CASE REPORTS

PEDIATRIC IDIOPATHIC MIDGUT VOLVULUS AND SHOCK IN THE INFANT

VÓLVULO INTESTINAL IDIOPÁTICO E CHOQUE NO LACTENTE

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ABSTRACT

Introduction: Intestinal volvulus is a surgical emergency in which a segment of the intestine twists over its mesenteric attachment, causing bowel obstruction. It usually presents with bilious vomiting and can progress to bowel necrosis and shock.

Case Report: A 40-days-old male infant presented with acute onset irritability, bilious vomiting, abdominal distention, and hematochezia. He rapidly evolved to shock with metabolic acidosis and coagulopathy, requiring fluid resuscitation, vasoactive agents, and invasive mechanical ventilation. The patient was submitted to urgent laparotomy, confirming midgut volvulus without malrotation. Partial reperfusion of the affected midgut was achieved, with no resection initially performed, but 48 hours later he was re-evaluated and partial enterectomy for midgut necrosis was performed. Despite the condition's severity, the patient had a good evolution with full recovery.

Discussion: Bilious vomiting in the infant is highly suggestive of intestinal obstruction. The authors emphasize the presence of midgut volvulus without malrotation, complicated with bowel necrosis and shock.

Keywords: idiopathic; infant; intestinal volvulus; shock

RESUMO

Introdução: O vólvulo intestinal é uma emergência cirúrgica que consiste na torção de uma ansa intestinal sobre o seu ponto de inserção, causando obstrução intestinal. Habitualmente apresenta-se com um quadro de vómitos biliares, podendo evoluir para necrose intestinal e choque.

Caso Clínico: Um lactente de 40 dias de idade foi observado por inicio súbito de irritabilidade, vómitos biliares, distensão abdominal e hematoquézia. Evoluiu para choque com acidose metabólica e coagulopatia, necessidade de volemização, aminas vasopressoras e ventilação mecânica. Foi submetido a laparotomia, com identificação de vólvulo intestinal sem malrotação e, recuperação da perfusão após desrotação, sem necessidade de resseção intestinal inicial. Após 48 horas, foi submetido a laparotomia de revisão, que demonstrou necrose intestinal com necessidade de enterectomia parcial do intestino delgado. Apesar da gravidade do quadro, o doente apresentou boa evolução clínica.

Discussão: A presença de vómitos biliares no lactente é sugestiva de obstrução intestinal. Os autores salientam a presença de vólvulo intestinal sem malrotação, complicado com choque e necrose intestinal.

Palavras-chave: choque; idiopático; lactente; vólvulo intestinal idiopático

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INTRODUCTION

Midgut volvulus can be defined as a complete bowel obstruction due to twisting of an intestinal segment over its mesentery.¹⁻⁴ Most cases occur in the neonatal period and first year of life, as a result of an abnormal embryologic rotation of the midgut, known as malrotation.^{1,4,5} Idiopathic midgut volvulus is very rare.^{2,3} It should be suspected in newborns and young infants with bilious vomiting and progressive clinical deterioration, characterized by abdominal distension, hematochezia, and hemodynamic instability from hypovolemia and/or septic shock.^{4,6,7} Urgent laparotomy is required after rapid resuscitation, to allow blood flow restoration and prevent intestinal necrosis and subsequent bowel resection.^{8,9}

CASE REPORT

A 40-days-old previously healthy male was admitted after five hours of persistent vomiting, abdominal distension, and irritability associated with hematochezia in the previous two hours. Parents denied sick contacts, history of fever, or any other symptoms on previous days. No history of surgery or digestive tract malrotation was identified in family medical history.

On physical examination at hospital admission, the patient had poor general condition, persistent groaning, labial cyanosis, and delayed capillary refill time. He was tachycardic, tachypneic, and hypotensive and had marked abdominal distention with decreased bowel sounds, but no rash or fever. Intraosseous access (IO) was placed after failure to establish peripheral venous access. Venous blood gas analysis showed mixed acidosis (pH 6.8, lactate > -15 mmol/L, pCO2 83 mmHg, HCO3- 7 mmol/L). Fluid resuscitation and sodium bicarbonate were administered and the patient was started on broad-spectrum antibiotic therapy after blood cultures collection. Despite initial therapy, he maintained hypotension and rapidly progressed to respiratory failure, requiring mechanical ventilation and vasopressor therapy. Blood tests showed anemia, thrombocytopenia, leukocytosis with neutrophilia, negative C-reactive protein, mild abnormal liver function, normal electrolytes and renal function, and abnormal coagulation study. The patient received red blood cell and fresh frozen plasma transfusions. Abdominal ultrasound excluded intussusception and standard abdominal X-ray revealed distended loops with thickened intestinal wall.

Due to suspicion of intestinal occlusion, the patient was submitted to urgent laparotomy, which confirmed small bowel volvulus with 50 cm of intestine with doubtful viability. Partial restoration of blood flow was achieved with derotation, and no resection was initially performed. No nonrotation or malrotation abnormalities were reported. After surgery, the patient was admitted to the Pediatric Intensive Care Unit, and maintained on inotropic support and invasive mechanical ventilation. He was submitted to a second-look laparotomy 48 hours later, showing multiple necrotic lesions (one of which perforated), spread over approximately 70 cm of small bowel, requiring segmental enterectomy and ileo-ileal anastomosis. The patient was extubated, stopped vasopressor support five days after surgery, and completed 12 days of antibiotic therapy, after two negative blood cultures. Electrocardiogram, echocardiogram, and transfontanellar ultrasounds were normal. He was discharged for outpatient care on the 12th day with adequate development and growth on seven-month follow-up.

DISCUSSION

Midgut volvulus commonly occurs secondary to intestinal malrotation.¹⁻¹² The resulting narrow mesenteric base allows the mesentery to twist around the superior mesenteric artery, causing small bowel ischemia.^{3,4,6,10} The condition may also be linked to other causes, namely duplication cysts, Meckel's diverticulum, tumors, and worm mass.^{2,6,7} Idiopathic small bowel volvulus, as seen in this case, is very rare.¹⁻³ The consequence of this condition is small bowel ischemic necrosis and intestine loss, resulting in an acute event with high mortality.¹¹ Most cases presents in the first year of life, 50% of which in the neonatal period, with bilious or nonbilious emesis.^{1,4,6,7} Over time, as small bowel blood supply is compromised, abdominal distension, hematochezia, and shock ensue.^{4,6,7} This rapidly progressive cardiovascular compromise is due to third-space fluid loss and sepsis, caused by the necrotic bowel.^{6,12} Clinical suspicion with rapid fluid resuscitation and emergent surgical intervention, as in the present case, are crucial for patient prognosis.6,9,10,12

The recommended approach for all patients with intestinal malrotation and volvulus or suspected volvulus and signs of systemic decompensation (e.g. hematemesis, hematochezia, abdominal distension, peritonitis, and shock) is to directly proceed to laparotomy.⁶

In the present case, the patient presented with shock and was submitted to abdominal ultrasound and radiograph, while waiting for pediatric emergency transport to a central hospital with Pediatric Surgery. In hemodynamically stable patients, diagnosis should be confirmed by radiologic evaluation, which normally begins with abdominal plain films to exclude intestinal perforation, followed by definitive diagnosis with upper gastrointestinal contrast series to identify rotation anomalies and the obstruction site.^{1,4,6,7,9,10} Ultrasonography can be a good screening tool, but is not as reliable as upper gastrointestinal contrast series.^{4,7}

Management of midgut volvulus consists in proceeding to exploratory surgery as soon as possible.1,6 Initial therapy includes cardiopulmonary and circulatory resuscitation and administration of broad-spectrum antibiotics, as performed in this case.^{1,4,6,7} During surgical exploration and after derotation, intestine of doubtful viability is often left in situ, as a measure to limit intestinal resection.^{6,11} A second look laparotomy is performed after 24 to 48 hours to reevaluate bowel viability.^{6,11} If necrotic bowel is present,

intestinal resection is required, as in this patient.

Possible postoperative complications include short bowel syndrome, adhesions, intra-abdominal abscess, and failure to thrive, none of which occurred in this case.^{1,6}

This case report emphasizes the importance of recognizing intestinal volvulus clinical presentation, enabling to start treatment in a timely manner, and minimize consequences. Additionally, this case reports a rarely described case of idiopathic midgut volvulus.

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