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Budd-Chiari Syndrome – Case Report

Síndrome de Budd-Chiari: Estudo de caso

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ABSTRACT

Budd-Chiari Syndrome (BCS) is a rare pathological condition characterized by an obstruction of the hepatic venous outflow. It can be classified as primary, due to an intra-luminal venous lesion, or secondary, resulting from extrinsic compression or invasion of the venous system. This study aims to report a case of Budd-Chiari Syndrome, emphasizing the importance of diagnosis and treatment in preventing progression to chronic liver disease. We report the case of a 47-year-old patient who presented to the emergency department with severe pain in the right hypochondriac region and abdominal distension. A total abdominal CT scan was performed and revealed thrombosis of the suprahepatic veins, confirming the diagnosis of BCS. The patient developed portal hypertension and signs of chronic liver disease. Treatment was administered with Spironolactone and Marevan. After six months of Marevan use, the patient experienced worsening of the esophageal varices and hematemesis, requiring esophageal variceal band ligation. Liver transplantation was not recommended as the patient's MELD score was 14. Therefore, an accurate diagnosis of BCS is crucial for appropriate therapeutic intervention in order to prevent the progression to chronic liver disease, with permanent damage to liver functionality and the patient's quality of life.

Keywords: Budd-Chiari; Thrombophilia; Portal Hypertension.

RESUMO

A Síndrome de Budd-Chiari (SBC) é uma condição patológica rara caracterizada por uma obstrução do fluxo venoso hepático, classificada em primária, devido a lesão venosa intraluminal, ou secundária, resultante de compressão extrínseca ou invasão do sistema venoso. Este estudo tem como objetivo relatar um caso de síndrome de Budd-Chiari, enfatizando a importância do diagnóstico e tratamento na prevenção da progressão para doença hepática crônica. Relata-se o caso de um paciente de 47 anos que deu entrada no pronto-socorro com quadro de dor intensa em região hipocondríaca direita e distensão abdominal. Foi realizada tomografia computadorizada de abdome total que revelou trombose de veias supra-hepáticas, confirmando o diagnóstico de SBC. O paciente desenvolveu hipertensão portal e sinais de doença hepática crônica. O tratamento foi administrado com espironolactona e marevan. Após seis meses de uso de marevan, o paciente apresentou piora das varizes esofágicas e hematêmese, necessitando de ligadura elástica de varizes esofágicas. O transplante de fígado não foi recomendado porque o escore MELD do paciente foi 14. Portanto, um diagnóstico preciso de SBC é crucial para uma intervenção terapêutica adequada, a fim de prevenir a progressão para doença hepática crônica, com danos permanentes à funcionalidade hepática e à qualidade de vida do paciente.

Palavras-chave: Síndrome de Budd-Chiari; Trombofilia; Hipertensão porta.

INTRODUÇÃO

Budd-Chiari Syndrome (BCS) is a rare pathological condition characterized by an obstruction of the hepatic venous outflow, extending from the supra-hepatic veins to the atriocaval junction (VALLA, 2017). It can be classified as primary or secondary. Primary BCS results from an intra-luminal venous lesion, most often due to thrombosis, and less frequently due to phlebitis and stenosis. The secondary one is caused by any extrinsic compression or invasion of the venous system, such as in cases of tumors (CATALINA-RODRÍGUEZ & DÍAZ-FONTENA, 2012; GAVIRIA et al., 2016).

This syndrome is a rare pathological condition in Western countries, with higher prevalence in Asian countries. In the West, it mainly affects women aged 30 to 40, while in Asian countries, it more commonly affects men around 45 years old (TRIPATHI et al., 2016; GRUS et al., 2019).

The initial pathophysiology of BCS consists of obstruction of venous outflow between the hepatic venules and the supra-hepatic segment of the inferior vena cava. This obstruction rarely affects all the hepatic veins simultaneously. In 10% of cases, only the inferior vena cava is obstructed, and in two-thirds of cases, three veins are occluded (CLAPAUCH, BARBERI, & LEAL, 2009; GAVIRIA et al., 2016). Continuous reduction in hepatic perfusion results in ischemic injury to hepatocytes, accompanied by regenerative changes, which may lead to hepatic cirrhosis (GRUS et al., 2019).

Thus, clinical manifestations of the disease may primarily consist of abdominal pain, ascites, and hepatosplenomegaly, as well as collateral circulation of the inferior cava type, nausea, and fever. Its chronic form includes cirrhosis, variceal bleeding, hepatic encephalopathy, and hepatocellular carcinoma. However, in many cases, it is asymptomatic (CATALINA-RODRÍGUEZ & DÍAZ-FONTENA, 2012; VALLA, 2017; OBLITAS et al., 2020).

Diagnosis is based on clinical suspicion obtained through history-taking and physical examination, followed by laboratory and imaging tests. The initial technique of choice is Doppler ultrasound, which can provide a diagnosis in approximately 75% of cases. Additionally, computed tomography and magnetic resonance angiography may be used, although they sometimes suggest non-specific findings of the syndrome. Occasionally, biopsies may be necessary in some cases (CATALINA-RODRÍGUEZ & DÍAZ-FONTENA, 2012; OBLITAS et al., 2020).

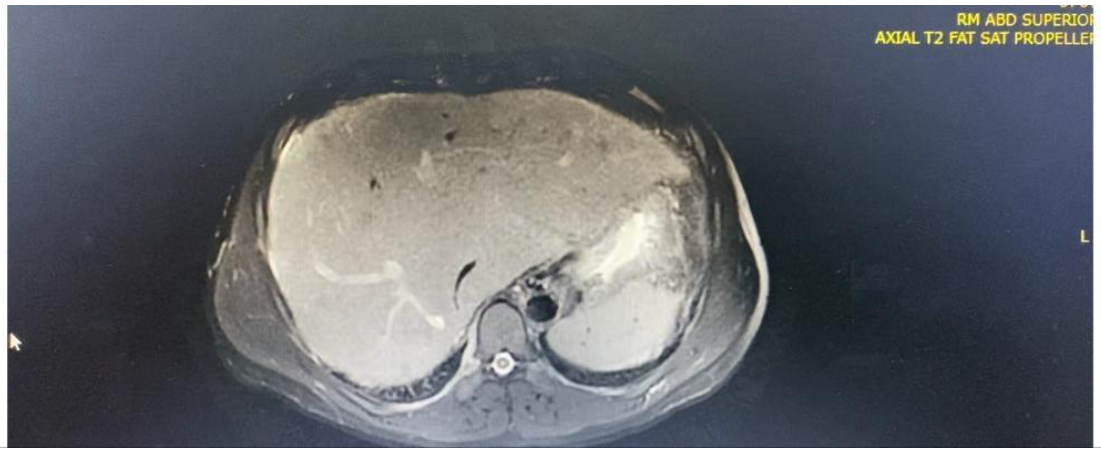
For the treatment of Budd-Chiari syndrome, anticoagulant therapy is initially used, even in asymptomatic patients. Other available options are: percutaneous angioplasty, transjugular intrahepatic portosystemic shunt, and liver transplantation. Asymptomatic cases have a good prognosis, while symptomatic cases have a heterogeneous course, with untreated patients estimated to die within three years (VALLA, 2017). According to Grus et al. (2017), five-year survival rates can range from 42% to 100%, depending on associated factors and the underlying cause. Therefore, this study aims to report a case of Budd-Chiari Syndrome, highlighting the importance of diagnosis and treatment in preventing progression to chronic liver disease.

CASE REPORT

A 47-year-old male patient was admitted to the hospital due to severe pain in the right hypochondrium region, which had started three days earlier, followed by increased abdominal volume. Along with pain and abdominal distension, the patient experienced dyspnea at rest. The patient was previously healthy, denied any prior morbidity, as well as chronic alcohol use and smoking. He had never undergone any surgical procedures and had a family history of cardiovascular diseases. On physical examination, the patient was in fair general condition, lucid, oriented, non-cyanotic, and mildly jaundiced (+/4+). Cardiac auscultation was normal, and lung auscultation revealed decreased vesicular breath sounds bilaterally at the bases, with dullness on percussion at the base of the right hemithorax. The abdominal exam showed tense ascites.

An imaging exam (magnetic resonance imaging), shown in figure 1, was performed and revealed thrombosis of the suprahepatic veins. A diagnostic and therapeutic paracentesis was performed to study the ascitic fluid, which showed a serum-ascites albumin gradient greater than 1.1, confirming the origin of the ascites (portal hypertension). With the aid of the imaging exam, the Budd-Chiari Syndrome diagnosis was confirmed, and treatment for ascites with Spironolactone was initiated. The patient maintained controlled ascites with this medication, and no further paracentesis was necessary. Due to the thrombosis of the suprahepatic veins, a hematology consultation was requested to evaluate the cause of the thrombosis. Tests for thrombophilia were required and confirmed the patient had Factor V Leiden deficiency. After collecting tests for thrombophilia screening, treatment with Marevan was started, and as the patient was stable, he was discharged for outpatient follow-up.

Figure 1 - Magnetic resonance imaging of the patient's upper abdomen. Signs of hepatic hypoperfusion and hepatomegaly characteristic of Budd-Chiari syndrome are observed.



Source: Personal archive.

Due to suprahepatic vein thrombosis, in addition to portal hypertension, the patient developed signs of chronic liver disease. During follow-up, an upper gastrointestinal endoscopy was requested to assess the presence of esophageal varices. In the initial examination, the patient had small-caliber varices. Treatment with Marevan was continued for six months, and after this period, a new imaging study was performed to evaluate the extent of thrombosis in the suprahepatic veins. There was an improvement in the extent of the thrombosis, but a worsening in the radiological appearance of the liver, which showed signs of chronic liver disease. A small amount of ascites was also noted.

After discontinuing Marevan, the patient experienced an episode of hematemesis, requiring esophageal variceal band ligation. Five sessions of band ligation were performed, and the patient is currently stable in outpatient follow-up, without episodes of bleeding or hepatic encephalopathy, and only using Spironolactone. Following evaluation by the transplant team, liver transplantation was not recommended at this time, as the patient had a MELD score of 14 (the MELD score is used to quantify the urgency for transplantation in patients over 12 years of age). The patient continues under follow-up, but has progressed to chronic liver disease, receiving continuous clinical treatment with Spironolactone to prevent the development of ascites. He shows no signs of hepatic encephalopathy and remains free of esophageal varices following eradication through elastic band ligation.

DISCUSSION

The described case presents clinical features similar to classic Budd-Chiari Syndrome, with an onset characterized by severe pain in the right hypochondrium region, the presence of large-volume ascites, hematemesis, and esophageal varices. However, the patient in question has a hereditary condition, a blood clotting disorder called thrombophilia, which consequently increases the risk of thrombosis. According to the literature, the clinical presentation of Budd-Chiari Syndrome is heterogeneous among patients, ranging from characteristic signs and symptoms to liver failure, which, if untreated, can lead to the patient's death (EUROPEAN ASSOCIATION FOR THE STUDY OF THE LIVER, 2016).

Upon the patient's admission to the hospital, an increase in the abdominal volume was identified, with physical examination confirming large-volume ascites. This sign, according to Dias et al. (2019), is one of the main manifestations of BCS. In Budd-Chiari Syndrome, with the blockage of blood flow out of the liver, blood accumulates in the organ, causing its enlargement. This blood accumulation (congestion) increases blood pressure in the portal vein, known as portal hypertension, which, combined with the enlarged and damaged liver, leads to ascites formation (ORFANIDIS, 2020). Thus, according to Junior Ramos et al. (2009), for a definitive diagnosis of ascites, it is essential to perform an abdominal paracentesis and analyze the ascitic fluid, as this is a way to determine portal hypertension as the pathophysiological mechanism.

Kuniyoshi et al. (2017) state that the development of esophageal varices (VEG) associated with Budd-Chiari Syndrome is a severe complication as well as a prognostic marker. The portal vein is crucial for blood circulation to abdominal organs, making a direct hepatic connection. Therefore, when its function is compromised, alternative pathways to decompress it are sought, aiming to transport blood to the systemic circulation. Thus, the emergence of esophageal varices is observed, consisting of dilated and tortuous veins in the esophagus as an attempt to alleviate portal overload (ORFANIDIS, 2020).

Normal portal venous pressure is below 5 mmHg. With the blockage of hepatic venous outflow, this pressure increases. Values above 10 mmHg pose a risk for varices development, and values above 12 mmHg pose a risk for variceal rupture (COELHO et al., 2014). In this case, the patient had fine-caliber esophageal varices, complicating his prognosis. Therefore, besides laboratory studies to assess liver function, periodic endoscopies should be performed to evaluate the presence of varices.

A complication arising from the formation of esophageal varices is upper gastrointestinal hemorrhage, which is one of the most frequent symptoms in cases of SBC. Its incidence is about 5% of patients, manifesting primarily as hematemesis. Therefore, hematemesis, which was also observed in the patient in question, results from the bleeding of these varices, formed by portal venous obstruction (ALVES et al., 2012). In this report, hematemesis was observed after the withdrawal of the medication used, Marevan. To address this bleeding, elastic band ligation of the esophageal varices was performed.

According to the literature, the main etiological factor of Budd-Chiari Syndrome is hematological abnormalities, particularly myeloproliferative disorders (MENON, SHAH, & KAMATH, 2004; OBLITAS et al., 2020). Other causes include thrombophilic states, such as inherited deficiencies of protein C, protein S, Factor V Leiden, and antithrombin III (MENON, SHAH, & KAMATH, 2004; VALLA, 2017). In this case, the patient has Factor V Leiden deficiency, which interferes with the action of the activated form of protein C, which, in the presence of protein S, acts in the proteolytic inactivation of factor Va and factor VIIIa (GODOY, 2005; RAMOS et al., 2006). Since protein C is an important pathway for anticoagulation, resistance to this protein results in hypercoagulability and, consequently, an increased risk of venous thrombosis (LEHMKUHL et al., 2012). This increased risk, in the cited case, led to the development of a suprahepatic vein thrombosis and subsequent portal hypertension, which characterizes Budd-Chiari Syndrome.

Among the treatments indicated for Budd-Chiari Syndrome is the use of anticoagulants. According to Valla (2017), this therapy should be initiated early, whether the prothrombotic disorder is identified or not. Later, after the cause is identified, its use should be assessed based on necessity. Anticoagulants are used to prevent further extension of venous thrombosis (MENON; SHAH; KAMATH, 2004).

Another treatment method is the implantation of a transjugular intrahepatic portosystemic shunt (TIPS), used to reduce portal system pressure by creating an intrahepatic shunt between a portal vein branch (usually the right) and the inferior vena cava (EUROPEAN ASSOCIATION FOR THE STUDY OF THE LIVER, 2016). According to Garrido et al. (2018) and Oblitas et al. (2020), TIPS is indicated when anticoagulant therapy is unsuccessful, as thrombolysis is limited. Additionally, it is recommended in cases of failed hepatic vein angioplasty and conditions such as acute liver failure and complete portal vein obstruction. The literature also indicates that its

results are effective in stopping hematemesis, controlling esophageal varices and ascites, which is also associated with the administration of small doses of diuretics (EUROPEAN ASSOCIATION FOR THE STUDY OF THE LIVER, 2016; OBLITAS et al., 2020).

Liver transplantation is the last treatment option for Budd-Chiari Syndrome. It is indicated in cases of fulminant liver failure, hepatocellular carcinoma development, or when percutaneous revascularization and TIPS have failed, progressing to hepatic deterioration (EUROPEAN ASSOCIATION FOR THE STUDY OF THE LIVER, 2016; KHAN et al., 2019). However, post-transplant, the possibility of recurrent BCS must still be considered. Thus, anticoagulant therapy should be maintained in most patients even after surgery (EUROPEAN ASSOCIATION FOR THE STUDY OF THE LIVER, 2016).

FINAL CONSIDERATIONS

Budd-Chiari Syndrome is a rare condition affecting hepatic blood flow. Portal hypertension resulting from this process can cause clinical manifestations such as ascites, esophageal varices, and hematemesis. Hematological abnormalities causing thrombophilic states are also factors that contribute to this condition. A precise diagnosis, based on the collection of anamnesis data, physical examination, and complementary tests, is essential. This way, appropriate therapeutic interventions can be promptly chosen, preventing the progression to chronic liver disease, which can cause permanent damage to the liver's structure and functionality, consequently affecting the patient's quality of life.

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