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CASE REPORT

Case report of splenic infarction in a child with biliary atresia

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Abstract

This case report is intended to describe a rare case of splenic infarction (SI) associated with biliary atresia, which contributed to the worsening of the prognosis. In this case study, a boy diagnosed with biliary atresia underwent the Kasai procedure at the age of 3 months. The patient presented an intense abdominal pain, ascites and fever at the age of 6 years. A computed tomography (CT) scan showed enlargement of the spleen and an image suggestive of SI. The symptoms improved with the administration of antibiotics, analgesics, while ascites decreased with the use of diuretics. Liver transplantation was indicated. SI is an extremely rare event characterized by vessel occlusion, parenchymal ischemia and tissue necrosis. The symptoms are non-specific and may be confounding factors with other causes of acute abdominal pain. Splenomegaly-inducing diseases may be a risk factor. There are few case reports of SI and cirrhosis of different etiologies in childhood. Abdominal CT scan with intravenous contrast is the imaging modality of choice and the treatment ranges from supportive care to splenectomy. Despite being a rare condition, its knowledge is extremely important, as it allows early clinical suspicion, with immediate institution of treatment, which may interfere with the prognosis.

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INTRODUCTION

Splenic infarction (SI) is an extremely rare event characterized by vessel occlusion and parenchymal ischemia, which can progress to tissue necrosis[1]. The two most common causes are hematological infiltrative diseases and thromboembolic diseases and, in children, sickle cell hemoglobinopathies.

Diseases that cause splenomegaly are considered risk factors for SI, such as chronic myeloid leukemia, myelofibrosis, lymphoma, Gaucher disease, malaria, Acquired Immunodeficiency Syndrome with mycobacterium avium[2] infection. Only one case of SI associated with biliary atresia (BA) has been described. The diagnosis of such a complication is often suggested based on the presence of underlying diseases more likely to cause it.

This report describes a case of splenic infarction in a pediatric patient with biliary atresia, reinforcing the relevance of suspecting this complication in cases of this disease that present with abdominal pain.

CASE REPORT

A Brazilian boy, with an uneventful pregnancy and birth, around 1 month of age, developed jaundice, fecal acholia, hepatomegaly and anemia. At 3 months of age, he was diagnosed with BA and underwent the Kasai procedure. Since then, total bilirubin (TB) has ranged from 2.34 to 5.47mg/dL using ursodeoxycholic acid.

Splenomegaly was seen at 13 months of age. Esophageal varices were detected by endoscopy after an episode of hematemesis at 2 years and 3 months, and endoscopic ligation (EL) treatment was performed. At the age of 6, the spleen measured more than 20 centimeters in its longest dimension upon ultrasound scan (upper limit for age of 9.5 cm)[3], and upon physical examination, it was palpable 15 cm below the left costal margin. At this point, according to data from the CDC/WHO (Centers for Disease Control and Prevention/World Health Organization) his bodily percentiles were: weight/age p34, height/age p3, body mass index/age p87. Laboratory tests showed: BT3.8; Albumin 3.5; INR 1.12.

At 6 years and 11 months of age, the patient presented intense pain in the upper left quadrant of the abdomen and developed a fever, reaching 39ºC. Within 2 days, the patient was admitted to the hospital presenting mild jaundice, distended abdomen with collateral circulation "in the shape of a medusa's head". The spleen was palpable 14cm below the left costal margin, and it was painful. Laboratory tests showed BT 3.55, Albumin 3.0 and INR 1.2 –Pediatric End-stageLiver Disease (PELD)[2]. An ultrasound scan (US) detected splenomegaly, measuring 20 centimeters at its longest dimension, with a hypoechogenic image in the anterior region of the middle third, measuring 1.9x 5.3 cm and in the upper region, measuring 7 cm in the longest axis, in addition to mild ascites. Computed tomography scan (CT) of the abdomen revealed an enlarged spleen, measuring up to 23 .2cm in its greatest extent, with regular contours, presenting an area of segmental hypocontrast in its superolateral portion, suggestive of infarction (Figure 1), in addition to punctate calcifications spread over the splenic parenchyma, with a sequelae appearance. Symptoms improved with the administration of antibiotics (ampicillin-sulbactam) and analgesics, and the ascites reduced with the use of diuretics (furosemide 1.5 mg/kg/day and spironolactone 3.5 mg/kg/day). He had pain relief, without new fever spikes, being discharged from the hospital on the 23rd day. Tests performed during the evolution are shown in Table 1. Follow-up US were performed on the 6th, 11th, 18th and 27th days after the onset of the disease, and showed no signs of complications. Liver transplantation was indicated.

This case report was approved by the Research Ethics Committee (CEP-Unicamp) with free and informed consent from the patient's legal representatives.

COMMENTS

Similar to the only case report of BA and SI described in a Japanese boy by Matsui et al[4], our patient was 7 years old when diagnosed. The Japanese boy had splenomegaly detected at 22 months of age, and esophageal varices detected at 4 years of age. In the present case, splenomegaly was detected very early, at 13 months, and esophageal varices were detected at 2 years and 3 months of age, after a gastrointestinal hemorrhage, showing a faster progression of the disease and its complications[4].



Figure 1. Computed tomography of the abdomen in the coronal plane demonstrating splenomegaly and segmental areas of hypocontrast in the splenic parenchyma, compatible with infarction (arrow).

Table 1. Laboratory tests according to the onset of the day (D) of symptoms.

	Reference value	D3	D5	D13
Hemoglobin	12.0-15.8g/dL	9.7	-	9.9
Hematocrit	35.4-40%	28.5	-	29.9
Leukocytes	3.540- 9.060/mm3	3,550	-	3,360
Platelets	150.000-415.000/mm3	45,000	-	54,000
Aspartate aminotranspherasis	5-40U/L	59	87	-
Alanine aminotranspherasis	7-31 U/L	40	47	-
Alkaline phosphatase	< 259U/L	503	-	-
Gama-glutamiltranspherase	5-43U/L	106	-	-
Albumin	3.2-4.5g/dL	3.0	-	-
International Normalized Ratio (INR)	< 1.25	1.2	-	-
Actvated partial thromboplastin time (APTT)	< 1.3	0.86	-	-
Total bilirrubin	0.2-1.0mg/dL	3.55	3.7	-
Direct bilirrubin	0-0.2mg/dL	2.15	2.23	-
Creatinine	0.5-1.2mg/dL	0.19	-	-
Urea	10.8-38.4 mg/dL	20	-	-
C-Reactive Protein	< 3.0 mg/L	-	58.2	19

Anoxia underlying splenic infarction can be produced by several factors: increase in the absolute mass of the spleen resulting in a greater demand for oxygen; compression of extrasplenic blood vessels by enlarged lymph nodes; decreased oxygen-carrying capacity of the blood caused by anemia; or blockage of the intrasplenic circulation by abnormal cells occluding the venous sinusoids or compressing the arterioles and venules[5]. The marked splenomegaly associated with portal hypertension may have corroborated the anoxia in our patient.

Symptoms of SI are not specific, requiring a broad differential approach with other causes of acute abdominal pain. Laboratory tests, such as liver function, bilirubin or lipase tests, can help investigate other causes of abdominal pain. Leukocytosis and elevated lactate dehydrogenase (LDH) can be found in SI, although they are not specific[2].

Abdominal CT performed with intravenous contrast is the imaging modality of choice in suspected spleen infarction, although US is generally used for initial assessment and subsequent follow-up[6]. In the hyperacute phase, US is less sensitive because there are no differences between the echogenicity of infarcted and normal tissue[7].

The formation of pseudocysts, abscesses, hemorrhages, splenic rupture and aneurysms may occur during the course of the condition[2]. The affected region may become infected and lead to the formation of abscesses, and may also undergo hemorrhagic transformation. These complications are an indication for urgent surgical management. In our case, ultrasound was used during follow-up, being performed 6, 11, 18 and 27 days after the onset of the disease, without signs of complications.

Treatment depends on the underlying condition, ranging from supportive care, such as hydration, analgesics and antiemetics, to splenectomy.

According to the case report by Matsuis[3], splenic infarction in patients with portal hypertension secondary to BA can lead to a significant acceleration of chronic liver failure, justifying the indication of liver transplantation in our case. Despite being a rare condition, its knowledge is extremely important, as it enables early clinical suspicion, with immediate treatment, which may interfere with the prognosis.

REFERENCES

1. Mortazavi AR, Joudi M, Zabolinejad N, Aelami MH, Farhangi H, Alamdaran SA. Massive Splenic Infarction with Specific Sonographic Feature: Two Case Reports with Rare Etiologies. Journal of Clinical Gastroenterology and Treatment. 2018;4(2):058.
2. Chapman J, Helm TA, Kahwaji CI. Splenic Infarcts. StatPearls [Internet] 2020. Disponível em: https://www.ncbi.nlm.nih.gov/books/NBK430902. [acessed in 2021 jul 4]
3. Rosenberg HK, Markowitz RI, Kolberg H, Park C, Hubbard A, Bellah RD. Normal Splenic Size in Infants and Children: Sonographic Measurements. AJR Am J Roentgenol. 1991;157: 119-121.
4. Matsui A, Shimada T, Sasaki N, Ishikawa T, Momoya T, Makino S. Splenic Infarction in a Child With Portal Hypertension Secondary to Biliary Atresia. J Pediatr Surg. 1997;32:648–649.
5. Hynes HE, Silverstein MN, Fawcett KJ. Spontaneous rupture of the spleen in acute leukemia. Report of 2 cases. Cancer. 1964;17:1356–60.
6. Ray S, Mridha AR, Ahammed M. Diffuse splenic infarction in a case of severe acute pancreatitis. Am J Surg. 2011;201:23–25.
7. Kianmanesh R, Aguirre HI, Enjaume F, Valverde A, Brugière O, Vacher B, et al. Spontaneous splenic rupture: report of three new cases and review of the literature. Ann Chir. 2003;128:303–309