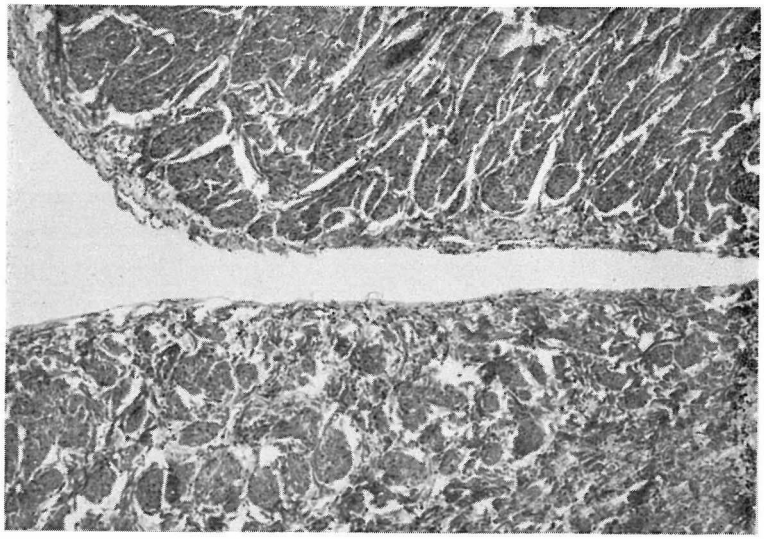

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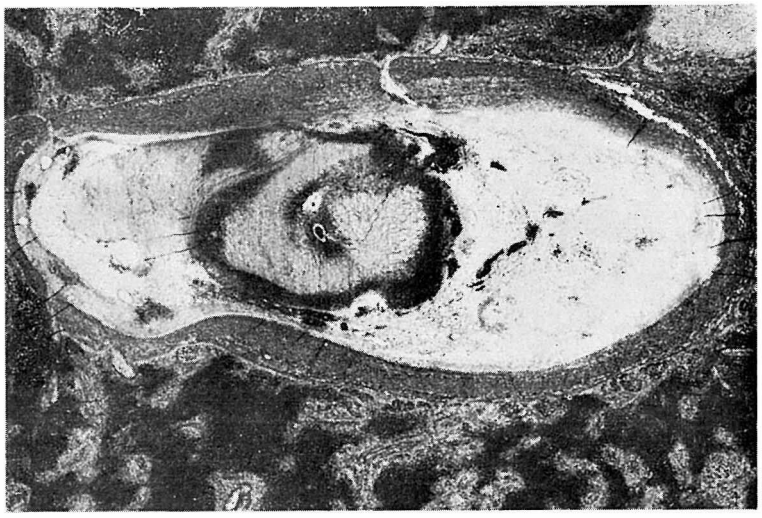
Fig. 3. a) Low-power microscopic photograph of the intrahepatic portion of the IVC. One of the hepatic veins is seen as a narrow channel. ×13
b) Osteal portion of the hepatic vein. ×40



a

b

Fig. 4. a) The large trunk of the right hepatic vein a few centimeters distal to the orifice. Note the organized thrombus with recanalization. Peripheral portion of the thrombus undertake a fibro-myxomatous change. ×7
b) Higher magnification of organized thrombus. ×40

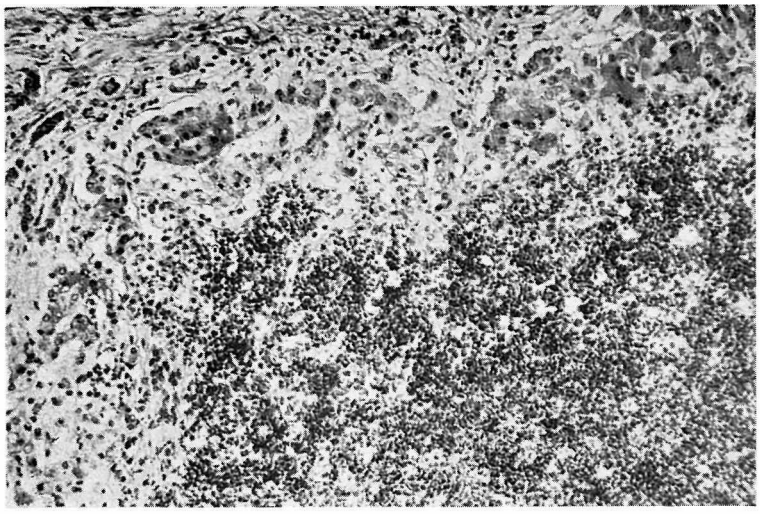


Fig. 5. The right lobe of the liver. Marked congestion with hemorrhage is seen in the centrilobular area. Liver cells are remained only in the periportal area. ×100

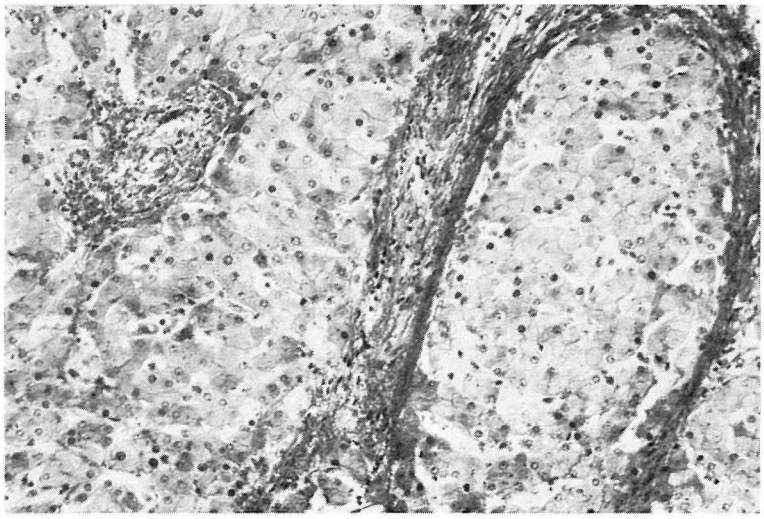


Fig. 6. The left lobe of the liver showed Laennec type cirrhosis. A portal area is included in one of the pseudolobules. ×100

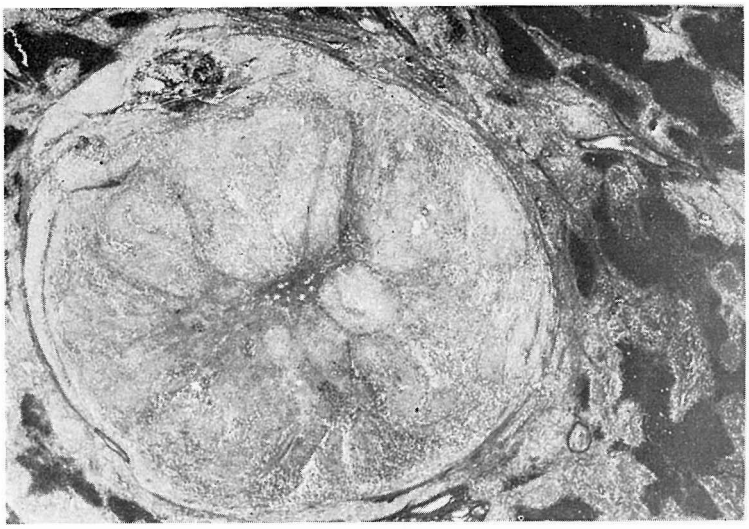


Fig. 7. Low-power microscopic photograph of the nodular hyperplasia in the right lobe. The nodule is well circumscribed and subdivided by centrally situated scar. ×7

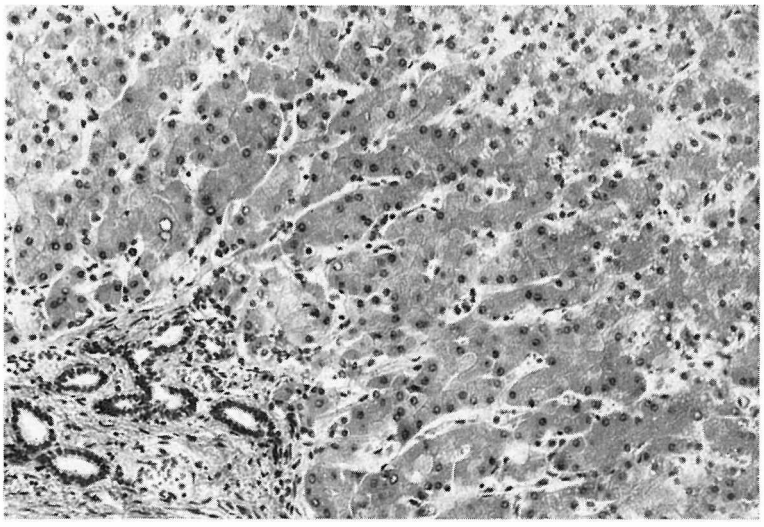


Fig. 8. The nodule consists of slightly hyperplastic liver cells and portal triad are visible in fibrotic area. ×100

intercostal space in the left side and 3rd rib in the right side. The parietal peritoneum and the serosa of the intestine were dark red and revealed marked dilatation of the tortuous veins which run longitudinally with close aggregation.

The liver weighed 1620 g. The right lobe was markedly swollen with congestion and hemorrhage. The left lobe was reduced in size and was occupied by miliary to rice-grain sized, hard nodular area (Fig. 1). There was a thumb-head sized concavity at the situation where the hepatic veins join to the IVC and on its concavity several small openings of hepatic veins were detected (Fig. 2). In the tributaries of these openings, hepatic veins were filled with numerous thrombi of varing stages. Intrahepatic portal vein was also obstructed by red thrombus from the hilus of the liver. In addition, thumb-head sized yellowish-gray nodules were scattered in both lobes. On cut surface, these nodules were subdivvided into small lobules by fibrous septa (Fig. 7).

The spleen weighed 180 g. Consistency was moderately hard with moderate amount of matter. Splenic vein was also obstructed by red thrombus.

Urinary bladder. The mucosa was partially desquamated and showed marked hydropic change associated with hemorrhage.

Microscopic Observation

The hepatic vein which could be traced into the lumen of the IVC were embedded in dense collagenous tissue and had the same structure as the veins of small caliver. Also its muscle layer continued to the muscular adventitia of the IVC at the orifice (Fig. 3a, 3b). There was no evidence of inflammation at the orifice of both hepatic veins. In the large trunks of the right hepatic vein, there were organized and recanalized thrombi without complete obstruction (Fig. 4a, 4b). The left hepatic vein showed only slit like stenosis surrounded by fibroelastic tissue.

The right lobe of the liver showed marked congestion and centrilobular necrosis with partial hemorrhagic lesion (Fig. 5). The left lobe revealed distinct pseudolobule formation with narrow fibrous band and in some part the portal area were contained in the center of the pseudolobule (Fig. 6). A large number of bile thrombi were detected in both lobes of the liver.

Nodules which were scattered in both lobes were composed of irregular collections of slightly hyperplastic liver cord cells and occasionally portal triad were noted in the nodules (Fig. 8). PAS stain and Sudan Ⅲ stain indicated that the hepatic parenchymal cells composing these nodules contained more glycogen and fat granules than surround-

ing liver cells. Silver stain showed the condensation of reticulum fibers at the periphery of the nodules. The cells in the nodule were sometimes binucleate and occasionally had a conspicuous nuclei, however no mitosis was seen.

The spleen showed marked congestion and hemosiderin laden macrophages were scattered in the red pulp. The white pulp was atrophic. The thrombus which obstructed the splenic vein was red thrombus and a part of which was organized by granulation tissue from the endothellium.

PATHOLOGICAL DIAGNOSIS

1. Budd-Chiari Syndrome
   1) Developmental anomalies of both hepatic veins in each osteal portion. Right; Several extremely narrow channels.
      Left; Single slit-like stenosis.
   2) Thrombosis of the hepatic vein, portal vein and paraumblical vein.
   3) Laennec type liver cirrhosis in the left lobe.
   4) Marked congestion with localized hemorrhage in the right lobe of the liver.
   5) Ascites (10 liters)
   6) Collateral circulations.
      Dilated paraumbilical and epigastric veins. Esophageal varices. Hemorrhoid.
2. Multiple nodular hyperplasia of the liver.
3. Hemorrhagic cystitis.
4. Postoperative condition of the portal venography and of appendectomy.

DISCUSSION

Findings of the hepatic vein orifices led us to suspect a developmental anomaly as a causative facter, for the reason that, microscopically, concaved wall of the IVC at which both hepatic veins were opened as several narrow channels has the same structure as the IVC, and at the orifice tunica media of the hepatic vein continues to the muscular adventitia of the IVC. Moreover, there was no evidence of inflammation in this portion.

We supposed that part of the both hepatic veins were patent until 3 years ago when she complained of abdominal pain and the onset of the symptom may be attributed to the thrombosis of the hepatic veins draining through the left lobe of the liver. This consideration benefits to explain the histological difference between the right and left lobe of the

liver, for the reason that, in the left lobe there is a histological finding of the fibrosis progressing from the center of the lobule to remain the portal triad in the pseudolobule. This finding may indicate the previous episode of the acute stage of the hepatic vein occlusion which was followed by the fibrosis. Furthermore, as Thompson et al.[5] describe in the case of thrombosis of the hepatic vein, with the reconstruction of the hepatic cells pseudolobules may be formed and ultimately take a histological pattern of the Laennec's cirrhosis. Right lobe of the liver showed marked congestion with collapsed centrilobular hepatocytes as a result of severe venous engorgement which can be attributed to the acute stage of the venous occlusion by the thrombi at the outlets of the right hepatic vein. Pathologically similar case was observed in Hashimoto N. et al.'s[9] literature in which the ostium of the right hepatic vein was extremely narrowed and one of the left hepatic veins was failed to be found on careful examination. In addition, liver showed cirrhotic change in the right lobe and marked congestion with hemorrhage in the left lobe. With the evidence that the narrow opening of the right hepatic vein had neither endothelial thickening nor inflammatory change, they thought the case of the disease on the basis of congenital anomaly.

Nodules which scattered in both lobes differed from adenoma and had a centrally situated scar containing bile ductules and blood vessels in the nodule. These histological patterns are consisted with the nodular hyperplasia of the liver. However, there is extensive confusion regarding to this entity[10-13]. Ishak[14] describe it as a well circumscribed nodule by mature collagen and characteristically subdivided into lobules by fibrous septa that converge toward a central stellate area of fibrosis. In this case, the nodular hyperplasia was thought to be an incidental finding associated with the liver of the Budd-Chiari syndrome at the time of autopsy.

SUMMARY

An autopsy case of Budd-Chiari syndrome which may be originated from developmental anomalies of both hepatic veins in each osteal portion was studied. Osteal portion of the right hepatic vein was made up of several narrow channels and that of the left hepatic vein showed single slit-like stenosis. In the tributaries of these openings, there were organized and recanalized thrombi without complete obstruction. The right lobe of the liver showed marked congestion and the left lobe revealed the Laennec's cirrhosis. In addition, there were several nodular hyperplasia of the hepatic parenchymal cells scattered in each lobe of the liver.

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