ABDOMINAL ULTRASONOGRAPHY IN HEPATOLENTICULAR DEGENERATION

A STUDY OF 33 PATIENTS

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Hepatolenticular degeneration (HLD) always evolves with liver involvement in any of the polar clinical picture [10]. Therefore, ultrasonography may yield new information on the degree of that damage and also afford the detection of complications sometimes unnoticed through the anamnesis and the physical examination, such as ascitis, splenomegaly, and cholelithiasis, complications that may occur during the evolution of whatever chronic hepatopathy. The present study was designed to evaluate by ultrasonography the structural changes of the liver, spleen, gall bladder and bile ducts of patients with HLD, comparing the findings with the clinical evolution.

MATERIAL AND METHODS

Thirty-three patients with HLD were studied. The diagnosis was based on the clinical manifestations and laboratory findings. The results were analyzed according to the clinical evolution after D-penicillamine therapy had been started. Twenty patients were males; ages varied from 12 to 49 years. In 4 instances ultrasonography was performed just when the diagnosis was made and before the onset of treatment; in all other patients abdominal ultrasonography was performed until 19 years after the diagnosis and until 30 years after the beginning of the symptoms.

The ultrasonographic examinations were made by means of longitudinal and transversal sections of the liver, gall bladder and spleen, using a real and static time equipment with a 3.5 MHz transductor (Toshiba Sac 12).

RESULTS

Table 1 summarizes the hepatic and splenic changes. The changes of hepatic echotexture showed an evolutional development, the mildest being the posterior atte-

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Changes	Number of cases	Percentage
Hepatomegaly	1	3.0
Hepatic shrinkage	10	30.3
Changes of hepatic echotexture	29	87.8
Splenomegaly	20	60.6
Splenectomy	1	3.0
Ascitis	1	3.0

Table 1 — Hepatic and splenic changes.

nuation of the acoustic beam (Fig. 1) progressing toward a pattern of diffuse increase of the echogenicity, assuming in advanced stages a coarse, reticulated echotexture, and culminating with a bosselated outline of the liver and shrinkage of the parenchyma, especially of the right lobe (Fig. 2). Assuming that the development of splenomegaly is secondary to the structural changes of the liver, especially fibrosis, and that this latter is responsible for the shrinkage of the parenchyma, the correlation between splenomegaly and hepatic dimensions was analyzed (Table 2). The patient submitted to splenectomy had a normal liver size. Table 3 shows the prevalence of cholelithiasis according to sex and age. No abnormality was found in the pancreas and the intra and extra-hepatic bile ducts.

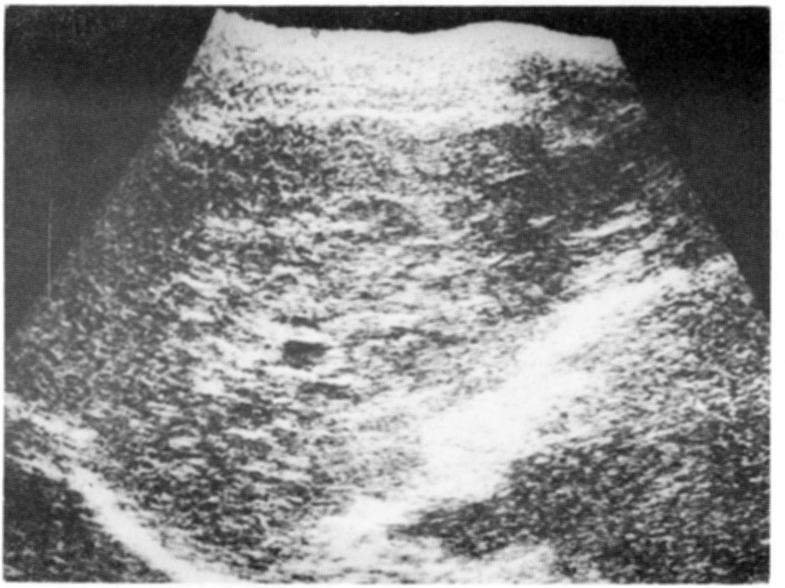

Fig. 1 — Ultrasonographic longitudinal section of the liver, showing increase of the echotexture, with a slight ondulation of the hepatic outline.

Fig. 2 — Ultrasonographic oblique section of the right costal ridge, showing the presence of ascitis and liver retraction with coarse echotexture and bosselated outline.

Hepatic dimensions	Number of cases	Splenomegaly	Percentage
Normal	22	10	45.5
Hepatic shrinkage	10	10	100.0
Hepatomegaly	1	0	0.0

Table 2 — Correlation between hepatic dimensions and splenomegaly.

Age range (years)	Patients with HLD (males / females)	Cholelithiasis (males / females)	Percentage (males / females)
11-20	4 (3/ 1)	1 (1/0)	25.0 (33.3/ 0.0)
21-30	17 (11/ 6)	3 (1/2)	17.6 (5.9/33.3)
31-40	7 (4/ 3)	2 (0/2)	28.5 (0.0/66.7)
41-50	5 (2/ 3)	2 (0/2)	40.0 (0.0/66.7)
Total	33 (20/13)	8 (2/6)	24.2 (10.0/46.2)

Table 3 — Prevalence of cholelithiasis according to sex and age.

COMMENTS

The same ultrasonographic abnormality may represent different morphologic changes, as well as a morphologic change may be expressed by several ways in the ultrasonography. Therefore, increased echogenicity, the attenuation of the posterior acoustic beam and the loss of details of vascular structures inside the hepatic parenchyma point indistinctly to the presence of steatosis or fibrosis [1,3,4,9,11,12]. Among abnormalities found in this study, changes of the hepatic echotexture were the most frequent. They indicate the presence of steatosis or fibrosis, but both may coexist in HLD. Only a patient showed hepatomegaly while 10 (30.3%) showed decrease of the liver size, especially of the right lobe. One could attempt to explain the small incidence of hepatomegaly emphasizing that the ultrasonography was made after the onset of D-penicillamine treatment, when the inflammatory and steatotic processes are in regression and the fibrosis must be established. However, the only patient with hepatomegaly was in treatment for 5 years, with stable clinical picture. In the 4 patients where ultrasonography was performed before the onset of therapy the liver size was reduced in two and was normal in the remainder. Perhaps the absence of hepatomegaly even in that stage of the disease could be due to delay in the diagnosis of HLD, usually four years after the symptoms started. On the contrary, in studies with prevalence of alcoholic liver disease — in spite that the time of evolution has not been considered — hepatomegaly was very frequent [3,12]. Consequentely other factors besides the time of diagnosis could be responsible for the rarity of hepatomegaly in HLD and the significant number of patients (30.3%) with decrease of the liver size. Among these factors is the possibility that copper has a greater fibrogenic activity than other hepatotoxic agents.

Ultrasonography is not infallible in the exclusion or in the diagnosis of an existing liver disease, be it acute or chronic [1,2,3,4,9,11,12]. It is usually admitted that an abnormal ultrasonography in a case with a clinical suspicion of chronic liver disease is a finding which tends strongly to confirm the illness. On the other hand, the absence of ultrasonographic changes in the liver does not exclude a hepatopathy, not even cirrhosis [9]. This can be illustrated by the finding of 3 absolutely normal ultrasonographies in our series, including a patient whose liver biopsy showed cirrhosis when the diagnosis of HLD was made, two years before.

Alterations of the splenic dimensions have been the second most frequent abnormality (62.6% of the patients, being 84.6% in females and 50% in males). If we admit that splenomegaly in HLD is related to the development of liver fibrosis, a significant correlation is found between liver atrophy and splenomegaly: all the patients with shrinkage of the liver parenchyma concomitantly showed splenomegaly. Out of 22 patients with normal hepatic dimensions, only in 9 (40.9%) the splenomegaly was present and one had been submitted to splenectomy because of upper digestive hemorrhage. In the patient with hepatomegaly the splenic dimensions were normal.

The incidence of cholelithiasis in cirrhotic patients is high, probably in view of the increase in the concentration of non-conjugated bilirubin in bile, either caused by an increase in its production by hemolysis, or by decrease of its conjugation in the hepatocyte. When the concentration of indirect bilirubin rises it would occur an imbalance among the solid constituents of bile, inducing the formation of a calcium bilirubinate calculus, a type of lithiasis related to that situation and to chronic hemolysis [7]. In the present study an increase of the prevalence of cholelithiasis was found in females, with an average of 46.2% and a trend to rise with age. In males the average incidence is the same as in the general population [6,8]; it must be emphasized, however, that the sole two male patients with cholelithiasis had 12 and 28 years of age. In other studies on cholelithiasis in cirrhotic patients the incidence varied from the absence of an increase [6,8] to rates around 32% [1,5,7]; the increase, however, was indistinct among both sexes [1,7] and in the several age ranges [1].

In conclusion, abdominal ultrasonography is an important propedeutic resource for the evaluation of patients with HLD, independently of its clinical form, and is also valuable in detecting possible abdominal complications of the disease.

SUMMARY

The ultrasonographic study of 33 patientes with hepatolenticular degeneration showed the following main changes: disorders of the hepatic echotexture (29 cases), changes of the splenic dimensions (21), liver shrinkage (10), cholelithiasis (8), hepatomegaly and ascitis (1). The disorders of liver echotexture exhibited different patterns, from slight to severe changes of the hepatic echogenicity, associated with anatomic distortions of the liver, such as alterations of outline and decrease of dimensions. Liver shrinkage was always accompanied by splenomegaly. The scarcity of hepatomegaly may be explained by the fact that the ultrasonography was performed after the onset of treatment with penicillamine, or by other factors still unknown, copper perhaps possessing a stronger fibrogenetic action than other hepatotoxic agents. Cholelithiasis was very frequent in females (6 of 13 patients), its incidence tending to increase with age. Regarding males there was no increase when the incidence was compared to the general population. The two sole males were young, an unusual finding in normal men.

RESUMO

Ultra-sonografia abdominal na degeneração hepatolenticular: estudo de 33 casos.

O estudo ultra-sonográfico de 33 pacientes com degeneração hepatolenticular revelou as seguintes alterações princpais: desordens da ecotextura hepática (29 casos), alterações das dimensões esplênicas (21), contração do fígado (10), colelitíase (8), hepatomegalia e ascite (1). As desordens da ecotextura hepática se apresentaram sob diversas formas, desde leves até graves alterações da ecogênese hepática, associadas com distorções anatômicas do fígado, tais

como modificações do perfil e redução das dimensões. A contração hepática sempre se acompanhou de esplenomegalia. A raridade da hepatomegalia pode ser explicada pelo fato de que a ultra-sonografia foi realizada após o início do tratamento com penicilamina, ou por outros fatores ainda desconhecidos, como a possibilidade de que o cobre possua uma ação fibrogênica maior do que a de outros agentes hepatotóxicos. A colelitíase foi muito freqüente no sexo feminino (6 de 13 pacientes) e sua incidência tendeu a crescer com a idade. Quanto aos doentes do sexo masculino, não se notou aumento da incidência em relação à freqüência na população geral. Os dois únicos pacientes do sexo masculino eram jovens, fato raramente observado nos homens normais.

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