Meckel diverticulum with ectopic pancreatic tissue in a 12-year-old boy with intestinal invagination. Case report and review of the literature

Divertículo de Meckel con tejido pancreático ectópico en un niño de 12 años con invaginación intestinal. Reporte de caso y revisión de la literatura

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Abstract

Introduction: Meckel's diverticulum is a remnant of the omphalomesenteric duct and its prevalence is 2%. **Clinical case:** A 12-year-old male with abdominal pain, accompanied by muco-sanguineous evacuations and hematemesis, with right lower quadrant mass. In the laparotomy, invasion was found; intussusception reduction and resection of the Meckel's diverticulum was performed. **Conclusion:** The association of Meckel's diverticulum with heterotopic pancreatic tissue in children is rare, its main manifestation is intussusception and ultrasound is an option for diagnosis.

KEY WORDS: Meckel diverticulum. Ectopic pancreatic tissue. Intussusception.

Resumen

Introducción: El divertículo de Meckel es un remanente del conducto onfalomesentérico y su prevalencia es del 2%. **Caso clínico:** Niño de 12 años con dolor abdominal, acompañado de evacuaciones mucosanguinolentas y hematemesis, con masa en cuadrante inferior derecho. En la laparotomía quirúrgica se encontró invaginación, y se realizó desinvaginación y resección del divertículo de Meckel. **Conclusión:** La asociación de divertículo de Meckel con tejido pancreático heterotópico en niños es poco frecuente. Clínicamente, su principal manifestación es la invaginación intestinal en la literatura revisada, y el ultrasonido constituye el mejor estudio para el diagnóstico de invaginación intestinal, no así para el divertículo de Meckel o el tejido pancreático heterotópico, en los que la tomografía sería una opción.

PALABRAS CLAVE: Divertículo de Meckel. Tejido pancreático ectópico. Invaginación intestinal.

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Introduction

Meckel's diverticulum is a remnant of the omphalo-mesenteric duct, located on the antimesenteric border of the ileum, at 40-60 cm from the ileocecal valve, and it is characterized for containing all the layers normally found in the ileum¹⁻³. The prevalence of Meckel's diverticulum is approximately 2% in the general population; of total cases, 76% are in adults and 24% in pediatric patients^{2,3}.

In 62% of cases heterotopic gastric mucosa can be found, in 6%, pancreatic tissue, in 5%, gastric and pancreatic mucosa, in 2%, jejunum mucosa and in 2% Brunner's tissue⁴. Clinical presentation can include intestinal bleeding, pain, intestinal occlusion or intussusception⁵. It can be diagnosed with technetium-99 scintigraphy when there is ectopic gastric mucosa, but computerized tomography is the method that has been shown to be the most specific, showing a tubular structure filled with fluid or intestinal intussusception images⁶.

The purpose of this paper is to present a case of Meckel's diverticulum with heterotopic pancreatic tissue in a 12-year-old boy with intestinal intussusception, as well as to perform a review of the literature, since this is a rare pathology and we did not find cases reported in the national literature.

Clinical case

Twelve-year-old boy with a history of laparoscopic appendectomy at 10 years of age. He attended the emergency department with an 8-hour history of abdominal pain, located at the mesogastrium with 10/10 intensity, accompanied by muco-sanguineous stools in seven occasions, which subsequently were melanic. There were episodes of vomiting on eight occasions, with gastro-alimentary content and then with blood. On physical examination, blood pressure of 129/48 mmHg, mean blood pressure of 73 mmHg, heart rate at 100 beats per minute, respiratory rate of 28 breaths per minute, temperature of 36 °C, 86% saturation and weight of 51 kg were observed. Dehydrated, conscious, oriented, pale oral mucosa, lung fields with air adequately flowing in and out. Precordium with heart sounds increased in intensity, without murmurs. Soft, depressible abdomen. With a diagnosis of digestive tract bleeding of etiology to be determined, management with omeprazole, acetaminophen and ondansetron was administered for 48 hours, with

no improvement. Laboratory tests reported hemoglobin 15.00 g/dL, platelets 193,000, white blood cells $10,600 \times 10^3 \mu\text{L}$, neutrophils 89.70%, lymphocytes 9.10%, monocytes 1%, basophils 0.20%, creatinine 0.82, glucose 128 mg/dL, sodium 139 mEq/L, potassium 4.1 mEq/L, calcium 9.40 mg/dL and C-reactive protein (CRP) 0.793 mg/dL. A pan-endoscopy did not reveal active bleeding in the stomach or esophagus. Since on abdominal palpation a mass was felt in the lower right quadrant, an ultrasound was carried out, which reported a fluid-free image with a "bull's eye" appearance, suggestive of invagination. The intussusception head was displayed at the hepatic angle, with a pediculate, rounded image dependent on a handle of the small intestine being identified at the center (Fig. 1).

With the diagnosis of intestinal intussusception with more than 48 hours of evolution, and considering the patient age, performing a surgical examination was decided, where double invagination was found: ileoileal and ileocolic intussusception (Fig. 2), with the first ileoileal segment harboring a Meckel diverticulum inverted on its axis and this segment doubly invaginated to the ascending colon. Desinvagination by taxis was carried out, together with intestinal resection of the Meckel diverticulum located at 40 cm of the ileocecal valve (Fig. 3), with 5 cm of intestine at each side and end-to-end anastomosis. Postoperative evolution was favorable, and the patient was discharged without complications, and at the moment of the report, he was in good health conditions.

On histopathological examination, macroscopically, the intussuscepted Meckel diverticulum was observed with its three layers and, at the center, solid-looking, light yellow tissue, reminiscent of pancreatic tissue (Fig. 4). On histological section, pancreatic acini and slightly dilated ducts could be appreciated; there were no Langerhans islets (Fig. 5).

Discussion

Heterotopic pancreas is defined as pancreatic tissue without a true anatomical or vascular connection with the pancreas, and was first reported by Jean-Schultz in 1729^{7,8}. Other terms used are ectopic pancreas, accessory pancreas and aberrant pancreas. In 1859, Zenker reported the first case of ectopic pancreas in a Meckel diverticulum⁹.

Heterotopic pancreas origin is still unclear, but it is believed to arise embryonically during rotation of the foregut and fusion of the dorsal and ventral pancreatic tissue, where some tissue separates from the

Figure 1. Abdominal ultrasound, where the intussusception head is visualized, with a pediculate, rounded image, dependent on a small bowel loop, is centrally identified in the internal lateral face.

Figure 2. Double intestinal intussusception: ileoileal and ileocolic.

Figure 3. Meckel's diverticulum with heterotopic pancreatic tissue.

pancreas and develops in any portion of the intestine. Another best known theory is based on the pancreatic metaplasia of endodermal tissue^{8,9}. Due to the proximity of embryonic pancreatic primordial buds and the foregut during development, 70-90% of ectopic pancreas occurs in the upper portion of the digestive tract¹⁰.

Most patients with Meckel's diverticulum are asymptomatic and the diagnosis is usually made by scintigraphy with technetium-99, tomography, endoscopy or during surgical examination motivated by other diseases. In children, most cases are detected due to the presence of intestinal intussusception. Heterotopic pancreatic tissue visualization is difficult, and we only found one report where heterotopic pancreatic tissue could be identified in a Meckel's diverticulum as a homogeneous, hypoechoic and well-defined mass on ultrasound¹¹.

According to Heinrich's criteria, heterotopic pancreas is classified as type 1 if it contains cells of exocrine glands, excretory ducts and islets of Langerhans, type 2 if it contains excretory glands and excretory ducts, and as type 3 if there are only excretory ducts. The most common is type 2, followed by type 1, while type 3 is uncommon¹²⁻¹⁴. Our case corresponded to type 2.

In our literature review, we were able to document 10 cases reported in children from 23 days to 15 years of age, 60% in children older than 2 years and 70%

Table 1. Documented cases of children from 23 days to 15 years of age with Meckel's diverticulum and heterotopic pancreatic tissue

Author, year	Age	Gender	Presentation	Histopathological classification
Guanà et al. ⁶ , 2014	7 years	Female	Intussusception	Type 1
Baysoy et al. ¹⁵ , 2010	5 years	Male	Intussusception	Unknown
Xiao et al. ⁵ , 2009	12 years	Male	Intussusception	Type 2
Huml et al. ¹⁶ , 2009	1 month	Male	Pain, nausea and vomiting	Type 1
Ogata et al. ¹⁷ , 2007	1 year	Female	Meckel's diverticulum, intestinal malrotation	Type 1
	4 years	Female	Intussusception	Type 2
	10 years	Male	Meckel's diverticulum	Unknown
	23 days	Male	Meckel's diverticulum, bile duct atresia	Unknown
Orri et al. ¹⁸ , 2006	7 months	Male	Intussusception	Unknown
Clog et al. ²⁰ , 1908	12 years	Male	Abdominal distension	Unknown
Current case	12 years	Male	Intussusception	Type 2

Figure 4. On the antimesenteric border, intussuscepted Meckel's diverticulum with its three layers is observed, and on the center, solid-looking, light yellow tissue, reminiscent of pancreatic tissue.

Figure 5. Histological section with pancreatic acini and slightly dilated ducts. There are no islets of Langerhans.

in males (Table 1). Around 50% had a presentation with intussusception, 30% with Meckel's diverticulum and, in two cases, exploratory laparotomy was

performed without an accurate diagnosis being established, and was based only on the presence of abdominal pain, vomiting or abdominal distension. In five cases, the type was reported according to Heinrich's histological classification, with types 1 and 2 being the most common^{5,6,15-20}.

We consider that heterotopic pancreatic tissue favors the presence of intestinal intussusception, and the literature also refers that if a patient develops acute pancreatitis in the "mother" organ, inflammation will also affect heterotopic foci, causing abdominal pain with elevation of pancreatic enzyme blood levels, which did not occur in our patient¹⁴.

Conclusions

Association of Meckel's diverticulum with heterotopic pancreatic tissue in children is rare. Clinically, its main manifestation is intestinal intussusception in the reviewed literature. Ultrasound is the best study for diagnosis of intestinal intussusception, but not so for Meckel's diverticulum and heterotopic pancreatic tissue, where tomography would be an option.

References

1. Levy AD, Hobbs CM. Meckel diverticulum: radiologic features with pathologic correlation. *Radiographics*. 2004;24:565-87.
2. Robustelli U, Manguso F, Armellino MF, Mannelli MP, Massa MR, Forner AL, et al. Acute symptomatic Meckel diverticulum management. Our experience on seven consecutive cases. *Ann Ital Chir*. 2014;85:129-35.
3. Elsayes KM, Menias CO, Harvin HJ, Francis IR. Imaging manifestations of Meckel's diverticulum. *AJR Am J Roentgenol*. 2007;189:81-8.
4. Burjonrappa S, Khaing P. Meckel's diverticulum and ectopic epithelium: evaluation of a complex relationship. *J Indian Assoc Pediatr Surg*. 2014;19:85-9.

5. Xiao WD, Chen W, Yang H. Heterotopic pancreas within Meckel's diverticulum with obscure then massive gastrointestinal bleeding in a 12-year-old child: case report and review of the literature. *J Int Med Res*. 2009;37:967-72.
6. Guanà R, Bucci V, Carbonaro G, Cerrina A, Ferrero L, Teruzzi E, et al. Heterotopic pancreas in Meckel's diverticulum in a 7-year-old child with intussusception and recurrent gastrointestinal bleeding: case report and literature review focusing on diagnostic controversies. *Afr J Paediatr Surg*. 2014;11:354-8.
7. Pearson S. Aberrant pancreas. Review of the literature and report of three cases, one of which produced common and pancreatic duct obstruction. *AMA Arch Surg*. 1951;63:168-96.
8. Tanaka K, Tsunoda T, Eto T, Yamada M, Matsuo S, Izawa K. Diagnosis and management of heterotopic pancreas. *Int Surg*. 1993;78:32-5.
9. Okamoto H, Fujishima F, Ishida K, Tsuchida K, Shimizu T, Goto H, et al. Intraductal papillary mucinous neoplasm originating from a jejunal heterotopic pancreas: report of a case. *Surg Today*. 2014;44:349-53.
10. Bromberg SH, Neto CC, Fernando A, Borger A, Franco MIF, Franca LCM, et al. Pancreatic heterotopias: clinicopathological analysis of 18 patients. *Rev Col Bras Cir*. 2010;37:413-9.
11. Monedero MD, Ripollés T, Nicolau MJ, Martínez-Pérez MJ. Pancreatic pseudotumor in Meckel diverticulum. *Abdom Imaging*. 2006;31:688-90.
12. Gunjača I, Mlinac-Lucijanić M, Pavlović A, Gunjača M. Inflammation of ectopic pancreatic tissue as unusual case of duodenal perforation — a case report. *Antropol*. 2010;3:1119-22.
13. Heinrich H. Ein Beitrag Zur Histologie des Sogenakzessorischen Pancreas. *Virchows. Arch Path Physiol*. 1909;198:392-401.
14. Zarand A, Bajtai A, Baranyai Z, Dede K, Jakab F. Inflammation of ectopic pancreatic tissue in a Meckel's diverticulum causing acute abdominal symptoms: a case report and review of the literature. *Int J Surg Pathol*. 2011;19:359-63.
15. Baysoy G, Balamtekin N, Uslu N, Karavelioğlu A, Talim B, Özen H. Double heterotopic pancreas and Meckel's diverticulum in a child: do they have common origin? *Turk J Pediatr*. 2010;52:336-8.
16. Huml M, Sýkora J, Kobl J, Pizingerová K, Šebor J, Skála V, et al. Heterotopický pankreas, ileokolická invaginace v kojeneckém věku" *Pediatr pro Praxi*. 2009;10:191-2.
17. Ogata H, Oshio T, Ishibashi H, Takano S, Yagi M. Heterotopic pancreas in children: review of the literature and report of 12 cases. *Pediatr Surg Int*. 2008;24:271-5.
18. Orri TO, Ingibjorg G, Ronald M. Diagnosis and treatment of gastric heterotopic pancreas. *World J Surg*. 2006;30:1682-9.
19. Ormarsson OT, Gudmundsdóttir I, Márvið R. Diagnosis and treatment of gastric heterotopic pancreas. *World J Surg*. 2006;30:1682-9.
20. Clog H. Acute Intestinal Obstruction. *Lancet*. 1908;1:639.