Hepatoportal Sclerosis: CT and MRI Appearance With Histopathologic Correlation

Pranay Krishnan[1]
M. Isabel Fiel[2]
Andrew B. Rosenkrantz[3]
Cristina H. Hajdu[4]
Thomas D. Schiano[5]
Irina Oyfe[1]
Bachir Taouli[1]

OBJECTIVE. The purposes of this study were to describe the spectrum of cross-sectional imaging findings of pathologically proven hepatoportal sclerosis and to compare the features of advanced and nonadvanced hepatoportal sclerosis.

MATERIALS AND METHODS. Eighteen patients with a histopathologic diagnosis of hepatoportal sclerosis who had concurrent MRI or CT images participated in the study. The following imaging features were assessed: presence of liver nodularity and liver lesions, portal vein patency, presence and degree of portal hypertension, liver volume, and caudate-to-right lobe ratio. These features were compared between patients who underwent transplant and those who did not.

RESULTS. The 18 patients (11 men and one boy, six women; mean age, 46.5 years) had hepatoportal sclerosis confirmed with liver biopsy (14 patients) or explant (four patients). Fourteen patients underwent contrast-enhanced MRI, and five underwent CT. The imaging findings were as follows: liver surface nodularity, five patients (all four transplant, one nontransplant) (p = 0.0016); evidence of portal hypertension, 17 patients; increased caudate-to-right lobe ratio, 16 patients; high periportal signal intensity on T2-weighted images, six patients; portal vein occlusion with cavernous transformation, five patients. The transplant patients had smaller pretransplant liver volume than did nontransplant patients ($p < 0.04$).

CONCLUSION. Hepatoportal sclerosis is characterized by caudate lobe hypertrophy and right hepatic lobe atrophy, preserved liver volume, and lack of the liver nodularity associated with portal hypertension. In advanced cases, liver nodularity and atrophy produce an imaging appearance indistinguishable from that of cirrhosis.

Keywords: CT, hepatoportal sclerosis, MRI, noncirrhotic portal hypertension

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[1]Department of Radiology, Mount Sinai Medical Center, 1 Gustave L. Levy Pl, Box 1234, New York, NY 10029. Address correspondence to B. Taouli (bachir.taouli@mountsinai.org).

[2]Department of Pathology, Mount Sinai Medical Center, New York, NY.

[3]Department of Radiology, New York University Langone Medical Center, New York, NY.

[4]Department of Pathology, New York University Langone Medical Center, New York, NY.

[5]Department of Medicine, Division of Liver Disease, Mount Sinai Medical Center, New York, NY.

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Hepatoportal sclerosis is a rare disease and a known cause of noncirrhotic portal hypertension [1]. The term was first proposed in 1965 by Mikkelsen et al. [2], but the entity has been referred to by multiple synonyms in the literature, including but not limited to noncirrhotic portal fibrosis [3, 4], idiopathic portal hypertension [5, 6], and intrahepatic noncirrhotic portal hypertension [7]. Common pathologic findings seen in hepatoportal sclerosis include phlebosclerosis (portal vein wall thickening with consequent luminal obliteration), megasinusoids (abnormally dilated sinusoids), and portal fibrosis [8]. The distribution of fibrosis only around the portal tracks differentiates it from cirrhosis. The typical clinical presentation of hepatoportal sclerosis is related to symptoms and complications of portal hypertension with preservation of hepatic synthetic function and only mild abnormalities in liver enzyme concentrations [1]. There are reports [7, 8], however, of progression of hepatoportal sclerosis to liver failure requiring liver transplant in patients with long-standing portal hypertension that was often presumed to be due to cirrhosis.

Imaging plays a limited role in the diagnosis of hepatoportal sclerosis. There have been few reports describing intraoperative or transhepatic portographic and hepatic venographic findings in idiopathic portal hypertension [9, 10] and a few sonographic studies of hepatoportal sclerosis [11, 12]. To our knowledge, however, there have been no published reports of the manifestations of hepatoportal sclerosis on cross-sectional images. Our objectives were to describe the spectrum of CT and MRI findings in pathologically confirmed cases of hepatoportal sclerosis and to compare advanced with nonadvanced hepatoportal sclerosis.

Materials and Methods

Patients

This dual-institution retrospective study was compliant with HIPAA and was approved by the lo-

cal institutional review boards at Mount Sinai and New York University Langone Medical Centers. Informed consent was not required. The cases of 18 patients who had a histopathologic diagnosis of hepatoportal sclerosis with concurrent abdominal MRI or CT images were identified in the pathology and radiology databases of both institutions (13 were identified at one institution and five patients at the other). The search was performed from 2000 to 2010. In cases with multiple imaging studies, only the contrast-enhanced MRI study closest to the time of histopathologic sampling was included in the review. Two patients received no IV contrast material for MRI, and thus concurrent contrast-enhanced CT scans obtained 2 days before and 37 days after the respective MRI studies were included in the review. Three patients did not undergo MRI, and contrast-enhanced CT scans were included instead.

Imaging Technique

MRI—Liver MRI was performed with different 1.5-T systems (Signa HDx, Signa Excite, GE Healthcare; Sonata, Avanto, Siemens Healthcare) and with a 3-T system (Trio, Siemens Healthcare). The routine liver protocol included the following sequences: coronal single-shot T2-weighted HASTE, transverse fat-suppressed fast spin-echo T2-weighted, dual-echo in- and opposed-phase T1-weighted gradient-recalled echo, time of flight, and a 3D T1-weighted fat-suppressed spoiled recalled-echo interpolated gradient-echo sequence before and after dynamic injection of extracellular gadopentetate dimeglumine (Magnevist, Bayer HealthCare). Images were obtained in three phases: arterial (timing with test bolus), portal venous (1 minute after contrast injection), and equilibrium (3 minutes after contrast injection). Slice thickness was 2.5–6 mm.

CT—Liver CT was performed with 6-MDCT (Emotion 6, Siemens Healthcare), 16-MDCT (Sensation 16, Siemens Healthcare), and 40-MDCT (Sensation 40, Siemens Healthcare) scanners. The protocol included acquisition of axial images before and after IV administration of 100 mL of nonionic radiopaque contrast agent (iopamidol, Isovue 370, Bracco). Images were obtained in the arterial and portal venous phases with the following parameters: 120–130 kV, 184–283 mAs, 0.6- to 1.25-mm collimation, and 5.0-mm reconstructed section thickness.

Image Analysis

Images were retrospectively reviewed by two observers (radiologist with 7 years' experience in abdominal MRI, fourth-year radiology resident). The observers were aware of the diagnosis of hepatoportal sclerosis but were blinded to the outcome (liver transplant versus no liver transplant).

Qualitative evaluation—Liver nodularity was graded as presence or absence of liver surface or internal liver nodularity. Any hepatic lesion other than cyst or hemangioma was characterized on the basis of signal characteristics on unenhanced T1- and T2-weighted images and pattern of contrast enhancement. Patency of the extrahepatic and intrahepatic portal veins (and hepatic veins) and any abnormal periportal signal was noted.

Semiquantitative and quantitative evaluation—In the evaluation of portal hypertension, the presence of varices or collateral vessels was assessed for the following five locations: gastroesophageal, paraesophageal, splenorenal, paraumbilical, and other [13]. Varices were graded on a scale of 0–3 (0, no visible varices; 1, one site involved; 2, two sites involved; 3, three or more sites involved). Severity of ascites was graded on a scale of 0–3 (0, no ascites; 1, minimal perihepatic and perisplenic fluid; 2, intraperitoneal fluid with no significant abdominal distention; 3, fluid causing marked abdominal distention) [14]. Splenomegaly was graded on a scale of 0–3 based on craniocaudal size (0, < 13 cm; 1, 13–15 cm; 2, 15–20 cm; 3, > 20 cm). In the one pediatric patient, an 8-year-old boy, splenomegaly was graded on the following scale: 0, < 11 cm; 1, 11–13 cm; 2, 13–15 cm; 3, > 15 cm) [15].

Caudate-to-right hepatic lobe ratio was measured by the two observers in consensus, as reported previously [16–18] (Fig. 1). This ratio is used to assess the degree of caudate lobe hypertrophy and right hepatic lobe atrophy.

A third experienced observer calculated whole liver, right hepatic lobe, left hepatic lobe, and caudate lobe volumes using an automated segmentation method (direct volume rendering) at a workstation (Aquarius version 3.7.0.13, TeraRecon). Volumes were calculated on transverse 3D T1-weighted gradient-echo MR images or contrast-enhanced CT images obtained during the portal venous phase.

Histopathologic Evaluation

To confirm the diagnosis of hepatoportal sclerosis, two hepatopathologists (15 and 6 years of experience) performed a detailed retrospective histopathologic examination of the needle and wedge liver biopsy specimens or total hepatectomy specimens from patients with the diagnosis of hepatoportal sclerosis established at the initial pathologic evaluation. For explanted total hepatectomy specimens, the initial gross examination included determination of liver weight and assessment for the presence of portal vein thrombus and established cirrhosis. Nodular regenerative hyperplasia (NRH) can be easily mistaken for cirrhosis owing to the presence of nodules on cut sections. The difference, however, is the absence of fibrous tissue that surrounds these nodules. It therefore was important to make this distinction during gross examination of the explant specimen. Standard H and E sections, trichrome stains to detect fibrosis, and reticulin stains to assess the hepatic architecture were evaluated. The degree of periportal fibrosis and inflammation was assessed, as was the presence of fibrous septa. Luminal size of portal venules and intimal hyperplasia and phlebosclerosis (thickening of portal vein radicles) were noted. The extent of periportal megasinusoid formation was determined.

Statistical Analysis

Age, sex, liver function test results, qualitative imaging findings (presence of liver nodularity, portal vein occlusion), and semiquantitative and quantitative imaging parameters (portal hypertension score, liver volume, caudate-to-right lobe ratio) were compared between transplant and nontransplant patients by Fisher exact test for noncontinuous variables and Student *t* test for continuous variables; $p < 0.05$ was considered to indicate significance.

Results

Demographic and Biologic Data

The cases of 18 patients (11 men and one boy, six women; mean age, 46.5 years; range, 8–69 years) were evaluated (Table 1). Hepatoportal sclerosis was confirmed with liver biopsy in 14 cases and examination of the explant in four cases. Fourteen patients underwent contrast-enhanced MRI, and five underwent CT. The mean age of the male patients was 43 years (range, 8–68 years), and that of the women was 53.5 years (range, 44–69

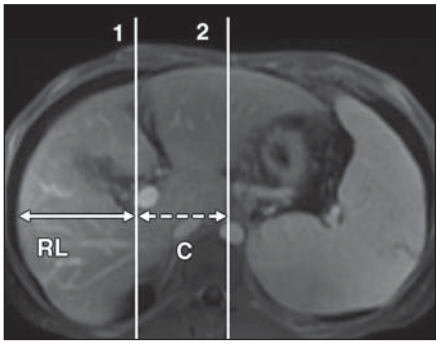

Fig. 1—23-year-old man with hepatoportal sclerosis. Contrast-enhanced T1-weighted portal venous phase MR image shows method used to calculate caudate-to-right lobe ratio. Line 1 is sagittal line tangential to right lateral wall of main portal vein. Line 2 is sagittal line through medial margin of caudate lobe parallel to line 1. Caudate and right lobe diameters are measured midway between main portal vein and inferior vena cava between lines 1 and 2 (C) and between line 1 and right lateral margin of liver (RL). C/RL is ratio of caudate lobe to right hepatic lobe diameters and equals 0.9 in this patient (reported values in controls vary between 0.37 and 0.43 [16–18]). Caudate lobe hypertrophy, splenomegaly, and ascites and lack of liver nodularity are evident.

years). Four patients needed liver transplant because of severe refractory ascites, and these patients underwent imaging a mean of 64 days (range, 28–112 days) before transplant. There was no significant difference in sex distribution between transplant and nontransplant patients ($p = 0.5686$). Transplant patients were older than nontransplant patients (mean age, 58.7 ± 8.1 [SD] years vs 43.0 ± 16.3 years; $p = 0.025$). Fourteen patients had a low platelet count (< $150 \times 10^3/\mu L$), transplant patients having a significantly lower platelet count than nontransplant patients (45 ± 9.2 vs 128.3 ± 67.6 × $10^3/\mu L$; $p = 0.001$). Seventeen patients had the following abnormalities in liver function test results at imaging: elevated prothrombin time (> 15.0 seconds), 10 patients; decreased serum albumin concentration (< 3.5 g/dL), 10 patients; elevated serum bilirubin concentration (> 1.12 mg/dL), eight patients; elevated alkaline phosphatase concentration (> 110 U/L), nine patients; elevated aspartate aminotransferase concentration (> 50 U/L), six patients; and elevated alanine aminotransferase concentration (> 50 U/L), five patients. In patients with elevated aspartate and alanine aminotransferase concentrations, the values were less than twice the upper limit of normal (< 100 U/L) in all but one case. Three patients had elevations in alkaline phosphatase concentration more than twice the upper limit of normal (> 220 U/L). There were no statistically significant differences between transplant and nontransplant patients in terms of quantitative laboratory values ($p < 0.228$ to $p < 0.943$).

Histopathologic Findings

Specimens for pathologic diagnosis were obtained by percutaneous needle biopsy in nine cases, transjugular needle biopsy in two cases, surgical wedge biopsy in three cases, and liver explant in four cases. The mean time between imaging and pathologic examination was 239 days (median, 51 days; range, 2–1436 days). This interval was less than 6 months in 14 cases. All patients had features of hepatoportal sclerosis, including phlebosclerosis (thickening

TABLE 1: Clinical, Pathologic, and Imaging Findings in 18 Patients With Hepatoportal Sclerosis

Patient No.	Sex	Age (y)	Specimen	Imaging	Time Between Imaging and Pathologic Examination (d)	Pathologic Finding	Imaging Finding
1	F	69	Explant	CT	112	Hepatoportal sclerosis	Nodular hepatic contour, portal hypertension
2	M	50	Explant	CT	67	Hepatoportal sclerosis and incomplete septal cirrhosis	Nodular hepatic contour, portal hypertension, portal vein occlusion
3	F	55	Explant	MRI	51	Hepatoportal sclerosis and incomplete septal cirrhosis	Nodular hepatic contour, portal hypertension, high periportal T2 signal intensity
4	M	42	Wedge biopsy	MRI	172	Hepatoportal sclerosis	Portal hypertension, portal vein occlusion, high periportal T2 signal intensity
5	F	47	Wedge biopsy	MRI	7	Hepatoportal sclerosis	Portal hypertension, portal vein occlusion
6	M	23	Needle biopsy	MRI	6	Hepatoportal sclerosis	Portal hypertension, high periportal T2 signal intensity
7	M	41	Needle biopsy	MRI	177	Hepatoportal sclerosis and nodular regenerative hyperplasia	Portal hypertension
8	F	62	Transjugular biopsy	CT, MRI	44	Hepatoportal sclerosis and nodular regenerative hyperplasia	Portal hypertension
9	M	8	Needle biopsy	MRI	1436	Hepatoportal sclerosis	Portal hypertension, portal vein occlusion, high periportal T2 signal intensity, liver lesions
10	M	68	Needle biopsy	MRI	299	Hepatoportal sclerosis	Portal hypertension, liver lesions
11	M	40	Needle biopsy	MRI	145	Hepatoportal sclerosis and nodular regenerative hyperplasia	Portal hypertension, high periportal T2 signal intensity
12	M	36	Wedge biopsy	MRI	34	Hepatoportal sclerosis	Mild splenomegaly
13	M	29	Transjugular biopsy	CT	2	Hepatoportal sclerosis	Portal hypertension
14	F	44	Needle biopsy	MRI	251	Hepatoportal sclerosis	Enlarged caudate lobe, otherwise normal MRI findings
15	M	63	Needle biopsy	MRI	1420	Hepatoportal sclerosis	Portal hypertension
16	F	44	Needle biopsy	MRI	43	Hepatoportal sclerosis	Portal hypertension
17	M	61	Explant	CT	28	Hepatoportal sclerosis and nodular regenerative hyperplasia	Nodular hepatic contour, portal hypertension
18	M	56	Needle biopsy	MRI	3	Hepatoportal sclerosis	Nodular hepatic contour, portal hypertension, portal vein occlusion, high periportal T2 signal intensity

of portal vein radicles), periportal megasinusoid formation, and dense portal fibrosis with partial or total obliteration of the portal vein radicles. Two of the explanted livers had incomplete septal cirrhosis evidenced by prominent portal fibrous septa and nodularity in sections from the hilum. Four patients had features of NRH (Table 1).

Qualitative Imaging Findings

Liver nodularity—Liver nodularity was found in five (27.8%) patients. It was found in all four transplant patients and in only one of the 14 (7.1%) patients who did not undergo transplant ($p = 0.0016$) (Fig. 2).

Hepatic lesions—Hepatic lesions were found in 2 of 18 (11.1%) patients. In one patient, multiple subcentimeter lesions were noted that were hyperintense on T1-weighted images and isointense on images obtained with all other sequences. Two additional lesions measured 2.0 and 2.6 cm and were hyperintense on T1- and T2-weighted images without arterial phase enhancement (Fig. 3). Follow-up MRI (after 18 months) showed a significant decrease in size of one of these lesions and resolution of the other, corroborating a presumed benign cause. In another patient, two 1.5-cm lesions with high signal intensity were identified on T2-weighted images. Both lesions were enhancing on arterial phase images and remained hyperintense on delayed phase images. No follow-up images were available. None of the lesions in either patient had histopathologic confirmation.

Portal vein patency and periportal signal intensity—Portal vein occlusion with cavernous transformation was found in 5 of 18 (27.8%) patients (Fig. 4). There was no difference between transplant and nontransplant patients in terms of presence of portal vein occlusion ($p = 1.0$). Increased periportal signal intensity on T2-weighted images was seen in 6 of 18 patients (33.3%) (Figs. 4 and 5) without associated macroscopic portal vein occlusion in three cases (Fig. 5).

Semiquantitative and Quantitative Imaging Findings

The results of semiquantitative and quantitative evaluation are shown in Table 2.

Portal hypertension—Portal hypertension was present in 17 of 18 patients (94.4%). Fourteen patients had splenomegaly, which was considered moderate or severe in 13 patients. Varices and collateral vessels were found in 16 patients and at more than one site in 13 patients. Ascites was found in 12 patients and was considered moderate or severe in eight patients. Patients who eventually underwent liver transplant had more severe portal hypertension than did patients who did not ($p < 0.001$).

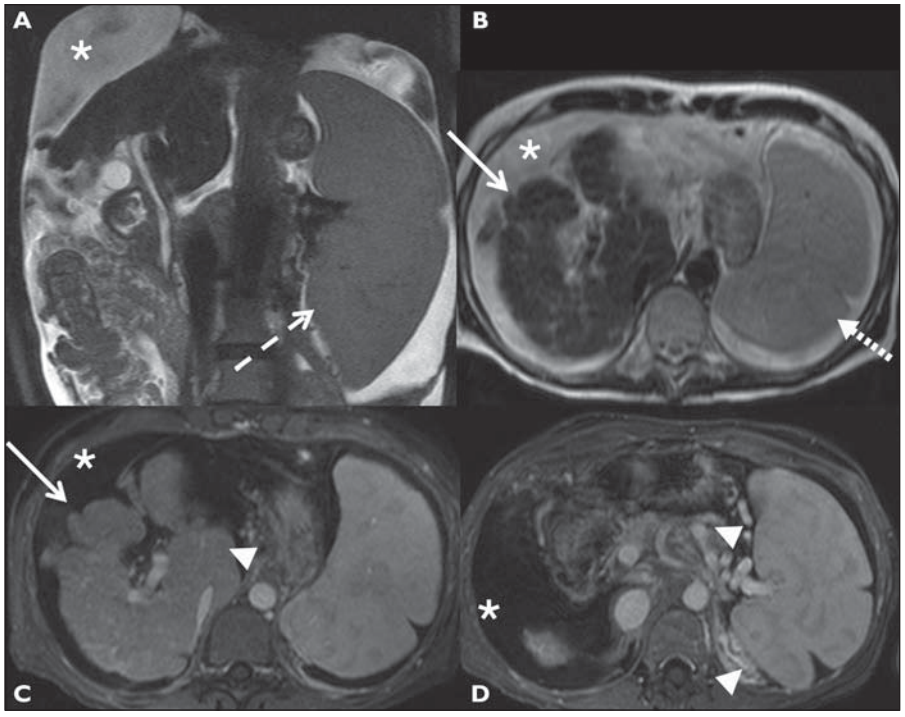

Fig. 2—55-year-old woman with advanced hepatoportal sclerosis and refractory ascites that required liver transplant.
A–D, Coronal (**A**) and transverse (**B**) single-shot T2-weighted HASTE and transverse contrast-enhanced fat-suppressed 3D portal venous phase T1-weighted images at different levels (**C** and **D**) show shrunken and nodular liver (*solid arrows*) with findings of portal hypertension, including splenomegaly (*dashed arrows*), ascites (*asterisks*), and varices (*arrowheads*). Imaging appearance is indistinguishable from that of cirrhosis. Pathologic examination of explant revealed hepatoportal sclerosis with incomplete septal cirrhosis.

Caudate lobe hypertrophy and right lobe atrophy—The caudate-to-right lobe ratio was greater than 0.37 in 16 of 18 patients (88.8%). The threshold value of 0.37 was reported in patients without cirrhosis in two previous studies [16, 18]. There was no difference in caudate-to-right lobe ratio between transplant and nontransplant patients ($p = 0.729$).

Liver volume—The mean hepatic volumes of the entire liver and right and left hepatic lobes are shown in Table 2. All liver volumes were significantly smaller in transplant than in nontransplant patients.

Discussion

Hepatoportal sclerosis is a rare clinicopathologic condition that causes noncirrhotic portal hypertension [1, 19]. The nomenclature for noncirrhotic portal hypertension is ambiguous, and various terms, such as noncirrhotic portal fibrosis [3, 4], idiopathic portal hypertension [5, 6], and intrahepatic noncirrhotic portal hypertension [7], have been used. The common pathologic findings of hepatoportal sclerosis include phlebosclerosis, megasinusoids, and portal fibrosis [8]. NRH and incomplete septal cirrhosis are also included in the spectrum of hepatoportal sclerosis. NRH is defined as the presence of multiple hyperplastic parenchymal nodules with minimal or no fibrosis. NRH often develops in patients with other conditions, such as myeloproliferative disease, collagen vascular disease, and drug toxicity [20]. Incomplete septal cirrhosis is characterized by slender fibrous septa that outline incomplete macronodules and by occlusive venous changes that can result in portal hypertension. The cause of hepatoportal sclerosis is unknown. Some authors [21] have proposed that an underlying prothrombotic state may be the root cause of portal venule obstruction. There have also been reports [22] of development of noncirrhotic portal hypertension secondary to hepatoportal sclerosis in patients with HIV infection undergoing antiviral therapy. The antiviral therapy and HIV

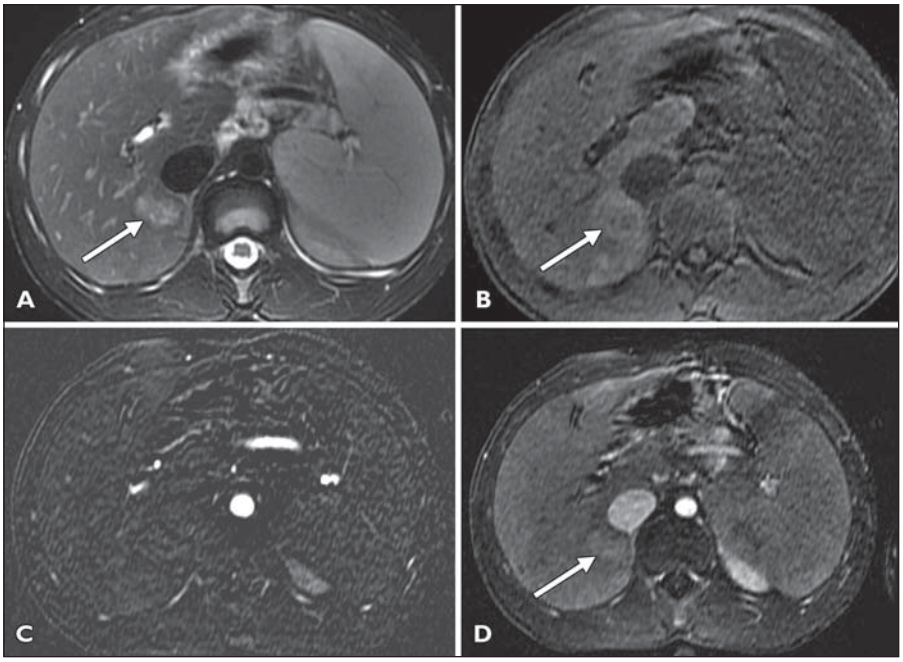

Fig. 3—8-year-old boy with hepatoportal sclerosis diagnosed with percutaneous liver biopsy. **A–D,** Transverse fat-suppressed fast spin-echo T2-weighted (**A**), fat-suppressed unenhanced (**B**), and contrast-enhanced subtracted 3D T1-weighted arterial (**C**) and portal venous phase (**D**) images show liver with smooth contour, enlarged caudate lobe, and splenomegaly. Right posterior lobe liver lesion (*arrows*) is T2 hyperintense with hypointense rim, slightly T1 hyperintense, and enhancing in portal venous phase. Patient had another lesion with same signal intensity and enhancement characteristics in right anterior lobe (not shown). Both lesions had decreased in size at 18-month follow-up MRI (not shown).

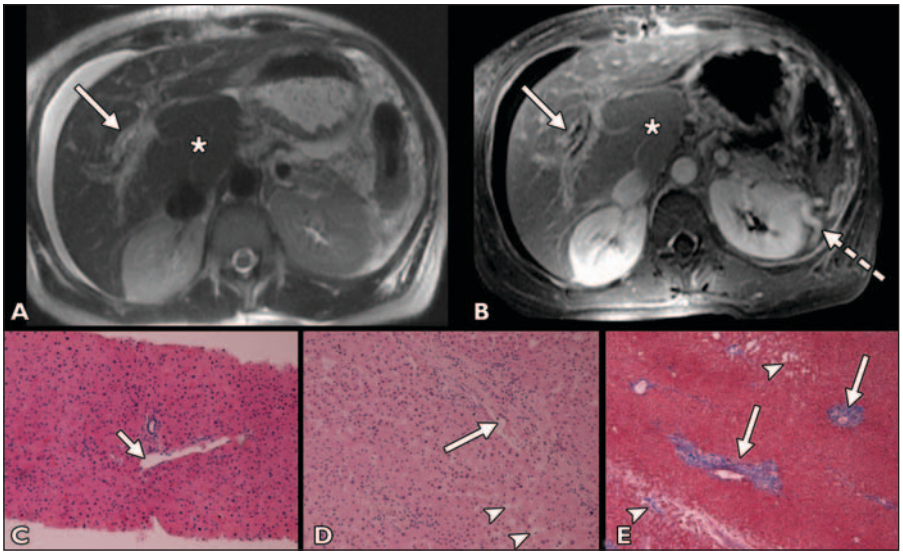

Fig. 4—47-year-old woman with hepatoportal sclerosis diagnosed with surgical wedge liver biopsy. **A** and **B,** Transverse single-shot T2-weighted HASTE (**A**) and transverse contrast-enhanced fat-suppressed 3D T1-weighted (**B**) portal venous phase images show liver with smooth contour and markedly enlarged caudate lobe (*asterisk*), portal vein occlusion, periportal cavernoma, increased periportal T2 signal intensity (*solid arrows*), and findings of portal hypertension, including ascites and perisplenic varices (*dashed arrow,* **B**).
C, Photomicrograph (H and E, ×40) shows dystrophic and herniated portal vein branch (*arrow*).
D, Photomicrograph (H and E, ×40) shows portal track without apparent portal vein lumen (*arrow*) and megasinusoids (*arrowheads*).
E, Photomicrograph (trichrome, ×40) shows densely fibrotic portal areas (*arrows*) and megasinusoid (*arrowheads*).

itself are considered potential sources of injury. The data on sex predominance in noncirrhotic portal hypertension are discordant. A large series of 150 cases in India had a female predominance (1.65:1) [23], but another large series of 75 cases in India had a strong male predominance (7:2) [24]. Although we observed a clear male predominance in our series, the sex predominance of hepatoportal sclerosis is unclear at this point.

We identified a spectrum of findings in patients with hepatoportal sclerosis, including features of portal hypertension and relative hypertrophy of the caudate lobe, that in the absence of liver nodularity and in the presence of preserved liver volumes with normal or minimally elevated aspartate and alanine transaminase concentrations should prompt consideration of hepatoportal sclerosis as a potential cause in the appropriate clinical situation. When liver nodularity and decreased liver volume are present, end-stage hepatoportal sclerosis is indistinguishable from cirrhosis on images, and liver biopsy is necessary for a definite diagnosis. Despite the rarity of hepatoportal sclerosis, we believe radiologists should be familiar with this entity because many cases are misdiagnosed as cirrhosis while liver function is generally preserved, and the treatment is mainly aimed at decreasing the risks of portal hypertension with variceal banding and a transjugular intrahepatic portosystemic shunt.

Imaging descriptions of hepatoportal sclerosis are limited. To our knowledge, there are no published reports of series of cases in which CT or MRI appearances are described. Futagawa et al. [10] performed a qualitative evaluation of portal vein and hepatic vein changes in hepatoportal sclerosis (called idiopathic portal hypertension in their article) compared with those of cirrhosis at intraoperative transhepatic

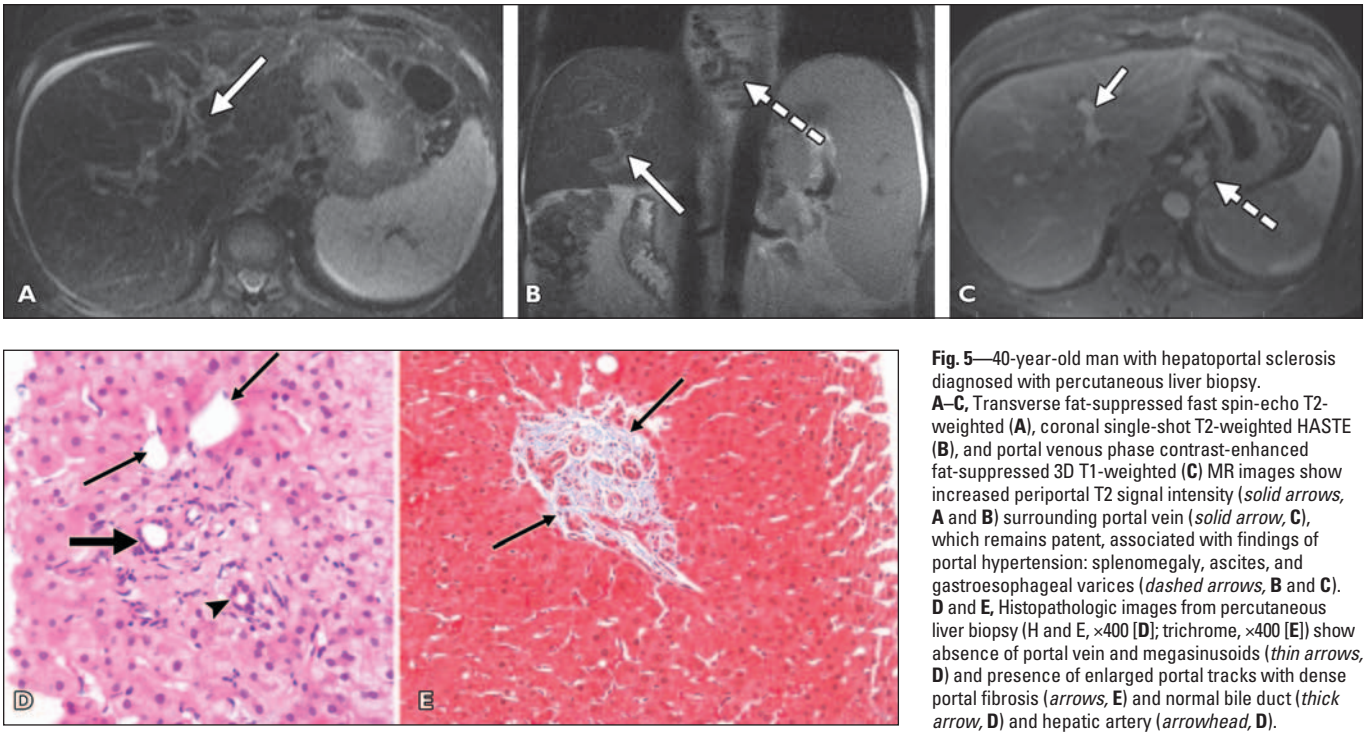

Fig. 5—40-year-old man with hepatoportal sclerosis diagnosed with percutaneous liver biopsy. **A–C,** Transverse fat-suppressed fast spin-echo T2-weighted (**A**), coronal single-shot T2-weighted HASTE (**B**), and portal venous phase contrast-enhanced fat-suppressed 3D T1-weighted (**C**) MR images show increased periportal T2 signal intensity (*solid arrows,* **A** and **B**) surrounding portal vein (*solid arrow,* **C**), which remains patent, associated with findings of portal hypertension: splenomegaly, ascites, and gastroesophageal varices (*dashed arrows,* **B** and **C**). **D** and **E,** Histopathologic images from percutaneous liver biopsy (H and E, ×400 [**D**]; trichrome, ×400 [**E**]) show absence of portal vein and megasinusoids (*thin arrows,* **D**) and presence of enlarged portal tracks with dense portal fibrosis (*arrows,* **E**) and normal bile duct (*thick arrow,* **D**) and hepatic artery (*arrowhead,* **D**).

TABLE 2: Quantitative Parameters for Imaging of 18 Patients With Hepatoportal Sclerosis

Patient Group	Portal Hypertension Score[a]	Whole Liver Volume (cm^3)	Right Lobe Volume (cm^3)	Left Lobe Volume (cm^3)	Caudate-to-Right Lobe Ratio
All (n = 18)	5.2 ± 2.6	1375.7 ± 724.0	756.0 ± 474.4	619.7 ± 280.4	0.66 ± 0.22
Transplant patients (*n* = 4)	8.25 ± 0.50	936.7 ± 185.7	484.2 ± 139.9	452.5 ± 88.5	0.70 ± 0.28
Nontransplant patients (*n* = 14)	4.3 ± 2.4	1501.1 ± 775.4	833.6 ± 510.4	667.5 ± 299.9	0.65 ± 0.20
p[b]	**< 0.001**	**0.025**	**0.038**	**0.033**	0.735

Note—Results stratified by outcome (liver transplant vs no transplant). Whole liver, right lobe, and left lobe hepatic liver volumes were significantly lower in transplant patients, and portal hypertension scores were higher in transplant patients.
[a]Maximum, 9.
[b]Student *t* test. Bold type indicates statistically significant difference.

portography and hepatic venography. The most common features of hepatoportal sclerosis were a paucity of medium-sized portal branches, irregular and obtuse-angled division of the peripheral branches, occasional abrupt interruptions, and lack of opacification of some of the large intrahepatic portal branches and their periphery. In another study by the same group [9], free and wedged hepatic venography were performed on 37 patients with hepatoportal sclerosis and on 88 patients with cirrhosis who acted as controls. Characteristic changes of hepatoportal sclerosis included frequent venovenous anastomoses, narrower angles between large veins and their tributaries, smooth and wavy middle-sized to large branches, homogeneous sinusoidal filling, and minimal to no filling of the portal venous system on wedged retrograde portographic images. To our knowledge, these findings have not been confirmed in other studies, and the methods described are invasive. In two previous studies [11, 12], investigators using sonography found hyperechoic bands surrounding the portal vein branches in patients with hepatoportal sclerosis. Similarly, we found increased periportal signal intensity on T2-weighted images of six patients. We speculate that this finding represents periportal fibrosis, although we did not clearly correlate the degree of periportal fibrosis and T2 signal intensity. Further prospective studies with explant correlation are warranted.

We also observed morphologic changes in most of our patients with hepatoportal sclerosis. We used caudate-to-right hepatic lobe ratio to measure caudate lobe hypertrophy and right hepatic lobe atrophy. An increased caudate-to-right lobe ratio reflects the morphologic changes of cirrhosis and other causes of liver disease, such as Budd-Chiari syndrome, end-stage primary sclerosing cholangitis, congenital hepatic fibrosis, portal cavernoma, and hepatoportal sclerosis. In congenital hepatic fibrosis, portal cavernoma, and hepatoportal sclerosis, however, liver nodularity is rarely observed, whereas it is commonly observed in cirrhosis [25]. Harbin et al. [16] initially reported a mean caudate-to-right lobe ratio of 0.37 ± 0.16 in healthy subjects. Subsequently, Awaya et al. [17] and Vilgrain et al. [18] reported caudate-to-right lobe ratios ranging between 0.373 ± 0.174 and 0.433 ± 0.112 in healthy subjects. The mean caudate-to-right lobe ratio of 0.66 ± 0.22 in our series not only is indicative of relative caudate lobe hypertrophy and atrophy of the right lobe but also is similar to the threshold cited by Harbin

et al. (0.65) [16] for the diagnosis of cirrhosis. The exact mechanism by which cirrhosis leads to caudate lobe hypertrophy relative to atrophy in the right lobe is unclear but is thought to reflect alterations in hepatic blood flow caused by fibrosis that lead to differential distributions of trophic factors [26]. Because hepatoportal sclerosis is characterized by periportal fibrosis with portal venule compression, similar alterations in portal flow with relative preservation of flow to the caudate lobe are expected. Although macroscopic portal vein occlusion was present on images of only five patients, portal venule obstruction was noted in all pathologic specimens. It is therefore not surprising that there is an overlap in the morphologic changes in patients with hepatoportal sclerosis and those with portal cavernoma. These findings further support the argument that alterations in portal venous flow may be driving this morphologic change.

The main limitation of this study was that it was retrospective. A prospective study of such a rare entity will be difficult. Analysis in some cases was also limited by the time between imaging and pathologic examination. In 14 of 18 cases, the time between imaging and pathologic examination was less than 6 months. In the other four cases, however, the gap was longer, as long as 2–3 years in two cases. Again the rarity of the entity and limited number of patients in the study precluded exclusion of these patients. In our series, parenchymal lesions were identified in only two patients; however, histopathologic correlation was not available in cases in which the diagnosis of hepatoportal sclerosis was made with needle biopsy. It is assumed that these nodules are benign because, to our knowledge, there have been no reports of neoplasms in patients with hepatoportal sclerosis.

Conclusion

In our series of 18 patients with hepatoportal sclerosis, a spectrum of imaging findings is evident. The dominant MRI and CT features of this condition include stigmata of portal hypertension, caudate lobe hypertrophy, preservation of liver volume, and absence of contour nodularity in nonadvanced cases. In advanced cases, ultimately requiring liver transplant, liver volume is decreased, liver nodularity is indistinguishable from that seen in cirrhosis, and the severity of portal hypertension is increased. Macroscopic portal vein occlusion, increased periportal T2 signal intensity, and parenchymal nodules were present in the minority of patients.

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