

A Case of Combined Intrahepatic Portosystemic Shunt, Midgut Malrotation, and Renal Rotational Anomaly in an Adult Patient

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An intrahepatic portosystemic venous shunt is a rare condition that was initially reported by Doehner et al. in 1956. In addition, the occurrence of midgut malrotation is rare in adulthood. The association of a portosystemic shunt along with a midgut malrotation and a renal rotational anomaly has not yet been described. We report a case of an intrahepatic portosystemic shunt, midgut malrotation, and renal rotation anomaly.

Index words : Portosystemic shunt

Portal vein

Intestine

Malrotation

Renal artery

Intrahepatic portosystemic venous shunt is generally a rare condition, except for cases of a dilated paraumbilical vein caused by portal hypertension, (1). Also, a midgut malrotation is rare in adulthood with a reported incidence of 0.2% (3). Midgut malrotation is usually an isolated abnormality, but it can be associated with congenital heart disease or situs problems (4). But, to our knowledge, there are no reports of associations of both abnormalities with kidney malrotation. We report, for the first time, a case of intrahepatic portosystemic shunt combined with midgut malrotation, and a renal rotation anomaly in a 44-year-old woman.

Case Report

A 44-year-old woman with frequent episodes of heart

palpitations when resting was referred for an evaluation. The palpitations lasted for up to 30 minutes and were accompanied by sweating and chest discomfort. The woman's vital signs and physical examinations were normal. A chest X-ray showed a non-specific finding except for mild cardiomegaly. A resting electrocardiogram (ECG) and serum cardiac markers like troponin, creatine phosphokinase, myoglobin, and lactate dehydrogenase were negative in testing for suspected heart disease. Coronary angiography results were almost normal. No atheroma was visualized throughout the coronary arteries, however a focal spasm was observed within the left circumflex artery (LCX). Thus, variant angina was suspected and medical therapy was initiated.

During admission, the patient requested that she receive a general health examination. She underwent an abdominal ultrasonography, which revealed a round cystic lesion ($\sim 4 \times 3$ cm) adjacent to the pancreatic body and a round heterogenous hyperechoic mass-like lesion (about $9 \times 7 \times 8$ cm) in the upper pole of the right kidney (Fig. 1).

At that time, a cystic lesion adjacent to the pancreatic body was regarded as a benign retroperitoneal lesion

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and the other mass-like lesion in right kidney was suspected as being a malignant mass.

The patient did not experience any trauma, nor had a

history of urinary tract infection or other underlying illness. Moreover, the patient was not a habitual drinker and a physical examination was unremarkable.

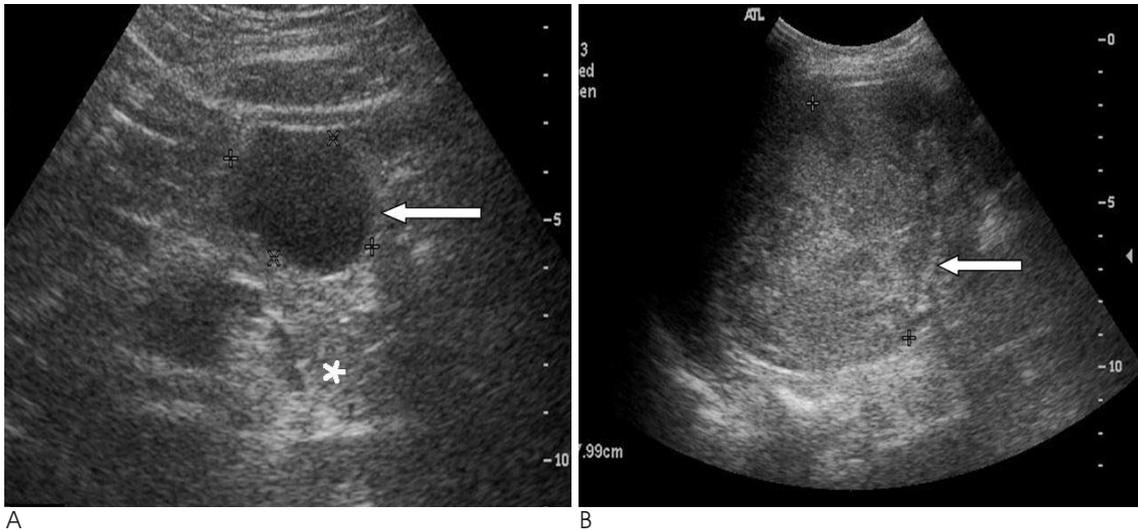


Fig. 1. A. Transverse sonogram demonstrates a round cystic lesion (arrow) (size: 4.2×3.0 cm) at the adjacent pancreatic body (*). B. Well-defined round heterogeneous hyperechoic mass-like lesion (arrow) ($\sim 9 \times 7 \times 8$ cm) in the upper pole of the right kidney.

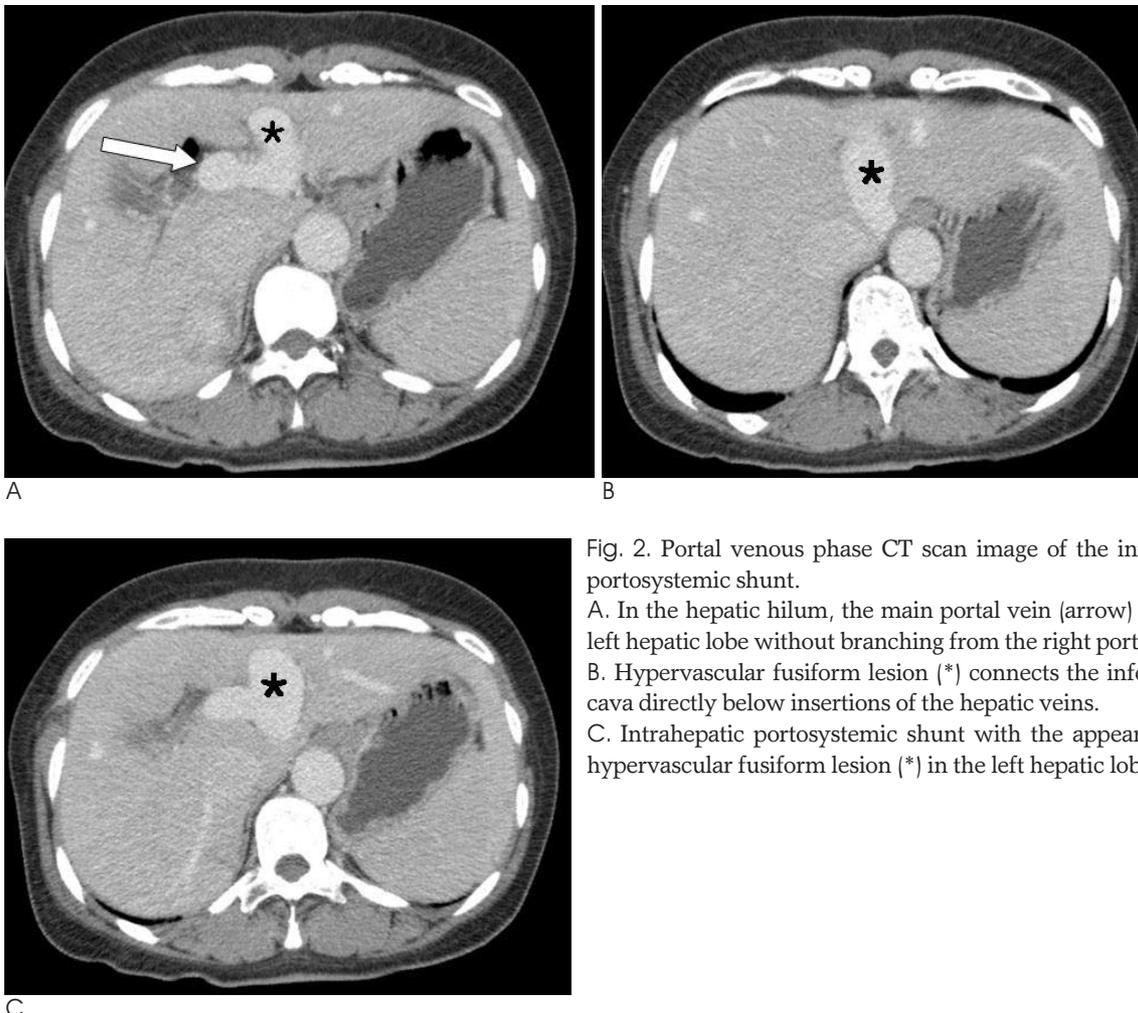


Fig. 2. Portal venous phase CT scan image of the intrahepatic portosystemic shunt.

A. In the hepatic hilum, the main portal vein (arrow) turns into left hepatic lobe without branching from the right portal vein.
B. Hypervascular fusiform lesion (*) connects the inferior vena cava directly below insertions of the hepatic veins.
C. Intrahepatic portosystemic shunt with the appearance of a hypervascular fusiform lesion (*) in the left hepatic lobe.

Laboratory data indicated that the patient had mild anemia; however, her serum BUN (blood urea nitrogen), creatinine, and urinalysis were all normal.

For further evaluation, she underwent an abdominal CT and digital subtraction portovenography. In summary, the CT imaging findings indicated that the entire portal vein was formed from the portosystemic shunt and the intrahepatic inferior vena cava. In the hepatic hilum, the main portal vein indicated fusiform dilatation, which turned into the left hepatic lobe without branching off from the right portal vein and entered the inferior vena cava directly, just below the point of the insertions of the hepatic veins (Fig. 2).

Moreover, a right ventrally malrotated kidney with a renal artery and vein which coursed ventrally to the kidney to enter the ventrally facing hilum was noted (Fig. 3).

A digital subtraction angiogram yielded a clear visualization of the portosystemic shunt. The portal vein was markedly dilated before entering the inferior vena cava. All portal blood flow was shunting to the systemic venous system through this shunt without supplying the liver (Fig. 4).

In addition, we had some findings on CT, UGI, and small bowel series, which were encountered by chance. Those examinations revealed the malrotation of the small bowel without evidence of the duodenum crossing the Treitz ligament. All small bowels were located on the right side of the abdomen and the "C" loop of the duodenum did not cross the midline (Fig. 5).

A renal artery angiogram showed a saccular form

aneurysm of right renal artery (10×15 mm), arising from branching point of the right main renal artery (Fig. 6).

The patient did not receive any treatment for the portosystemic shunt, intestinal malrotation, or renal artery aneurysm because she did not present with any symptoms consistent with shunt-like encephalopathy or intestinal malrotation. In addition, her renal function was normal.

Discussion

A congenital intrahepatic portosystemic shunt is characterized as an abnormal intrahepatic connection be-



Fig. 4. Digital subtraction portovenogram demonstrates the portosystemic shunt. The portal vein markedly dilated (arrow) before entering the inferior vena cava.

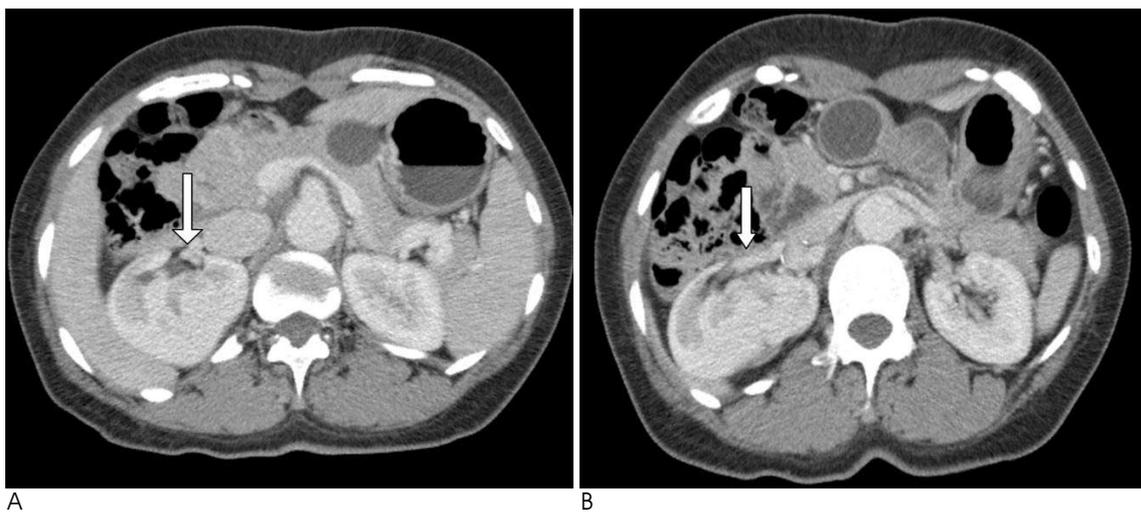


Fig. 3. Abdominal CT scan showing a malrotation of the right kidney.

A. Renal pelvis (arrow) is anteriorly located away from the midline.

B. Renal artery and vein (arrow) coursed ventrally to the kidney to allow entry into the ventrally positioned hilum.

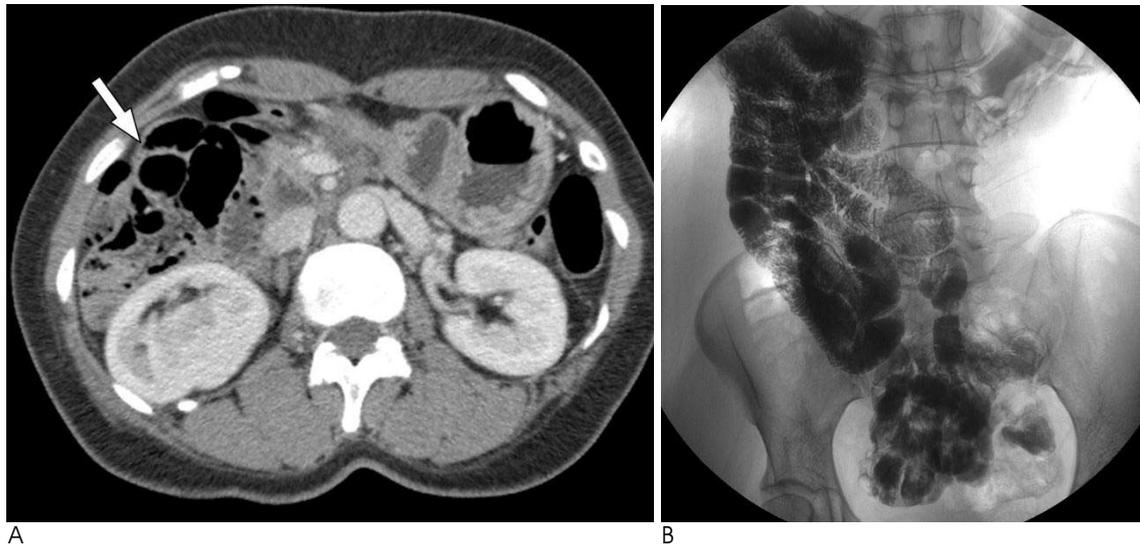


Fig. 5. A. CT scan thorough mid-abdomen with intravenous administration reveals the small bowel present only on the right side (arrow). B. Image from upper gastrointestinal series reveals the entire small bowel present in the right side of the abdomen without evidence of the duodenum crossing the lumbar spine.

tween the branches of the portal and hepatic veins. It is a rare condition that was first reported by Doehner et al. in 1956 (1, 2).

The cause of intrahepatic portosystemic venous shunts remains unknown. However, some authors postulate that persistent embryonic venous anastomoses such as the patent ductus venosus (5) or right vitelline vein (6) may be the contributing factors. Others advocate that the cause is a result of the rupture of a portal venous aneurysm into the hepatic vein (7), or from a dilated portal vein communicating with the inferior vena cava through the inferior phrenic or suprarenal vein (8).

Portohepatic venous shunts can be categorized into four different morphologic types (1): Type I - Single large shunt with a constant diameter that connects the right portal vein to inferior vena cava. Type II - localized peripheral shunt in which single or multiple communication is found between peripheral branches of the portal vein and hepatic veins in one hepatic segment. Type III - peripheral portal and hepatic veins are connected through an aneurysm. Type IV - multiple communications exist between portal and hepatic veins in both the lobes. Of the four types, type I is most common. Also, persistent ductus venosus could be considered as a type V portosystemic shunt.

Treatment options include dietary control, transcatheter embolization and surgical correction (1, 6). Our patient did not undergo treatment for the portosystemic shunt because she did not present with en-



Fig. 6. Renal artery angiogram shows a saccular aneurysm (*) arising from right main renal artery.

cephalopathy.

However, when an intrahepatic portosystemic venous shunt is presented with hepatic encephalopathy, the correct diagnosis is required prior to appropriate treatment (1).

Midgut malrotation is defined as an anomaly of fetal intestinal rotation and fixation of the midgut. It is usually diagnosed in the first month of life, but in rare cases, the diagnosis is not made until adulthood. The reported incidence of malrotation in adulthood is 0.2% (3).

Treatment remains as it was originally described by Ladd in 1936: mobilization of the right colon and duode-

num, division of Ladd's bands, division of adhesion around the superior mesenteric artery, and appendectomy (9).

Midgut malrotation is usually an isolated abnormality, but it can be associated with congenital heart disease or situs problems (4). To our knowledge, no syndromic or sporadic associations of a combination of abnormalities with intrahepatic portosystemic shunt and kidney malrotation has been described. Also, we encountered other findings, such as a renal artery aneurysm and a mild case of cardiomegaly. We presume these conditions were caused by the intrahepatic portosystemic shunt.

In summary, we report the case of an intrahepatic portosystemic shunt combined with midgut malrotation and a renal rotational anomaly.

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**성인에서 발생한 중장회전이상과 신장회전이상을 동반한
간내 문맥정맥단락: 증례 보고¹**

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김보미 · 한윤희 · 서정욱 · 차순주 · 허 감

선천성 문맥정맥 단락은 매우 드문 이상으로, 1956년 Doehner등에 의해 처음 보고되었다. 그리고 중장회전이상 또한 성인에서 보기 드문 현상이다. 중장회전이상과 신장회전이상을 동반한 문맥정맥단락은 현재까지 보고된 바 없는 이상현상으로 저자는 문헌 고찰과 함께 보고하는 바이다.