ORIGINAL ARTICLE

Idiopathic portal hypertension and related ultrasound findings

Hipertensão portal idiopática e suas alterações ultrassonográficas

João Paulo Kawaoka Matushita[1], José de Laurentys-Medeiros[2], Julieta Sebastião Matushita[3], Cristina Sebastião Matushita[4], João Paulo Kawaoka Matushita Jr.[5]

DOI: 10.5935/2238-3182.20130047

ABSTRACT

Objective: to characterize idiopathic portal hypertension clinically and by ultrasound (US). Patients and methods: 60 patients with clinical diagnosis of chronic liver disease and portal hypertension were retrospectively assessed by color Doppler US, from January 1996 to January 2006, in a privately owned clinic. Two cases of idiopathic portal hypertension were found. **Comments:** in young patients with splenomegaly, recurring hematemesis, anemia, ascites, normal liver function and portal hypertension without evidence of cirrhosis, US revealed livers of usual size, heterogeneous texture and flat surface, without deformities in the branches of the intra-hepatic portal veins. The portal vein walls were also thickened starting at 3 mm and marked by an abrupt narrowing of the secondary intra-hepatic portal veins, accompanied by splenomegaly.

Key words: Hypertension, Portal; Liver Diseases; Ultrasonography, Doppler, Color.

RESUMO

Objetivo: *caracterizar clínica e pela ultrassonografia (US) a hipertensão portal idiopática.* **Pacientes e métodos:** *foram avaliados, retrospectivamente, por US com doppler colorido, 60 pacientes com diagnóstico clínico de hepatopatia crônica e hipertensão portal, de janeiro de 1996 a janeiro de 2006, em clínica privada, encontrando dois casos de hipertensão portal idiopática.* ***Comentários:*** *em jovens com esplenomegalia, hematêmese recorrente, anemia, ascite, funções hepáticas normais e hipertensão portal sem evidência de cirrose, foram observados à US fígado de tamanho usual, textura heterogênea e superfície lisa, ausência de deformidades das ramificações das veias portais intra-hepáticas, espessamento da parede da veia porta maior ou igual a 3 mm e estreitamento abrupto das veias portais secundárias intra-hepáticas acompanhado de esplenomegalia.*

Palavras-chave: *Obesity; Pregnant Women; Public Health; Women's Health.*

[1] Associate Professor, School of Medicine, Universidade Federal de Minas Gerais. Radiologist at the Diagnostic Image Center Dr. Matsushita Ltda. Belo Horizonte, MG – Brazil.
[2] Full Professor of Medical Semiology, Faculdade de Ciências Médicas de Belo Horizonte. Belo Horizonte, MG – Brazil.
[3] Radiologist. Diagnostic Image Center Dr. Matsushita Ltda. Belo Horizonte, MG – Brazil.
[4] Nuclear medicine physician. Diagnostic Image Center Dr. Matsushita Ltda. Belo Horizonte, MG – Brazil.
[5] Physician. Graduate student in Radiology and Image Diagnostics, School of Medicine, Universidade Federal do Rio de Janeiro-FM-UFRJ. Rio de Janeiro, RJ – Brazil.

INTRODUCTION

Idiopathic portal hypertension (IPH), also known as Banti's syndrome, was first described by Banti in 1889[1] and researched in India by Ramalingaswami et al[2]. in 1962. Many synonyms are used to identify it, such as: hepatoportal sclerosis,[3] non-cirrhotic portal fibrosis,[4] obliterative portal venopathy of the liver,[5] non-cirrhotic intrahepatic portal hypertension,[6] benign intrahepatic portal hypertension,[7] and idiopathic portal presinusoidal hypertension.[8]

Submitted: 08/23/2012
Approved: 01/21/2013

Institution:
Diagnostic Image Center Dr. Matsushita Ltda
Rua dos Caetés 530/216, Centro
Belo Horizonte, MG – Brasil. CEP 30120-080

Corresponding Author:
João Paulo Kawaoka Matushita
E-mail: jpauloejulieta@gmail.com

Little is known of its etiopathogenesis. Sherlock[8] believed that lesions in the branches of the intrahepatic portal venous system and sinusoidal endothelial cells were predisposing factors. Its etiology can be infectious, toxic, immunologic,[9] and, most of the time, unknown.[8]

IPH is more frequent in women, in the ratio of 2: 1 to 4: 1,[10-12] and the average age of occurrence ranges from 25 to 35 years.[9]

This work aims to present some clinical characteristics of IPH under ultrasound.

PATIENTS AND METHODS

From January 1996 to January 2006, sixty patients with a clinical diagnosis of chronic liver disease and portal hypertension were examined retrospectively in a private clinic using color Doppler ultrasound, and two cases of IPH were found.

The device used to perform the ultrasound was a Toshiba PowerVision 6000 with broadband probe, 3-5 mHz multi-frequency convex probe and a linear array probe of 7-10 mHz with color Doppler.

The exams were documented using the SISMED image capturing system.

RESULTS

The results are presented in Tables 1 to 3 and Figures 1 to 4.

DISCUSSION

IPH can be characterized primarily by prolonged splenomegaly, recurrent hematemesis, anemia, small or moderate ascites that respond well to diuretics, hepatocellular functions close to normal, and portal hypertension without evidence of cirrhosis or portal vein obstruction (Table 4).

Non-cirrhotic hepatic fibrosis can be differentiated from cirrhosis as the liver function is preserved.

Table 1 - Age, sex, and clinical picture

Case	Age	Sex	AL	PH	DH	SP	PH	A
1	26 Y	M	No	No	Yes	Yes	Yes	Yes
2	24 Y	F	No	No	Yes	Yes	Yes	Yes

AL: alcoholism; PH: previous hepatitis; DH: digestive hemorrhage; SP: splenomegaly; PH: Portal hypertension, A: anemia.

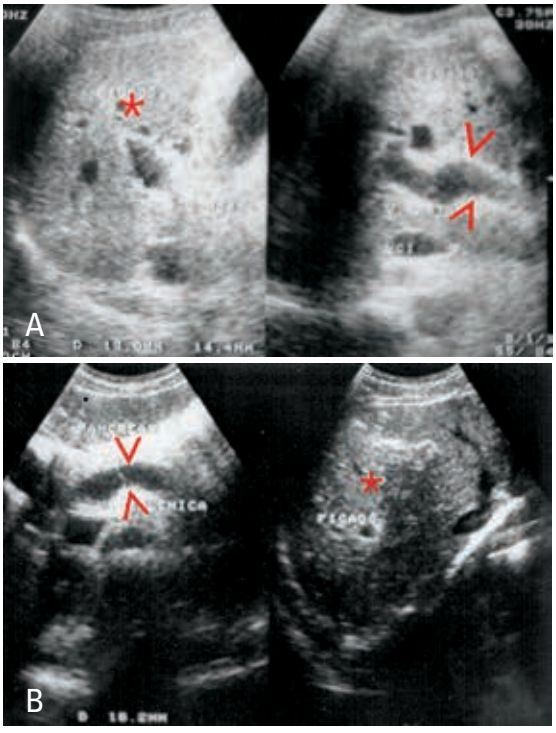
Table 2 - Laboratory exams

Case	Anemia	HbsAg	AST/SGPT	Anti Hbs	Anti Hbc IgC	Anti HIV through Elisa	L
1	Hypochromic	–	N	–	–	–	Leukopenia
2	Hypochromic	–	N	–	–	–	Leukopenia

L: leucograma.

Table 3 - Ultrasound aspects of the liver and spleen

Parameters	Case 1	Case 2
Liver, size	Normal	Normal
Liver, texture,	Heterogeneous	Heterogeneous
Liver, surface	Smooth	Smooth
homogeneous thickening of the periportal wall intrahepatic Diffusion	3.2 mm	3.0 mm
Abrupt thickening of secondary ramifications	Yes	Yes
Flux	Hepatofugal	Hepatofugal
Dilated hepatic portal vein	14.4Mm	14.7Mm
Dilated splenic vein	16.2Mm	16.0Mm
Gallbladder with varices	Thickened walls	Thickened walls
Gastric/perivesicular, esophageal varices, hepatic and/or splenic hilum	Yes	Yes
Spleen	Splenomegaly	Splenomegaly



Figures 1 A e B - A and B - Liver of seemingly normal size, heterogeneous texture (*), with the portal and splenic veins dilated and thickened (red arrows).

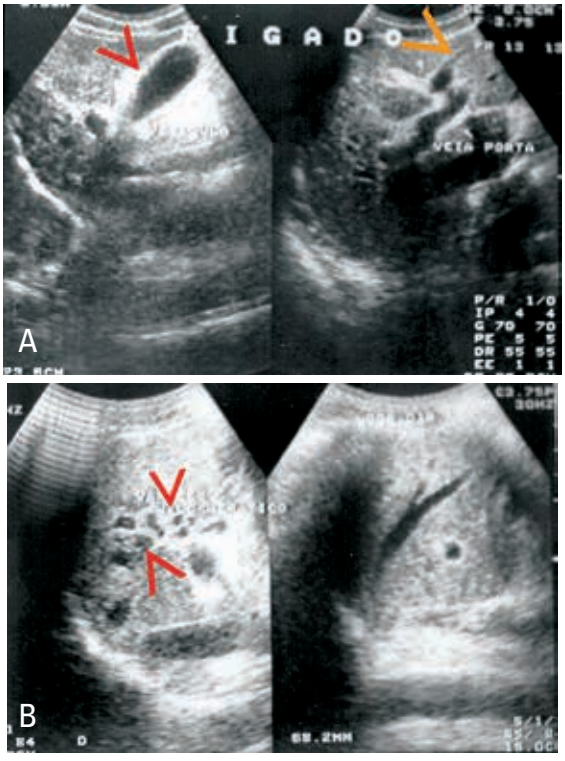


Figure 2 - A: Intense thickening of bladder wall (red arrow) and portal vein (orange arrow). Recanalization of the paraumbilical vein. Left. B: Varicose veins in the hepatic hilum (red arrows).

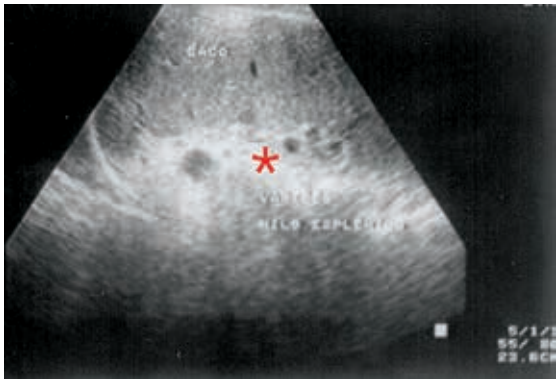
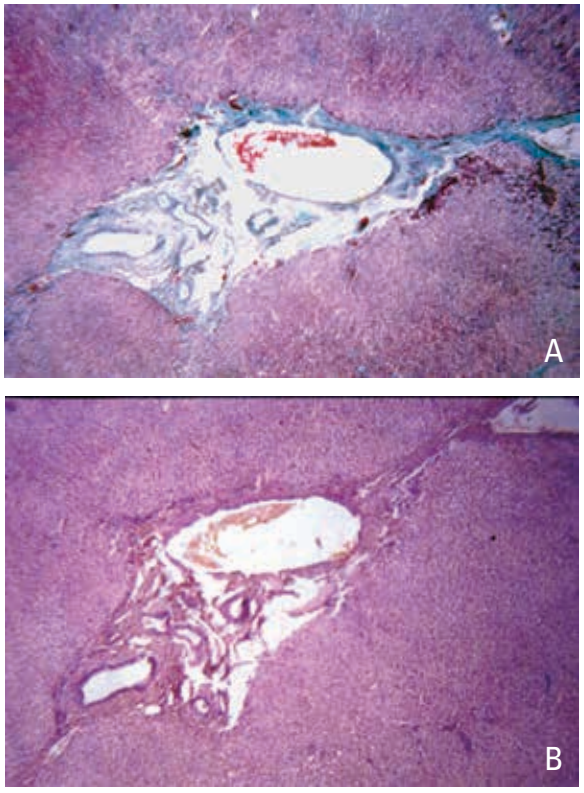


Figure 3 - Splenomegaly with varicose veins in the splenic hilum (*).



Figures 4 A e B - Liver Biopsy. Fragments of the liver showing hepatocyte with polyploidy and absence of inflammatory infiltrate. Major thickening of the portal veins. Sclerosis of some of the vessels of the intrahepatic venous network with minimal fibrosis.

The ultrasound findings that distinguish IPH from cirrhosis include: liver with usual size, heterogeneous texture and smooth surface, absence of deformities in the branches of intrahepatic portals veins, thickening of the portal vein wall larger than or equal to 3 mm, and abrupt narrowing of secondary intrahepatic portal veins, followed by splenomegaly.

Table 4 - A comparison of the various parameters that help distinguish IPH from cirrhosis in Brazil.[14]

Parameters	IPH	Liver cirrhosis
Etiology	Unknown	Viral hepatitis, alcohol, drugs, schistosomiasis
Incidence	Infrequent	Common
Sex and age	Middle age Female sex	Middle age Male sex
Clinical symptoms	Most do not change over many years	Most are progressive
Pathological processes	Intense periportal fibrosis	Appearance of diffuse pseudo-nodules
Formation of intrahepatic shuts	No	Very evident
Arterial flux in the liver	Decreased	Increased
Association with hepatocellular cancer	No	Common

The natural history of IPH is unknown and seems to have a benign evolution, provided that recurrences of esophageal varices are controlled. Progressive hepatic fibrosis, however, can lead to liver damage and death.

CONCLUSION

IPH liver disease is characterized by variations of the degrees of fibrosis and fibro-sclerotic changes in the portal venous system. However, the pathological changes of this syndrome in the liver are not pathognomonic and most changes seen can be the result of long portal venous circulatory insufficiency.

REFERENCES

1. Banti G. Splenomegalie mit Leberizirrhose. Beitrage zur pathologischen. Anat Allgemeinea Pathol. 1889; 24:21-33.
2. Ramalingaswami B, Wig KL, Sama SK. Cirrhosis of the liver in northern India. A clinico pathological study. Arch Intern Med. 1962; 110:350-8.
3. Mikkelsen WP, Edmondson HA, Peters RL, Reynolds TB. Extra and intrahepatic portal hypertension without cirrhosis (hepatoportal sclerosis). Ann Surg. 1965; 162:602-20.
4. Sarin SK. Progress report. Non- cirrhotic portal fibrous. Gut. 1989; 30:406-15.
5. Nayak NC, Ramalingaswami B. Obliterative portal venopathy of the liver. Arch Pathol Med. 1969; 87:359-69.
6. Levison DA, Kinghan JCG. Liver pathology in benign intra hepatic portal hypertension. In Okuda K, Omata M. (eds). Idiophatic portal hypertension. Tokio: University of Tokio Press; 1983. p. 81-4.
7. Levison DA, Kinghan JCG, Dawson AM, Stansfeld AC. Slow cirrhosis or no cirrhosis. A lesson causing benign intra hepatic portal hypertension. J Hepatol. 1982; 137:253-72.
8. Polish E, Christie J, Cohen A. Idiopathic presinusoidal portal hypertension (Banti's syndrome). Ann Intern Med. 1962; 56:624-27.
9. Sherlock S, Dooley J. The portal venous system and portal hypertension. In: Diseases of the liver and biliary system. 11. ed. Milan: Blackwell; 2002. p. 168-9.
10. Habibullah CM, Rao GN, Murthy DK. Non portal fibrosis in Andtora Pradesh. J Assoc Phys India. 1978; 26:379-82.
11. Sama SK, Bhargava S, Gopi N. Non cirrhotic portal fibrosis. Am J Med. 1971; 51:160-9.
12. Sarin SK, Sachdev G, Nanda R. Follow–up of patient after varical eradication. A Comparison of patient with cirrhosis, non–cirrhotic portal fibrosis and extrahepatic Obstruction. Ann Surg. 1986; 204:78-82.
13. Dhiman RK, Chawla Y, Vasishta RK, Kakkar N, Dilawari JB, Trehan MS, *et al.* Non – cirrhotic portal fibrosis (idiopathic portal Hypertension): Experience with 151 patientes and a review of the literature. J Gastroenterol Hepatol. 2002 Jan; 17(1):6-16.
14. Okudaira M, Ohbu M, Okuda K. Idiopathic portal hypertension and its pathology. Sem Liver Dis. 2002; 22(1):59-70.