ACG CASE REPORTS JOURNAL

CASE REPORT | SMALL BOWEL

Unusual Upper Gastrointestinal Bleeding Due to a Neuroendocrine Tumor Arising in Meckel Diverticulum

Katherine Elizabeth Eberly, MD[1], Jasmin Martinez, BS[2], and Nitin Sardana, MD[1]

[1]*Inova Fairfax Medical Campus, Fairfax, VA*
[2]*College of William and Mary, Williamsburg, VA*

ABSTRACT

Meckel diverticulum is the most common congenital variation of the gastrointestinal tract arising from incomplete obliteration of the vitelline duct during gestation. In most cases, individuals are asymptomatic. This is a case of a 38-year-old patient with hematochezia in whom Meckel diverticulum was diagnosed. A mass was identified within the diverticulum. Histopathological and immunohistochemical studies revealed a well-differentiated neuroendocrine tumor. The development of tumors in Meckel diverticulum is rare, and when identified, only 0.5%–3.2% of these tumors are found to be malignant. Furthermore, gastrointestinal bleeding is an infrequent clinical feature of neuroendocrine tumors. A unique aspect of this case was that numerous imaging studies and endoscopic procedures were unable to definitively identify the presence of Meckle diverticulum and underlying neuroendocrine tumor. Through a high clinical suspicion and collaboration with surgical colleagues, an exploratory laparotomy was performed, which ultimately led to the identification and diagnosis of the underlying pathology.

KEYWORDS: gastrointestinal bleeding; Meckel diverticulum; neuroendocrine tumor

INTRODUCTION

Carcinoids are the most common neuroendocrine tumors of the gastrointestinal tract; however, they rarely appear in a Meckel diverticulum.[1,2] In fact, they occur only in 0.74% of patients.[3] These tumors typically manifest as a long history of vague abdominal symptoms and rarely cause gastrointestinal bleeding. We present a case of a carcinoid tumor arising from a Meckel diverticulum that manifested as upper gastrointestinal bleeding. It was managed with resection of the small bowel, including the diverticulum, with resolution of the bleeding.

CASE REPORT

A 38-year-old man presented to the hospital with 1 day of painless hematochezia, lightheadedness, and dyspnea. The night before presentation, he experienced 4 episodes of bright red blood per rectum. He denied abdominal pain, nausea, vomiting, dysphagia, or anorexia. Six weeks before admission, he underwent a complete physical examination and basic laboratory evaluation, including a complete blood count and basic metabolic panel, which were unremarkable. He had no history of anemia or gastrointestinal bleeding. The patient noted a history of acid reflux for which he occasionally took omeprazole. He denied nonsteroidal anti-inflammatory drug use. He had not undergone an endoscopic evaluation before presentation. He denied alcohol or tobacco use. On arrival, the blood pressure was 92/54 mm Hg and heart rate was 98 beats per minute. Physical examination was notable for pallor and bright red blood visible on rectal examination. Laboratory tests demonstrated a hemoglobin of 8.8 g/L, hematocrit of 25.9%, and mean corpuscular volume of 92.2 fL. Abdominal and pelvic computed tomography (CT) with contrast did not demonstrate any significant abnormality. A colonoscopy was then performed, which showed blood in the terminal ileum and maroon blood throughout the entire colon. An esophagogastroduodenoscopy did not reveal a source of bleeding. Anterograde balloon enteroscopy showed multiple non-bleeding petechiae in the jejunum. A technetium Tc 99 m pertechnetate scintigraphy to assess for Meckel diverticulum was negative. A video capsule endoscopy (VCE) did not show active bleeding, although it suggested a possible Meckel diverticulum in the distal ileum. The patient continued to have maroon blood per rectum and required 4 units of blood during his hospital stay. There was high suspicion for Meckel diverticulum despite numerous unrevealing imaging studies, so an exploratory

ACG Case Rep J 2024;11:e01345. doi:10.14309/crj.0000000000001345. Published online: April 18, 2024

Correspondence: Katherine Elizabeth Eberly, MD (katherine.eberly@inova.org).

laparotomy was performed. Intraoperatively, a Meckel diverticulum with an intraluminal mass was found. A segment of the small bowel including the diverticulum was resected. Macroscopic evaluation of the mass showed a 1.4 × 1.2 × 0.9 cm tumor at the base of the Meckel diverticulum. Surgical pathology revealed a well-differentiated, low-grade neuroendocrine tumor (carcinoid) within the Meckel diverticulum. There was no further evidence of bleeding, and the patient was discharged after an uneventful postoperative recovery.

DISCUSSION

Meckel diverticulum is a vestigial remnant of the vitelline duct that fails to obliterate around the eighth week of gestation. It is the most common congenital variant of the gastrointestinal tract, present in around 2% of the population. Generally, these diverticula are asymptomatic incidental findings requiring no intervention. The development of tumors within Meckel diverticula is rare, and when identified only 0.5%–3.2% of these tumors are malignant.[1,2] Carcinoid tumors make up 33% of these malignant tumors.[4,5] This was supported by a study conducted by Modlin et al, which examined 13,715 patients diagnosed with carcinoid tumors between 1950 and 1990. They found that only 0.74% of these carcinoid tumors developed within the Meckel diverticulum.[6] In a 1997 literature review, Sutter et al identified only 111 reported cases of carcinoid tumor arising in the Meckel diverticulum.[7] The rarity of this tumor makes it challenging to predict the clinical presentation and outcome. Furthermore, gastrointestinal bleeding is an infrequent clinical feature of neuroendocrine tumors in adults. While gastrointestinal bleeding is a known complication associated with Meckel diverticulum (11.8% of complications), this is usually caused by heterotopic mucosa, specifically gastric mucosa.[8] Symptoms of intestinal carcinoid tumors can include vague abdominal pain, intestinal obstruction, and, as in this case, gastrointestinal bleeding. In 10%–20% of carcinoid tumor cases associated with Meckel diverticulum, patients develop carcinoid syndrome with episodes of skin flushing, diarrhea, hepatomegaly, and cardiac lesions. Given the nonspecificity of these symptoms, the average time between symptom onset and diagnosis ranges from 2 to 20 years.[9] It is notable that multiple imaging studies did not conclusively reveal the Meckel diverticulum present in this patient. The role of VCE in diagnosing Meckel diverticulum has not been well studied, and there are no clear data on the sensitivity of this modality for this purpose. Slobodan et al examined 157 patients from China with occult bleeding who received VCE as part of their workup and were able to identify the source of bleeding in 70 of 157 patients (44.6%). Of those 70 patients with an identified bleeding, 13 were diagnosed with Meckel diverticulum by the presence of a double-lumen sign or visible blood. The study suggests that VCE has a positive predictive value of 84.6% for the diagnosis of Meckel diverticulum.[10] Technetium Tc 99 m pertechnetate scintigraphy is the first-line imaging study for diagnosis of Meckel' diverticulum, revealing an accumulation of isotopes in the right lower quadrant of the abdomen when ectopic gastric mucosa is present within the diverticulum. Radioisotope scintigraphy was negative in this patient as his gastrointestinal bleeding was associated with the carcinoid tumor as opposed to ectopic gastric mucosa. Alternative diagnostic methods for Meckel diverticulum include Tc99m red blood cells labeled scan and CT enterography, though these modalities are limited by low sensitivity (>0.1 and 0.5 mL/min) unless a brisk bleeding is present. On CT, they can present as a fluid or gas-filled pouch that is continuous with small bowel loops. In uncomplicated patients, it is difficult to distinguish Meckel diverticulum from normal small bowel. Ultimately, when imaging is unrevealing and a high suspicion for Meckel diverticulum remains, exploratory laparotomy may be the best option as was the case for this patient.

DISCLOSURES

Author contributions: KE Eberly wrote the initial draft and is the article guarantor. J. Martinez and N. Sardana wrote and edited the manuscript. All authors approved the final version.

Financial disclosure: None to report.

Informed consent was obtained for this manuscript.

Received September 26, 2023; Accepted March 19, 2024

REFERENCES

1. Thirunavukarasu P, Sathaiah M, Sukumar S, et al. Meckel's diverticulum—A high-risk region for malignancy in the ileum. Insights from a population-based epidemiological study and implications in surgical management. *Ann Surg.* 2011;253(2):223–30.
2. Dumper J, Mackenzie S, Mitchell P, Sutherland F, Quan ML, Mew D. Complications of Meckel's diverticula in adults. *Can J Surg.* 2006;49(5):353–7.
3. Modlin IM, Lye KD, Kidd MA. A 5-decade analysis of 13,715 carcinoid tumors. *Cancer.* 2003;97(4):934–59.
4. Dixon AY, McAnaw M, McGregor DH, Keushkerian S, Miller LK, Pierce PD. Dual carcinoid tumors of meckel's diverticulum presenting as metastasis in an inguinal hernia sac: Case report with literature review. *Am J Gastroenterol.* 1988;83(11):1283–8.
5. Weistein EC, Docherty MB, Waugh JM. Neoplasms of Meckel's diverticulum. *Int Abstr Surg.* 1963;116:103–11.
6. Modlin M, Sandor A. An analysis of 8305 cases of carcinoid tumors. *Cancer.* 1997;79(4):813–29.
7. Sutter PM, Canepa MG, Kuhrmeier F, Marx A, Martinoli S. Carcinoid tumor in Meckel's diverticulum: Case presentation and review of the literature. *Schweiz Med Wochenschr Suppl.* 1997;89:20S–24S.
8. Kabir SA, Raza SA, Kabir SI. Malignant neoplasms of Meckel's diverticulum; an evidence based review. *Ann Med Surg (Lond).* 2019;43:75–81.
9. Carpenter SS, Grillis M. Meckel's diverticulitis secondary to carcinoid tumor: An unusual presentation of the acute abdomen in an adult. *Curr Surg.* 2003;60(3):301–3.
10. Slobodan NK, Martinov JB, Sokic-Milutinovic AD, et al. Capsule endoscopy is a useful diagnostic tool for diagnosing Meckel's diverticulum. *Eur J Gastroenterol Hepatol.* 2016;28:702–7.