Persistent Primitive Hypoglossal Artery Complicated by Atrial Septal Defect and Congenital Intrahepatic Shunts
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During early embryogenesis, anastomoses are formed between the carotid artery and the basilar or the vertebral artery, and subsequently, these anastomoses regress. In some cases, these anastomoses remain as persistent carotid-basilar or carotid-vertebral anastomoses. Atrial septal defect (ASD), a communication between the atria at the septal level, is a congenital heart anomaly. Intrahepatic venous shunts between the portal and hepatic veins are very rare and only some are considered congenital. We present the first case report of a patient with an ASD, a persistent primitive hypoglossal artery, and congenital portahepatic shunts. (Internal Medicine 37: 60–64, 1998)

Key words: persistent carotid-basilar anastomoses, portahepatic venous shunts

Case Report

The patient was a 67-year-old woman who had undergone surgery for direct closure of an atrial septal defect (ASD) (central fossa ovalis defect) at the age of 60. She was admitted to the Nippon Medical School First Hospital in February of 1996, with somnolence and left hemiparesis. Neurologic examination revealed hypotonia of her left upper and lower extremities. Deep tendon reflexes were increased in her left extremities. Pathologic reflexes were in both lower extremities. The right pupil measured 5.0 mm and the left, 6.5 mm. Light reactions were normal. There were no spontaneous movements of the left extremities. Her blood pressure was 138/70 mmHg, and her pulse rate was 88/min and irregular. A pansystolic heart murmur (Levine II/VI) was heard maximally at the cardiac apex. The liver and spleen were not palpable. A chest radiograph demonstrated moderate cardiomegaly. An electrocardiogram revealed atrial fibrillation and incomplete right bundle branch block. An echocardiogram revealed severe mitral regurgitation, moderate tricuspid regurgitation, severe pulmonic valve regurgitation, and dilatations of the left atria, right atria, and right ventricle. Laboratory studies including serum total protein, serum glutamic oxaloacetic and glutamic pyruvic transaminases, alkaline phosphatase, creatinine phosphokinase, electrolytes, blood urea nitrogen, and serum creatinine were normal on admission. Total bilirubin and blood sugar were slightly elevated (total bilirubin, 1.6 mg/dl). The hematocrit was low. Serum ammonia values were high on the day after admission (ammonia, 128 µg/dl). An electroencephalogram showed sporadic triphasic waves with diffuse theta activity.

Three days after admission the patient became alert, and the left hemiparesis resolved. Neurologic examination found no abnormality. Magnetic resonance imaging (MRI) of the brain was normal. A computed tomogram (CT) of the liver was normal. We suspected hepatic encephalopathy with or without a superimposed reversible ischemic neurological deficit (RIND). Therefore angiography of the brain and liver was performed.

An aortography and selective angiographies were performed via a percutaneous transfemoral approach. The aortography found no vertebral arteries (Fig. 1). In an oblique lateral view, a left carotid angiography showed a carotid-basilar anastomosis (Fig. 2). In the anteroposterior view (Fig. 3) the left vertebral artery was absent, and both posterior inferior cerebellar arteries arose from the carotid-basilar anastomosis. The lateral view showed the carotid-basilar anastomosis arising from the left internal carotid artery at C2 and joining the lower portion of the basilar artery. The carotid-basilar anastomosis entered the skull through the hypoglossal canal (Fig. 4). The anastomosis ascended near the anterior aspect of the atlas, with a small dorsal curve. The anastomosis was the sole arterial supply to the posterior circulation of the brain.

We also carried out angiography of the portal system. With an opaque catheter we injected a bolus of radio-opaque contrast material into the superior mesenteric artery. An early venous phase of the angiogram demonstrated a fistula between a portal vein and a right hepatic vein (Fig. 5). The venous phase of the
Figure 1. Aortography shows no vertebral artery.

Figure 2. The oblique-lateral view of the left common carotid artery by subtraction angiography shows a left carotid-basilar anastomosis.

Figure 3. The anteroposterior view of the left common carotid artery by subtraction angiography. A basilar artery is visualized through a persistent anastomosis with the left common carotid artery.

Figure 4. Lateral view of the cranial portion of the left common carotid artery. A hypoglossal artery arises from the internal carotid artery at the C2 level and enters the skull through the hypoglossal canal.

celiac angiogram demonstrated a fistula from the portal vein to a left hepatic vein (Fig. 6). Both angiograms also demonstrated saccular portal venous aneurysms in the right lobe of the liver.

Discussion

At the 4-mm stage of development, anastomoses between the internal carotid arteries and paired longitudinal neural arteries (which subsequently fuse to form the basilar artery) form the primitive trigeminal and optic arteries [Fig. 7(1)], which later regress. Anastomoses between the paired dorsal aortae and longitudinal neural arteries include the primitive hypoglossal, proatlantal intersegmental, and cervical intersegmental arteries (which also regress later). The cervical intersegmental arteries subsequently fuse to form the vertebral arteries (1). At the 18-mm stage, a caudal division of the internal carotid
Figure 5. An early venous phase of the superior mesenteric artery. Venous aneurysms in the right lobe of the liver, a portal vein, and a fistula between the portal vein and a right hepatic vein are visualized.

Figure 6. An early venous phase of the celiac artery. Venous aneurysms in the right lobe of the liver, a portal vein, and a fistula between the portal vein and a right hepatic vein are visualized.

artery has formed an anastomosis with the longitudinal neural artery at the mesencephalon and has thereby become a posterior communicating artery.

Lie stated (2) that a persistent primitive hypoglossal artery arises from an internal carotid artery at the level of C1-3, enters the skull through the hypoglossal canal, and anastomoses with the basilar artery. In these cases no posterior communicating artery is visualized by angiography. The carotid-basilar anastomosis of this patient is such a primitive hypoglossal artery, that it enters the skull through the hypoglossal canal rather than passing through the foramen magnum (3). The angiographic incidence of primitive hypoglossal arteries is approximately 8 to 10 times less than that of primitive trigeminal arteries (4), varying from 0.023% to 0.091% (4–6). Brismar has described hypoplasia of the posterior communicating arteries as a prerequisite for the persistence of a primitive hypoglossal artery (7). Moreover, Gilmartin (5) proposed that when vertebral arteries are hypoplastic or aplastic, a hypoglossal artery is present. In
Blood from the portal vein enters the hepatic vein through the portal hepatis in right and left branches, one to each lobe. The portal vein enters the medial third of the splenic vein. The portal vein enters the head of the pancreas. The inferior mesenteric vein usually enters the portal venous system includes all veins from the abdominal portion of the alimentary tract, the gall bladder, pancreas, and spleen. The portal vein is formed by the union of the superior mesenteric vein and the splenic vein posterior to the head of the pancreas. The inferior mesenteric vein usually enters the medial third of the splenic vein. The portal vein enters the portal hepatis in right and left branches, one to each lobe. Blood from the portal vein enters the hepatic vein through the hepatic sinusoids in the portal lobules, with no direct anastomoses. In the present patient, encephalopathy appeared to result from shunting of superior mesenteric venous blood into the systemic circulation via the hepatic veins, given that the incidence of encephalopathy in operative patients with surgically created total portosystemic shunts is 29% to 52% (16).

Raskin et al (17) reported a middle-aged man who had multiple intrahepatic shunts between the portal and hepatic veins as demonstrated by portography. A needle biopsy specimen of the liver was histologically normal, thus they concluded that the shunts represented a congenital malformation. Kozuka et al has reported an autopsy case of a massive shunt between the intrahepatic portal vein and hepatic vein in a middle-aged man with a 6-year history of Inose’s type hepatocerebral disorder (18). The wall of the shunt consisted of a single layer of endothelium and fibrous tissue without a muscular layer or elastic lamina. The fibrous wall of the shunt resembled the fibrous tissue seen in hepatic cirrhosis. The author felt that the shunt was an acquired abnormality arising from dilatation of hepatic sinusoids.

In the fourth week of embryogenesis, rapidly proliferating cords of liver cells from the hepatic bud extend laterally to subdivide the paired vitelline veins into networks of sinusoids which are incorporated into the expanding right and left hepatic lobes (19). At the 5-mm stage, the distal segments of the paired vitelline veins communicate by anastomoses (a part of which subsequently becomes the portal vein). Some distal remnants of the paired intrahepatic vitelline veins persist as portal outlets. The proximal segment of the left vitelline vein disappears, and the right vitelline vein becomes the hepatic veins. As the primitive liver expands, it comes into contact with the paired umbilical veins which then are diverted by way of the hepatic sinusoids to the heart.

Because the present patient had multiple shunts in the liver in combination with other cardiovascular malformations, the intrahepatic shunts may be congenital. Angiography demonstrated saccular aneurysms in the right lobe of the liver. Chagnon et al (20) state that a high-flow communication between the omphalomesenteric venous system (which then becomes the ductus venosus) and the right horn of the sinus venosus can retard sinusoid formation, resulting in aneurysms in the right lobe. The shunts in our patient are unlikely to have resulted from aneurysms of the right portal vein rupturing into the hepatic veins because the shunts were multiple. A communication may have arisen between the portal and hepatic veins from a defect in late embryogenesis after hepatic sinusoid formation. Alternatively, liver cells may have focally failed to proliferate during early embryonic stages.

Chagnon et al (20) suggested that mature brains are less tolerant of toxic metabolites, such that hepatic encephalopathy begins in middle age in patients with hepatic shunts. A change in the systemic circulation after the ASD operation may have brought on the encephalopathy in the present patient. After the cardiac surgery, the venous flow through the shunts from portal to hepatic veins may have increased because of the decrease in hepatic venous pressure caused by a decrease in the pressure.
within the vena cava. Therefore, encephalopathy may often occur in this patient in the future.

It is interesting that congenital cardiac, hepatic and cephalic vascular malformations coexist in this patient. There may be a common mechanism of vascular malformations (including ASD) in liver, brain, and heart. An abnormality in the regulation of vessel fusion may cause these malformations (i.e. resorption of the vessel walls) and may result in the junction of vessel lumens.

**Conclusion**

A 67-year-old woman presented with a left hemiparesis and somnolence. She had undergone surgery for an atrial septal defect (central fossa ovalis defect) at age 60. An angiogram revealed a persistent left primitive hypoglossal artery without vertebral arteries. Angiograms found portahepatic venous shunts and portal venous aneurysms in the right lobe of the liver. We propose that a common mechanism in the liver, brain, and heart may have caused these congenital malformations. This is the first case report of a persistent primitive hypoglossal artery accompanying an ASD and congenital interhepatic shunts.

**References**