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CASE REPORT

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Intestinal lymphangiectasia in a young infant: Clinical case

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Abstract

Introduction: *Intestinal lymphangiectasia is a disease characterized by an abnormal dilation of the intestinal lymphatic vessels, causing alterations in the intestinal mucosa with the subsequent loss of lymph to the intestinal lumen, causing: hypoproteinemia, hypoglobulinemia, lymphopenia and edema.*

Clinical case: *A 4-month-old infant who arrives at the emergency department with intussusception and severe dehydration. The emergency management was done with parenteral fluid resuscitation, taking the patient to stable conditions and antibiotics, deciding surgical intervention. Intussusception with ileal perforation was found intraoperative, requiring intestinal diversion. In the postoperative period, he presented a poor clinical evolution, with a hospital stay with hypoproteinemia, hypoglobulinemia, lymphopenia and hypocalcemia, despite corrections and continuous parenteral intake. The patient required a new surgical intervention due to clinical and radiological data of intestinal perforation and a new perforation was found in a site that was not involved with intussusception, as well as multiple serous hematomas in different intestinal portions. The outcome was fatal and intestinal lymphangiectasia was established as a final diagnosis.*

Discussion: *In this case, there is a poor evolution with persistent metabolic alterations despite management with high intakes of protein, calcium and albumin, as well as corrections when warranted. Conclusions. We found a case of early-onset intestinal lymphangiectasia where, due to its presentation, it was aggressive, with a poor prognosis and difficult to manage, leaving it inconclusive if it was a primary case and this was the cause of intussusception or secondary to intussusception.*

Keywords: *lymphangiectasia, intestinal occlusion, intestinal perforation, pediatrics*

Introduction

Intestinal lymphangiectasia is a disease characterized by an abnormal dilation of the intestinal lymphatic vessels, which cause alterations in the intestinal mucosa with the subsequent loss of lymph to the intestinal lumen, causing hypoproteinemia, hypoglobulinemia, lymphopenia and edema [1]. The disease is usually primary; it was described for the first time by Dr. Thomas A. Waldmann in 1961, the disease that bears his name and the most frequent presentation [2]. It can be secondary, due to intestinal inflammatory processes, tumors or post-chemotherapy [3-5].

The most frequent clinical presentation is with edema and chronic diarrhea which is characterized by being a protein loser. In turn, patients present with hypoproteinemia, hypoglobulinemia, hypocalcemia, hypocholesterolemia, and lymphopenia, usually in children under 3 years of age.

The diagnosis is established by radiological, endoscopic, and histopathology studies. Treatment is dietary, with medical management and sometimes by surgery, when resecting lesions located in the intestine.

Case presentation

A A 4-month-old female infant who begins her current condition with rejection of the oral route, later vomits of gastric content, goes to a private doctor who administers Diphenidol in an unspecified dose without presenting improvement, approximately 12 hours after having started with vomiting, he presented stools with mucus and blood, so he went to the emergency department. Upon admission to the emergency department, he was found to be an infant weighing 6.2 kg, heart rate 130 ‘, respiratory rate 40’, temperature 36.4oC, capillary filling in 2-3 seconds, hypoactive, crying and irritability in intermittent periods . At abdominal level with Dance sign (+), blood sausage sign (+) at epigastric level. A digital rectal examination was performed and the presence of a “tench mouth” sign (+) was found on auscultation with peristaltic noise present in the upper abdomen and absence in the lower abdomen. Laboratory studies are taken which are reported in table 1. Due to the clinical findings and presentation of the symptoms, the diagnosis of intussusception was established, it was managed with parenteral water intake, initiation of antibiotics with spectrum for gram (+) and gram (-) and an emergency surgical intervention is decided. As a relevant finding, there is ileocecocolic invagination to sigmoid, invaginating by taxis, finding perforation at 6 cm of the ileus cecal valve and necrosis of the ascending colon, for which a resection of the terminal ileus and right hemicolectomy + intestinal diversion at the ileal level at 2 cm was decided. Perforation site where viable intestine was found (Figures 1-5).

After surgery, the patient presented a poor evolution, with the presence of generalized edema, pulmonary rales, abdominal wall edema, diarrheal evacuations and a tendency to hypocalcemia and hypoalbuminemia (Table 1) despite protein, albumin and calcium intake in total parenteral nutrition. On day 5, a control abdominal film was taken and data on free peritoneal air were found, so it

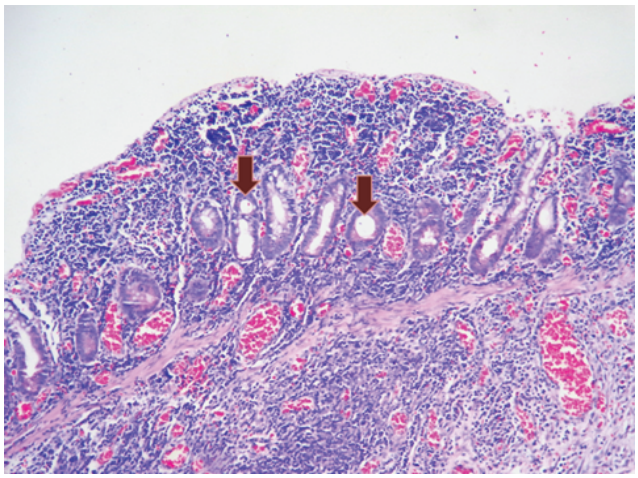

***Figure 1**. Dilated lymphatic vessels in the lamina propria of intestinal velocities.*

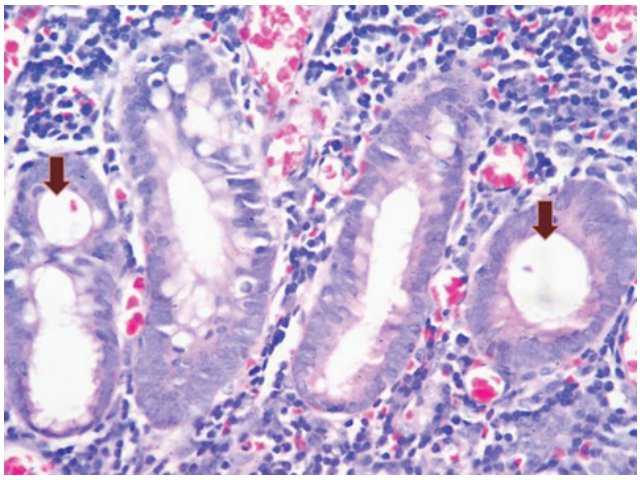

***Figure 2**. The dilated lymphatic vessels do not have inflammatory cells inside.*

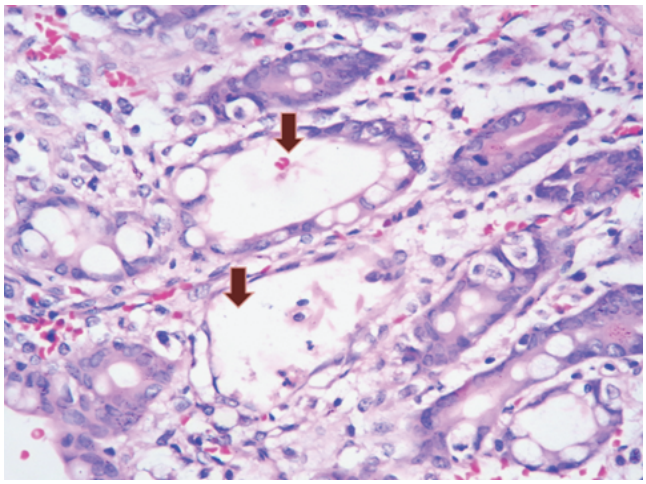

***Figure 3.** These findings are nonspecific. The diagnosis is made considering the clinical and endoscopic context.*

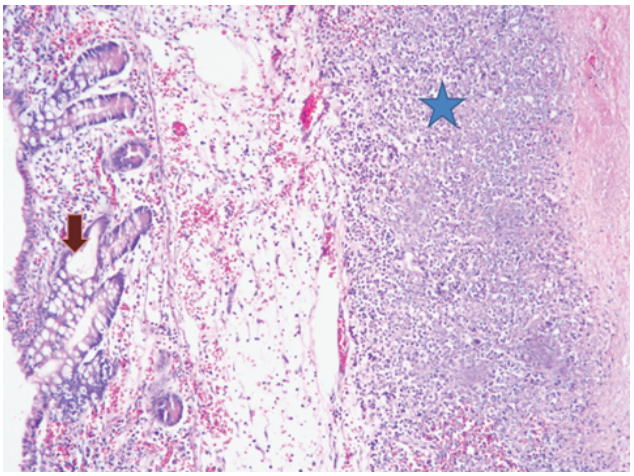

***Figure 4.** Severe transmural acute inflammatory process with ischemic-hemorrhagic injury secondary to intestinal perforation / obstruction.*

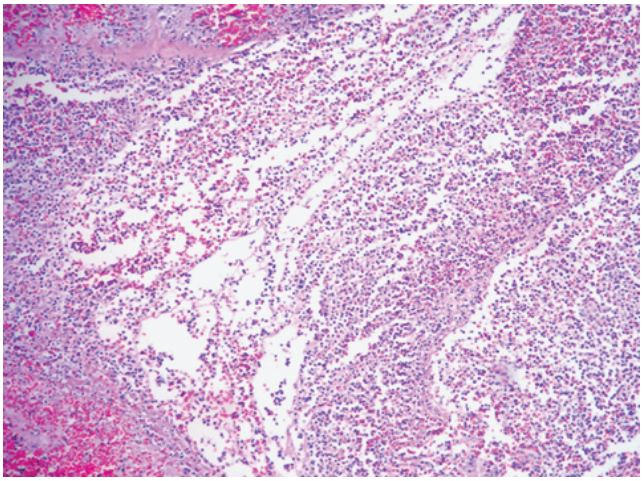

***Figure 5**. Acute Inflammation, fibrin, and hemorrhage*

was decided to perform a new emergency surgical exploration. During the intraoperative period, intestinal perforation was found 24 cm from the stoma, generalized intestinal serous edema and multiple macroscopic hematomas including jejunum, resection was performed with 2-cm borders free from the perforation site and end-to-end anastomosis, and the specimen was sent for histopathological study. The evolution continues to be torpid with the same characteristics of diarrhea, pulmonary rales, edema of the abdominal wall, hypocalcemia, hypoalbuminemia and hypoglobulinemia, despite high contributions and corrections made as management. As a relevant added data, the patient remained afebrile. Finally, the patient died from multiple organ failure on day 9 of the stay. The day after death, intestinal lymphangiectasia was reported by histopathology.

Discussion

Intussusception is rare in newborns, occurring in only 0.3 to 1.3% of all cases. Its etiology is unclear and may be related to intestinal hypoperfusion, dysmotility, or stenosis. Clinically it is indistinguishable from necrotizing enterocolitis and its diagnosis is generally established during surgery. In our case, the clinical presentation began as a severe intestinal intussusception [6]. Lymphangiectasia has been classified within the group of rare

Table 1. *Laboratory studies*

	Hospital stays (Days)							
Laboratory	Entry	2º	3º	4º	5º	6º	7º	9º
Hemoglobin (g/dl)	9.9	11.1	7.4	10.7	10.8	8.5	10.5	11.4
Hematocrit (%)	30.4	33.7	21.1	31.7	32.8	26.2	30.2	33.6
Leucocytes	16200	12900	9900	13700	1200	7800	9500	11800
Neutrophils/ lymphocytes (%)	68/20	66/23	52/25	71/22	80/19	54/37	36/45	66/25
Platelets/µL	349000	38000	61000	13000	17000	7000	32000	42000
ESR (sec)	16							
Procalcitonin (ng/ml)						1.8		
PCR (mg/dl)	12							
Na (mmol/l)	132	139	141	137		140	139	141
Cl (mmol/l)	99	113	115	110				106
K (mmol/l)	4.6	2.8	2.5	4.6	3.6	3.7	3.9	
Ca (mmol/l)		7	8.9	5.1		1.6/4.6	1.7/8.8	2.3/9
Glucose (mg/dl)	151		107	76		88	95	
Urea (mg/dl)	30		14	20		31	47	
Creatinin (mg/dl)	0.26		0.34	0.33		0.35	0.39	
AST (U/l)				27		21		
ALT (U/l)				17		16		
TB (mg/dl)				1.8		2.2		
DB (mg/dl)				1.1		1.1		
IB (mg/dl)				0.7		1.1		
AP (U/l)				55				
A/TP (g/dl)			1.5	1.6/3.3		2.1/4.2		1.9/4.7
Globulin (g/dl)				1.7				
PT (sec)	12.5				48.5		13.5	
PTT (sec)	25.4				35		30	
INR	1.15				1.7		1.2	
pH			7.45	7.37		7.44		
PCO2 (mmHg)			21.8	32		35.8		
PO2 (mmHg)			87	50		45.8		
HCO3 (mmol/l)			15.6	18		25		
Base excess (mEq/L)			-7.1	-5		1.8		
O2 Saturation (%)			97	84		86		

ESR: Erythrocyte sedimentation rate; PCR: C-reactive protein; TB: Total bilirrubin; DB: Direct bilirrubin; IB: Indirect bilirrubin; AP: Alkaline phosphatase; ATP=Albumin/Total proteins; PT: Protrombin time; PTT: Partial thromboplastin time.

diseases, so there are few reports about this entity. It is described more frequently in the early ages of life, as happened in our case, although some reports of incidence in adults have been described.

Primary and secondary causes are described, among which are indicated with greater significance the presence of lymphomas, systemic lupus erythematosus, tuberculosis intestinal, retroperitoneal fibrosis, post radiation effects, parasitic infections, liver cirrhosis and constrictive pericarditis and it is suspected that 73% of cases are idiopathic or primary [7].

This entity can generally be diagnosed before the age of five and progress to adulthood with appropriate treatment and its management is carried out through the restriction of long-chain fatty acids and supplements of the dietary treatment of IPL with acid restriction long-chain fatty acids and dietary supplements with medium-chain triglycerides. That allows clinical control and adequate development in most cases. The immunological deficit secondary to the loss of chyle does not condition a special susceptibility to suffer infections [8]. Aycardi Valverde, et al., report a case in an 8-month-old girl who underwent a diagnosis by endoscopy and taking a biopsy, being possible its diagnosis and management with satisfactory evolution [9,10].

Alshikho et al. [11], they report a case in an adult woman who presented edema in the lower limbs, cellulitis and repeated infections, making the corresponding diagnosis and management with good results.

In this case, there is a poor evolution with persistent metabolic

alterations despite management with high intakes of protein, calcium, and albumin, as well as corrections when warranted.

On questioning the family members, there was no history of infectious, respiratory or gastrointestinal symptoms, as usually occurs in cases of intussusception. It was also established that the infant's feeding was adequate for her age and she had not manifested diarrhea or edema as the literature describes it as common in the clinical presentation of primary intestinal lymphangiectasia. We are faced with a case where the cause of the dilation of the intestinal lymphatic vessels functioned as an "invagination head", being an early presentation of the disease, or also, that the dilation of the vessels was secondary to the intestinal inflammatory process and obstruction lymphatic drainage caused by intussusception. The postoperative evolution with the clinical presentation of abdominal wall edema, rales, and protein and calcium disorders described, coincide with the description of the disease, which was corroborated by histopathology, which is the determining diagnostic study.

Conclusions

We found a case of early-onset intestinal lymphangiectasia where, due to its presentation, it was aggressive, with a poor prognosis and difficult to manage, remaining inconclusive if it was a primary case and this was the cause of intussusception or secondary to intussusception.

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