A rare presentation of gastrointestinal stromal tumor arising from a meckel’s diverticulum: A case report

Aditya Patel, Meenakshi Yeola, Pankaj Gharde

Department of General Surgery, Jawaharlal Nehru Medical College, Sawangi (Meghe), Wardha, Maharashtra, India

Abstract

The most common congenital gastrointestinal tract abnormality is Meckel’s diverticulum, affecting 1% to 2% of the population. Meckel’s diverticulum can sometimes house a gastrointestinal stromal tumor (GIST), which is a rare malignant tumor. A 63-year-old man presented with occult melena. It turns out to be a GIST developing from Meckel’s diverticulum after an exploratory laparotomy. Occult melena is one of the causes of unexplained and intractable anemia. Every family and general physician should keep in mind the possibility of occult blood loss via the gastrointestinal tract when treating the patient.

Keywords: Bleeding, diverticula, GIST, Meckel's diverticulum, melena, occult

Introduction

Meckel's diverticulum is the most common congenital gastrointestinal tract malformation that affects 1% to 2% of the population.[1] Majority of it remains asymptomatic.[3] Surgical removal indicated only on complication or malignant conversion. Malignant tumors developing from Meckel’s diverticulum are extremely uncommon, accounting for about 0.5% to 3.2% of all Meckel diverticula. Among these, 12% of malignant tumors are gastrointestinal stromal tumors (GIST).[1] Malignant tumor of Meckel's diverticulum is one of the causes of GI bleed leading to intractable anemia. General and family physicians are the first line of contact for anemia and any unexplained and intractable anemia should raise suspicion. We are reporting such a rare incidence, where we got GIST arising from the Meckel diverticula.

Case Presentation

A 63-year-old male presented with 15 days history of melena. On admission, physical examination showed a pulse rate of 90/min, blood pressure of 90/66 mmHg, and respiratory rate of 18/min. Abdominal examination revealed no significant findings. The hemoglobin was 5 g/dl. He had a packed red cell transfusion of 5 units. An ultrasound of the abdomen revealed a suprapubic tumor of unclear origin. There was no sign of upper gastrointestinal hemorrhage on upper GI endoscopy, and colonoscopy revealed normal mucosa throughout the colon and a normal ileocecal junction with terminal ileum. The patient was next exposed to the CECT abdomen pelvis, which revealed an e/o 6.6 × 4.4 × 5.3 cm well-defined heterogeneously enhancing lesion emanating mostly from the ilium, with a substantial extra-luminal component and a little intra-luminal component [Figure 1].

The patient had a laparotomy, and the intraoperative observations were s/o large brownish tumor, with irregular lobulated surface, arising from diverticulum of ileum which was situated 60 cm from the ileocecal junction [Figure 2]. The patient underwent resection and primary anastomosis with a...
The post-operative time went very smoothly.

The histopathological examination revealed a submucosal tumor with well-defined margins that had spread to the serosa, with negative vascular and lymphatic invasion. Fascicules of spindly cells with epithelioid morphology make up the tumor [Figure 3a]. Mitotic figures were spotted on rare occasions, with an average count of 5/50 high power fields [Figure 3b]. The immunohistochemical (IHC) stain CD-117 was positive. It was classified as a mixed type GIST in Meckel’s diverticulum.

**Discussion**

During the 5<sup>th</sup> to 7<sup>th</sup> week of fetal life, the omphalomesenteric (vitelline) duct is generally obliterated. The Meckel diverticula are the outcome of the obliteration failing. This is a real diverticulum, with all three layers of the intestinal wall present.<sup>[3]</sup> Meckel diverticula are usually seen on the small bowel’s antimesenteric border around 70 to 100 cms from the ileocecal valve.<sup>[2]</sup> Heterotopic mucosa such as gastric, duodenal, ileal, colonic, pancreatic, Brunner’s gland, and hepatobiliary system line Meckel diverticula. The gastric mucosa is the most prevalent, followed by pancreatic mucosa.<sup>[1,2]</sup> We found gastric mucosa as heterotrophic mucosa in our case.

The majority of it remains asymptomatic and may incidentally found intra-op. Bleeding, blockage, diverticulitis, perforation, intussusception, ulceration, and in rare cases, the presence of cancer inside the Meckel diverticulum can all cause symptomatic diverticula. Malignant tumors developing from Meckel’s diverticulum are extremely uncommon, accounting for about 0.5% to 3.2% of all Meckel diverticula.<sup>[1,4]</sup> Among them, most are benign like leiomyoma, lipoma, adenoma, angiomat, and malignant including adenocarcinoma, sarcoma, carcinoid tumor, lymphoma, and GIST.<sup>[2,5]</sup> In our case, the patient came with a complaint of melena.

GISTs are very rare and account for 0.1% to 1% of gastrointestinal tumors.<sup>[5]</sup> GISTs are described as pleomorphic mesenchymal tumors of the gastrointestinal tract that exhibit the tyrosine kinase receptor (KIT) proteins CD 117 and CD 34 on immunohistochemistry.<sup>[6]</sup> GISTs originate from the Cajal’s interstitial cells, which are the digestive tract’s pacemakers.<sup>[1,7]</sup>

The stomach is the most common location for GIST, followed by the small intestine, colon, rectum, and esophagus. Malignant GIST arises from the mesenchymal tissue of the GI tract which constitutes approximately 20% of the malignant neoplasm of the small intestine.<sup>[1,8]</sup> When we talk about the intestine, jejunum and ileum are the most common sites for these tumors. These tumors are typically diagnosed around 5<sup>th</sup> and 6<sup>th</sup> decades of life.<sup>[8]</sup> GIST occurrence is slightly male predominant.<sup>[9]</sup> In 80% of patients, the malignant GISTs were bigger than 5 cm at the time of diagnosis.<sup>[10]</sup> We had similar findings as the patient being 63-year-old male with 6 cm of GIST at Meckel diverticulum.

GISTs are most commonly seen in the muscularis propria and grow extramurally. Bleeding per rectum and obstruction are the most common indications for surgical intervention. When hemorrhagic necrosis occurs in large tumor masses, it leads to free
perforation. GISTs often invade locally and spread through direct extension into surrounding tissues and hematogenous to the liver, lungs, and bone; lymphatic metastases are uncommon. For the survival and the risk for metastasis, the most useful indicators are the size of tumor at the time of presentation, evidence of tumor invasion into the lamina propria, and mitotic index. The mucosal ulceration causes occult GI bleeding which is the most common clinical presentation of symptomatic GIST.

Computed tomography is usually an adequate investigation to diagnose and locate the intra-abdominal tumor. Gastrointestinal schwannomas are the most common differential diagnosis that show homogeneous attenuation on CT scan, making them easy to distinguish from large benign or malignant GIST, which shows heterogeneous enhancement owing to necrosis, hemorrhage, and intracellular cystic alterations.

GIST becomes symptomatic when it reaches a large size or is placed at a critical anatomical location like small bowel. However, GIST from Meckel diverticula comes with the presentation which includes abdominal pain, gastrointestinal bleeding, acute bowel obstruction, weight loss, weakness, and very rare with perforation. In our case, the patient came with a complaint of passing black stool associated with on and off pain in the abdomen.

Surgical resection is the therapy of choice for symptomatic Meckel’s diverticulum. When there is palpable ectopic tissue, intestinal ischemia, perforation, or any tumor like GIST, this should be done either by diverticulectomy or segmental bowel resection and anastomosis. Because these tumors do not exhibit any lymph node metastases, lymphadenectomy is typically not necessary.

GIST presents a wide range of histopathological findings, with epithelioid (70%) and spindle cell histology (30%). In this case, the tumor composed of a mixed variety of spindle cells with epithelioid morphology.

Tumor size <10 cm, the absence of metastases, a low mitotic index, and full excision of the tumor with a negative microscopic margin and no intraperitoneal spread are all prognostic markers. Regardless of the mitotic index, the tumor’s placement in the small intestine and its size of more than 5 cm indicates that it has a moderate malignant potential.

The tyrosine kinase receptor (KIT) protein functions as a transmembrane receptor tyrosine kinases (RTK) in general; immunohistochemical (IHC) staining can establish the CD117 antigen as a KIT protein marker. It is evident from IHC studies that KIT mutations are found in GIST in greater than 90% of cases. Imatinib is a tyrosine kinase inhibitor that has been authorized to treat high-risk GIST patients and is also a drug of choice for unresectable or metastatic GIST.

Computed tomography scans should be performed every 3 to 6 months on individuals who had their GIST surgically removed to monitor for metastatic or recurring illness. In patients with clearly unresectable or metastatic disease, CT should be performed within 3 months after starting tyrosine kinase inhibitor as neoadjuvant therapy.

We began adjuvant imatinib 400 mg daily for 3 years and followed up every 3 months with a CT scan every 6 months for monitoring.

**Conclusion**

Occult melena is one of the causes of unexplained and intractable anemia. Every family and general physician should keep in mind the possibility of occult blood loss via the gastrointestinal tract when treating the patient. Sometimes, patient presenting with a pelvis or abdominal mass of suspected bowel origin in which imaging technique could not point out the diagnosis, neoplasm arising from the Meckel diverticula should be kept in mind as a differential diagnosis.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Chandramohan K, Agarwal M, Gurjar G, Gatti RC, Patel MH, Trivedi P, et al. Gastrointestinal stromal tumor in Meckel’s diverticulum. World J Surg Oncol 2007;5:50.
2. Koppad SN, Sonawane SR, Kapoor VB, Deshmukh AM, Borole KA. A rare case of gastrointestinal stromal tumor from Meckel’s diverticulum: Images for surgeons. ANZ J Surg 2013;83:184-5.
3. Manerikar K, Ali I, Patil C, Dholakia M, Mody P. A rare entity of gastrointestinal stromal tumour arising from a Meckel’s diverticulum: A case report. J Gastrointest Cancer 2018;49:351-4.
4. Soltero MJ, Bill AH. The natural history of Meckel’s diverticulum and its relation to incidental removal. Am J Surg 1976;132:168-73.
5. Yamaguchi M, Takeuchi S, Awasu S. Meckel’s diverticulum. Am J Surg 1978;136:247-9.
6. Mohanraj T, Hanif H, Zainal AA. Bleeding Jejunal GIST: An uncommon cause of gastrointestinal bleeding. Med J Malaysia 2015;70:31-2.
7. Mazur MT, Clark HB. Gastric stromal tumors reappraisal of histogenesis: Am J Surg Pathol 1983;7:507-20.
8. DeMatteo RP, Lewis JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF. Two hundred gastrointestinal stromal tumors: Recurrence patterns and prognostic factors for survival. Ann Surg 2000;231:51.

9. Mitura K, Blicharz P, Romaszcuk M. Perforated gist of Meckel's diverticulum. Pol J Surg 2012;84:258-61.

10. Sabiston Textbook of Surgery. 20th ed. Available from: https://www.elsevier.com/books/sabiston-textbook-of-surgery/townsend/978-0-323-29987-9. [Last accessed on 2021 Dec 06].

11. Daldoul S, Moussi A, Triki W, Baraket RB, Zaouche A. Jejunal GIST causing acute massive gastrointestinal bleeding: Role of multidetector row helical CT in the preoperative diagnosis and management. Arab J Gastroenterol 2012;13:153-7.

12. Sagar J, Kumar V, Shah DK. Meckel's diverticulum: A systematic review. J R Soc Med 2006;99:501-5.

13. Demetri GD, Benjamin RS, Blanke CD, Blay J-Y, Casali P, Choi H, et al. NCCN task force report: Management of patients with gastrointestinal stromal tumor (GIST)—update of the NCCN clinical practice guidelines. J Natl Compr Canc Netw 2007;5:S-1-29.