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Semra DURAN[1]
Betül AKDAL[1]
Alper DİLLİ[2]

Case Report

INTRAHEPATIC PORTOSYSTEMIC VENOUS SHUNT

Abstract

Portosystemic shunts can be classified as intrahepatic and extrahepatic according to their anatomic features. Intrahepatic portosystemic shunts are rare vascular anomalies occurring between the portal vein and hepatic vein/ inferior vena cava, and can be congenital or acquired. With recent advances in diagnostic imaging techniques, the number of reports of intrahepatic portosystemic venous shunts identified incidentally in patients without symptoms is increasing. This is clinically important because intrahepatic portosystemic venous shunt can lead to hepatic encephalopathy. The rate of hepatic encephalopathy increases with age because of decreasing tolerance of the brain to toxic metabolites. The correct radiological diagnosis and proper treatment of this unusual abnormality is important. We present a case of noncirrhotic patient with a diagnosis of intrahepatic portosystemic venous shunt, accompanied by imaging findings.

Key Words: Patent Ductus Venosus; Ultrasonography, Doppler; Tomography, Spiral Computed; Magnetic Resonance Imaging.

Olgu Sunumu

İNTRAHEPATİK PORTOSİSTEMİK VENÖZ SANT

Öz

Portosistemik santlar anatomik özelliklerine bağlı olarak intrahepatik ve ekstrahepatik olarak sınıflandırılabilir. İntrahepatik portosistemik şant portal ven ve hepatic ven/ inferior vena cava arasında meydana gelebilen nadir rastlanan vasküler anomalilerdir ve konjenital veya kazanılmış olabilir. Tanısal görüntüleme yöntemlerindeki gelişmeler ile, semptomu olmayan hastalarda insidental olarak saptanan intrahepatik portosistemik venöz şantların sayısı giderek artmaktadır. Bunlar klinik olarak önemlidir çünkü intrahepatik portosistemik venöz şantlar hepatik ensefalopatiye neden olabilir. Hepatik ensefalopati oranı yaş ile artar çünkü beynin toksik metabolitlere olan toleransı azalır. Bu vasküler anomalilerin radyolojik tanısı ve uygun tedavisi bu açıdan önemlidir. Biz intrahepatik portosistemik venöz şant tanısı alan karaciğer sirozu bulunmayan olguyu görüntüleme bulguları eşliğinde sunmayı amaçladık.

Anahtar Sözcükler: Portosistemik Venöz Şant; Ultrasonografi, Doppler; Tomografi, Spiral Komputerize; Manyetik Rezonans Görüntüleme

İletişim *(Correspondance)*

Sema DURAN
Ankara Numune Education and Research Hospital, Clinic of Radiology ANKARA

Tlf: 0312 508 48 71
e-posta: sduran@isnet.net.tr

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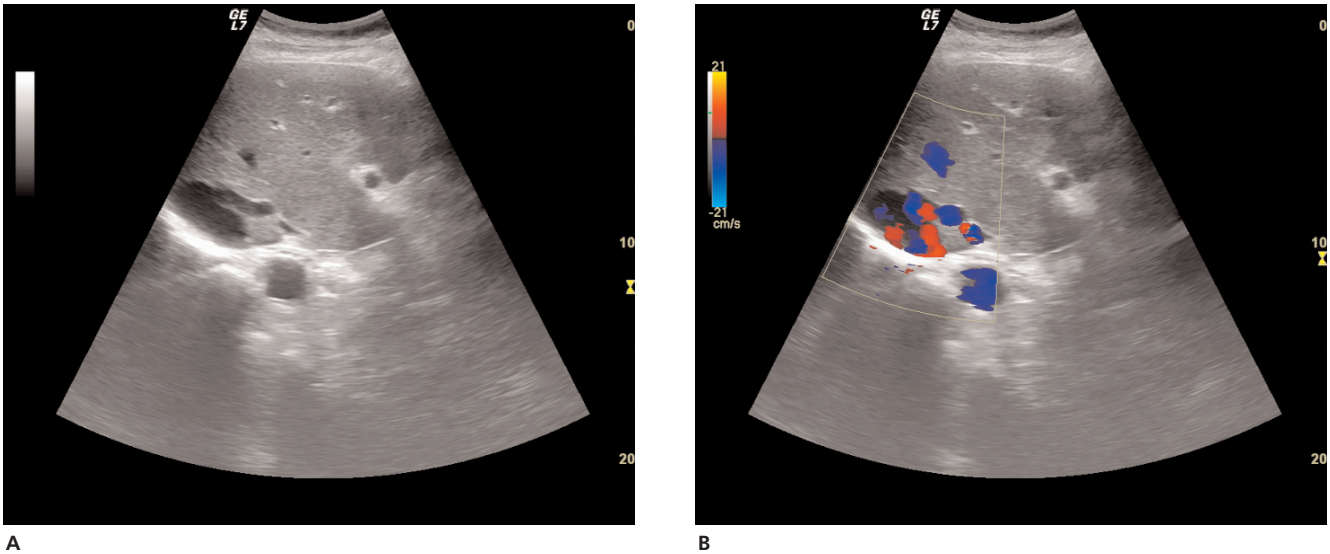
[1] Ankara Numune Education and Research Hospital, Clinic of Radiology ANKARA
[2] Ankara Dıskapı Yıldırım Beyazıt Education and Research Hospital, Clinic of Radiology ANKARA

Introduction

Portosystemic shunt are classified into two types, according to their anatomical features, as intrahepatic venous type and extrahepatic venous type (1). Extrahepatic shunts, which provide communication between the systemic vein and portal vein in cases with portal hypertension, occur with significant frequency. Coronary vein, esophageal varices and retroperitoneal collaterals are among the sites of extrahepatic shunts (2, 3). Intrahepatic portosystemic venous shunts (IPSVS) are abnormal communications between branches of the portal vein and the hepatic vein. They are rarely- encountered vascular anomalies which can be congenital or acquired (from trauma, cirrhosis, portal vein aneurism rupture, or percutaneous biopsy) (3, 4). Asymptomatic cases are increasingly detected with more frequent use of diagnostic modalities such as ultrasonography (US), computerized tomography (CT), and magnetic resonance imaging (MRI). The correct radiological diagnosis and proper treatment are clinically important, as these lesions may cause hepatic encephalopathy (HE) (3,4-8). Here we present a case of noncirrhotic patient in whom IPSVS was incidentally detected , accompanied by imaging findings.

Case

A 71 year-old female patient was admitted to our hospital following the identification of a hyperintense lesion in liver in T2- weighed images obtained by surrenal MRI, at another institution she had attended for hypertension. She did not have any history of trauma, operational procedure such surgery or biopsy , or alcohol use. Physical examination of the patient revealed hypertension (TA=160/100 mmHg). The liver was not enlarged , there was no evidence of ascites or symptoms of encephalopathy. No abnormalities were detected in the laboratory findings (aspartate aminotransferase (AST): 27U/L, alanine aminotransferase(ALT):25 U/L, gamma glutamyltranspeptidase (GGT): 35 U/L, alkaline phosphatase (ALP): 72U/L, lactate dehydrogenase: 166 IU/L, total bilirubin: 0.4mg/dL, albumin: 41 g/L). Tumor and hepatitis markers were negative (AFP:2.2 ng/mL, CEA: 1.57 ng/mL, HBs Ag:0.00(-), HCV Ab: (-)) and ammonia level was normal (20 microg/ml). In the abdominal US, a cystic lesion 3 cm in diameter at the 7th segment of the liver, and a tubular anechoic structure connected to the right portal vein in the vicinity, were observed. In the color Doppler US, color coding in the lesion (Figure 1A-B) and non-phasic current with low speed in the aneurismal section were detected. The dynamic abdominal MRI visualized the vascular communication between the right portal vein and the hepatic vein, while the aneurismatic lesion was detected in the liver by US. In dynamic images, the aneurysmal lesion showed simultaneous filling with the portal vein (Figure 2A-C). A finding of intrahepatic portosystemic venous shunt was considered for this patient. Neither cirrhotic nodules nor malignant mass lesions were detected in the liver. Collateral structures were observed



A B

Figure 1— Blood flow is observed **(A)** in US, in the cystic lesion detected in the 7th segment of the liver, and in the tubular structure connecting right portal vein to the lesion **(B)** in Color Doppler US.

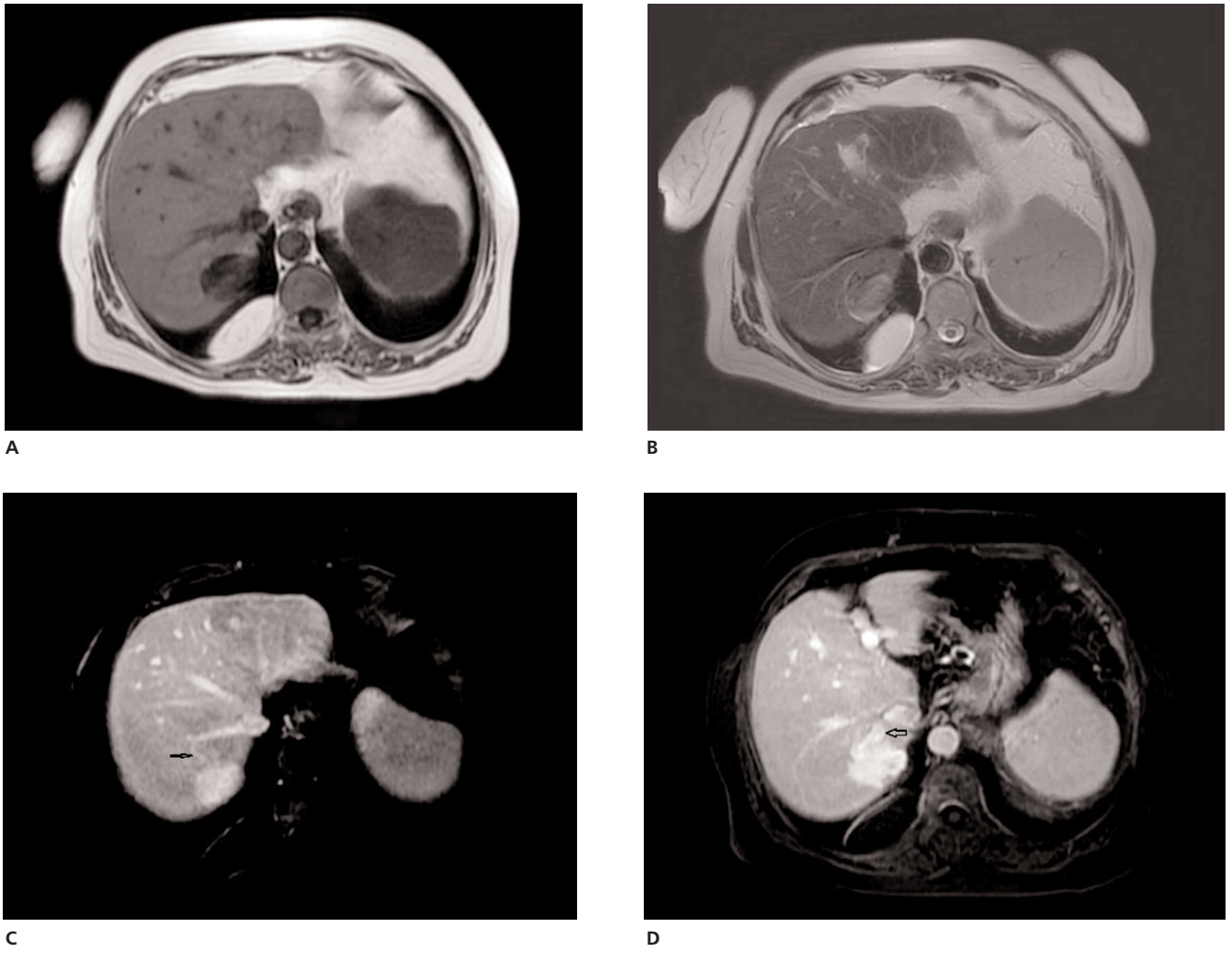


Figure 2— The communication of the aneurismatic lesion detected in Doppler US with the right portal vein (thick arrow) and the right hepatic vein (thin arrow) is seen (Park Type III) in **(A)** T1 weighted axial **(B)** T2 weighted axial **(C)** post-contrast fat suppression T1 weighted axial (portal venous phase) images.

in the splenomegaly and splenic hillus. The umbilical vein was open. Grade 2 esophageal varices were found in the endoscopy. The patient was assigned to follow-up with a dietary arrangement.

Discussion

The portosystemic venous collaterals observed in patients with portal hypertension are mostly extrahepatic (3). IPSVS is a rare entity; in approximately 40% of cases the shunts have been associated with liver cirrhosis and portal hypertension, and in 50-60% of cases with cerebral manifestations related to portohepatic encephalopathy (3,4,6,9). The pathogenesis of these vascular abnormalities is controversial. When a portal vein- hepatic vein communication is seen in a patient without liver disease or a history of trauma, it is presumed to be spontaneous or congenital in origin. The theory of congenital development of IPSVS suggests that an anastomosis exists between the subcardinal venous system and vitelline venous system in an early stage of embryologic development (3,4,6,9-11). Others suggest that a rupture of the portal vein aneurysm into the hepatic vein is the cause (3, 4).

Park et al. classified IPSVS into 4 groups (12). In Type I, a single wide vein is present between the right portal vein and

inferior cava. In Type II, single or multiple communications are observed between the peripheral branches of portal and hepatic veins in a single hepatic segment. In Type III, peripheral portal and hepatic vein branches are connected to each other through an aneurysm. In Type IV, there are multiple communications between peripheral portal and hepatic veins in both lobes of the liver. Our case was consistent with Type III of the Park classification. Although Park et al. mentioned Type I as the most frequent type of shunt, Tsitouride et al. detected Type III shunts in 5 of the 8 cases in their study (6). Remer et al. found Type III shunts in 13 of their series of 22 cases (3).

The prevalence of IPSVS as an incidental finding in asymptomatic patients is unknown. Some authors report that the prognosis of IPSVS depends on the shunt ratio and patient's age (5-7). In most reported cases, patients were over 50 years old (3,6,9). This is clinically important because IPSVS can lead to HE. Although many young patients do not develop HE when the shunt rate is low, the cerebral tolerance for hepatotoxic substances decrease gradually with age and increase the patient's risk for HE, particularly in elderly patients (1,3,4-7,11) .The pathophysiology of HE caused by portosystemic shunts involves substances that, instead of being normally metabolized by the liver, are shunted directly into the systemic circulation (11). Radiological diagnosis of these lesions is important in treatment planning (3,4,6). IPSVS are usually detected on sonographic examination of the liver, as cystic or tubular anechoic structures and their vascular nature can be inferred by continuity with the intrahepatic vessels (9). Small aneurismal lesions may be mistakenly diagnosed as hepatic cysts in US (2, 11). Color and power Doppler are diagnostic since they demonstrate the vascular features of the abnormalities detected sonographically, showing the presence of blood within the cystic or tubular lesion. Furthermore, spectral tracings and velocity measurements can be used to evaluate the hemodynamics of the shunt (2, 9,10). When the shunt ratio exceeds 60%, the risk of developing HE is increased, and a portal shunt ratio more than 60%, even without encephalopathy, is an indication for therapeutic intervention in noncirrhotic cases (4). In our case, the blood flow in a cystic lesion associated with the portal vein detected in US was shown using Doppler US and hepatic vein communication was found in dynamic MRI, applied after pre-diagnosis of the portal vein aneurysm.

Multiphase helical CT and dynamic MRI represent advances in cross-sectional imaging that allow evaluation of the liver during arterial and portal venous phases of contrast enhancement (9,11). The Type III IPSVS observed in our case may be identified through the hepatic vein branch that provides early venous drainage to the hepatic vein and dilated portal vein branch associated with venous aneurism in dynamic MRI (2, 3,6,9,13). The major advantage of this modality is multiplanarity, and MR angiography seems useful in evaluating the relationships of the shunt to the portal and hepatic vessels (3,6,9). Angiography is preferred in cases where endovascular treatment would be used (9). The selection of treatment in IPSVS is controversial. In the treatment of symptomatic shunts, diet control and coil embolization by angiographic intervention or surgical methods may be preferred. Among these surgical methods are liver resections and shunt ligation. Surgical methods have higher morbidity and mortality rates compared to coil embolization (4,6,10).

In conclusion, noncirrhotic IPSVS is a rare vascular abnormality. The elderly patients with HE can be misdiagnosed as dementia, psychiatric disorders or irrational liver damage and to make matter worse, ineffective therapies such as medications and dietary restrictions are given or advised. The accurate diagnosis of IPSVS and an awareness of this disease are important.

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