

Cruveilhier-Baumgarten syndrome: sonographic aspects

Síndrome de Cruveilhier-Baumgarten: aspectos ultrassonográficos

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DOI: 10.5935/2238-3182.20130010

ABSTRACT

Objective: The purpose of this paper is to analyze the sonographic changes that characterize the Cruveilhier-Baumgarten syndrome and its clinical characteristics. Cruveilhier-Baumgarten syndrome is characterized by a prominent paraumbilical vein, with dilated superficial veins of the abdominal wall radiating from the umbilicus of patients with chronic liver disease and portal hypertension. **Patients and methods:** In a sonographic evaluation of 60 patients diagnosed with chronic liver disease with portal hypertension we found 4 cases of Cruveilhier-Baumgarten syndrome, which we studied with color doppler sonography. **Results:** All patients were male, in their third decade of life, with a clinical history of splenomegaly and varicose abdominal wall veins. Patients had schistosomiasis and micronodular cirrhosis on histopathological examination. We also evaluated two patients with esophageal varices without hematemesis. All patients had ultrasound signs of splenomegaly and chronic liver disease, with hepatopetal flow in all segments of the portal vein and hepatofugal flow in the efferent veins of the liver (paraumbilical vein in the falciform ligament and the anterior abdominal wall surface veins). **Conclusion:** CBS is characterized by the association of signs of chronic liver damage with splenomegaly, distention of paraumbilical veins (portal hypertension) and blood flow towards the superficial abdominal wall veins, which are dilated in the umbilical region, associated with an audible bruit in both clinical and sonographic examination.

Key words: Hypertension, Portal; Cruveilhier-Baumgarten Syndrome, Liver Diseases; Ultrasonography, Doppler, Color.

RESUMO

Objetivo: este trabalho analisa as alterações ultrassonográficas que caracterizam a síndrome de Cruveilhier-Baumgarten (SCB), com suas alterações clínicas. A SCB caracteriza-se pela veia paraumbilical proeminente, com dilatação das veias superficiais da parede abdominal de aspecto radiado provenientes do umbigo dos pacientes com hepatopatia crônica e hipertensão portal. **Casística e métodos:** em avaliação ultrassonográfica de 60 pacientes com diagnóstico de hepatopatia crônica com hipertensão portal foram encontrados quatro casos com a SCB em que foi realizado o estudo por meio do doppler colorido. **Resultados:** todos os pacientes avaliados eram do gênero masculino, estavam na terceira década de vida, tinham história de esplenomegalia, varizes na parede abdominal e eram portadores de esquistossomose e aspecto histopatológico de cirrose micronodular. Foram analisados também dois pacientes com varizes do esôfago sem clínica de hematêmese. Ultrassonograficamente, todos os pacientes tinham sinais de hepatopatia crônica com esplenomegalia, com fluxo hepatopetal em todos os segmentos da veia porta e fluxo hepatofugal nas veias que deixam o fígado (veia paraumbilical no ligamento falciforme e nas veias superficiais da parede abdominal anterior). **Conclusão:** a SCB caracteriza-se pela associação de sinais de hepatopatia crônica com esplenomegalia, veias paraumbilicais com aumento de calibre (hipertensão porta) e com fluxo para as veias superficiais da parede abdominal, que se acham

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Submitted: 04/09/2012
 Approved: 01/21/2013

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dilatadas na região umbilical e com sopro à ausculta pelo ultrassom e ao exame físico.

Palavras-chave: Hipertensão Portal; Síndrome de Cruveilhier-Baumgarten; Hepatopatias; Ultrassonografia Doppler em Cores.

INTRODUCTION

Cruveilhier-Baumgarten Syndrome (CBS) was first described by Pégot in 1833, by Cruveilhier in 1852, and by Von Baumgarten in 1907.¹

It is characterized by the presence of very a prominent paraumbilical vein or recanalized umbilical vein associated with portal hypertension, and compensated cirrhosis or normal liver in patients with congenital patency of the umbilical vein.² The diagnosis is suggested by engorged superficial veins in the abdominal walls, radiating from the navel, whose appearance earned it the denomination of “caput medusae”.³⁻⁵ It is also associated with bruit in the umbilical region (the Cruveilhier-Baumgarten sign),⁶ which can be heard by placing the stethoscope over the engorged vessels, and is a result of the increased and turbulent blood flow in these vessels.

There is no distinction between CB syndrome and disease.⁷ The only difference relates to onset of early portal hypertension.⁸

The purpose of this paper is to enumerate the clinical and sonographic changes in CBS.

PATIENTS AND METHODS

A total of 60 patients diagnosed with chronic liver disease and portal hypertension underwent color Doppler ultrasound examinations from January 1996 to December 2006.

Among those patients we found 4 cases of Cruveilhier-Baumgarten Syndrome, which we further studied using sonography.

The ultrasound equipment used was a Toshiba Power Vision 6000, with 3-5 mHz convex multi-fre-

quency and 7-10 mHz linear color Doppler broadband probes.

Examinations were documented using Sismed captured images.

RESULTS

All patients were male, in their third decade of life, with a clinical history of splenomegaly and varicose abdominal wall veins. Patients had schistosomiasis and micronodular cirrhosis upon hystopathological examination. We also evaluated two patients with esophageal varices without hematemesis (see Tables 1 and 2 and Figures 1 to 7).

DISCUSSION

The combination of splenomegaly and portal hypertension is often found in patients with cirrhosis or portal vein thrombosis. CBS diagnosis is made upon finding the paraumbilical vein.¹ A recanalized paraumbilical vein observed using color Doppler ultrasonography is a specific sign of portal hypertension of hepatic or post-hepatic origin.⁹

The sonographic features found in this study were: coarse echotexture and nodular surface; liver of irregular contours and coarsely serrated; liver with increased left lobe and reduced right lobe; thickening of the gallbladder wall; echogenic periportal thickening extending from the hepatic hilum to the periphery of the liver; engorged portal and splenic veins; increased caliber of the portal vein, especially to the left; hepatopetal flow in the right and left portal veins; hepatofugal collaterals in the right and left portal veins⁸; engorged paraumbilical vein following the falciform ligament, connecting the left portal vein to the anterior abdominal wall veins to the umbilicus and from the umbilicus to the upper or lower epigastric veins or through the subcutaneous veins in the anterior abdominal wall, the “caput medusae”, to achieve the IVC.

Table 1 - Clinical findings of all patients with portal hypertension upon ultrasound examination

Cases	Sex	Age (Years)	Clinical history	Etiology	Biopsy
Cases	Sex	Age (Years)	Clinical history	Etiology	Biopsy
2	Masc	34	Esplenomegalia, varizes na parede abdominal	Esquistossomose	Cirrose micro-nodular
3	Masc	31	Esplenomegalia, varizes na parede abdominal	Esquistossomose	Cirrose micro-nodular
4	Masc	39	Esplenomegalia, varizes esofágicas e na parede abdominal	Esquistossomose	Cirrose micro-nodular

Color Doppler ultrasound examination of 55 patients with recanalized paraumbilical vein found that 39 displayed the classic CBS hepatic venous flow, that is, hepatopetal flow in all segments of the portal vein and hepatofugal flow from the liver via the paraumbilical vein to the falciform ligament to the veins in the abdominal wall surface.

Table 2 - Sonographic findings in all patients assessed by conventional color Doppler ultrasonography

1	Coarse echotexture and nodular surface
2	Irregular and coarsely serrated liver contours
3	Increased left liver lobe and reduced right liver lobe
4	Dilated splenic and portal veins
5	Thickened gallbladder wall
6	Echogenic periportal thickening from hepatic hilum to liver periphery
7	Engorged portal vein, especially to the left
8	Flow in the right and left portal veins, of hepatopetal aspect
9	Collaterals in the right and left portal veins, of hepatofugal aspect
10	Engorged umbilical vein following the falciform ligament, connecting the left portal vein to the superficial veins of the anterior abdominal wall to the umbilicus and from the umbilicus to the upper or lower epigastric veins or through the caput medusae subcutaneous veins of the anterior abdominal wall to reach the inferior vena cava (IVC)



Figure 1 - Cirrhotic portal hypertension (Photo donated by Prof. Dr. José de Laurentys-Medeiros).



Figure 2 - Hepatic sonography with color Doppler. Marked hyperechogenicity of periportal wall. Engorged portal vein, with hepatopetal flow.

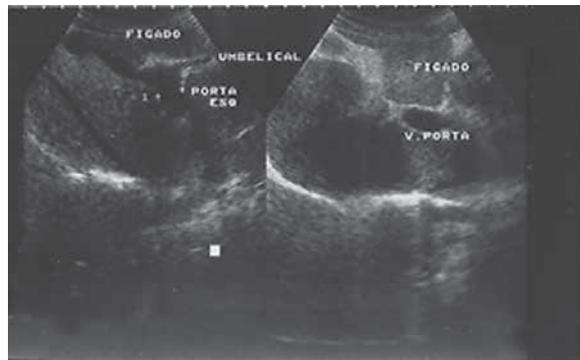


Figure 3 - Hepatic sonography. Livers showing texture changes of chronic aspect. The dilated paraumbilical vein following the falciform ligament.



Figure 4 - Hepatic sonography with color Doppler. Texture changes of chronic aspect in the liver, and paraumbilical vein connecting the superficial veins of the abdominal wall, with hepatofugal flow.

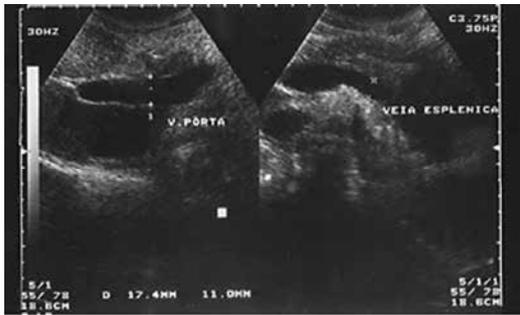


Figure 5 - Hepatic sonography. Engorged portal vein (17.4 mm) and splenic vein (11.0 mm).



Figure 6 - Hepatic sonography with color Doppler. Showing the left paraumbilical vein connecting the superficial veins of the abdominal wall.



Figure 7 - Hepatic sonography with color Doppler. Showing the left paraumbilical vein circling the liver.

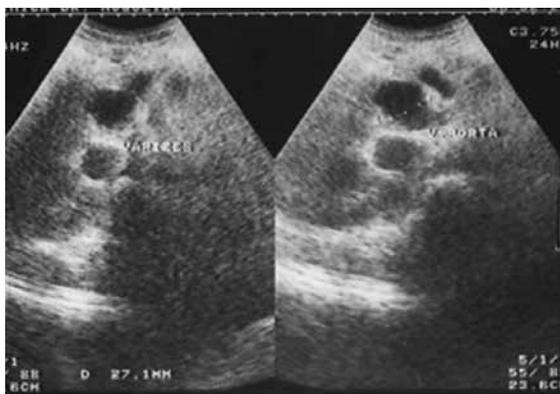


Figure 8 - Superficial abdominal sonography. Varices at the umbilicus level.

The presence or absence of an intra-hepatic afforestation depends on the functional value of the paraumbilical veins. The larger the caliber of the vein, the lower the intrahepatic afforestation tends to be. Absence of afforestation can initially be felt in the left branch of the portal vein, also reaching, upon the swift development of the umbilical vein, the right branch.

In CBS, the left branch is more voluminous because it receives the largest volume of blood from the portal vein, blood that reaches the IVC via the paraumbilical vein. The left portal branch functions as the de facto continuation of the portal vein trunk, a consequence of the large volume of portal blood passing through the paraumbilical vein, which originates in the left portal branch. However, this bifurcation in the portal vein is almost always visible. It seems that the presence of the pervious umbilical vein prevents esophageal varices in patients with CBS⁹ because it decompresses the portal system.¹⁰

The 1982 study by Aagaard *et al.*¹¹ with 107 patients with cirrhosis and portal hypertension detected through splenoportography 28 patients (26%) with recanalized paraumbilical vein. The authors concluded that recanalization of the paraumbilical vein often occurs in patients with cirrhosis and portal hypertension. Even the engorged paraumbilical veins and the massive spontaneous portosystemic communication were unable to relieve portal hypertension, avoid esophageal varices or protect esophageal varices from bleeding and from ascites. In our study, we found two patients with esophageal varices without hematemesis in their clinical evolution.

Color Doppler ultrasound enabled us to assess the presence and direction of the flow in the porto-systemic collateral route connecting the paraumbilical vein to the left portal vein and to the veins in the anterior abdominal wall surface, forming the so-called “caput medusae”.

CONCLUSION

Signs of chronic liver disease with esplenomegalia and engorged paraumbilical vein are findings that indicate portal hypertension. However, when these findings are associated with the presence of engorged superficial veins in the abdominal wall near the umbilical region and bruit upon auscultation (as observed by ultrasound and physical examination) we call it CBS.

Classic CBS is characterized by the hepatopetal flow in all segments of the portal vein and the hepatofugal flow in the veins leaving the liver (paraumbilical vein in the falciform ligament and in the veins of the abdominal wall surface).

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