

Massive lower intestinal bleeding due to GIST in a Meckel's diverticulum

Sangrado digestivo bajo masivo por tumor de GIST en un divertículo de Meckel

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Keywords:

Meckel's
diverticulum,
gastrointestinal
bleeding,
hypovolemic shock,
gastrointestinal
stromal tumor.

Palabras clave:

Divertículo de
Meckel, sangrado
gastrointestinal,
choque hipovolémico,
tumor del estroma
gastrointestinal.

ABSTRACT

The presence of tumors in Meckel's diverticula is rare. Most of them are benign, but among the malignant ones we may find adenocarcinomas, sarcomas, and carcinoids. Also, the presence of gastrointestinal stromal tumors (GIST), which can be benign or malignant, has been reported. We were motivated to this work by the clinical presentation of a GIST in a Meckel's diverticulum with severe lower gastrointestinal bleeding and hypovolemia, which led to an urgent surgical intervention. **Case report:** We present the case of a 42-year-old woman with severe lower gastrointestinal bleeding and hypovolemic shock; she underwent emergency surgery because the cause of the bleeding was a gastrointestinal stromal tumor in a Meckel's diverticulum. Surgical treatment consisted of intestinal resection with oncologic limits. Histopathological study confirmed the presence of the malignant tumor. **Conclusions:** The presence of a gastrointestinal stromal tumor in a Meckel's diverticulum, although rare, has been reported in the medical literature, but not the clinical presentation of a GIST in a Meckel's diverticulum with severe hemorrhage leading to hypovolemic shock.

RESUMEN

La presencia de tumores en los divertículos de Meckel es poco frecuente. La mayoría es benigna, pero entre los malignos encontramos adenocarcinomas, sarcomas y carcinoides. También, ha sido informada la presencia de tumores del estroma gastrointestinal (GIST), los cuales pueden ser benignos o malignos. Nos motivó a este trabajo la presentación clínica de un divertículo de Meckel con una hemorragia digestiva baja severa e hipovolemia, la cual provocó una intervención quirúrgica urgente. **Caso clínico:** Se presenta el caso de una mujer de 42 años con hemorragia digestiva baja severa y choque hipovolémico; ella fue operada de urgencia, porque la causa del sangrado era un tumor del estroma gastrointestinal en un divertículo de Meckel. El tratamiento quirúrgico consistió en una resección intestinal con límites oncológicos. El estudio histopatológico confirmó la presencia del tumor maligno. **Conclusiones:** La presencia de un tumor del estroma gastrointestinal en un divertículo de Meckel, aunque es poco frecuente, ha sido informada en la literatura médica, pero no la presentación clínica de un GIST en un divertículo de Meckel con una hemorragia severa que conduce a un choque hipovolémico.

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Received: 07/11/2018
Accepted: 11/20/2018

INTRODUCTION

The presence of Meckel's diverticulum was first mentioned in 1598 by Guillelmus Fabricius Hildanus or Fabricius von Hilden or Fabricius Hildamus, who is considered the "father of German surgery". In 1671, Lavater also reported this same finding, but the most complete description was made by Johann Friedrich Meckel in 1809.¹

Meckel's diverticulum is associated with another congenital abnormality (tracheoesophageal fistula) in 30% of cases. It is located between 30 and 100 cm at the antimesenteric border of the terminal ileum.^{1,2} Cases have been reported up to 180 cm from the ileocecal valve, although this varies according to age; in children it is located on average 30-35 cm from the ileocecal valve



How to cite: Mederos-Curbelo ON, Barrera-Ortega JC, López-Sotelo A, Mederos-Trujillo ON, González-Lara J. Massive lower intestinal bleeding due to GIST in a Meckel's diverticulum. Cir Gen. 2020; 42(1): 45-49.

and in adults 60-80 cm. It measures between 1-8 cm and has an incidence of 1-4% with a higher frequency in males.² In 50% of cases there is gastric heterotopic tissue; other tissues found have been pancreatic, duodenal, jejunal, colonic, hepatobiliary, and endometrial tissue. Symptoms are related to hemorrhage, obstruction, diverticulitis, perforation and tumors.^{1,3}

Lower gastrointestinal bleeding is more frequent in children under two years of age; it presents with heterotopic gastric tissue, which exhibits peptic ulcers in the mucosal transition from the diverticulum to the ileum. In 55% of the cases, this presentation occurs in children under 18 years of age.^{1,4,5} The frequency of a tumor in a diverticulum ranges from 0.5-3.2%; if they are benign, they present as leiomyomas, angiomas and lipomas, but if they are malignant, carcinoids, adenocarcinomas, sarcomas, and gastrointestinal stromal tumor (GIST) may be found. The latter occurs in 12% of cases.⁴⁻⁶

GIST is a subepithelial tumor that was previously classified as mesenchymal tumor (previous nomenclature), such as leiomyoma, nerve sheath tumor, schwannoma, and lipoma, even, these can be benign or behave as malignant. Some characteristics of malignancy are mucosal or epithelial involvement, i.e., ulceration, hemorrhage, and presence of clots. Of all cases of GIST, 30 to 35% are found in the small intestine. Foreign bodies such as enteroliths and gallstones have also been described in Meckel's diverticula. The clinical presentation of a GIST in a Meckel's diverticulum with massive lower gastrointestinal bleeding and hypovolemic shock -which led to urgent surgical intervention- is exceedingly rare.

CLINICAL CASE

The case of a 42-year-old woman with a history of constipation is presented. She was admitted to the "Comandante Manuel Fajardo" University Hospital in Havana, Cuba for presenting with bloody, wine-colored stools. Her blood pressure was 110/70 mmHg with a heart rate of 105 per minute. On rectal examination, the rectal ampulla was found to be occupied by stool, and the glove was stained

with dark pasty stool and with the presence of clots. Volume replacement was started with colloid and crystalloid solutions. The following complementary analyses were performed:

Hemoglobin: 9 g/dl, hematocrit 28%.

Blood group: O negative.

Upper endoscopy: erosive edematous gastritis with no signs of bleeding.

Abdominal ultrasound: a segment of a loop dilated located in the right lower quadrant with concentric thickening of the walls suggestive of inflammatory process was seen.

Less than an hour later, the patient presented a bleeding of more than 1,000 ml. She appeared pale, sweating, and her body temperature was cold. She reported "dark vision"; her mucous membranes were also pale. She had a heart rate of 130 beats per minute, a blood pressure of 90/60 mmHg, and a respiratory rate of 24 per minute.

It was discussed as a group and it was concluded that this was a patient with a negative blood group, who presented an active lower gastrointestinal bleeding of great severity with signs of hypovolemic shock. Based on these criteria and the results of the complementary tests, emergency surgery was decided. The preoperative diagnosis was massive lower gastrointestinal bleeding due to possible Meckel's diverticulum with heterotopic gastric mucosa. An exploratory laparotomy was performed using an infraumbilical median incision.

The intraoperative finding was a small bowel tumor located 90 cm from the ileocecal valve measuring 15 centimeters in height and a diameter of 10 centimeters at the tip (*Figure 1*). Surgical resection of the affected segment was performed with an oncologic margin on both sides. A continuous extra-mucosal end-to-end anastomosis was also performed in one plane using Vicryl suture 00. There were no lymphadenopathies in the mesentery; the rest of the intestine and abdominal organs were seen as normal. Two units of red blood cells were transfused in the operating room. There were no complications, and the patient was discharged from the hospital on the sixth day.

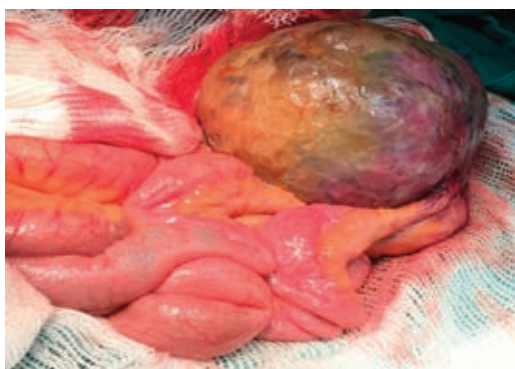


Figure 1: Tumor in a Meckel's diverticulum 90 cm from the ileocecal valve.

The pathological anatomy was as follows:

- Macroscopic examination of the surgical specimen: segment of small intestine (ileum) 16 centimeters long, surgical limits of 7 centimeters on each side of the tumor. The tumor reached a main diameter of 10 cm.
- Histopathology examination: GIST, weakly positive. Actin negative (meaning negative for leiomyoma or leiomyosarcoma). Tumor cells positive in 30%. Mitosis grade of less than 5 per 50 fields.
- Conclusion: GIST in a Meckel's diverticulum.

Despite being a localized and intermediate risk disease (size between 5-10 cm and mitosis of less than 5 per 50 fields of higher magnification), it was decided to give adjuvant treatment with tyrosine kinase inhibitors (imatinib mesylate). The indicated dose was 400 mg every 24 hours orally for three years; 12 months after surgical resection, the patient was free of oncologic disease.

DISCUSSION

GIST (gastrointestinal stromal tumor) is a term introduced by Mazur and Clark in 1983; it is identified as the most common mesenchymal tumors of the digestive tract and represents 0.1-1% of malignant tumors. In order of frequency, it affects the stomach (60-70%), small intestine (20-30%), rectum (3%), colon (2%), and esophagus (1%). It is seen more

infrequently in the appendix, gallbladder, omentum, mesentery, peritoneum, liver, pancreas, ovaries, uterus, retroperitoneum, and in paravaginal and periprostatic tissues.^{7,8} In most patients they occur between 50 and 80 years of age, being more frequent in male patients. Our case is different since it occurred in a young woman.

Because their growth is usually extraluminal and their consistency is soft, these tumors do not produce intestinal transit obstruction, so they are asymptomatic at the onset of the disease.^{6,8} Symptoms appear in tumors larger than 5 cm. The clinical presentation varies according to the anatomical location of the tumor, its size, and degree of malignancy. The most common symptom is mild abdominal pain that worsens later. Other symptoms are nausea, vomiting, anorexia, weight loss, presence of a palpable abdominal mass, weight loss, hemorrhage, tumor perforation, and peritonitis.

Gastrointestinal bleeding can be acute (melena or hematemesis), occult, or chronic producing anemia, due to ulceration of endoluminal growth tumors. In our patient the lower gastrointestinal bleeding was of great severity leading to hypovolemic shock. This was probably caused by its location in a Meckel's diverticulum. When reviewing the literature, no similar case combining both elements was found. The presence of mesenteric nutritional vessels crossing the ileum to distribute themselves in the tumor mass -which was observed in the case presented- is characteristic of malignant tumors of the small intestine (Figure 2).

Meckel's diverticulum is seen on a computerized tomographic (CT) scan as a cul-de-sac image, usually with fluid, air or particulate material. On an ultrasound study, in patients with bleeding, a hyperechogenic tubular structure like a cyst is usually seen.^{2,4,5} In the case presented, the ultrasound performed assisted in a possible preoperative diagnosis.

Treatment of GIST depends on the extent and prognosis of the disease. This is based on the risk classification described in 2002 by C.D. Fletcher,⁹ according to the size of the primary tumor and its mitosis rate. This is distributed as follows:

- Very low risk: less than 2 cm, mitotic rate less than 5 per 50 fields of higher magnification.
- Low risk: between 2-5 cm, and mitotic rate less than 5 per 50 fields of higher magnification.
- Intermediate risk: size less than 5 cm, mitoses 6-10 per 50 fields of higher magnification, or size between 5-10 cm and mitoses less than 5 per 50 fields of higher magnification.
- High risk: larger than 5 cm and mitosis rate less than 5 per 50 fields of high power, or 10 or more cm and any mitotic rate, or any size and mitoses of more than 10 per 50 fields of higher magnification.

Other factors would be histologic subtypes, the degree of cellular pleomorphism, and the patient's age. For a well localized tumor at low risk, surgical resection would be sufficient. However, for the rest of the tumors, adjuvant treatment is recommended to prevent the action of the KIT receptor, as well as to stop cell division and prevent dissemination.⁷⁻¹¹ Although in our case it was a localized malignant disease without extension to other organs and a local surgical resection was performed with a good oncologic margin (2 centimeters accepted) and a low rate of proven mitosis, it was a bulky tumor, which was diagnosed during a complication (digestive bleeding). Consequently, it was classified as high risk due to its size, so adjuvant therapy with imatinib mesylate administration was



Figure 2: *The mesenteric nutritional vessels crossing the ileum towards the tumor are observed.*

decided. The good evolution of the case accredited this decision.

CONCLUSION

The presence of a GIST in a Meckel's diverticulum, although rare, has been reported in the medical literature, but not in a Meckel's diverticulum with severe lower gastrointestinal bleeding and hypovolemic shock. Timely diagnosis with radical surgical-oncologic treatment without the presentation of postoperative complications, complemented with adjuvant treatment with imatinib mesylate has allowed a recurrence-free survival.

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Ethical considerations and responsibility:

Data privacy. In accordance with the protocols established at the authors' work site, the authors

declare that they have followed the protocols on patient data privacy and preserved their anonymity. The informed consent of the patient referred to in the article is in the possession of the author.

Funding: No financial support was received for this study.

Disclosure: The authors declare that there is no conflict of interest in this study.

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