Polysplenia Syndrome: A Case Associated with Transhepatic Portal Vein, Short Pancreas, and Left Inferior Vena Cava with Hemiazygous Continuation

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Abstract. This paper presents a 57-year-old asymptomatic patient with the polysplenia syndrome with abdominal manifestations. In addition to multiple spleens, the abdominal findings included a left inferior vena cava with hemiazygous continuation, a striking portal anomaly with the portal vein following an “intraperitoneal-left transhepatic” route to reach the porta hepatis, and a congenitally short pancreas.

Key words: Spleen, abnormalities—Spleen, CT—Portal vein, CT—Portal vein, abnormalities—Vena cava, abnormalities—Pancreas, abnormalities—Polysplenia syndrome.

Polysplenia syndrome is a complex of congenital anomalies, and is frequently associated with cardiopulmonary, and abdominal disorders. Major abdominal vascular anomalies associated with the syndrome include interruption of the inferior vena cava with azygous or hemiazygous continuation, and the so-called “preduodenal” portal vein [1–7]. Herein we present an asymptomatic adult patient affected with the abdominal manifestations of the syndrome, in whom the presence of an abnormal portal vein following a left “transhepatic” route before reaching the porta hepatis appears to be a unique portal anomaly. Additionally detected was a congenitally short pancreas, a recently recognized entity [1, 2, 3].

Case Report

The patient is a 57-year-old female without signs and symptoms of cardiac disease. An abdominal CT study was requested because a mass other than the spleen was palpated during a physical examination. CT revealed situs solitus (normal position) of the liver and spleen; however, multiple spleens were present. An abnormal portal vein traversed the abdomen in a posteroinferior direction, suggesting an intraperitoneal location (Fig. 1A), and entered into the left hepatic lobe (Fig. 1B), continuing as a tubular structure (Fig. 1C) and finally reaching the porta hepatis (Fig. 1D). A congenitally “short” pancreas with a small head and absence of the body and the tail was evident (Fig. 1B). The gallbladder showed an abnormally posterior location, and the caudate lobe appeared thinned and pointed (Fig. 1C). The normal (right) inferior vena cava was absent, and instead a vascular structure was evident to the left of the aorta, representing a dilated hemiazygous vein, evidenced by its retrocrural location (Fig. 1C). A venographic study revealed that the vascular anomaly was a left inferior vena cava with hemiazygous continuation (Fig. 2). In the thorax, this vein first drained into a dilated azygous vein, and then to the superior vena cava. Further angiographic study revealed no cardiac pathology. The hepatic veins drained independently to the right atrium via a common channel. Sonography revealed that the “transhepatic” portal vein flowed toward the porta hepatis; however, several branches of this vein were evident, supplying the left hepatic lobe. It had a normal caliber and, at the porta hepatis, a right portal vein originated from it to supply the right hepatic lobe. These are diagrammatically shown in Fig. 3. Sonographically, the common hepatic vein had a normal caliber, and drained directly to the right atrium, and the biliary system was normal. Conventional radiographs and CT of the thorax were within normal limits, except for prominence of the azygous vein in the right paratracheal region.

Discussion

Although the polysplenia syndrome is a congenital disorder, it may be first seen in adulthood, particularly when congenital heart disease is absent [4]. Therefore, an abdominal sonographic or CT study may be the first imaging method to detect changes with the polysplenia syndrome, as in the current patient. The intraabdominal manifestations of the syndrome consist of partial situs inversus, polysplenia (two or more spleens), preduodenal portal vein, duplicated or interrupted vena cava with azygous/hemiazygous continuation, short pancreas and
other pancreatic abnormalities, intestinal malrotation and malformations, symmetric lobulation of the liver, gallbladder abnormalities, biliary atresia, genitourinary anomalies such as renal agenesis, and several others [1–7].

With regard to the vascular malformations, interruption of the vena cava with azygous/hemiazygous continuation is seen almost consistently with the syndrome [4, 5]. In the current patient, a “left” inferior vena cava continued with the hemiazygous vein. The association of preduodenal portal vein (anterior to the superior mesenteric artery and aorta) has been recognized with increasing frequency in recent years in autopsy series and imaging studies [1, 2, 6]. In the current patient, however, the portal vein was positioned far anterior to these organs, and this suggested an intraperitoneal location. More interestingly, it followed a left transhepatic route (passing through the left lobe of the liver) to the porta hepatis. For these reasons we would suggest the terms “transhepatic,” “preduodenal-transhepatic,” or “intraperitoneal-transhepatic” portal vein, for this anomaly.

Short pancreas is a recently recognized anomaly, which was first discovered in an autopsy series [3] and recently reported in the radiological literature [1, 2]. It has been postulated that the congenitally short pancreas embryologically may be caused by dysgenesis of the dorsal pancreatic bud which gives rise to a portion of the pancreatic head, and the body and tail. Moreover, because both the spleen and the dorsal pancreas develop...