PORTAL VEIN PATHOLOGY

CASE REPORT: Congenital absence of the portal vein associated with nodular hyperplasia in the liver

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Abstract Congenital absence of the terminal portion of the portal vein with visceral venous return to the suprahepatic inferior vena cava, a rare malformation, was demonstrated in an 18-year-old Japanese woman. She had nodular hyperplasia in the liver and a non-functioning pancreatic endocrine tumour. It is generally believed that reduction of portal venous flow causes atrophic changes and, subsequently, nodular hyperplasia occurs in a well-perfused area in the liver. However, the liver was not perfused by the portal vein in this case. It is suggested that nodular hyperplasia can occur without portal blood flow.

Key words: congenital absence of the portal vein, congenital anomaly of the portal tract, nodular hyperplasia of the liver.

INTRODUCTION

Congenital absence of the portal vein (CAPV) is an extremely rare malformation of the splanchnic vein system. To our knowledge, only 15 cases of CAPV have been reported in the literature. Most of them were complicated by cardiac, skeletal and visceral anomalies. Hepatic tumours or tumour-like lesions are also associated frequently with CAPV. In this report, we describe the case of an 18-year-old Japanese woman with CAPV who had nodular hyperplasia in the liver. She congenitally lacked portal venous flow in the liver. Both the superior mesenteric and splenic vein flowed into the suprahepatic inferior vena cava (IVC).

According to Wanless et al., various forms of nodular hyperplasia in the liver such as focal nodular hyperplasia (FNH), nodular regenerative hyperplasia (NRH) and partial nodular transformation (PNT) are formed in the presence of disturbed portal venous flow. Poorly perfused areas undergo atrophy and, in compensation, well-perfused areas, hypertrophy. The liver of this patient, however, was not supplied with portal blood from the portal trunk or collaterals. These blood vessels could not be detected by angiography.

Clearly, arterial blood supply without portal blood can cause hepatocellular nodular hyperplasia resembling ordinary nodular hyperplasia.

CASE REPORT

An 18-year-old Japanese woman was admitted to Numazu City Hospital because of an abrupt epigastric pain. Physical examination did not reveal any abnormal items. Laboratory data showed a mild elevation of leucine aminotransferase (50 IU/L, normal 25-45) and γ-glutamyl-transpeptidase (27 IU/L, normal 0-25). Serum ammonia (NH₃) was also mildly elevated (95 μg/dL, normal 0-79). Serological markers for hepatitis B and C virus were negative.

Abdominal ultrasonography (US) showed several nodular lesions in the left lobe of the liver. Swelling of the tail of the pancreas was also found. The lesions revealed by liver sonographically consisted of two hyperechoic nodules in segment IV (41 mm, 18 mm) and a slightly hyperechoic nodule in segment VII (37 mm). Dynamic incremental computed tomography (CT) showed multiple nodules in segments IV and VI of the liver (Fig. 2). On magnetic resonance imaging, T1-weighted images demonstrated only one nodule in segment IV. They were almost isointense. The other nodules were not clearly visualized. However, T2-weighted images of MRI revealed multiple low-intensity nodules which were not detected by CT or US (Fig. 3).

On angiography, venous phases of superior mesenteric and splenic arteriography did not show any portal

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venous flow or collaterals going into the liver (Fig. 4a–c). It revealed a continuation of superior mesenteric and splenic venous flow into the IVC. On abdominal US, splenic vein and superior mesenteric vein joined before entering the IVC. The portal trunk was detectable, but it went up through the back of the liver as an abnormal vessel. The ligamentum venosum showed a high echoic line, suggestive of fibrotic change. These findings were not consistent with patent ductus venous Arantii. Based on these findings, we diagnosed this case as congenital absence of the portal vein (CAPV). Celiac arteriography revealed dilatation of the celiac trunk and tortuous change of hepatic arteries. Ultrasonography with intra-arterial infusion of carbon dioxide microbubbles (US angiography) showed a hypervascular tumour in segment IV and other tumours were isovascular.

To clarify the exact nature of the multiple liver nodules and pancreatic lesion, we performed ultrasonically guided needle biopsies. The two liver specimens (segments IV and VII) did not show apparent atypia suggestive of malignant tumour. Part of this liver specimen, shown in Fig. 5, demonstrated atrophic trabeculae with dilated sinusoids suggesting circulatory disturbances in the liver and the other part showed hyperplasia. In this area, large hepatocytes showed a low nuclear cytoplasmic (N/C) ratio. This hyperplastic area gradually changed into an atrophic area, where a reduced size of hepatocyte showed a high N/C ratio. Because this high N/C ratio was caused by liver cell atrophy, the specimen was not diagnosed as hepatocellular carcinoma. Unfortunately, the biopsy specimen did not contain portal tracts.

The specimen from the tail of the pancreas was diagnosed as a pancreatic endocrine tumour. Serum levels of insulin, glucagon, somatostatin and pancreatic polypeptide were within normal ranges.

**DISCUSSION**

Congenital absence of the portal vein is defined as a congenital vascular anomaly that consists of an absence of the portal vein with abnormal drainage of mesenteric and splenic veins into the systemic circulation. In the process of embryological development, the normal portal vein begins developing at 4–10 weeks after fertilization. The portal vein derives from selective involution and persistence of the peri-intestinal vitelline venous loop. Abnormal patterns of involution and persistence may result in either a duplicated, pre-duodenal, prebiliary or absence of the portal vein.

Congenital absence of the portal vein is frequently associated with a cardiac anomaly1–5,7–9,13,15,16 as well as visceral3,12–14,16 and skeletal7,9,14,15 anomalies. Association of hepatic tumours has also been reported5,11,14 Hepatic tumours were described in 6/15 cases in Table 1. Three of these tumours were focal nodular hyperplasia (FNH),6,8,14 one was an adenoma,10,11 one a well-differentiated hepatocellular carcinoma9 and one a hepatoblastoma.5

It is well known that various forms of nodular hyperplasia (nodular transformation) are frequently
Nodular hyperplasia in CAPV

Figure 4  The venous phase of superior mesenteric (a) and splenic (b) arteriography revealed continuation of both veins into the inferior vena cava. Neither of the angiograms delineated the portal vein. (c) An illustration of the vasculature based on these two angiograms.

Figure 5  A needle biopsy specimen from the tumour of segment IV. The liver tissue did not show any malignant feature. The hyperplastic area (H) gradually changed into an atrophic area (A) with dilated sinusoids (HE x 64). Inset (i) high magnification view of the atrophic area. Sinusoids are dilated. Owing to the reduced cell size caused by atrophy, the nuclear cytoplasmic (N/C) ratio is increased without nuclear enlargement. (HE x 320). Inset (ii) high magnification view of the hyperplastic area. Because of the large cell size, the N/C ratio is low. (HE x 320).

associated with portal venous abnormalities. Those lesions include FNH, nodular regenerative hyperplasia (NRH) and partial nodular transformation (PNT). Wanless et al. proposed a new classification for all these nodular hyperplasias, classifying them by the number, location, size and distribution of the nodules. The presence and degree of fibrosis and the degree of contrast between nodular and inter-nodular tissues are also considered in the classification. Wanless et al. reported nine cases of NRH associated with haematological disorders. A morphometric method showed obliteration of portal vein radicles in all cases. They suggested that obliteration of the portal vein led to the formation of hyperplastic nodules in well-perfused acini and atrophic change in ischaemic acini. Wanless et al. reviewed 18 cases of PNT reported in the literature between 1909 and 1985. Five of these cases had old portal vein thrombosis (PVT). The authors speculated that PVT occurred first and that abnormal portal venous distribution at the hilum of the liver produced PNT. They also speculated that NRH was caused by a similar mechanism. The size and distribution of obliterated portal veins were different in the cases of NRH. Portmann et al. reported a case with FNH and NRH-like lesions within the liver. Terayama et al. reported a case of PNT associated with portal vein thrombosis and malignant lymphoma. In this case, the main trunk and large branches of the portal vein were thrombosed. Histological studies revealed recanalization of PVT. They proposed that obstruction and recanalization of PVT might be aetiologically related to PNT.

In our case, however, portal venous flow was congenitally absent and there were no collateral veins. This unusual vasculature was proved by angiography and US. Therefore, recanalization or abnormal perfusion of the portal blood flow in the liver cannot be the cause of the nodular hyperplasia. The hepatocellular nodular hyperplasia in our case histologically resembled nodular hyperplasia as seen in FNH, NRH and PNT. However, as stated by Wanless, uneven perfusion of arterial blood supply instead of portal blood could cause nodular hyperplasia. Arterial hypervascularity perhaps compensates for the lack of portal flow and the angiographic findings support this hypothesis. Clearly, in some cases, abnormal portal venous flow and/or PVT is not required for the development of
Table 1  Reported cases of congenital absence of the portal vein (Matsuoka et al., modified)

<table>
<thead>
<tr>
<th>Case</th>
<th>Author</th>
<th>Age/Sex</th>
<th>Drainage site</th>
<th>Hepatic tumour</th>
<th>Cardiac anomaly</th>
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<tbody>
<tr>
<td>1</td>
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PDA, patent ductus arteriosus; PFO, patent foramen ovale; VSD, ventricular septal defect; ASD, atrial septal defect; Co-A, coarctation of the aorta; TR, tricuspid regurgitation; FNH, focal nodular hyperplasia; HCC, hepatocellular carcinoma; IVC, inferior vena cava.

+, Cardiac anomaly existed, however, the name of anomaly was not described.

REFERENCES

Nodular hyperplasia in CAPV


