Polysplenia syndrome with hepatic artery of superior mesenteric artery origin and a circumaortic renal vein

N. Çağla Tarhan, A. Muhteşem Ağıldere, Fatih Boyvat, Mehmet Coşkun, Hale Ersoy
Department of Radiology, Başkent University Faculty of Medicine, Ankara, Turkey

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An $8^1/_2$-month-old girl with biliary atresia and polysplenia syndrome having multiple vascular anomalies without cardiac anomalies is reported. Interruption of the inferior vena cava with azygous continuation, which is a common anomaly, was seen in conjunction with origin of the common hepatic artery from the superior mesenteric artery and with a circumaortic renal vein. The case has particular importance in that no hepatic artery or renal vein variations have been described with biliary atresia and polysplenia syndrome in the literature thus far to our knowledge. The anomalies were shown using different radiological examinations including computed tomography, echocardiography, angiography, venography and magnetic resonance imaging.

Key words: polysplenia syndrome, biliary atresia, vascular anomalies.

Polysplenia syndrome involving biliary atresia is a rare phenomenon to occur in conjunction with other congenital anomalies[1-2]. The most commonly associated anomalies are cardiovascular, but the syndrome is also seen coupled with intestinal and respiratory malformations[1]. The vascular anomaly most frequently noted in polysplenia syndrome is interruption of the inferior vena cava (IVC) with azygous or hemiazygous continuation, seen in 50-85 percent of patients[2]. Cardiac anomalies are common, but the other vascular abnormalities described as part of this syndrome are rare[3].

The patient described here, with polysplenia syndrome and biliary atresia, had not only the common anomalies such as IVC interruption with azygous continuation, but unusual vascular variations as well. The common hepatic artery originated from the superior mesenteric artery (SMA), and the left renal vein was circumaortic. We present these malformations in this report, with special attention given to the computerized tomography (CT), magnetic resonance imaging (MRI), and angiography findings.

Case Report

An $8^1/_2$-month-old girl was admitted to hospital with jaundice and failure to thrive. She had jaundice since birth and had undergone a liver biopsy which was consistent with biliary atresia. Physical examination revealed icteric skin, hepatomegaly, a high-arched palate, ascites, and abdominal venous collaterals. Blood test results were consistent with anemia. Other laboratory tests indicated abnormal liver function. A chest x-ray showed cardiomegaly and increased bronchovascular markings in the hilar region. The right main bronchus was longer and had a wider angle than normal which was symmetric with the left bronchus.

On CT, the liver was located slightly to the left side and had irregular contours with hypodense nodules in the parenchyma (Fig. 1). Multiple, rounded splenules of variable size were situated in the right hypochondrium (Fig. 2). The stomach was located on the right side, and the cardiac apex was shifted to the left. These findings were consistent with abdominal situs inversus and left isomerism. The intrahepatic segment of the IVC was not visible, and an enlarged azygous vein was identified to the right of the aorta (Fig. 1). The left renal vein (Fig. 2) could be seen in a circumaortic configuration.

Echocardiography showed left atrial isomerism and narrowed hepatic veins draining directly into the right atrium (Fig. 3a). MRI demonstrated narrowed hepatic veins in cirrhotic liver as well (Fig. 3b).

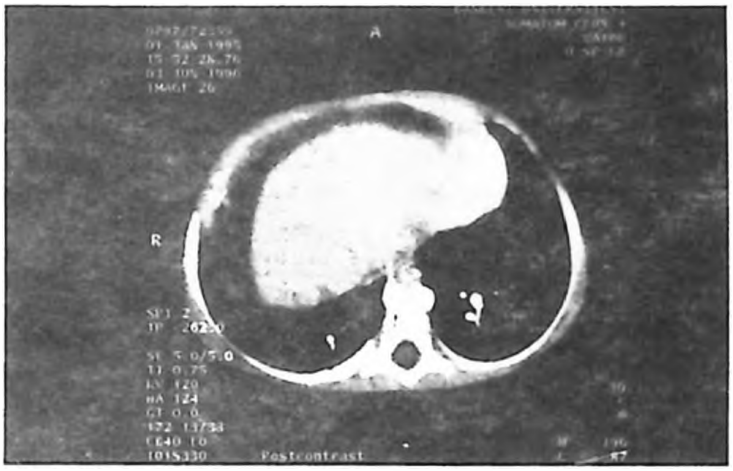

Fig. 1. Contrast-enhanced CT scan of the upper abdomen shows an enlarged liver with irregular contours and hypodense nodules. Also note the absence of the intrahepatic segment of the IVC and the dilated azygous vein (arrow) to the right of the aorta.

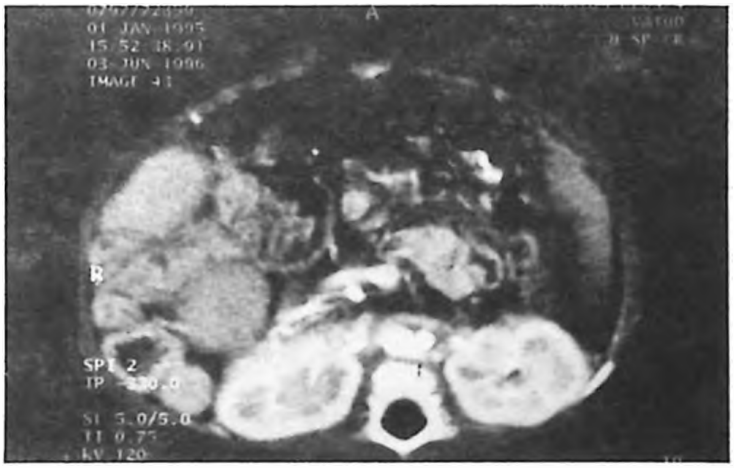

Fig. 2. Contrast-enhanced CT scan of the upper abdomen shows multiple, rounded splenules of variable size located in the right hypochondrium. The circumaortic left renal vein (arrow) is also visible.

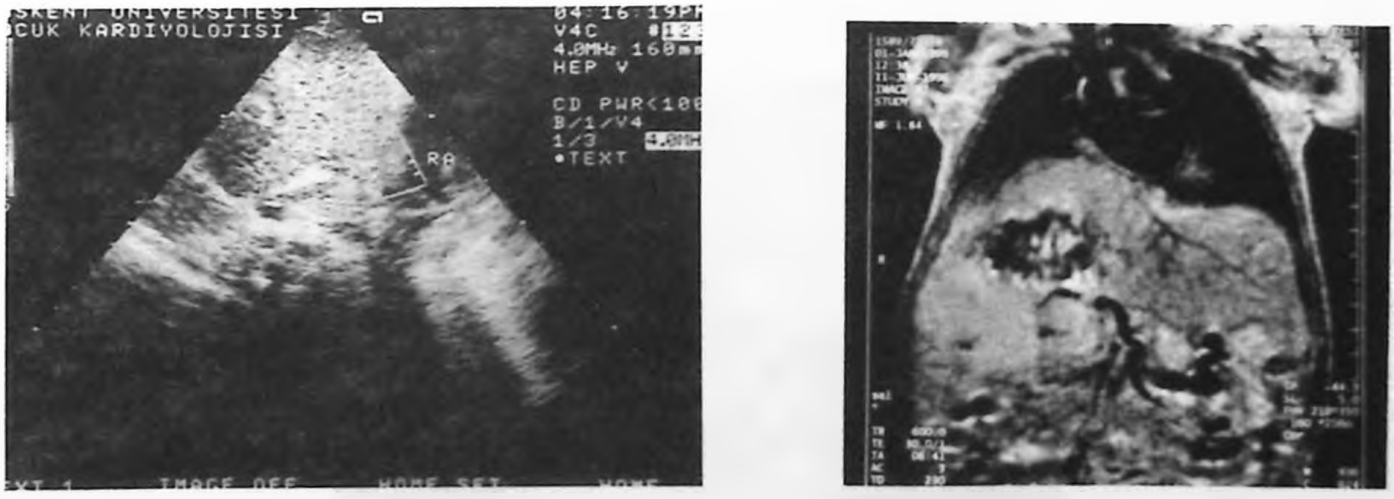

Fig. 3. Echocardiography shows one of the narrowed hepatic veins draining into the right atrium (a). Narrowed hepatic veins also noted on MRI [TR: 600 msec, TE: 30 msec, Acq: 3] (b).

Inferior vena cavography and visceral angiography were performed in order to confirm the vascular anomalies seen on CT, and to identify whether additional vascular anomalies were present. Vena cavography demonstrated interruption of the IVC at the level of the right renal vein, as well as azygous continuation. The azygous vein was dilated and was observed to be draining into the superior vena cava (Fig. 4). Visceral angiography examinations revealed that the common hepatic artery originated from the SMA, which was not demonstrated on CT (Fig. 5). MRI also showed the enlarged azygous vein, and bilateral bronchial symmetry with a longer right main bronchus, traits also consistent with left isomerism.

Discussion

Polysplenia syndrome describes a condition wherein more than two spleens of variable size are present, and where visceral heterotaxia and bilateral left sidedness are involved[1-2]. Polysplenia syndrome in conjunction with biliary atresia is a rare phenomenon, with only 10-20 percent of biliary atresia cases having such an association[2,3]. A condition seen more commonly with the syndrome is abdominal situs inversus, which usually includes cardiovascular, intestinal, and respiratory anomalies[1-3]. Recent studies have labeled polysplenia syndrome an acquired intrauterine anomaly of unknown etiology, stating that it

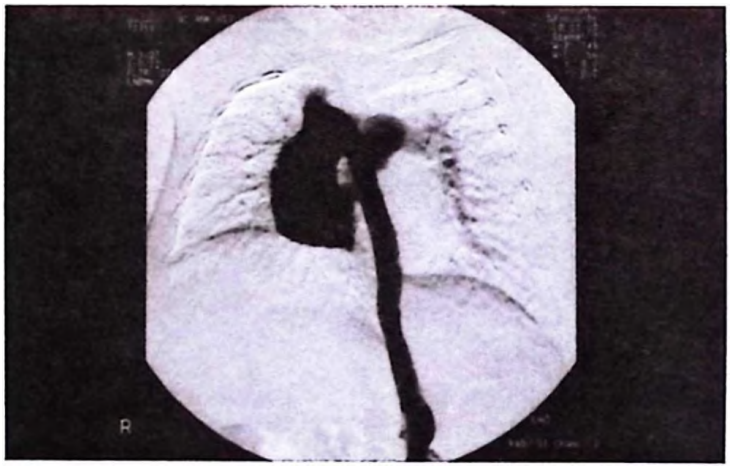

Fig. 4. Left anterior oblique inferior vena cavography showing dilated azygous vein continuation and drainage into the superior vena cava.

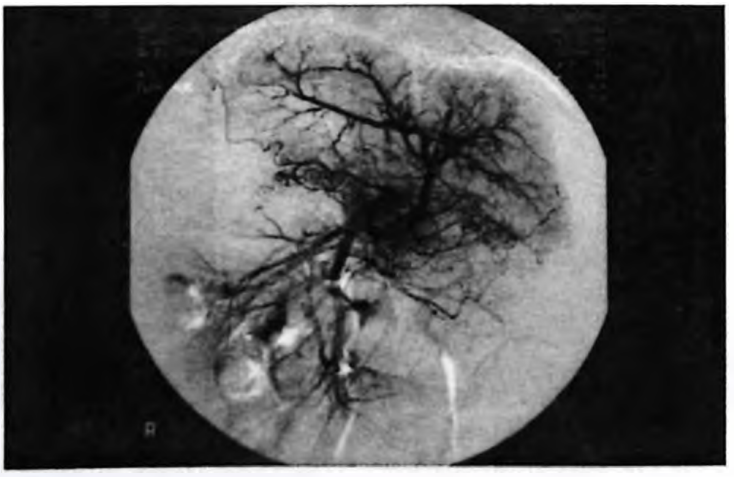

Fig. 5. Selective superior mesenteric artery (SMA) angiography shows the common hepatic artery originating from the SMA. Notice the slightly left-sided orientation of the liver.

occurs due to defective lateralization of abdominal organs and duplication of the left-sided ones[2,3]. The condition is thought to be the result of an intrauterine insult since no known cases of biliary atresia and polysplenia syndrome have been documented in autopsied stillborn infants[1]. It is also known that embryogenesis of the bile ducts, spleen, bronchial tree, cardiac septa and endocardial cushions, and formation of the IVC through the union of the subcardinal and hepatic veins occurs at roughly the fifth week of gestation[2,3].

Cardiac anomalies are more common than vascular types in polysplenia syndrome[1]. The most frequently seen vascular anomaly is interruption of the IVC with azygous continuation[2]. Other, less commonly described, vascular anomalies include preduodenal portal vein, bilateral superior vena cava[1,2], hepatic veins draining directly into the right atrium[2], pulmonary venous anomalies and anomalous origin of the right subclavian artery[3]. Anomalous hepatic arterial supply was mentioned in the study of Karrer and his collegues[3], but it was neither described in detail nor shown in any case.

Circumaortic renal vein is a vascular variation in which a two-limbed renal vein is present. The upper limb passes as normal in front of the aorta to join the IVC, and the lower limb passes behind the aorta. This malformation is seen in up to 17 percent of otherwise normal individuals[4]. Renal veins develop around the fifth to seventh week of gestation, which is around the same time as embryogenesis of the structures mentioned above[5]. The midline anastomosis of the supracardinal vein with the left renal vein when persists can be at the level of the left renal vein or more caudally located. In our case, the circumaortic renal vein was at the same level, with one limb passing in front of the aorta and the other passing behind (Fig. 2). Hepatic artery variations are also seen in normal individuals, at an incidence of 10 percent. In only 2.5 percent of these cases, the common hepatic artery originates entirely from the SMA[4]. To our knowledge, a case where the common hepatic artery originates at the SMA and a circumaortic renal vein is present as not been identified to date in connection with polysplenia syndrome. The case presented here included some of the more common anomalies of polysplenia syndrome: abdominal situs inversus, polysplenia, bilateral bronchial symmetry, and IVC interruption with azygous continuation. However, unique in terms of the literature were the patient's other vascular anomalies. Together with hepatic and renal vein variations, polysplenia syndrome and biliary atresia, being an uncommon anomaly as mentioned above, become more distinct as a case. These vascular anomalies were suspected on CT scans, and were confirmed by echocardiography, MRI, venacavography and abdominal angiography.

REFERENCES

1. Chandra RS. Biliary atresia and other structural anomalies in the congenital polysplenia syndrome. J Pediatr 1974; 85: 649-655.
2. Gagner M, Munson JL, Scholz FJ. Hepatobiliary anomalies associated with polysplenia syndrome. Gastrointest Radiol 1991; 16: 167-171.
3. Karrer FM, Hall RJ, Lily JR. Biliary atresia and the polysplenia syndrome. J Ped Surg 1991; 26: 524-527.
4. Kadir S. Esophago-gastrointestinal angiography, angiography of the kidneys. In: kadir S (ed). Diagnostic Angiography. Philadelphia: W.B. Saunders Company; 1986: 338-376, 445-495.
5. Trigaux JP, Vandroogenbroek S, De Wispelaere JF, Lacrosse M, Jamart J. Congenital anomalies of the inferior vena ccava and left renal vein: evaluation with spiral CT. JVIR 1998; 9: 339-345.