Macroscopic intrahepatic portosystemic venous shunts is defined as communications between the portal and the systemic venous circulation, measuring more than 1 mm in diameter, and at least partially located inside the liver. Portosystemic shunts are expected to be associated with portal hypertension, trauma, surgical intervention or liver biopsy, but incidentally seen otherwise normal individuals. These spontaneous intrahepatic portosystemic shunts (SIPSVS), except for dilated paraumbilical veins caused by portal hypertension, have generally been considered to be rare even though modern imaging modalities have successfully demonstrated these abnormalities (5,10,12,14,18,19,21,23,27,28). We describe a case with a SIPSVS incidentally diagnosed in check-up investigations.

**Case Report**

A 74-year-old female admitted to clinic for health screening. She had no history of trauma, liver biopsy and surgical operation. There had been no episode of hepatic encephalopathy or hypoglycemic symptom. Laboratory data showed normal blood counts, liver and renal function tests, glucose and lipid levels. Ultrasonography, an examination for screening, delineated a snail-like anechoic area in the liver, its connection with the portal and hepatic veins, and marked dilatation of the veins connecting with the shunt in an area with 7 cm diameter in the left lobe of the liver. Parenchymal echogenicity of the rest of the liver was normal and there was no focal parenchymal lesion. The main portal vein was patent and the course of the right and left portal veins was normal. Colour Doppler imaging showed turbulent flow in the portosystemic venous shunt. CT-angiography demonstrated an aneurysmatic SIPSVS of 23 mm diameter, between the left hepatic vein and the left portal vein in left lobe. Adjacent to that lesion, a couple of small (3–5 mm) SIPSVS were also seen (Figure 1). She was informed and reassured about that vascular abnormality. But, no intervention was performed because she did not have any symptom or finding.

One year later, the sonographic examination was repeated and no difference was found from the previous one. Then, she was undertaken follow-up.

**Discussion**

Although recent advances in USG and CT have detected asymptomatic SIPSVS in an increasing number of patients, SIPSVSs are rarely seen disorder. Park et al. categorized them into four different morphologic types (27). The most common type is a single large tube of constant diameter that connects the right portal vein to inferior vena cava. Type I includes patent paraumbilical veins, located in the liver. Shunts of this type are considered to be collateral pathways which develop in the hepatic parenchyma as a result of portal hypertension. The second type is localised peripheral shunt in which single or multiple communications
are found between peripheral branches of portal vein and hepatic veins in one hepatic segment. The third type is aneurysmal with peripheral portal and hepatic veins being connected through an aneurysm. The fourth type has multiple communications between peripheral portal and hepatic veins diffusely in both lobes. According to this classification, our patient had an aneurysmatic type 3 connection between left hepatic vein and left portal vein. Chevallier et al. did another classifications as follows: Type I includes patent paraumbilical veins, located in the liver, and commonly encountered in portal hypertension. Types II, III include shunts, unique or multiple, between the portal branch and the hepatic vein, located either in two adjacent liver segments (type II) or in non-adjacent liver segments (type III). Type IV corresponds to any tubular communication developed between the right portal branch and the inferior vena cava. The exceptional patent ductus venosus or a patent umbilical vein should not be considered as IPSVS since their course is strictly extrahepatic (4).

The diagnosis of intrahepatic portosystemic venous shunts can be established by color Doppler, CT-angiography, MRI or conventional angiography (8,10,25,27,28). Color Doppler imaging demonstrate a direct communication of color flow signals between the portal vein and hepatic vein, in addition to the characterization of the Doppler spectrum at each sampling point from a continuous waveform signal (portal vein) to a turbulent signal (aneurysmal cavity), and finally, to a biphasic waveform signal (hepatic vein). Color Doppler imaging is useful in the diagnosis of an intrahepatic portosystemic hepatic venous shunt, and the measurement of shunt ratio may be useful in the follow-up and determining the therapeutic option (19). Magnetic resonance imaging also can clearly demonstrate the portal-hepatic venous shunt due to “flow void”. Multiple diffuse shunts or a solitary shunt can be visualized. The solitary shunt can be either tubular, focally dilated or racemose in configuration (2,14,25,28). Color Doppler imaging and CT angiography are relatively non-invasive in diagnosis of SIPSVS.

The vascular malformation we present is believed to be a congenital anomaly, as no signs of cirrhosis or trauma were found. But, there are still many arguments about it. Even though certain embryological events remain obscure, certain developmental aberrations may cause this vascular connection. Some authors postulates a persistent venous anastomosis such as ductus venosus and right vitelline vein. Others advocate an acquired cause from rupture of a portal venous aneurysm into the hepatic vein or from a dilated hepatic vein communicating with the inferior vena cava via inferior phrenic and suprarenal vein (5,10,14,19,21,28,29). Persistent ductus venosus could be a remote possibility in our case. Rupture of the portal vein aneurysm is more probable mechanism. Another case was reported of a newborn with a shunt with spontaneous resolve in first year of age (8,13). SIPSVS may be demonstrated in any age of life between 20-day-old and late seventies (3,5,12,14,18,19,20,22,24,26,31).
Intrahepatic shunts may cause neurocognitive abnormality. A 23-year-old female patient with complaint of fatigue was reported to have right portal vein aneurysmal communication in otherwise healthy woman. Neuropsychological testing, imaging, and MR spectroscopy revealed changes similar to that described in patients with liver cirrhosis and subclinical hepatic encephalopathy. T1-weighted MRI showed a hyperintense globus pallidus, a feature seen in subjects with an without portal-encephalopathy. Portal systemic shunting in the absence of parenchymal liver disease reproduces neurological features described in cirrhosis (6). Because, she did not accept, we have not performed cerebral investigations in our patient.

This vascular abnormality accounts for the blood glucose problems: the portocaval shunt explains the early hypoglycemia by defective liver uptake of glucose and secondary hyperinsulinism because of the reduced hepatic degradation of the insulin secreted in normal quantity. The late hyperinsulinism then leads to secondary hypoglycemia. These cases may benefit dietetic treatment (7,9,12,18).

The size of SIPSVS has been reported to probably increase in time. However, in a year we have not seen a significant change in the lesion of our patient. But, this issue remains to be defined with prospective studies.

In the literature, there are several reports of distinctive association with SIPSVS such as congenital biliary atresia, polycystic ovary syndrome, huge pelvic myoma, coronary artery fistulas, leptospirosis, hemangiomas and membranoproliferative glomerulonephritis. There are cases of SIPSVS associated with focal nodular hyperplasia or hereditary hemorrhagic telangiectasia (HHT) (1,15,16,30,32). HHT is an autosomal dominant disorder characterized by telangiectasias and arteriovenous malformations of potentially every organ (3). Hepatic involvement occurs in 8–31% of cases. Hepatic involvement is shown by biopsy, sonography, Doppler sonography, CT, and MR imaging. This pictorial essay illustrates the broad spectrum of abnormalities of hepatic vessels and collaterals in HHT that are detectable by imaging techniques even in the early or clinically silent stages of the diseases. The association between the hepatic vascular lesions and HHT is varied, ranging from telangiectasias to large shunts between three vascular channels. In an advanced stage of involvement, large portovenous shunts may be present. In another case, development of pulmonary hypertension was accepted to be due to SIPSVS. Because, it was assumed that vasoconstrictive agents which should be metabolized by the liver in normal subjects, passing through the intrahepatic shunt into the lung (11). Our patient had no abnormality that have been demonstrated. She had no complaint. Our patient underwent no treatment for the portosystemic shunt because she did not present with hepatic encephalopathy. That’s why she was undertaken follow-up. One year later, there was nothing changed. Then we planned to follow her with intervals of two years.

In conclusion, clinicians should not be confused in incidental diagnosis of SIPSVS. However, when intrahepatic portosystemic shunt does present with hepatic encephalopathy, the correct diagnosis is required prior to appropriate treatment.

References

1. Aiba N, Moroka J, Myazono T, Okita H, Yata Y, Okuda K, Nambu S, Iznini R. Case Report: intrahepatic portal-hepatic venous shunts associated with a large gastric lesion. J Gastroenterol Hepatol 1998;13(2):158–62.
2. Araki T, Otomo K, Kachi K, Monzawa S, Hikara T, Ohba H, Aionod T, Kumagai H. Magnetic resonance imaging of macroscopic intrahepatic portosystemic venous shunts. Gastrointest Radiol 1991;16(3):221–4.
3. Buscarini E, Buscarini L, Cirvadi G, Arruzolla S, Bosiaini G, Pandiania M. Hepatic vascular malformations in hereditary hemorrhagic telangiectasia: imaging findings. AJR Am J Roentgenol 1994;163(5):1055–10.
4. Chevalier P, Oelde F, Souci J, Duine B, Pavodani B. Macroscopic intrahepatic portosystemic venous shunt. Pediat Radiol 1991;21(7):529–30.
5. Cosme-Jimener A, Bujianda Fernandez de Perialo L, Poch Zapirain M, Ordoloca Alba R, Ojeda Perez E, Arenas Mrica JI. Congenital intrahepatic venous shunt as a cause of hepatic encephalopathy. Gastroenterol Hepatol 1995;18(9):460–3.
6. Crepin J, Nemeck A, Reikemper G, Blei AT. Intrahepatic portal-hepatic venous anastomosis: a portal-systemic shunt with neurological repercussions. Am J Gastroenterol 2000;95(6):1568–71.
7. Dupre J, Gounin B, Benazet MF, Le Gal J. Glucol intolerance and post stimulatory hypoglycemia secondary to a probably congenital intrahepatic portocaval anastomosis. Ann Med Intern 1985;116(8):655–8.
8. Gitzelmann R, Forster I, Willy UV. Hyperglicaglossum in a newborn: self limiting intrahepatic portosystemic venous shunt. Eur J Pediatr 1997;156(9):719–22.
9. Gounin B, Le Gal J, Dupre J, Sanson J. Congenital intrahepatic portocaval anastomosis: analysis of manifested glucose abnormalities. Gastroenterol Clin Biol 1984;8(5):464–8.
10. Grattagliano A, Rapaccini GL, Camaldo G, Pompili M, Marino P, Mastromatteo AM, Cotroneo AR, Gubarlari G. Spontaneous intrahepatic portosystemic venous shunt in a patient with cirrhosis: diagnosis by combined color Doppler and pulse Doppler ultrasonography. Liver 1997;17(6):307–10.
11. Hondo T, Teragawa H, Mizumori M, Morishima N, Watanabe H, Ogata S, Ohe H, Yoshikawa M, Ohbayashi M. Portal hepatic venous shunt through a portal aneurysm complicated by hepatic encephalopathy and pulmonary hypertension. Intern Med 1997;36(11):790–1.
12. Horiguchi Y, Kitano T, Imul H, Ohshiki M, Yamauchi M. Intrahepatic portal-systemic shunt: its etiology and diagnosis. Gastroeneterol Jpn. 1987; 22(4):496–502.
13. Jabra AA, Taylor GA. Ultrasonographic diagnosis of congenital intrahepatic portosystemic venous shunt. Eur J Pediatr 1997;21(7):529–30.
14. Kakitsubata Y, Kakitsubata S, Kiyomizu H, Ogawa T, Kato T, Watanabe K. Intrahepatic portal-hepatic venous shunts demonstrated by US, CT, and MR imaging. Acta Radiol 1994;35(7):680–4.
15. Kantarcio F, Milmanian I, Kora B, Cantadídmir M, Adáiller I. Spontaneous intrahepatic portosystemic venous shunt in leptospirosis: is it rare association or coincidence? Eur Radiol 2000;13 Suppl 4:L235–6.
16. Karashima S, Hattori S, Nakazato H, Awata H, Séguchi S, Reda S, Sera Y, Endo F. Membranoproliferative glomerulonephritis in congenital portosystemic shunt without liver cirrhosis. Clin Nephrol 2000;53(3):206–11.
17. Kim IO, Cheon JE, Kim WS, Chung JW, Yeon SJ, Seo JK, Choi JH. Congenital intrahepatic portosystemic venous shunt: treatment with coil embolization. Pediatr Radiol 2000;30(5):336–8.
18. Kousa S, Sassa R, Kakumu S. An enormous intrahepatic shunt between portal and hepatic veins. Angiology 1975;26(4):365–71.
19. Kudo M, Tomita S, Tochio H, Minowa K, Todo A. Intrahepatic portosystemic venous shunt: diagnosis by color Doppler imaging. Am J Gastroenterol 1993;88(5):723–9.
20. Kudo M. Intrahepatic portosystemic venous shunt in liver cirrhosis: is it congenital or acquired? AJR Am J Roentgenol 1993;160(2):421–2.
21. Maeda T, Mori H, Aikawa H, Komatsu E, Kagawa K. Therapeutic embolization of intrahepatic portosystemic shunts by retrograde transcaval catheterization. Cardiovasc Intervent Radiol 1993;16(4):245–7.
22. Materne R, Van Beers BE. Images in clinical radiology. Spontaneous intrahepatic portosystemic venous shunt and focal nodular hyperplasia. J Belge Radiol 1998;81(4):180.
23. Matsumoto R, Izutsu M, Kobayashi S, Kusano S. A case of multiple intrahepatic portosystemic venous shunts associated with multiple hemangioma-like lesions of the liver. Rinsho Hoshasen. 1990;35(9):1085–8.
24. Mori K, Dohi T, Yamamoto H, Kamada M. An enormous shunt between the portal and hepatic veins associated with multiple coronary artery fistulas. Pediatr Radiol. 1990;21(1):66–8.
25. Oguz B, Akata D, Balkanci F, Akhan O. Intrahepatic portosystemic venous shunt: diagnosis by color/power Doppler imaging and three dimensional ultrasound. Br J Radiol. 2003;76(907):487–90.
26. Paley MR, Farrant P, Kane P, Heaton ND, Howard ER, Karani JB. Developmental intrahepatic shunts of childhood: radiological features of management. Eur Radiol 1997;7(9):1377–82.
27. Park JH, Cha SH, Han JK, Han MC. Intrahepatic portosystemic venous shunt. Am J Roentgenol. 1990;155(3):527–8.
28. Santamaria G, Pruna X, Serres X, Inaraja L, Zuasnabar A, Castellote A. Congenital intrahepatic portosystemic venous shunt: sonographic and magnetic resonance imaging. Eur Radiol. 1996;6(1):76–8.
29. Satoh M, Yokoya S, Hachiya Y, Hachiya M, Fujisawa T, Hoshino K, Saji T. Two hyperandrogenic adolescent girls with congenital portosystemic shunt. Eur J Pediatr 2001;160(5):307–11.
30. Serrien B, Rigauts H, Marchal G, Lambrechts P. Congenital intrahepatic portosystemic shunt. J Belge Radiol 1992;75(6):492–4.
31. Tanour S, Kyouss H, Komatsu E, Hori Y. Symptomatic intrahepatic portosystemic venous shunt: embolization with an alternative approach. AJR Am J Roentgenol 2003;181(1):71–8.
32. Yamagami T, Nakamura T, Tokiwa K, Ohno K, Inoh H, Maeda T. Intrahepatic portosystemic venous shunt associated with biliary atresia: case report. Pediatr Radiol. 2000;30(7):489–91.

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