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Primary sclerosing cholangitis: MR cholangiopancreatography and T2-weighted MR imaging findings

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PURPOSE
To present MR cholangiopancreatography (MRCP) findings and to determine the hepatic morphological changes of primary sclerosing cholangitis (PSC) seen on T2-weighted fast spin echo (FSE) images.

MATERIALS AND METHODS
Twenty-three patients (15 women, 8 men) with ages ranging from 17 to 80 years (median, 45.1 years) were included in the study. MR imaging was performed on a 1 Tesla MR unit using a phased-array coil. Heavily T2-weighted images were obtained with single-shot fast spin echo technique for MRCP. Morphological changes encountered in livers were evaluated with coronal and axial T2-weighted fast spin echo images.

RESULTS: Irregularities, multifocal strictures, and dilatations in different levels of the biliary channels were seen in all patients. T2-weighted images showed lobulated hepatic contours in 21.73%, atrophy in both anterior and posterior segments of the right lobe in 21.73%, atrophy in the anterior segment in 13.04%, atrophy in both medial and lateral segments of the left lobe in 17.39%, atrophy in the medial segment in 8.69%, atrophy in the lateral segment in 4.34%, hypertrophy in the posterior segment of the right lobe in 4.34%, global hypertrophy in the left lobe in 4.34%, hypertrophy in the lateral segment of the left lobe in 4.34%, and caudate lobe hypertrophy in 21.73% of the patients. In addition, periportal edema was noted in 39.13%, increased parenchymal signal on T2-weighted images in 26.08%, periportal and/or portocaval lymphadenopathy in 34.78%, and portal hypertension in 34.78% of our patients. In one patient (4.34%), the liver had a round shape characteristic of PSC.

CONCLUSION: MR imaging is a useful method for establishing the changes in biliary ducts specific to PSC, and for identifying long-standing cases complicated with cirrhosis.

Key words: ● *cholangitis, sclerosing* ● *magnetic resonance imaging*

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Primary sclerosing cholangitis is an uncommon disease of unknown etiology which is characterized by chronic inflammation and fibrosis of bile ducts. Progressive and obliterative fibrosis of small, medium, and large bile ducts causes secondary biliary cirrhosis and cholestasis, which results in hepatic insufficiency (1-4). In most cases, the only treatment option for primary sclerosing cholangitis accompanied with inflammatory intestinal disease is liver transplantation. Above and beyond clinical and laboratory findings, imaging methods are used for diagnosis. Endoscopic retrograde cholangiopancreatography (ERCP) is defined as the standard reference for the evaluation of bile ducts, and magnetic resonance cholangiopancreatography (MRCP) is a method with proven efficiency in the evaluation of asymptomatic patients and in the early diagnosis of complications, especially cholangiocellular carcinoma, which is highly likely to develop in patients who have primary sclerosing cholangitis (1-4). Both morphological changes, which can be seen in the biliary system, and changes in the hepatic parenchyma, which are accompanied by primary sclerosing cholangitis, can be evaluated by magnetic resonance (MR) imaging in a non-invasive manner.

In this study, MRCP findings related to intrahepatic and extrahepatic bile ducts, and morphological changes detected by T2-weighted MR images of patients diagnosed as primary sclerosing cholangitis are presented together.

Materials and methods

Twenty-three patients (15 women, 8 men) with ages ranging from 17 to 80 years (median, 45.1 years) were included in the study. Primary sclerosing cholangitis was diagnosed by clinical findings, which supported anamnesis and ERCP findings. In six cases, inflammatory intestinal diseases (ulcerative colitis, 5; Crohn's disease, 1) were noted. MRCP examinations were performed on a 1.0 Tesla MR unit (Signa LX Horizon, GE Medical Systems, Milwaukee, WI, USA) using a phased-array coil. Heavily T2-weighted images were obtained with the single-shot fast spin-echo (SSFSE) technique for MRCP. Patients were asked not to eat at least 5-6 hours before the examination. During the examination, neither oral nor intravenous contrast media was used. Scans were performed in the axial plane, by identifying the common bile duct on the coronal scout image starting from a few cm above the porta hepatis up to the ampulla Vateri, without leaving slice interval and with 5 mm slice thickness. Axial plane source images were obtained as thick slices, ranging from 35-70 mm thickness, on coronal or coronal-oblique plane by considering the common bile duct as the center without respiration. In addition, construction source images were obtained during the expiratory phase of respiration with respiratory triggering with SSFSE technique using 3 mm slice thickness without slice interval. By using a

maximum intensity projection (MIP) algorithm on source images in coronal and oblique planes, reformatted images were obtained. Reformatted images and thick slice images were evaluated in addition to the axial and coronal source images. T2-weighted fast-spin echo (FSE) (TR: changes according to patient's respiration behavior/TE: 102 msec, echo train length: 4-18) coronal (n=23 cases) and axial (n=11 cases) images were evaluated as well.

Results

In all of our cases, irregularities, multifocal strictures, and dilatations were found at different levels of the intrahepatic bile ducts (Figure 1-5).

In three cases, a common bile duct stone was noted and in one case, diverticula-like structures in relation to the intrahepatic bile ducts were noted. Liver parenchymal changes that were seen are as follows: lobulated hepatic contours in 5 cases (21.73%); atrophy in both anterior and posterior segments of the right lobe in 5 cases (21.73%); atrophy in the anterior segment in 3 cases (13.04%); atrophy in both medial and lateral segments of the left lobe in 4 cases (17.39%); atrophy in the medial segment in 2 cases (8.69%); atrophy in the lateral segment in 1 case (4.34%); hypertrophy in the posterior segment of right lobe in 1 case (4.34%); global hypertrophy in the left lobe in 1 case (4.34%); hypertrophy in the lateral segment of left lobe in 1 case (4.34%), and caudate lobe hypertrophy in 5 cases (21.73%). Additionally, the following observations were noted: periportal edema (Figure 6) in 9 cases (39.13%), increased parenchymal signal on T2-weighted images (Figure 7) in 6 cases (26.08%), portal and/or portocaval lymphadenopathy in 8 cases (34.78%), and portal hypertension findings (splenomegaly, collateral vascular structures, and/or ascites) in 8 cases (34.78%). In 6 cases (26.08%), morphology of the liver was regular. In one patient (4.34%), the liver had a round shape (Figure 8). Results are summarized in Table.

Table. Findings on T2-weighted coronal or transverse MR images in primary sclerosing cholangitis

Ages and gender of the patients (n=23)	Liver changes
58, F	CL, atrophic right lobe, PHT, PPE
39, F	CL, atrophic right lobe, hypertrophic left and caudate lobe, PHS, PPE, PPLN
18, M	PPE
43, F	CL and square-spheric shape, hypertrophic left and caudate lobe, atrophic right and left lobe, PHS, PPE, PHT, PPLN
60, F	Atrophic right lobe anterior segment
46, F	Hypertrophic left lobe lateral segment, atrophic left lobe medial segment, PHS, PPE, PPLN
45, F	Atrophic left lobe, atrophic right lobe anterior segment, hypertrophic right lobe posterior segment, hypertrophic caudate lobe, PHT
35, M	N
39, F	N
29, M	N
63, F	CL, atrophic right lobe, PHS
80, F	Atrophic left lobe medial segment, PCLN
35, F	Atrophic left lobe, hypertrophic caudate lobe, PHT
54, M	CL, atrophic left lobe lateral segment, PHT, PPLN
27, F	Liver is N, PHT
70, F	Atrophic right lobe, hypertrophic caudate lobe, PHS
47, F	N
54, F	Atrophic right lobe anterior segment, PHS, PPE, PHT, PPLN
55, M	PPE, PPLN
55, M	N
33, M	Atrophic left lobe
35, M	PPE, PCLN
17, F	PPE, PHT

CL: contour lobulation, PHS: parenchymal high signal, PHT: portal hypertension, PPE: periportal edema, PPLN: periportal lymph node, PCLN: portocaval lymph node, N: normal

Discussion

Primary sclerosing cholangitis is an idiopathic disease characterized by chronic inflammation and fibrosis of intra- and extrahepatic bile ducts. In addition to the clinical findings, diagnosis of this disease should be supported by both histopathological and biochemical studies, as well as imaging (1-4). Bile duct obliteration, which appears as the disease progresses, leads to cholestasis, and in 49% of symptomatic patients, to biliary cirrhosis and liver damage. Inflammatory intestinal disease, in particular, is seen together with ulcerative colitis in 70% of cases (1-3). Although the etiology is unknown, as it appears together with diseases such as retroperitoneal fibrosis, mediastinal fibrosis, and Sjögren's syndrome, it entails an autoimmune period (1). The risk of developing cholangiocarcinoma is 10-15% among primary sclerosing cholangitis patients. Although various medical and invasive methods are used in treatment, the definite treatment is orthotopic liver transplantation. Secondary causes such as stricture, stones, or bacterial cholangitis secondary to earlier surgery, parasitic infections, ischemia, or cholangitis secondary to chemotherapy should be eliminated before diagnosing primary sclerosing cholangitis (1-6).

Patients can be asymptomatic, whereas in 55% of the cases chronic fatigue, itching, jaundice, and stomach pain can be observed (1). In order to confirm the diagnosis, biochemical analysis (increase in the levels of serum bilirubin and alkaline phosphatase), imaging findings, and for equivocal cases histopathological evaluation should be obtained in addition to the clinical findings. The histopathological findings in the portal region and the increase in the levels of bilirubine and alkaline phosphatase, which can be recorded in most cholestatic diseases,

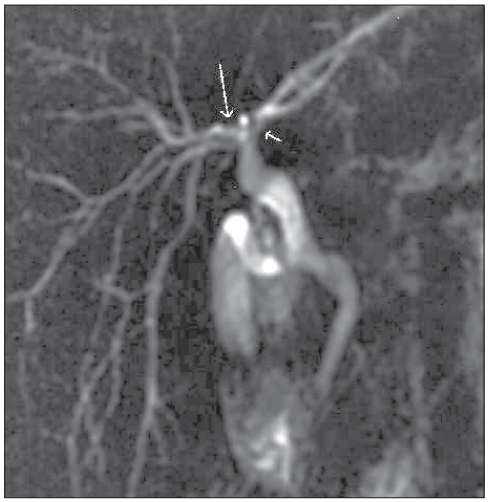

Figure 1. MRCP image shows that the right hepatic duct is irregular *(long arrow)* and the left hepatic duct is relatively thin *(short arrow)*. There is also dilatation in the intrahepatic bile ducts. Note that the angle between the central hepatic ducts has expanded.

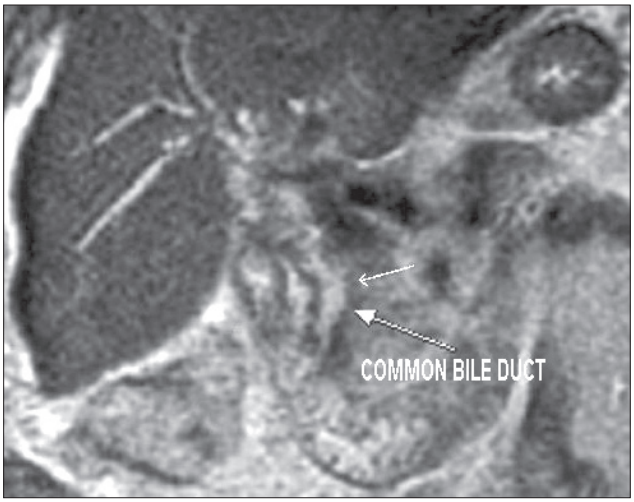

Figure 2. Coronal T2-weighted MR image shows focal narrowing in the right hepatic duct and dilated ducts proximal to it. Additionally, irregularities in the common bile duct contours and pseudodiverticula formation among the wall are noteworthy *(arrow)*.

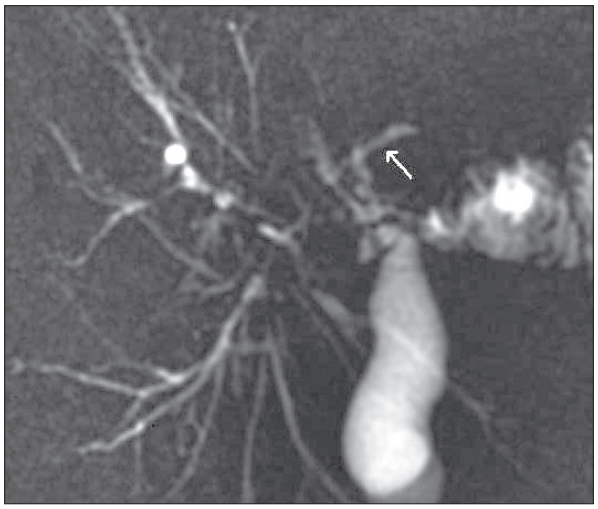

Figure 3. MRCP image shows multifocal strictures and dilatations in the central intrahepatic bile ducts. Moderate expansion in more peripheral ducts is also observed.

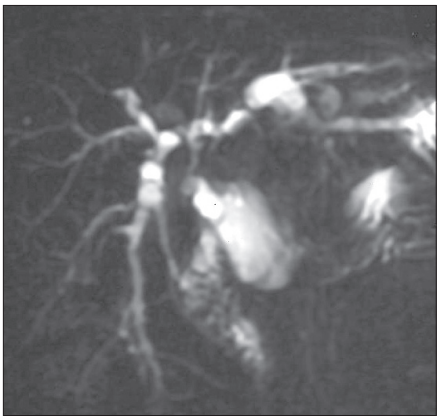

Figure 5. Beaded appearance of the bile ducts. MRCP image shows focal luminal dilatation and narrowing evident in the left intrahepatic bile ducts.

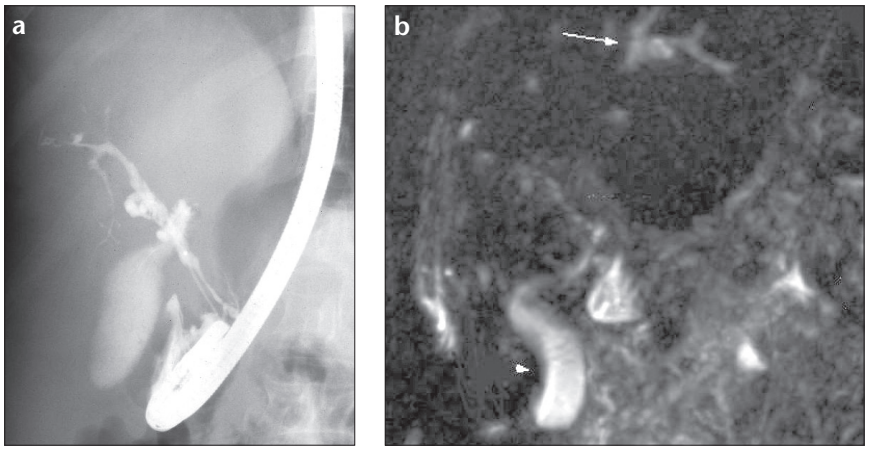

Figure 4. a, b. ERCP image **(a)** shows narrowing and irregularities in the central ducts. MRCP image **(b)** of the same patient shows that thin central ducts can not be visualized as they do not contain a sufficient amount of bile, but focal expansion in the left peripheral bile ducts *(arrow)* marks narrowness that prevents drainage at this level *(arrowhead: gallbladder)*.

are not specific to primary sclerosing cholangitis (1, 6).

ERCP is a standard-reference imaging technique used in the diagnosis of primary sclerosing cholangitis (1, 3, 5). Although dilatation of the bile duct and wall thickening can be imaged by ultrasonography and computed tomography, these findings are insufficient for the diagnosis of primary sclerosing cholangitis (6). MRCP is a non-invasive alternative technique to ERCP for imaging the biliary anatomy and pathology of the bile ducts (1, 7). Although ERCP has some advantages such as high sensitivity in pointing out the pathology of the peripheral intrahepatic ducts, possibility of mechanical dilatation of strictures, stent implantation, and the ability to perform a biopsy concurrently with the examination session, it can cause serious complications such as infectious cholangitis in primary sclerosing cholangitis patients as a result of biliary stasis and sepsis, hemorrhage, the risk of perforation related to excess rigidity of ducts, and pan-

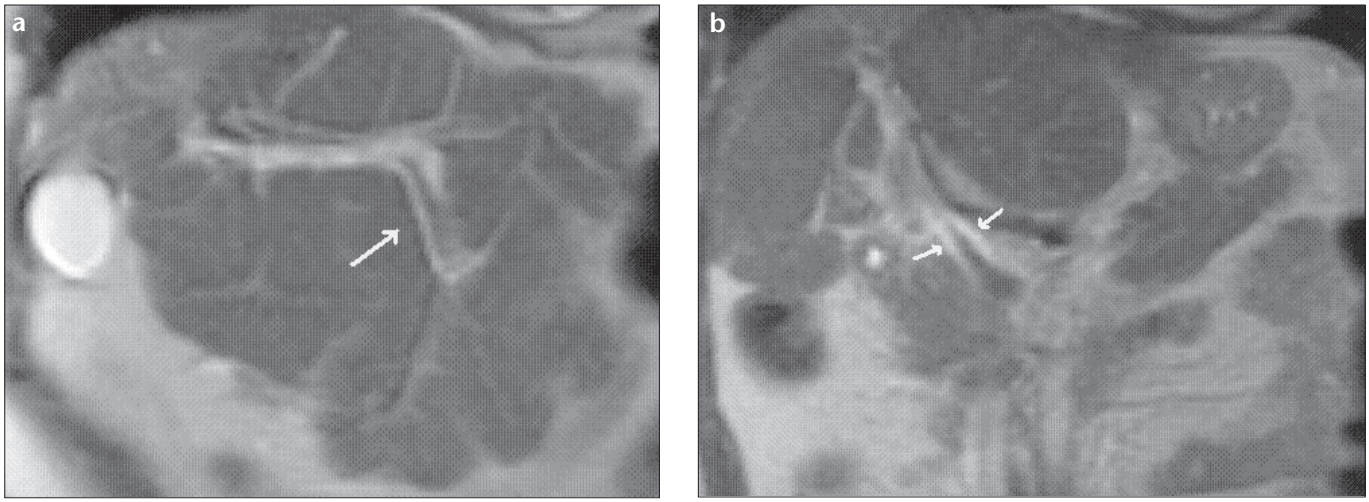

Figure 6. a, b. Periportal edema and contour lobulation. Coronal T2-weighted MR images show increased band shaped intensity compatible with edema, which is in the vicinity of the portal vein *(arrows),* and evident lobulation on the liver surface.

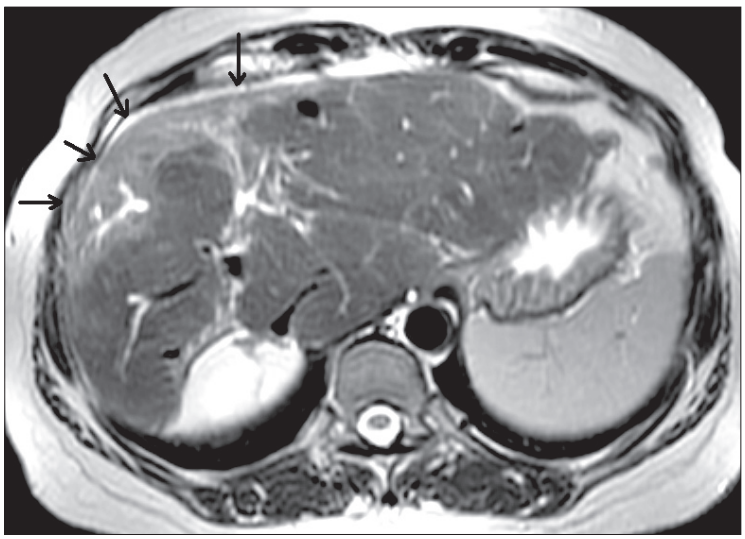

Figure 7. Increased hepatic parenchymal MR signal. Transverse T2-weighted MR image shows increased signal in the peripheral, atrophic parenchymal regions of the right liver lobe (probably secondary to perfusion failure) *(arrows).*

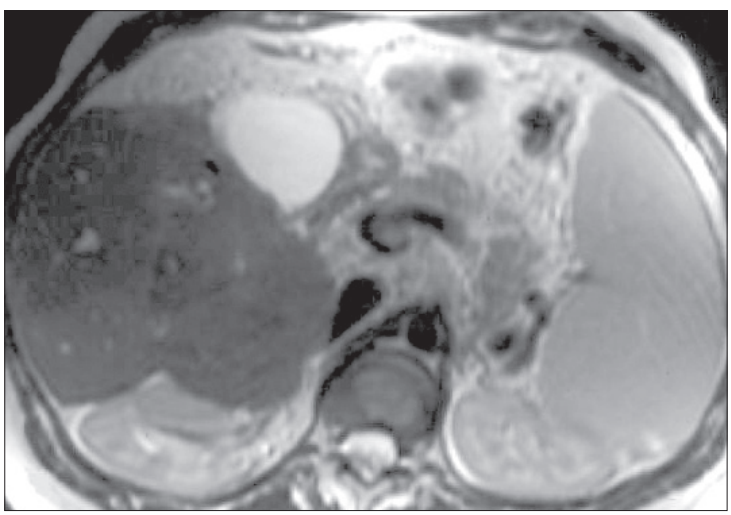

Figure 8. Spherical shape of the liver. Transverse T2-weighted MR image shows lobulation in liver contours and a spherical liver shape. This shape was observed in the cases where left lobe atrophy and caudate lode hypertrophy were evident.

creatic and intestinal perforation (1, 5, 6). The probability of observing these complications in primary sclerosing cholangitis patients is higher than in other patients (5). Furthermore, it was reported that as the disease progresses, ERCP could cause the development of cholestasis (6).

While measuring duct diameters, there is a problem associated with ERCP and techniques used in sectional imaging, namely the pressure caused by the injection of contrast media (1). It has been reported that since MRCP images have a somewhat low spatial resolution, and unlike ERCP, distension does not form as a result of injection of contrast media into ducts, MRCP is not always sufficient for imaging the minor pathology of ducts. In recent studies related to primary sclerosing cholangitis, high specificity and high sensitivity of MRCP has been reported (1, 3, 4, 6). For instance, in a study by Fulcher et al. (6), sensitivity and specificity of MRCP in the diagnosis of primary sclerosing cholangitis were 83%-89% and 92%-99%, respectively, whereas in a smaller group of patients Ernst et al. (4) determined that the sensitivity and specificity were 100%.

Cholangiographic findings change according to the degree of disease. The most important finding is randomly dispersed annular strictures, which are not proportional to dilatation proximally (1, 2). Total or segmentary-subsegmentary involvement can be seen in the liver. Early in the course of the disease, randomly distributed, short, annular intrahepatic strictures alternating with normal or slightly dilated

segments, produce a beaded appearance (1). As the fibrosing process worsens, strictures increase and the ducts become obliterated. Moreover, the peripheral ducts can not be visualized to the periphery of the liver with ERCP, producing a "pruned tree" appearance. In addition, the acute angles formed with the central ducts become more obtuse. With further progression, strictures of the central ducts prevent peripheral ductal opacification in ERCP. Whereas ductal dilatation proximal to strictures can cause obstruction, they can be observed with MRCP (1). Slightly dilated peripheral ducts and central ducts are not in continuity, and this is a characteristic MRCP finding for primary sclerosing cholangitis (1-4). Formation of mural nodes and thickening in the duct wall, diverticula, and webs, although not pathognomonic for primary sclerosing cholangitis, can be observed. Up to 27% of patients with primary sclerosing cholangitis have diverticula. Primary pigmented stones occur in 30% of patients with primary sclerosing cholangitis secondary to bile stasis (1). In this study, the formation of cystic structures compatible with diverticula related to the intrahepatic bile duct was observed in one case (4.34%).

In healthy individuals, in contrast to central ducts, which are wider in diameter and contain sufficient bile in order to be observed in MRCP, it is difficult to image minor ducts with low signal intensity because they contain a negligible amount of bile (3). However, in primary sclerosing cholangitis patients, peripheral ducts can be imaged more effectively with MRCP in terms of dilatation, which is secondary to strictures in central ducts, and bile stasis and strictures can be identified easily in MRCP as well (3). Difficulty during injection of contrast media due to strictures, low infusion pressure, improper catheter positioning, insufficient experience of the person who assists in the endoscopic procedure, and poor patient cooperation may result in non-opacification of the ducts in ERCP and hence causes underestimation of the disease (3, 5).

For primary sclerosing cholangitis patients, MRCP has an important role in following-up the progression of the disease and complications in a noninvasive manner, thereby lowering the mortality and morbidity rates. Complications such as the development of cholangiocarcinoma can be detected in the early phase by including T1- (without and with contrast media) and T2-weighted conventional images in the examination protocol (3, 6).

Various difficulties may be encountered while evaluating bile ducts during MRCP examination. Biliary enteric anastomosis, stent, or air-filled bile ducts arising from recent ERCP, and concentrated bile cause signal loss or divergence from optimal visualization of ducts. Diffuse strictures of central and peripheral ducts prevent visualization of ducts and evaluation of disease in MRCP. In minor ductal pathologies, examination of ducts in a physiological, non-dilated state decreases the sensitivity of MRCP. In cirrhotic livers, visualization of bile ducts is blocked because of the extrinsic pressure of regeneration nodes (3, 6). In addition, MRCP delays percutaneous or endoscopic therapeutic intervention for obstructive pathology of bile ducts during cholangiography (1).

Morphology of the liver may be distorted in the late phase of primary sclerosing cholangitis. In the central regions of the liver, which are not affected by cholestatic parenchymal damage, compensatory hypertrophy and regeneration nodes develop (2). Hypertrophy in the caudate lobe is similar to other types of cirrhosis. In studies by Revelon et al. (8) and Ito et al. (9), caudate lobe hypertrophy was reported at the rate of 23% and 68%, respectively, whereas in our study this rate was found to be 21.73%. When anterior and left lobe medial segments were atrophic, we observed serious atrophy in the left lateral segment, which caused spherical and square like shapes in the liver. To the best of our knowledge, there is no information in the literature related to this finding. Lobulation is dominant on the hepatic surface. In addition to these findings, primary sclerosing cholangitis is a rare cirrhotic disease in which dilatation of bile ducts is also observed (10). Observation of intrahepatic duct stones may be useful while distinguishing other types of cirrhosis. In the atrophic regions of hepatic parenchyma, bile ducts are collected together. Regular contoured intensity changes, probably secondary to perfusion failure, can be observed in the peripheral regions of atrophic parenchymal segments characterized by low signals on T1-weighted and high signals on T2-weighted MR images (2, 8, 9). In our study, the rate of signal increase observed in the peripheral parenchymal regions was 26.08%; in various other studies, it was reported that this rate varies between 23%-72% (2, 8, 9). The rate of signal increase related to edema in the periportal region, which was observed on T2-weighted images in the studies by Revelon et al. (8) and Ito et al. (9), was 40% and 68%, respectively. In our study, this rate was found to be 39.13%.

In some cases, enlarged lymph nodes are observed in the abdomen (9-11). In the study by Ito et al. (9), the rate of periportal lymphadenopathy was 77%, whereas in our study it was 34.78%. It is clear that this rate would have been higher if we had set the sequences for monitoring lymph nodes more accurately in our study, which only utilized MRCP and T2-weighted MR imaging.

In conclusion, MRCP is an alternative method to invasive cholangiographic techniques for the evaluation of intra- and extrahepatic bile ducts, observing disease progression, and for monitoring complications (such as portal hypertension and cholangiocarcinoma) in primary sclerosing cholangitis. Furthermore, MRCP does not carry the risk associated with ionizing radiation, it is a fast imaging method and there is no need for contrast media. T2-weighted images can be obtained at the same time as MRCP, and this has some benefits such as defining the typical liver morphology in primary sclerosing cholangitis patients and detetecting possible portal hypertension findings together with the changes in the bile ducts.

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