

Imaging Findings of Osler-Weber-Rendu Disease Involving the Liver¹

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A 62-year-old woman with hereditary hemorrhagic telangiectasia(HHT) or Osler-Weber-Rendu disease involving the liver is presented. Imaging findings including color Doppler sonography and CT findings are described.

Index Words : Liver, CT
Liver, US
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Liver, diseases

Osler-Weber-Rendu disease or hereditary hemorrhagic telangiectasia(HHT) has been known to be an autosomal dominant disorder characterized by telangiectases and arteriovenous malformations of multiple organ systems(1). Hepatic involvement occurs in 8—31 % of cases with HHT and includes telangiectases or arteriovenous fistulas associated with fibrosis and cirrhosis(2, 3). Recent reports demonstrate various imaging findings including Doppler sonography with color flow signals, CT, and MR findings of the liver involvement in patients with HHT(2-4).

A few cases with HHT had reported in Korea(5). We describe the findings of conventional and Doppler sonography, and computed tomogram(CT) in a case with HHT involving the liver.

CASE REPORT

A 62-year-old woman was admitted because of hematemesis and melena for 4 days prior to the admission. She also had a history of intermittent epigastric pain with a duration of 30 years. Physical examinations showed facial telangiectasia and macules on the tongue and oral mucosa. Liver was palpable with three finger breadths. Liver function tests were normal except for an albumin value of 2.7 g/dl. On gastrofiberscopic examination, multiple telangiectases were noted in the gastric body and antrum, and active ulcer

crater was seen in the duodenal bulb. Her mother also had telangiectases in the face and oral mucosa.

Sonogram showed that celiac trunk and common hepatic artery were dilated with numerous tortuous intrahepatic branches as well as distension of hepatic veins. There was a suspicion of cystic dilatation in the posterior superior segmental branch of right hepatic artery(Fig. 1) The portal vein appeared normally. Duplex sonogram revealed a high velocity signal in the hepatic artery and arterialized pattern of flow signal in the hepatic veins, suggesting the communication between hepatic arteries and veins(Fig. 2). The portal flow was normal. Color Doppler imaging showed high color signal in the tortuously dilated intrahepatic arteries(Fig. 3). Spiral volumetric CT scans obtained with 3 mm slice thickness and 4 mm table feed after administration of contrast material at a rate of 3 ml/s for 40 s with scanning initiated 30 s after the start of injection. CT scans demonstrated dilated intrahepatic arteries and simultaneous opacification of hepatic veins in the arterial dominant phase(Fig. 4). There were also inhomogeneous perfusion of the hepatic parenchyma and an saccular dilatation of hepatic artery branch in the right posterior superior segment. CT angiogram obtained by maximum intensity projection and multiplanar reformation showed dilated, tortuous hepatic arteries, aneurysm in the posterior superior segment of right lobe of the liver, and simultaneous visualization of hepatic veins(Fig. 5).

Hepatic arteriogram with subtraction(Fig. 6) confirmed aneurysm, diffuse telangiectasia, and arteriovenous shunts by showing simultaneous enhancement of the arteries and the hepatic veins with inhomogeneous staining of hepatic parenchyma.

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Fig. 1. Sonogram of right lobe of the liver shows tortuously dilated tubular structures parallel to portal vein. These structures communicate with regional cystic lesion(arrow).

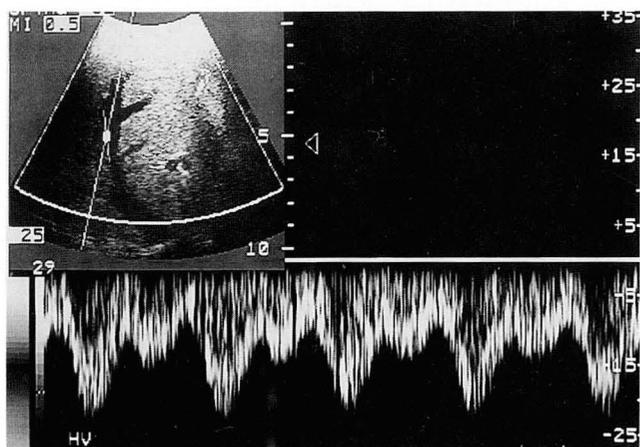


Fig. 2. Duplex sonogram of the hepatic vein shows "arterialized" high-velocity signal.

DISCUSSION

Osler-Weber-Rendu disease or hereditary hemorrhagic telangiectasia is a rare vascular disease with multiple organ manifestations. It occurs most commonly in the oral mucosa(79%) or skin(61%), but digestive tract(34%), lung(26%), liver(15%), or brain(6%) can be involved as well(7). Various forms of vascular abnormalities can be developed, which include angiodysplasia, telangiectasia, arteriovenous malformation, and aneurysm formation(8). Hepatic involvement of the lesion results in fibrosis or cirrhosis. The angiodysplasias in the liver are thought to be dilated capillary and venous channels lined by a single endothelial layer with absent muscular and elastic tissue(8).

Classic diagnostic triad of HHT is a family history, tel-

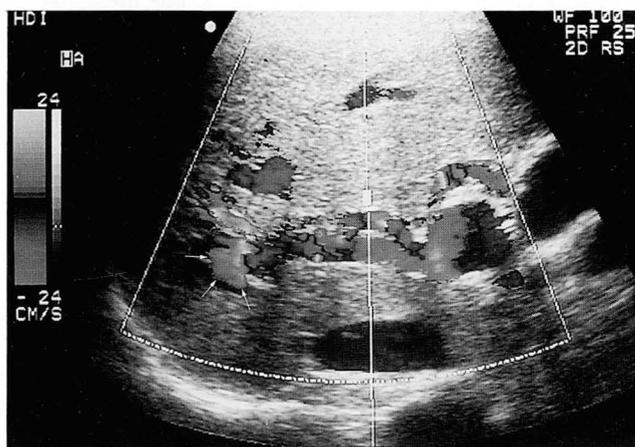


Fig. 3. Color Doppler sonogram shows turbulent high-velocity flow within tortuous tubular structure shown in Fig. 1. Aneurysm formation is suspected in the posterior superior segment of right liver(arrow).



a



b

Fig. 4. a. Spiral CT scan shows dilated intrahepatic arteries(straight arrows), and simultaneous visualization of hepatic veins(curved arrows) in early arterial phase. Aneurysm formation is suspected in posterior superior segment of right liver(arrowhead). b. At a level of pancreas, CT shows dilated celiac trunk and very inhomogeneous perfusion of liver parenchyma.



Fig. 5. Coronal CT angiogram with maximum-intensity-projection reconstruction technique clearly shows anomalous hepatic artery as well as saccular aneurysm (large arrow) and simultaneous visualization of hepatic veins (small arrows).

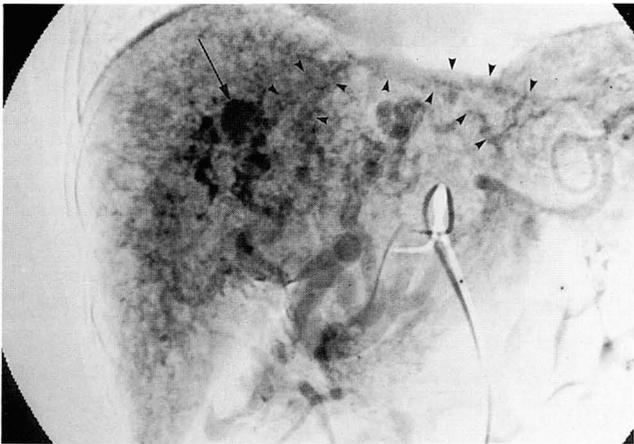


Fig. 6. Celiac angiogram at arterial phase confirms hepatic vascular abnormalities including diffuse telangiectasia and saccular aneurysm formation (arrow). There are early filling of hepatic veins (arrowheads).

angiectasia, and recurrent episode of bleeding(4). The most common episode is epistaxis with onset in childhood although our case did not have such history. Telangiectases are friable vessels that easily rupture and bleed(5). In our case, hematemesis and melena caused by either rupture of gastric telangiectases or duodenal ulcer were confirmed on gastrofiberscopy.

Imaging findings of the liver involvement of HHT depends largely on the severity of vascular malformations. In cases with arteriovenous malformation are demonstrated large feeding arteries, pulsating and ectatic vascular structures, and large draining veins in sonography(4-6). Doppler sonography with color signals can directly visualize the arteriovenous connections between hepatic artery branches with hepatic veins or portal veins(2). In the cases with absence of discrete arteriovenous fistulas, arteriovenous communications can be suggested by the dilated hepatic veins

with arterialized pattern of flow signal on Doppler sonography, or enlarged hepatic arteries with simultaneous enhancement of the hepatic veins on CT and hepatic angiography(2, 3). In our case, arteriovenous shunting seemed to be present distally because discrete arteriovenous fistula was not directly visualized, and so we expected that the type of arteriovenous shunting was capillary angiodysplastic lesions.

Pulsed and color Doppler sonograms have a great advantage of simultaneous visualization of the gray scale image and flow signals. It is also useful in distinguishing dilated vessels from biliary tree as in this case and otherwise, the diagnosis might be missed as biliary tract obstruction. Color Doppler sonogram easily confirmed dilated tubular structures as hepatic arteries.

Although presence of vascular malformations and arteriovenous shunts on Doppler sonogram may be enough for the diagnosis of the liver involvement of HHT, CT with dynamic scans could confirm vascular abnormalities in these patients. Recently, spiral volumetric CT can maximize the effect of contrast enhancement and is useful in easily performing the dynamic scans at various vascular phases and CT angiograms. In this patient, spiral CT and CT angiogram (Fig. 4 & 5) obtained with post processing clearly demonstrated tortuous, dilated extra and intrahepatic artery, inhomogeneous parenchymal enhancement (diffuse telangiectasia), and early visualizations of hepatic veins (arteriohepatic shunting). Those findings are nearly same with celiac arteriography (Fig. 6).

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유전성 출혈성 혈관확장증:간병변의 방사선학적 소견¹

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Osler-Weber-Rendu 씨병 또는, 유전성 출혈성 혈관확장증(hereditary hemorrhagic telangiectasia)은 매우 드문 질환으로서 체내 여러 장기에 혈관기형을 일으킨다. 저자들은 간을 침범했던 유전성 출혈성 혈관확장증환자 1 예를 경험하여 보고하는 바이다.