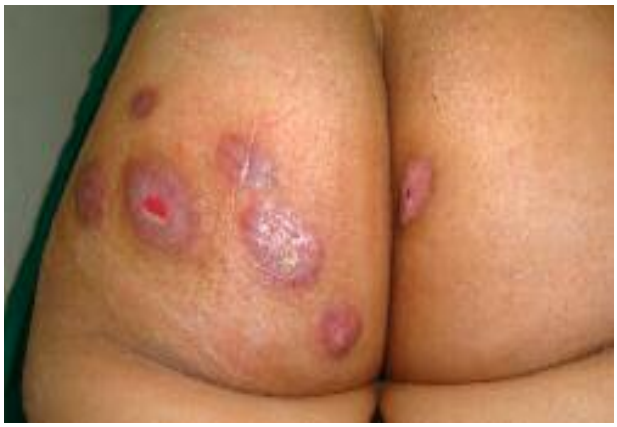
Figure 3 : Good results 1 month after CO2 laser treatment



Conclusion

Unlike xanthelasma treatment, few data are available on therapeutic modalities for TX in familial hypercholesterolemia as this illness is extremely rare. In some cases treatment of the underlying medical disorder may cause regression of xanthomas. But, as observed in this case, the patient continued to develop new cutaneous xanthomas although she was receiving *simvastatin* and *cholestyralamin*.

We used CO2 laser in our patient because it allows treatment of widespread TX in one session under local anesthesia. To our knowledge this is the first reported case of TX treated by CO2 laser with a good cosmetic result. CO2 laser appears as safe and effective for the treatment of TX and should be considered a good alternative to surgery for widespread lesions. Further studies including more patients should be performed to confirm such results.

References

1. Raulin C, Schoenermark MP, Werner S, Greve B. Xanthelasma palpebrarum: treatment with the ultrapulsed CO2 laser. Lasers Surg Med 1999;24:122-7.
2. Alster TS, West TB. Ultrapulse CO2 laser ablation of xanthelasma. J Am Acad Dermatol 1996;34:848-9.
3. Lieb WE, Klink T, Münnich S. CO2 and erbium YAG laser in eyelid surgery. A comparison. Ophthalmologe 2000;97:835-41.
4. Carpo BG, Grevelink SV, Brady S, Gellis S, Grevelink JM. Treatment of cutaneous lesions of xanthoma disseminatum with a CO2 laser. Dermatol Surg 1999;25:751-4.
5. Basar E, Oguz H, Ozdemir H, Ozkan S, Uslu H. Treatment of xanthelasma palpebrarum with argon laser photocoagulation. Argon laser and xanthelasma palpebrarum. Int Ophthalmol 2004;25:9-11.
6. Karsai S, Czarnecka A, Raulin C. Treatment of xanthelasma palpebrarum using a pulsed dye laser: a prospective clinical trial in 38 cases. Dermatol Surg 2010;36:610-7.
7. Park EJ, Youn SH, Cho EB, et al. Xanthelasma Palpebrarum. Treatment with a 1,450-nm-Diode Laser. Dermatol Surg 2011;37:791-6.
8. Fusade T. About the treatment of xanthelasma palpebrarum using a 1,064 Q-switched neodymium-doped yttrium aluminum garnet laser. Dermatol Surg 2011;37:403-4.

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Liver cirrhosis localized in the left lobe: an unusual presentation of small duct primary sclerosing cholangitis.

Liver cirrhosis is defined as a diffuse process characterized by fibrosis and the conversion of normal liver architecture into structurally abnormal nodules [1]. To our knowledge, there is no report of cirrhosis localized only in a part of the liver. However, cirrhosis is a heterogeneous condition with differing clinical manifestations and prognosis depending on the etiology and the severity of hepatic architectural distortion [2]. Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disorder characterized by inflammation, obliteration and fibrosis of the intra-hepatic and/or extra-hepatic biliary ducts [3]. The course of PSC is often progressive, leading to biliary cirrhosis. Small duct PSC is a variant with normal cholangiogram but typical biochemical and histological features of PSC [3.4].

We report herein the case of a woman with small duct PSC at the stage of cirrhosis localized in the left lobe.

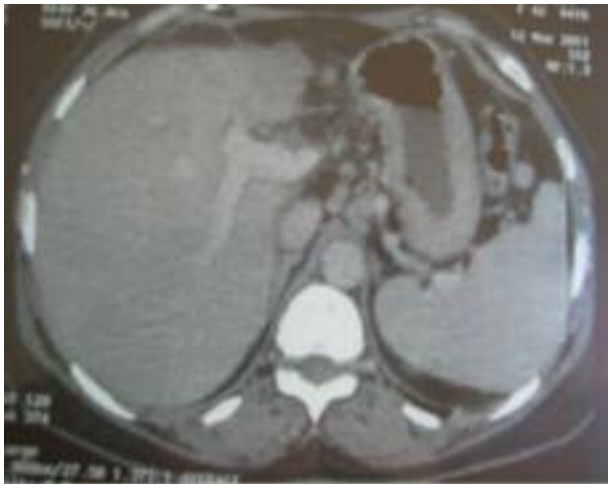
Case-report

A 42 year-old woman was referred to our department for a 2 year history of right upper quadrant pain. There was no pruritus, no jaundice, no chills, and no fever or weight loss. She complained of peripheral arthritis and inflammatory low back pain. In her past medical history, she reported 2 spontaneous abortions and one pre-eclampsia in the last delivery 8 years earlier. She was on no medication and did not consume alcohol. A part from a mild back stiffness, physical examination was normal and revealed no signs of chronic liver disease. Her body mass index was 21 kg/m2. Laboratory data disclosed anicteric cholestasis with elevated ɣglutamyl transpeptidase level of 107 IU/l (normal range (NR) <35 IU/l), alkaline phosphatase of 245 UI/l (NR<110 IU/l) and normal bilirubin level. Serum aspartate aminotransferase was 35 IU/l and alanine aminotransferase 77 IU/l (NR< 45 IU/l). The same cholestatic biochemical picture was present one year ago. The erythrocyte sedimentation rate was 94mm (first hour) and protein C reactive 39 mg/l. Microcytic anemia and elevated ferritinemia was present, hemoglobin was 10 g/dl, while reticulocyte count was low, consistent with an inflammatory origin. Leucocytes count and platelet count were normal. Total cholesterol was 6.85 mmol/l (NR<6 mmol/l), albumin was 32.3 g/l with ɣglobulins 26g/l (IgG = 21.2g/l). Prothrombin time was 91%, factor V 100%. Serological tests for hepatitis B and C were negative. Antismooth muscle, antimitochondrial, anti nuclear, anti liver kidney mocrosome1, anti liver cytosol1, anti GP210, anti PML and anti SLA antibodies were not detected. Human leucocytes antigen was A31 B50.

Abdominal ultrasound and computed tomography (CT)

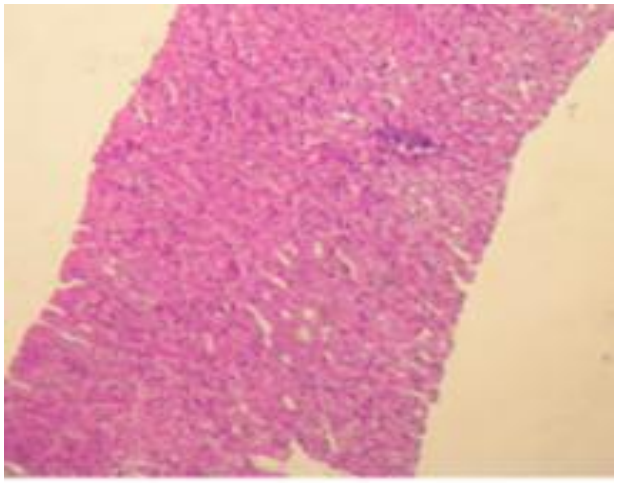
disclosed left portal vein thrombosis and atrophy of the left lobe of the liver while hepatic artery was permeable (Figure 1). There were neither signs of portal hypertension nor bile duct dilatation. Esophagogastroduodenoscopy was normal. Magnetic resonance cholangiography (MRC) was normal.

Figure 1: Abdominal CT scan showing atrophy of the left lobe and left portal vein thrombosis.



Percutaneous liver biopsy was performed on the left and the right lobe and repeated 2 times in order to confirm findings. On the right lobe, it disclosed normal parenchyma without fibrosis and nodularity, even with picrosirius and Masson trichrome coloration, there was only a slight inflammatory infiltrate in one portal triad (Figure 2).

Figure 2: Right liver biopsy showing normal parenchyma.



On the left lobe, there was a diffuse annular fibrosis delimitating regenerative nodules (Figure 3). In increased magnification, there were inflammatory cells in the portal triad, predominantly lymphocytes, infiltrating the interlobular bile duct. In some portal triads, fibrosis surrounded bile ducts and ductular epithelial cell were dystrophic (Figure 4). Elsewhere, proliferation of bile ducts was seen without ductopenia. These findings were consistent with a fibrosing inflammatory cholangitis and cirrhosis localized in the left lobe. Thrombophilia screening (including antiphospholipids antibodies, protein C, S, antithrombin, homocysteinemia, protein C resistance and JAK 2 mutation) was negative. Joint ultra sound did not reveal any signs of synovitis nor enthesitis. Lombar CT scan disclosed features of advanced bilateral sacroiliitis and spondylitis localized at vertebrae L3 and L4 (Figure 5). Coloscopy with biopsies showed no signs of inflammatory bowel disease.

Figure 3: Left liver biopsy showing fibrosis and regenerative nodules.

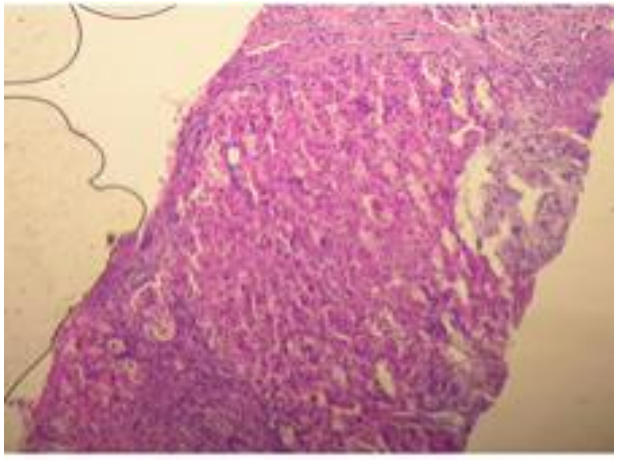
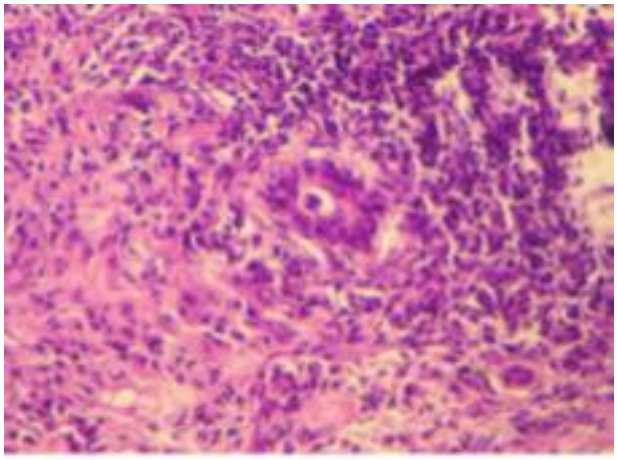


Figure 4: Left liver biopsy in increased magnification showing inflammatory fibrosing cholangitis.



Based on the clinical, biochemical, imaging and histopathological findings, diagnosis of small duct PSC at the stage of cirrhosis localized in the left lobe, associated with ankylosing spondylitis (modified new York criteria) was made. She was started on ursodeoxycholic acid in attempt to slow the progression of liver disease, for the spondylitis, she had excellent response to non steroidal-inflammatory drug therapy.

Conclusion

This report had many diagnosis challenges. First of all, it was unusual to have a picture of cirrhosis localized in a part of the liver. On the other hand, etiology of the cirrhosis was difficult to find. As imaging excluded biliary obstruction, a disease affecting small intra hepatic bile duct was the most likely. Histologic findings were consistent with fibrosing inflammatory cholangitis which is a classic finding in PSC [3,4]. Moreover, ischemic cholangitis was ruled out since hepatic artery was permeable, also, modifications of liver architecture induced by portal obstruction usually don't lead to inflammatory cholangitis. Furthermore, in PSC, involvement of bile ducts can be heterogenous and cases of PSC localized in a small intra hepatic bile duct have been reported [4-8]. Therefore, the diagnosis of small duct PSC at a stage of cirrhosis localized in the left lobe associated with ankylosing spondilitis was the most likely.

Références

1-Hytiroglou P, Snover DC, Alves V, et al. Beyond «cirrhosis»: a proposal from the International Liver Pathology Study Group. Am J Clin Pathol 2012;137:5-9.

2-Pinzani M, Rosselli M, Zuckermann M. Best Practice and Research Clinical Gastroenterology 2011;25:281-90.

3-Portincasa P, Vacca M, Moschette A, et al. Primary sclerosing cholangitis: updates in diagnosis and therapy. World J Gastroenterol 2005;11:7-16.

4- Chapman R, Fevery J, Kalloo A, et al. Diagnosis and management of primary sclerosing cholangitis. AASLD practice guidelines. Hepatology 2010;51:660-78.

5-Kawasaki T, Ueo T, Itani T, et al. A case of localized primary sclerosing cholangitis mimicking intahepatic bile duct cancer. Hepatoloy Research 2001;20:259-64.

6-Kojima T, Shimura T, Takahashi M, et al. A localized primary sclerosing cholangitis preoperatively diagnosed as hilar carcinoma. Hepatogastroenterology 2004;58:961-3.

7-Matsumoto T, Ajiki T, Matsumoto I, et al. Intrahepatic segmental primary sclerosing cholangitis. Surg Today 2006;36:638-41.

8-Sokal EM, Ville de Groyet J, Buts JP, et al. Unifocal stricture of the common bile duct in two children : a localized form of primary sclerosing cholangitis. J Pediatr Gastroenterol Nutr 1990;11:268-74.

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