Acute pancreatitis and abdominal vaso-occlusive crisis: a difficult differential diagnosis in sickle cell anemia

Pancreatite aguda e crise vaso-oclusiva abdominal: um diagnóstico diferencial difícil na anemia falciforme

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ABSTRACT

Vascular occlusion in sickle cell disease is a complex process and is responsible for most of the clinical manifestations of the disease. Abdominal pain is an important component of the painful vaso-occlusive crisis. Abdominal pain due to sickle cell vaso-occlusive crisis is often indistinguishable from an acute intra-abdominal disease process such as acute cholecystitis, acute pancreatitis, hepatic infarction, ischemic colitis and acute appendicitis. We describe a case of abdominal pain in sickle cell disease and discuss differential diagnosis between vaso-occlusive crisis and acute pancreatitis.

Keywords: Vascular occlusion; sickle cell anemia; abdominal pain; pancreatitis.

RESUMO

A oclusão vascular na doença falciforme é um processo complexo e responsável pela maioria das manifestações clínicas da doença. A dor abdominal é um componente importante da crise dolorosa vaso-oclusiva. A dor abdominal devido a crise vaso-oclusiva falciforme é muitas vezes indistinguível de um processo de doença intra-abdominal aguda, como colecistite aguda, pancreatite aguda, infarto hepático, colite isquêmica e apendicite aguda. Descrevemos um caso de dor abdominal na doença falciforme e discutimos o diagnóstico diferencial entre crise vaso-oclusiva e pancreatite aguda.

Descritores: Oclusão vascular; anemia falciforme; dor abdominal; pancreatite.

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INTRODUCTION

Sickle cell disease (SCD) is the most common monogenic disorder, characterized by chronic hemolytic anemia and painful vaso-occlusive crisis (VOC) [1-4]. Vascular occlusion in SCD is a complex process and is responsible for most of the clinical manifestations of the disease. Stress, infection or dehydration cause deoxygenation and consequently falcization of red blood cells that make vaso-occlusion, tecidual ischemia and infarct. Abdominal pain is an important component of the painful VOC. The etiology of abdominal pain in SCD is often difficult to diagnose clinically. Despite the frequent occurrence, diagnostic dilemma, and the need for an accurate, early diagnosis, abdominal pain in sickle cell disease has not been rigorously studied. Pancreatitis is an inflammation of pancreas due to gallstones, alcohol abuse, connective tissue disease, vasculitis or shock. The most common cause is gallstones (40-50%) and the prevalence in general population is 40:100.000 patients. In SCD patients, gallstone has an incidence of 13-45/100.000 patients, with a 10% of hospitalization for abdominal pain in this group. Acute pancreatitis (AP) is rarely included as a cause of abdominal pain in patients with VOC. When it occurs, it may result from biliary obstruction, but in other cases, it may be a consequence of microvascular occlusion that causes ischemia. We describe a case of abdominal pain in SCD and discuss differential diagnosis between VOC and AP [1-3, 5].

CASE REPORT

A 58-year-old male patient with SCD (HbSS), with abdominal pain in a band like pattern, afebrile, associated with inappetence, nausea and vomiting, progressive worsening and improvement with antalgic position. Patient presented similar pain for 8 days, but with less intensity and improvement with hydration and absence of alcoholic or drug intake. Blood tests showed Hemoglobin 5.6 g/L, hematocrit 16,1%, white blood count $8.0x10^3$/L without left shift, platelets $200.0x10^3$/L, lactic dehydrogenase 2205 U/L, alanine aminotransferase 25 U/L, aspartate aminotransferase 70 U/L, alkaline phosphatase 123 U/L, gamma globulin transpeptidase 207 U/L, amylase 1119 U/L and lipase 2313 U/L, urea 223 mg/dL and creatinine 5.3 mg/dL, C-reactive protein 0.34 mg/dL, total bilirubin 3.15 mg/dL and indirect bilirubin 2.04 mg/dL. Doppler ultrasonography of the abdomen showed biliary sludge without acute inflammatory signs and absence of arterial or venous thrombi. Computed angiography of the abdomen and pelvis excluded mesenteric ischemia and showed diffuse densification of the peripancreatic adipose planes without areas of necrosis or intra-pancreatic collection (Figure 1). The patient was treated with conservative treatment, receiving hydration and dialytic support according to demand with excellent resolution of the acute condition.

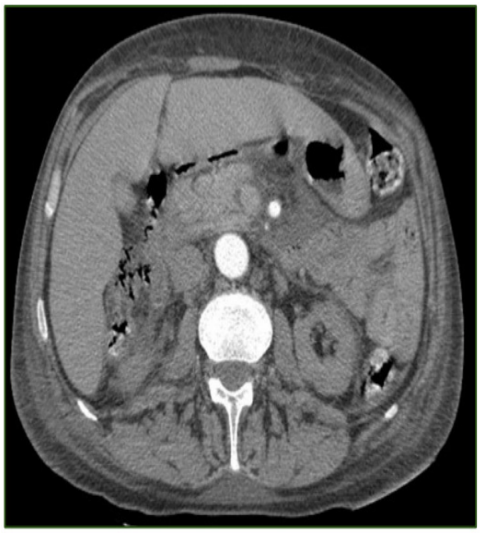

Figure 1 - Computed angiography of the abdomen showing diffuse densification of the peripancreatic adipose planes, without areas of necrosis or intra-pancreatic collection.

DISCUSSION

Abdominal pain is an important component during VOC and AP in SCD, being attributed to sickling, hypercoagulability, capillary engorgement and stasis. The most frequent

causes are acute cholecystitis, opioid constipation, renal papillary necrosis, hepatic sequestration, splenic sequestration, urinary tract infection, peptic ulcer and intestinal ischemia. Acute ischemic pancreatitis is a very rare complication of VOC. The clinical features are indistinguishable from other causes of acute abdomen and represent a diagnostic challenge 3,5 [3, 6].

Bile sludge, as in the case described, can cause cholestasis or sphincter of Oddi injuria, causing edema and biliopancreatic reflux. This process triggers intracellular activation of pancreatic digestive enzymes, although the mechanism is not well understood. Increased intraductal pressure plays a role in the pathophysiology of the disease, since the extent of pancreatic injury is directly related to the duration of ampullary obstruction. In gallstone pancreatitis, the pancreatic homeostasis is altered and the organ damage is mediated by inflammatory cytokine es that worsen parenchymal damage and potentially initiate a systemic inflammatory response [5].

On the other hand, the role of hypoxemic injury and microvessel occlusion in the pathophysiology and clinical manifestations of the intra-abdominal disease process, as in the case of acute pancreatitis, is not well known [6].

The diagnosis is established based on clinical suspicion, biochemical evidence and radiological findings. In addition to abdominal pain, our patient had elevated serum lipase and CT findings of acute pancreatitis [3, 6]. In this case, there was no evidence of drug, alcohol, trauma, toxin or obstructive etiology and especially the absence of bile duct dilation, suggesting therefore that pancreatitis was probably caused by sickle cell ischemic etiology.

Treatment of pancreatitis as well as VOC is based on hydration, pain control and electrolytic balance [3].

Treatment of cholelithiasis is still controversial. Therefore, prophylactic cholecystectomy should be considered, after crises, for patients with cholelithiasis who are at high risk of developing complications [2].

Blood transfusion is indicated when hemoglobin in low. When the hemoglobin is high or in critical cases is necessary change transfusion to low HbS under 30% or to avoid hemoglobin above 100 g/L because it can elevate the viscosity and worsen the VOC [1-2].

CONCLUSION

Abdominal pain due to sickle cell vaso-occlusive crisis is often indistinguishable from an acute intra-abdominal disease process such as acute cholecystitis, acute pancreatitis, hepatic infarction, ischemic colitis and acute appendicitis. Acute pancreatitis of ischemic etiology should be considered as an important differential diagnosis in any patient with SCD presenting abdominal pain, since the early therapeutic approach is crucial for the good evolution of these conditions.

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