



Septic shock: an atypical complication of alimentary tract duplication

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ABSTRACT

Alimentary tract duplications are heterogenous congenital anomalies of the digestive tract. Their form of presentation is varied, and they may lead to different complications, depending on their natural course. Infection is a rare complication, but it cannot be ignored because of its severity.

Here we describe the case of an otherwise healthy 2-year-old girl with an atypical complication of alimentary tract duplication: septic shock. She initially consulted due to abdominal distension and pain associated with a palpable abdominal mass. The imaging studies showed a partial fluid septation in the right side of the abdomen. During hospitalization, an intratumoral infection developed, which progressed to septic shock. The patient responded favorably to medical treatment for shock, and surgical resection was subsequently performed. The pathology report confirmed the presence of alimentary tract duplication.

Keywords: *gastrointestinal tract; septic shock; pediatrics.*

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INTRODUCTION

Alimentary tract duplications (ATD) are heterogenous congenital anomalies that may be found adjacent to the digestive tract.¹ They may go unnoticed and be found incidentally or, on some occasions, they are palpable abdominal masses that are referred for consultation.^{1,2} During their natural course, they may develop some type of complication, such as hemorrhage, obstruction, volvulus, etc.²⁻⁴

Here we report an atypical form of presentation in a patient presenting with severe systemic infection.

CASE REPORT

This was a 2-year-old female patient with no relevant medical history who consulted due to abdominal distension and pain associated with a hypogastric tumor. On physical examination, the patient was in good general condition, with normal body weight and no fever. On inspection, marked abdominal distension was observed; palpation revealed a palpable mass predominantly in the right side of the abdomen which was mobile, painful and had a hard-elastic consistency. The rest of the abdomen was soft and depressible. Lab tests showed anemia with hemoglobin levels of 6.3 g/dL, which required red blood cell

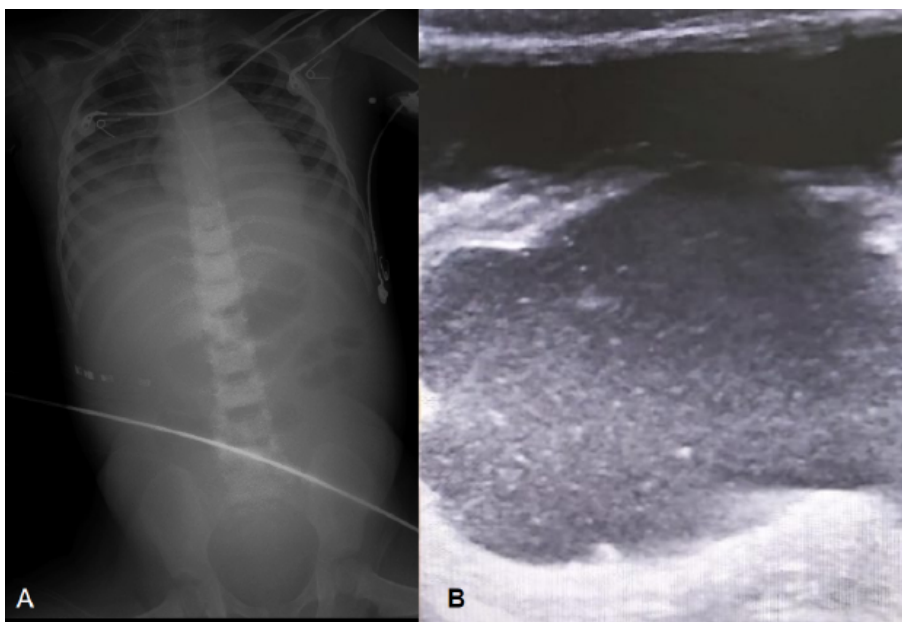
transfusion, and no other pathological laboratory findings.

The abdomen X-ray (*Figure 1A*) showed an important abdominal “silence,” consistent with the palpable mass, with no bowel sounds or other pathological signs. The ultrasound (*Figure 1B*) showed a cystic image with hyperechogenic walls that measured 98 mm × 59 mm and contained small septa with finely particulate fluid inside that extended from the mesogastrium and hypogastrium into the right side of the abdomen, displacing the intestinal loops. The magnetic resonance imaging findings were similar to those of the ultrasound (*Figure 2*).

Tumor markers were negative.

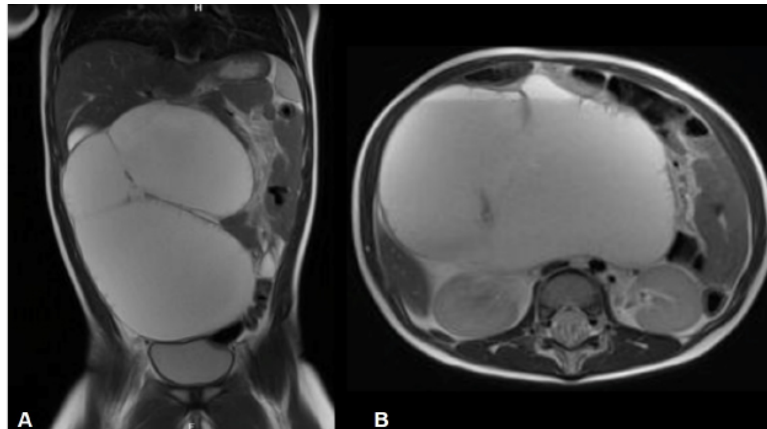
Since the findings showed a septated cystic tumor, a macrocystic lymphatic malformation was suspected and surgical treatment was planned (aspirate and injection of sclerosing agent). While awaiting the procedure, the patient developed hyperthermia (maximum temperature of 40 °C) associated with blood pressure of 75/53 mmHg, heart rate of 170 bpm, respiratory rate of 58 bpm, slow capillary refill (4 to 5 seconds) with bounding pulses and altered sensorium. Intravascular volume expansion was induced with crystalloids at 20 mL/kg, but with no favorable response.

FIGURE 1. X-ray and ultrasound of abdomen



A. X-ray of abdomen. Important “silence” in right side of abdomen.

B. Ultrasound. Tumor with echogenic wall and fine particulate content.

FIGURE 2. T2 sequence of magnetic resonance imaging

Partial fluid septation located from the epigastric to the hypogastric region, predominantly in the right side of the abdomen and displacing adjacent structures.

A: Coronal section. B: Axial section.

Since the condition was refractory to treatment, she was transferred to the Intensive Care Unit, where she was placed on mechanical ventilation and started receiving vasopressors (epinephrine and norepinephrine) and broad-spectrum antibiotics (vancomycin, amikacin, and piperacillin-tazobactam). Specimens for blood and urine cultures were also collected. Lab tests showed leukopenia (1400 leukocytes/mm³) and increased acute phase reactants (PCR: 147 mg/dL) and lactic acid level of 2.7 mg/dL. An ultrasound-guided percutaneous puncture of the tumor was also performed; cloudy hemorrhagic fluid was collected and sent for cytochemical analysis and culture. An intracystic drain was not placed to reduce the risk of overinfecting a tumor that was originally not communicated with the exterior and whose etiology had not yet been confirmed. The cytochemistry showed abundant red blood cells and leukocytes with a predominance of polymorphonuclear cells, with low glucose levels. The culture was positive for *Pseudomonas aeruginosa*, as was the blood culture. The isolation of the microorganism allowed to reduce the antibiotic therapy to amikacin and piperacillin-tazobactam.

After the treatment, the patient responded favorably and, on the third day, was in optimal condition for the surgery, with no fever or need for vasopressors. An exploratory laparotomy was performed through a median infraumbilical entry; a cystic tumor was found that involved the mesentery of the terminal ileum and the wall of the cecum and ascending colon, irrigated by arteries parallel to the ileal arteries, which was difficult to interpret. An en bloc resection was

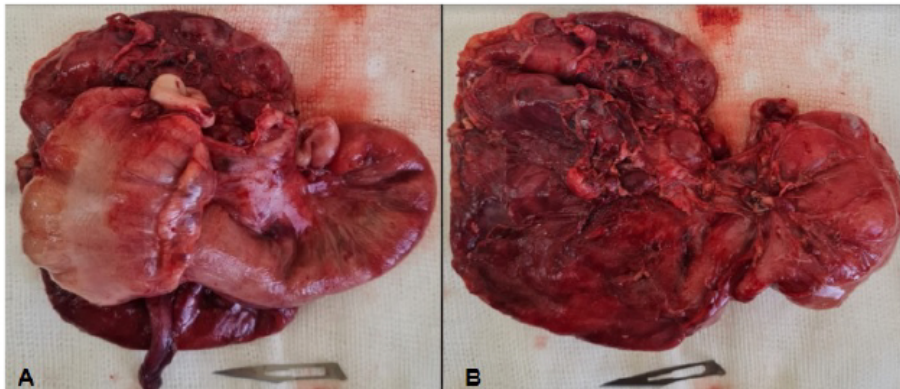
performed including the tumor, terminal ileum, cecum, and ascending colon (Figure 3), and a shotgun tube ostomy was performed between the terminal ileum and ascending colon. The deferred pathological study was compatible with a cystic duplication of the terminal ileum, type 1B as per the vascular classification by Li et al.⁵ (Figure 4). The patient had an adequate postoperative course.

DISCUSSION

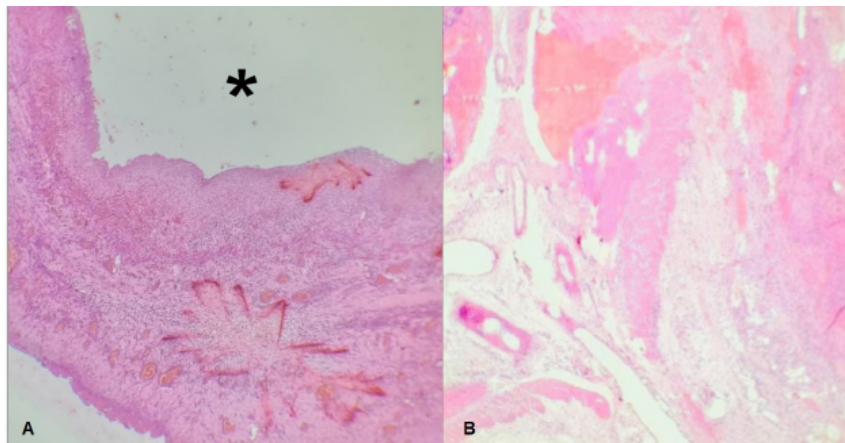
As described for this case, ATD may lead to severe complications, such as infection with systemic involvement. Although intratumoral hemorrhage has been well described as a complication in ATD² and was suspected in our patient as a probable cause of hypotension, the development of hyperthermia, leukopenia, increased levels of acute phase reactants, refractoriness to fluid replacement therapy, and the need for vasopressor therapy suggested a severe infection that triggered septic shock.⁶

This is not a common complication; the case series published by Guerin et al.⁷ included 114 patients and none of them were diagnosed in the context of an infection. This is consistent with other series, such as those published by Erginel et al.⁸ and Sujka et al.⁹, who included 40 and 35 patients, respectively. We believe that bleeding could be a predisposing factor for infection, as observed in our patient.

Other abdominal cystic tumors, such as macrocystic lymphatic malformations, ovarian cysts, cystic teratomas, and hydatid or mesenteric cysts, should be considered as part of the

FIGURE 3. Surgical specimen

A: Anterior view. B: Posterior view.

FIGURE 4. Pathological study

The histology was compatible with alimentary tract duplication: internally covered by differentiated intestinal epithelium with atrophic villi, smooth muscle arranged in longitudinal and smooth layers, and externally covered by serosa.
 A: Transperietal view (*inside the cyst). B: Amplification of the mucosa.

differential diagnosis.¹⁰ The signs and symptoms of these tumors are usually similar, which renders the initial suspected etiology difficult, and although imaging studies may guide the treating team, the definitive diagnosis is always based on the pathological study.² An ultrasound should be the first study requested in case of an abdominal cystic tumor.¹¹ Cross-sectional imaging studies, such as a computed tomography or a magnetic resonance imaging, are reserved for large or complicated cystic tumors;¹² they will provide information on tumor location, size, shape, walls, and relationship to adjacent structures. In the differential diagnosis, an ATD shows a characteristic ultrasound pattern with 3 layers in the cyst wall corresponding to the mucosa, submucosa, and muscle;¹¹ however, when found

in relation to an inflammatory process, such pattern is lost and makes diagnosis difficult. In our patient, the intracystic inflammation resulted in the loss of this pattern. The same experience was described by Di Serafino et al.¹² in their series.

Our case was also notable due to the large size of the tumor, with a maximum diameter of 98 mm. This characteristic made the initial suspected etiology difficult to establish, as an ATD is usually less bulky. In the series published by Guerin et al.,⁷ the largest ATD measured 42 mm, with a mean diameter of 35 mm in the postnatal diagnosis group and a diameter of 30 mm in the prenatal diagnosis group. Most likely, the inflammatory process could have been a facilitating factor for the increase in tumor size, as observed in our case.

Most authors agree that the definitive surgical treatment of choice is enucleation or resection with primary anastomosis.^{13,14} In our patient, these options were not viable due to her clinical condition, so it was decided to perform an intestinal resection of the affected segment and an ostomy. Some publications suggest an expectant management in asymptomatic patients.¹⁵ However, we do not believe this is an appropriate recommendation because an ATD left to its natural course may cause severe and potentially fatal syndromes, as in our patient.

To conclude, we present the case of an atypical presentation of ATD, which progressed to an intracystic infection. Although the incidence of this complication is low, we believe it is necessary to note such onset condition due to its potential severity. An elective surgery at the time of diagnosis would be the most appropriate course of action. ■

REFERENCES

- Spătaru RI, Lupuşoru MOD, Şerban D, Ivanov M, Iozsa DA. Alimentary tract duplications in children - a 15 years' experience. *Rom J Morphol Embryol*. 2021;62(3):751-6.
- Xiang L, Lan J, Chen B, Li P, Guo C. Clinical characteristics of gastrointestinal tract duplications in children: A single-institution series review. *Medicine (Baltimore)*. 2019;98(44):e17682.
- Iyer CP, Mahour GH. Duplications of the alimentary tract in infants and children. *J Pediatr Surg*. 1995;30(9):1267-70.
- Sangüesa Nebot C, Llorens Salvador R, Carazo Palacios E, Picó Aliaga S, Ibañez Pradas V. Enteric duplication cysts in children: varied presentations, varied imaging findings. *Insights Imaging*. 2018;9(6):1097-106.
- Li L, Zhang JZ, Wang YX. Vascular classification for small intestinal duplications: experience with 80 cases. *J Pediatr Surg*. 1998;33(8):1243-5.
- García PCR, Tonial CT, Piva JP. Septic shock in pediatrics: the state-of-the-art. *J Pediatr (Rio J)*. 2020;96 Suppl 1(Suppl 1):87-98.
- Guérin F, Podevin G, Petit T, López M, et al. Outcome of alimentary tract duplications operated on by minimally invasive surgery: a retrospective multicenter study by the GECl (Groupe d'Etude en Coeliochirurgie Infantile). *Surg Endosc*. 2012;26(10):2848-55.
- Erginel B, Soysal FG, Ozbey H, Keskin E, et al. Duplication Cysts in Children: A Single-Institution Series with Forty Patients in Twenty-Six Years. *World J Surg*. 2017;41(2):620-4.
- Sujka JA, Sobrino J, Benedict LA, Alemayehu H, et al. Enteric duplication in children. *Pediatr Surg Int*. 2018;34(12):1329-32.
- Fernández Ibieta M, Rojas Ticona J, Martínez Castaño IM, Reyes Ríos P, et al. Quistes mesentéricos en la edad pediátrica: ¿qué son en realidad? *An Pediatr (Barc)*. 2015;82(1):e48-51.
- Barr LL, Hayden CK Jr, Stansberry SD, Swischuk LE. Enteric duplication cysts in children: are their ultrasonographic wall characteristics diagnostic? *Pediatr Radiol*. 1990;20(5):326-8.
- Di Serafino M, Mercogliano C, Vallone G. Ultrasound evaluation of the enteric duplication cyst: the gut signature. *J Ultrasound*. 2015;19(2):131-3.
- Rasool N, Safdar CA, Ahmad A, Kanwal S. Enteric duplication in children: clinical presentation and outcome. *Singapore Med J*. 2013;54(6):343-6.
- Holcomb GW 3rd, Gheissari A, O'Neill JA Jr, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. *Ann Surg*. 1989;209(2):167-74.
- van Zitteren LM, Ruppert M, Op de Beeck B, Wojciechowski M. Infected enteric duplication cyst. *BMJ Case Rep*. 2017;2017:bcr2017222391