Gastrointestinal Stromal Tumor Arising from Meckel's Diverticulum: A Cause of Obscure GI Bleeding

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Abstract: <u>Background</u>: Meckel's Diverticulum is the most commonly encountered congenital anomaly of the small intestine, occurring in approximately 2% of the population. Occasionally Meckel's diverticulum harbors neoplasms. <u>Case presentation</u>: A 65 year old male, presented with complaints of intermittent malena since 4 months. On exploratory laparotomy, it turned out to be gastrointestinal stromal tumour (GIST) arising from Meckel's diverticulum. Short history and review of literature are discussed. <u>Conclusion</u>: A Gastrointestinal Stromal Tumor occurring from Meckel's diverticulum, even though rare, should be considered as differential diagnosis of Obscure Gastro Intestinal bleeding arising from bowel, wherever imaging modalities fail to give a definitive diagnosis.

Keywords: GIST -GASTRO INTESTINAL STOMAL TUMOUR

1. Background

Meckel's diverticulum, the most commonly encountered congenital anomaly of the small intestine, affects 2% of the population [1,2]. The vast majority of Meckel's diverticulae are incidentally discovered during autopsy, laparotomy, or barium studies [3]. Meckel's diverticulum is surgically removed only when a complication arises or a neoplasia develops. The tumors are infrequent and observed only in 0.5-3.2% of the Meckel's diverticula. Of these, 12% tumors are GIST. We are reporting one such incidence, where we came across a Meckel's diverticulum harboring GIST.

2. Case Presentation

- A 65 Yr old male presented with complaints of intermittent Malena on &off since 3mnths.Last epidose 3 days back.
- c/o colicky lower abdominal pain since 3 days.H/o loss of appetite , No hematemesis,jaundice,fever.No medical comorbidities.
- Known Alcoholic .Not a known smoker.
- Patient obese, pallor with tachycardia.
- Abdominal Examination un remarcable.
- PR –Examination –No e/o Hemorrhoids /Fissures, No Active bleed.

Investigations done

- Hb -6.4 %, Pcv -25%.
- RFT NORMAL
- VIRALS –Non reactive.
- Endoscopy and Colonscopy were Normal
- CECT ABD 3.5 x 5.1 x 4.4 cms peripherally enhancing thick walled hypodense lesion of fluid attenuaton noted in pelvis ,indenting the distal ileal loops with peri-lesional fat stranding . Loss of fat planes with sigmoidal colon with minimal wall thickening is noted. Fat planes with rt lateral wall of Urinary Bladder well maintained . Wall of lesion is thin at the site of indentaton with ileum.



Figure 1

Case posted for surgery

- Exploratory laportomy done
- Intra op findings –Dense adhesions between Terminal Ileum, Sigmoid Colon & Urinary bladder noted.
- Careful Adhesiolysis done.
- A 5x2 cm diverticulum identified in theTerminal Ileum about 2 feet from IC Junction in Antimesenteric side.
- Segmental resection of Ileal segment with Diverticulum with Primary anastamosis done.
- Specimen sent for HPE
- 2Units of packed red blood cell transfusion done.
- Patient was discharged on 6 th post op day.
- Preoperative CECT scan showing Chronic abscess due to sealed off ileal perforation/inflammatory bowel disease.



Figure 2

Intraoperative picture showing tumour arising from Meckel's diverticulum; Tumour was located in Meckel's diverticulum

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Figure 3

Intraoperative picture showing tumour and Meckel's diverticulum; Picture of Meckel's diverticulum with part of tumour, which was delivered out and primary anastamosis • HPE -- Report -- GIST /GANT.

- The tumour shows fragments of viabe ileal mucosa with spindle cell tumor involving the muscularis propria. (figure -4).



Figure 4

High power view of the histopathology slide; Tumour cells shows fragments of viable ileal mucosa with spindle cell tumor involving muscularis propria .(figure-5) and tumour exhibits fascicular growth pattern with interlacing of bundles and whorling at areas (figure-6).mitosis sparse.most of the tumor part of has under gone necrosis with superadded secondary inflammation (figure-7).



Figure 5

- Advice Immuno-hisochemistry for confirmation includes
- Infammatory myofiboblastic tumor (IMT)
- GIST -CD 117
- GANT-S100
- IMT -Vimentin

Histopathology slide after Immunostaining for C- 117, which suggests GIST.



CD 117 -negative

Figure 6

3. Discussion

Meckel's diverticulum is the most commonly encountered congenital anomaly of the small intestine, occurring in approximately 2% of the population [<u>1,2</u>]. Meckel's diverticulum is located on the antimesentric border of the ileum approximately 45 to 60 cm proximal to the ileocecal valve and results from incomplete closure of the omphalomesentric, or viteline duct. An equal incidence is found among men and women. Heterotopic mucosa is present in Meckel's diverticulum, the most common of which is gastric mucosa (present in 50% of all Meckel's diverticula). Pancreatic mucosa is encountered in approximately 5% of diverticula. Similarly these diverticula may habour colonic mucosa.

The vast majority of Meckel's diverticula are incidentally discovered during autopsy, laparotomy, or barium studies. The most common clinical presentations of the Meckel's

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diverticulum are gastrointestinal bleeding (from chronic acid-induced ulcer in the ileum adjacent to a Meckel's diverticulum that contains gastric mucosa), intestinal obstruction, and diverticulitis. Incidence of tumours within the Meckel's diverticulm is 0.5 to 3.2% [4-6]. Most of them are commonly benign tumours like leiomyomas, angiomas, and lipomas. Malignant neoplasms include adenocarcinoma (which commonly originate from the gastric mucosa), sarcoma, carcinoid tumour and GIST.

GISTs are rare neoplasms which account for 0.1-1% of gastrointestinal malignancies. The term itself was first used in 1983 by Mazur and Clark [7] to identify a heterogeneous group of tumours, all of them histologically characterized by hyperplastic fused cells, not necessarily leiomuscular ones, but even neural ones. Gastrointestinal stromal tumours arise from the interstitial cells of Cajal, pace maker cells of Gastrointestinal tract [8].

GIST occurs predominantly in adults at a median age of 58 years. The majority of GISTs (60% to 70%) have been reported to arise in the stomach, whereas 20% to 30% originate in the small intestine, and less than 10% in the esophagus, colon and rectum. GISTs also occur in the extra-intestinal abdominopelvic sites such as the omentum, mesentery, or retroperitoneum [9-11]. GISTs arising from Meckel's diverticulum are extremely rare [12-16].

For many patients, the detection of GIST may be an incidental finding during evaluation of nonspecific symptoms. Symptoms tend to arise only when tumours reach a large size or are in critical anatomic location. Most symptomatic patients present with tumours larger than 5 cm in maximal dimension. Symptoms at presentation may include abdominal pain, abdominal mass, nausea, vomiting, anorexia, and weight loss. The vast majority of metastatic GISTs are located intraabdominal, either in the liver, in the omentum, or in the peritoneal cavity [9]. Metastatic spread to lymph nodes and to other regions via lymphatics is very rare.

CT is usually an adequate technology to diagnose tumours arising from Meckel's diverticulum as long as appropriate techniques for both noncontrast and intravenous contrast administrations are used [13]. Histopathologically, disease exhibits a wide variety of appearances with characteristics of either epitheloid (approximately 70%) or spindle cell histology (remaining 30%). Normally, the KIT protein serves as a transmembrane RTK; the CD117 antigen can be detected by immunohistochemical staining as marker for the presence of the KIT protein. With the use of sophiscated technology, it has become clear that KIT mutations can be noted in more than 90% of GIST cells.

CD34 expression is not specific for GIST, because it can also be noted in desmoid tumours, and approximately 60% to 70% of GIST lesions are positive for CD34 [<u>17-19</u>]. Expert analysis of the KIT and PDGFRA genotype may be useful to define with certainty the group of rare patients with CD117-negative GISTs in the future. GIST is considered to be potentially malignant tumours. [<u>19</u>].

Most reliable prognostic factors are the size of the primary tumour and the mitotic index which measure proliferative activity of the cells. Other prognostic factors are specific histologic subtypes (epitheloid vs. spindle cell), the degree of cellular pleomorphism and age of the patient. Recurrence and survival rates have also been reported to correlate with the location of the primary GIST lesion, with small bowel tumours showing a somewhat worse prognosis. The functional imaging in GISTs with FDG-PET can give additional information that can assist clinicians in the management of patients. Definitive surgery remains the mainstay of treatment for patients with localized, primary GIST.

4. Conclusion

Neoplasms arising from Meckel's diverticulae are differential diagnoses in patients presenting with Obscure Gastro intestinal bleeding of bowel origin, in which imaging modalities doesn't pinpoint a definitive diagnosis. Eventhough it is not so common, one should keep in mind this differential diagnosis in pelvic masses of suspected bowel origin.

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