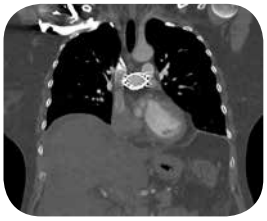

Acute Budd-Chiari Syndrome Secondary to Alcoholic Cirrhosis. Interventional Management and Description of a Complication

Síndrome de Budd-Chiari agudo secundario a cirrosis alcohólica. Manejo intervencionista y descripción de una complicación

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Summary

Budd-Chiari syndrome (BCS) is an infrequent and potentially fatal disease if not diagnosed and treated early. We describe a case of BCS secondary to obstruction of intrahepatic inferior cava vein and left and middle suprahepatic veins, with interventional management (stent placement in the cava vein) and subsequent complication with stent migration to the pulmonary artery.

Resumen

El síndrome de Budd-Chiari (SBC) es una patología poco frecuente y potencialmente fatal si no se diagnostica y se trata a tiempo. Se describe un caso de SBC secundario a obstrucción de la vena cava inferior intrahepática y venas suprahepáticas izquierda y media, con manejo intervencionista (colocación de endoprótesis en la vena cava) y posterior complicación con migración de la endoprótesis a la arteria pulmonar.

Introduction

Budd-Chiari syndrome (BCS) was described by Budd in 1845 and by Chiari in 1899. Its clinical and paraclinical manifestations show liver disease secondary to obstruction of the venous drainage in the suprahepatic veins (SHV), in the inferior vena cava (IVC) intrahepatic, in its union with the right atrium or postsinusoidal veins (1,2). Early diagnosis has important implications in the treatment and prognosis of the patient. We present a case of BCS of acute manifestation due to obstruction of intrahepatic IVC and left and intermediate SHV.

Description of the case

A 55-year-old woman who consulted for five days of emesis, coluria and abdominal pain. As a relevant background, he reported heavy alcoholism for 30 years and high blood pressure. The physical examination is icteric, with pain on palpation of the right hypochondrium, hepatomegaly, without signs of peritoneal irritation. Leukocytosis (12,800, 79% neutrophils) and platelets in 166,000, total bilirubins 5.8, direct 4.7, albumin 2.5, and hyperbilirubinuria. The ultrasound showed hepatomegaly with diffuse increase in echogenicity, without focal lesions, and signs of portal hypertension (enlarged portal, splenomegaly and ascites) (figure 1).

Abdominal tomography (CT) confirmed the signs of portal hypertension and showed intrahepatic IVC occupation by low density eccentric material (thrombus), which decreased its caliber by 80%; signs suggestive of hepatic vascular involvement (heterogeneous density with peripheral areas of low density) were found in the liver, which led to the suspicion of BCS (figure 2). The patient received full anticoagulation with low molecular weight heparin and antiaggre-

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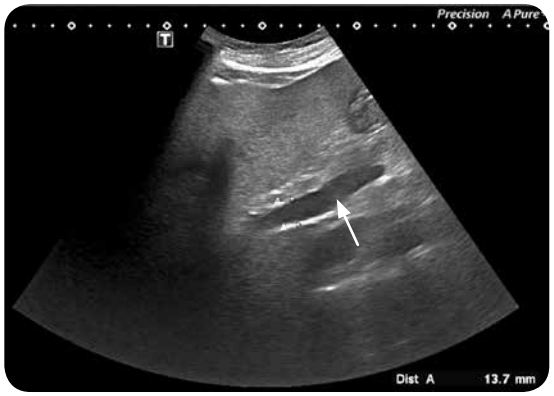

Figure 1. Liver ultrasound: diffuse increase in liver echogenicity without focal lesions. Increase in the diameter of the portal vein (arrow).

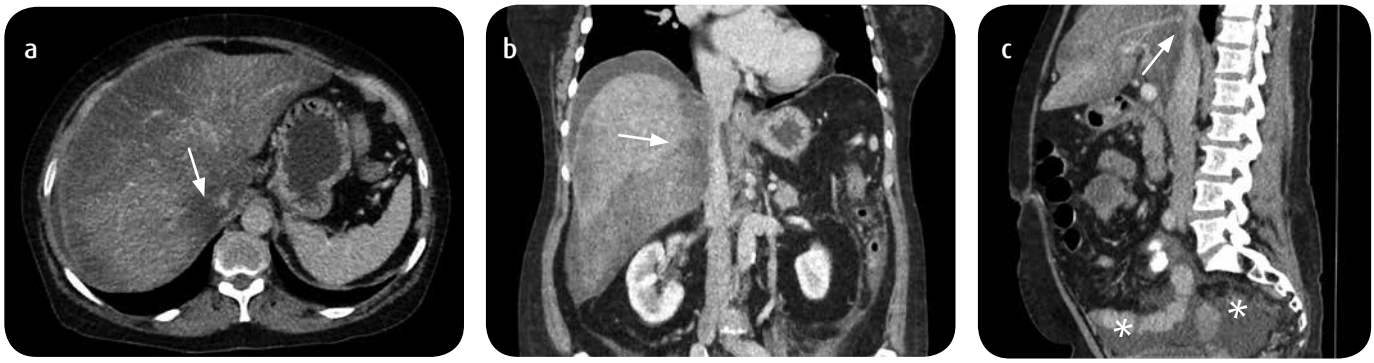

Figure 2. CT with contrast medium. a) Delineated with white line, irregular IVC is observed by exophytic thrombus in its lumen. The liver density is heterogeneous. b) Obstruction of the IVC (white arrow). c) IVC occlusion area (arrow) and free fluid in the abdominal cavity (stars).

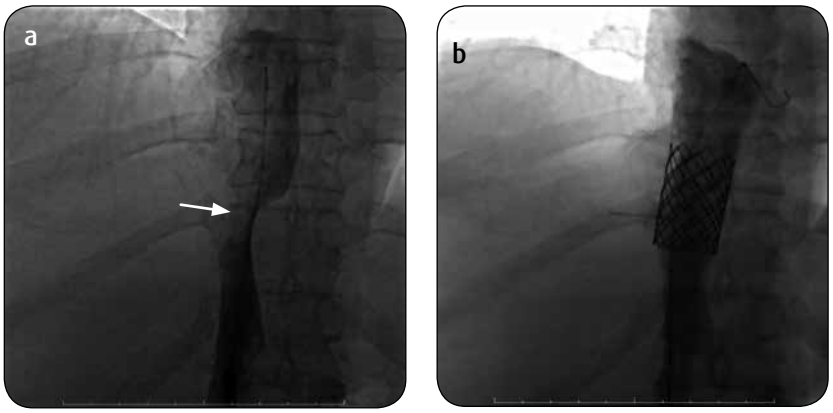

Figure 3. Fluoroscopy for stent placement in IVC. a) Partial obstruction of the intrahepatic IVC evidenced by the passage of the contrast medium (arrow). b) Proper contrast step after stent placement. There is no evidence of stenosis or residual occlusion.

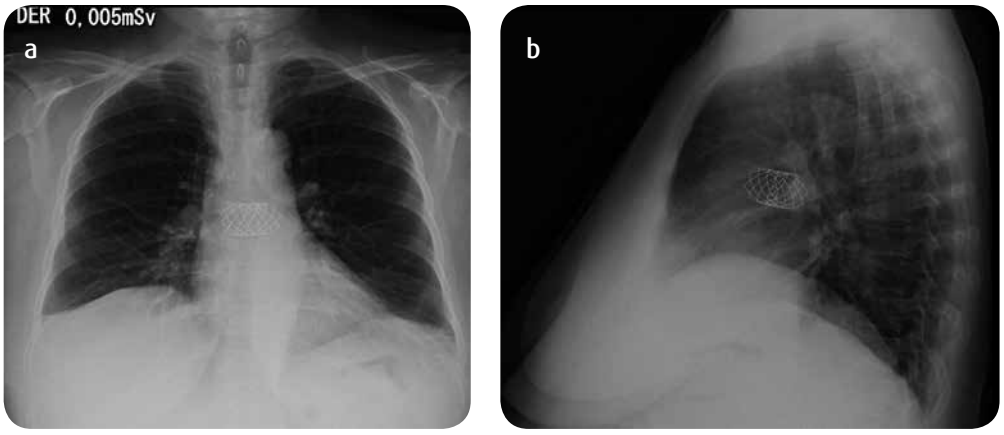

Figure 4. Chest radiography. a) Radiopaque element that projects in cardiac area, and corresponds to endoprosthesis. b) Endoprosthesis at the level of the main pulmonary vessels. It is not possible to estimate the exact location of the stent on the radiograph.

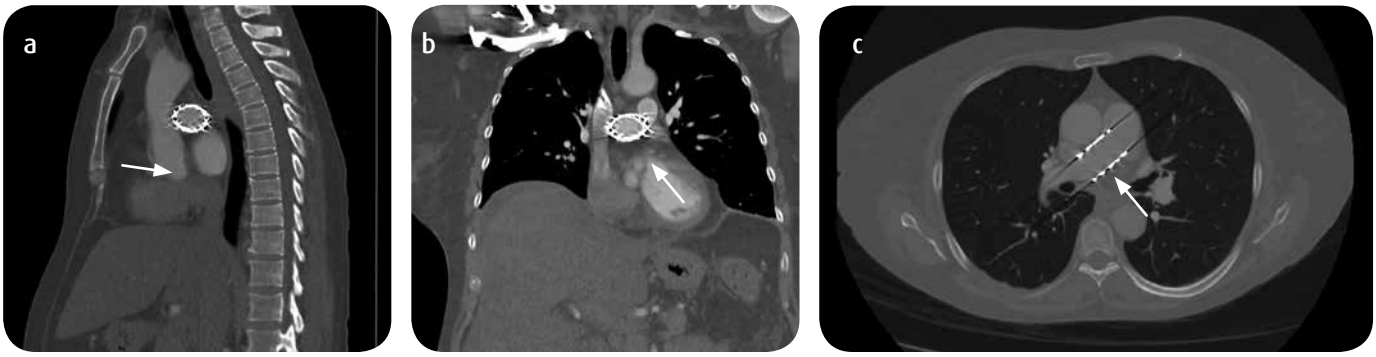

Figure 5. Chest CT angiography. Indicated initially due to suspicion of TEP. a-c) The embolized endoprosthesis is evidenced in the pulmonary trunk with projection towards the right AP (arrow). Proper alignment and apposition of the stent with respect to the pulmonary vessels.

gation. Interventional radiology performed selective angiography of SHV and IVC that showed critical stenosis greater than 90% of the lumen of the IVC in its intrahepatic segment, and focal stenosis in the drainage of the left and middle suprahepatic confluent, with satisfactory drainage of the right suprahepatic confluent. Given the symptomatic critical stenosis, angioplasty and stent implantation of caliber 12-24 × 45 in IVC was performed, with a subsequent satisfactory passage of the contrast medium, without reflux, to the hepatic venous system (figure 3).

The patient presented progressive clinical and paraclinical improvement; however, two days after the procedure he manifested chest pain and dyspnea; it was considered intermediate probability thromboembolism. The chest radiograph (figure 4) showed the stent projected on the cardiac silhouette. CT angiography confirmed the location of the endoprosthesis in the trunk of the pulmonary artery (PA), projected towards the right pulmonary artery, longitudinally with respect to the axis of the vessel and with its edges in adequate apposition with the walls of the artery. (figure 5). It was decided not to perform more interventions, either to remove the stent (high surgical risk) or to place a new one (high risk of embolism). Full anticoagulant therapy was continued. Clinical improvement and hepatic profile continued, so they were discharged. The thrombophilia study was performed on an outpatient basis and with negative results. There was complete resolution of the thoracic symptoms without signs of pulmonary embolism

Discussion

BCS is a rare and heterogeneous condition secondary to obstruction of the hepatic venous flow at any point between the hepatic venules and the right atrium, either primary (intrinsic) or secondary (extrinsic). The incidence varies from 0.33-0.88 cases / year and the prevalence is 1.4-7.6 cases / million inhabitants, with the largest series reported in China. There is a higher incidence among women and young adults (2-4). Clinical manifestations occur because of hepatocyte dysfunction, secondary to decreased perfusion. There is increased sinusoidal pressure and hepatic congestion. Depending on the duration of the commitment, there may be necrosis, progressive fibrosis, nodular regenerative hyperplasia, cirrhosis (CH) and liver failure (5). The most frequent clinical picture is abdominal pain, ascites and hepatomegalia (2,3), a triad that the patient reported in this case.

In the series described, in Asia and Africa the chronic form is more frequent due to the involvement of the IVC with or without extension to the ostium of the SHV; In the western world, acute presentation is more frequent due to the exclusive commitment of SHV (2,4). In this case, an acute BCS was described due to obstruction of the IVC with extension to the SHV, studied in the literature as the least frequent type (2). The acute form can generate fulminant hepatic failure (duration less than 4 weeks), without time to form collateral circulation or develop compensatory mechanisms. Also, acute cases with variable hepatic dysfunction and splenomegaly that depend on collateral circulation have been described. The patient's alcoholic cirrhosis (diagnosed de novo) is a risk factor that increases the likelihood of fulminant hepatic failure of acute BCS. On the other hand, the chronic form shows mild symptomatology and relative conservation of liver function, with changes in progressive CH (2,6).

Regarding the cause, about 75% of patients with BCS have one or more predisposing factors. The main ones are hematological disorders (myeloproliferative disorders), congenital or acquired coagulation disorders (protein C deficiency, protein S, antithrombin, pregnancy or cancer) and other uncommon causes, which include Behçet syndrome, aspergillosis, trauma, inflammatory bowel disease and tumor invasion, mainly of hepatocellular, renal and adrenal origin. When it is not possible to identify a factor, we speak of idiopathic BCS (3,5).

In this case, the only identified factor associated with the development of BCS was CH of alcoholic origin, which was considered to be the trigger of thrombosis. The pathophysiology is related to the coagulopathic effect secondary to cirrhosis, due to the involvement of various coagulation proteins, including those that participate as anticoagulants, such as antithrombin III, and proteins C and S (7,8).

The migration of the stent is an infrequent complication. It is described that it occurs in <2.5% of the procedures. Less frequent is the migration of the IVC stent to the PA, which occasionally has serious consequences, such as pulmonary infarction and tricuspid valve damage, and there are very few studies in the literature (9). In this case, for the implantation of the endoprosthesis, the IVC diameter and the degree of stenosis were taken into account, which was greater than 90%. The initial apposition was satisfactory; nevertheless, the continuation of the anticoagulant treatment produced dissolution of the thrombus that kept the device in position, caused recanulation of the

IVC and subsequent dislodgment and migration of the endoprosthesis. Before implanting a stent, it is advisable to complete an adequate anticoagulation scheme and perform balloon angioplasty; If the clinical and imaging results are not satisfactory, the implantation of the metal endoprosthesis may be considered. This behavior will reduce the risk associated with the implant and give the opportunity for resolution with medical management of the occlusive lesion.

Although prior management was considered with removal of the migrated endoprosthesis from its abnormal location, recent case reports have described the observe-wait behavior. The risks of the procedure vs. observation (10). In the case described, it was taken into account that the risk of intervening exceeded the benefit and clinical and imaging follow-up was performed, without evidence of valvular damage or pulmonary parenchyma. In addition, appropriate apposition of the endoprosthesis to the PA was found without evidence of an obstructive effect to the passage of the contrast medium into the pulmonary vessels in the chest CT angiography practiced.

Conclusions

Although CH of alcoholic origin is not described as a risk factor for the development of BCS, in the case described it was the only factor that was identified.

Although angioplasty with stenting is the treatment of choice for a significant number of cases, mainly when thrombosis with critical stenosis of the IVC is demonstrated, this procedure is not free of complications. The management of stent migration should be carefully evaluated based on clinical evolution, diagnostic images, prognosis and independent factors for each case.

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