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Combined surgical and medical treatment of a retroperitoneal lymphatic-venous malformation presenting with significant haemorrhage

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ABSTRACT

Lymphatic-venous malformations are rare congenital defects resulting from the abnormal communication between lymphatic vessels and systemic circulation, integrating the group of combined vascular malformations, with low-flow. They affect both genders equally and present preferably in the head and neck (> 90%), rarely in retroperitoneal location. Clinical diagnosis can be challenging since presentation is highly variable. Surgical resection is the gold standard but, in complex malformations or when facing incomplete surgical resection due to inaccessible location, combined medical and surgical approaches can improve the prognosis. The authors report a case concerning a three-month-old infant with a retroperitoneal lymphatic-venous malformation, with an uncommon life-threatening presentation and a successful combined medical and surgical approach. In the light of this case report, a specific literature review is discussed.

1. Introduction

Lymphatic-venous malformations or hemolymphangiomas (old classification), integrate a group of vessel morphogenesis anomalies called vascular malformations, according to the 2014 ISSVA Classification of Vascular Anomalies [1,2]. Vascular malformations are present at birth, generally subtle, and become progressively more obvious (expand proportionally to overall growth), becoming infiltrative and, occasionally, destructive [1,3]. Moreover, factors such as local trauma, infection, changes in blood or lymphatic flow and hormonal variations lead to increasing ectasia [1,3].

Vascular malformations can be divided into simple, when affecting only one vessel type; combined, defined as two or more vascular malformations found in one lesion; of major named vessels and associated with other anomalies [2]. They can also be classified according to flow characteristics, into low flow (capillary, venous, lymphatic, or combination) and high flow (arterial, arteriovenous) [1]. Lymphatic-venous malformations are combined low flow malformations involving lymphatics and veins [2]. Venous malformations are the most common type of congenital vascular malformation with an incidence higher than 50%. Lymphatic malformations are less common, and the combination of venous and lymphatic malformations has even a lower incidence [4], varying from 1,2 to 2,8 per 1000 newborns [5], with both genders equally affected [6]. As congenital malformations, they can be diagnosed at any age, however, 90% are identified in children under two-years of age (60% of those in newborns) [7,8].

We describe a case report concerning an infant with a rare retroperitoneal lymphatic-venous malformation, with an uncommon life-threatening presentation and a successful combined medical and surgical approach.

2. Case report

A three-month-old male infant, with an uneventful pregnancy, was born at 40 weeks by vaginal delivery, with an Apgar Index of 9/10/10 and a weight of 3530 g. Besides a left hypochondrium haemangioma, no other abnormalities were evident at first examination. Perinatal period was without incidents except for frequent colic pain episodes. He was admitted at emergency department for alternating periods of irritability and prostration, groaning, anorexia, an episode of vomiting and mucus in his stools, for 72 h. On physical examination, he was prostrated, had abdominal distention with diminished bowel sounds on the left and high-pitched bowel sounds on the right quadrants. Abdominal ultrasound (US) showed significant quantity of intraperitoneal free fluid with echogenic content, and an anechoic image with 1,5 cm in diameter along the intra-hepatic inferior vena cava; bowel intussusception was excluded. In the context of acute abdomen, he was submitted to urgent laparotomy, that revealed moderate intraperitoneal fluid (negative on

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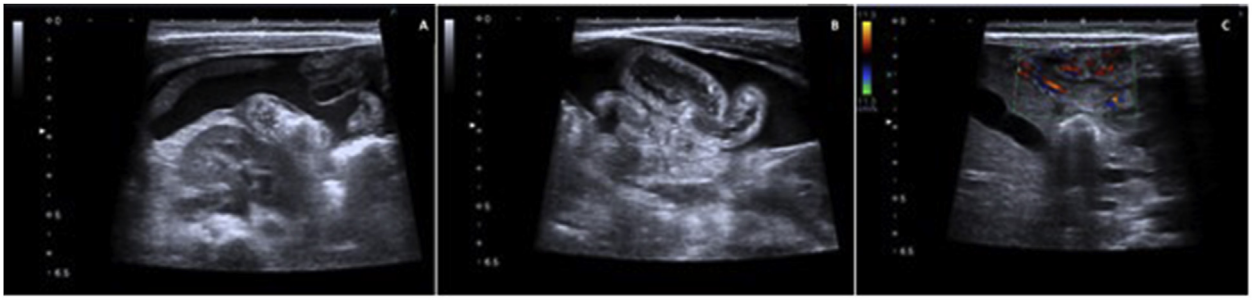

Fig. 1. Ultrasound images: A – high-volume intraperitoneal echogenic free fluid, B – distended bowel loop with thickened wall, C – irregular prominent mesocolon vessels.

microbiology analysis) and inflammatory findings on the ascending and transverse colon without any vascular compromise. Assuming an invagination with spontaneous reduction, he was discharged, asymptomatic.

Readmitted after 24 h, due to high-volume intraperitoneal echogenic free fluid (Fig. 1A), distended bowel loop with thickened wall (Fig. 1B) and irregular prominent mesocolon vessels on the right quadrant (Fig. 1C) on an US re-evaluation. Later, he started episodes of irritability, groaning, colic pain, abdominal distension and mucous in stools. Laboratorial data revealed a normocytic normochromic anaemia (7.9–8.1 g/dL) requiring blood transfusion, consumption of the factors of the extrinsic pathway of blood coagulation and fibrinogen, D-dimers elevation, lipase elevation with normal amylase and a maximum CRP of 4.55 mg/dL. Cystic fibrosis test was negative. Bacterial and viral investigations were negative. An abdominal computerized tomography (CT) angiography showed generalized intraperitoneal free fluid, enlarged irregular mesenteric vessels, some distended bowel loops, thin septum cysts with hypointense content, at the hepatogastric ligament, adjacent to the hepatic border and retroperitoneal, adjacent to the inferior vena cava (Fig. 2 A, B e C), at the intrahepatic segment and peripancreatic, without secure exclusion of a multiloculate lymphatic malformation.

One month after the first surgery, a new laparotomy was performed due to hemodynamic instability associated with sudden increase in abdominal free fluid and cystic volume (maximum diameter of 29 mm) (Fig. 3). An hemoperitoneum was found associated with multiple cysts on the supramesocolic compartment which were resected from perigastric, duodenal, epiploic and mesocolon locations. Additionally, a duodenal perforation with active bleeding was identified and repaired. Intraoperative blood loss was 300 ml, requiring blood transfusion during the procedure.

After surgery, he remained hemodynamically stable, with sporadic colic episodes, resuming normal intake and stools by the 5th postoperative day. Lipase, fibrinogen and D-dimers normalized. An abdominal US on the 8th postoperative day revealed cystic dimensions reduction, adjacent to the inferior vena cava and the left hepatic border, maintaining echogenic content, and no intraperitoneal cysts. An increased retro-epiploic cyst was identified, with 55 × 26 mm, communicating with a cyst medial to the spleen, elongating behind the pancreatic tail. An abdominal magnetic resonance imaging (MRI) was made for better characterization and found no lesions on the liver, pancreas, spleen and kidneys, a cystic image close to the inferior vena cava, with blood content (hyperintense sign at T1 and T2), and another posterior-inferior to the stomach with liquid content (hypointense at T1 and hyperintense at T2) (Fig. 4 A e B).

Histologic analysis confirmed a venous and lymphatic malformation, and malignancy was excluded. Consequently, he initiated oral treatment with propranolol (3 mg/kg/day), after normal cardiac evaluation, with notorious reduction of the cutaneous haemangioma and progressive cyst reduction on ultrasound follow-up. Discharged on the 32nd postoperative day, asymptomatic, under oral propranolol, and remained on close follow-up on the daily clinic. Sustained ultrasound cystic reduction until seven months after starting oral propranolol, when all cystic images disappeared. Accordingly, oral propranolol was suspended.

After three years of follow-up, he remains asymptomatic, with adequate growth height and weight (above 50th percentile) and without cystic images on abdominal US.

3. Discussion

Lymphatic-venous malformations result from an obstruction of the venolymphatic communication between the dysembryoplastic vascular tissue and systemic circulation [9]. These rare malformations, located over 90% in the head and neck [6,8,10], are very rare in intra-abdominal or retroperitoneal locations [8,10]. Among retroperitoneal tumours, lymphatic malformations and, particularly lymphatic-venous malformations, are also rare [11]. A literature review in PubMed without language restriction, of published data until November 2017, with search terms including Retroperitoneal/Retroperitoneum Hemolymphangioma and Lymphatic-Venous/Lymphovenous Malformation, found five relevant articles, but only four were full-text, with

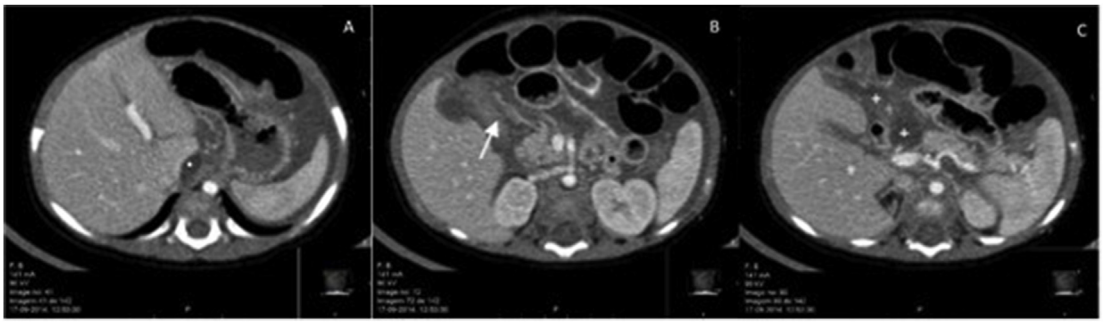

Fig. 2. CT angiography images with generalized free fluid: A – cyst with hypointensive content adjacent to the inferior vena cava (signed by *), B – enlarged irregular mesenteric vessels (signed by an arrow), C – cysts with hypointensive content adjacent to the hepatic border (signed by +).