Gastrointestinal Stromal Tumour of Meckel's Diverticulum Causing Perforation and Peritonitis: A Case Report

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Abstract A 23-year-old man presented with history of fever and pain in the right lower abdomen. He was diagnosed to have acute appendicitis with perforation and was taken up for emergency laparotomy after resuscitation. There was pus in the right iliac fossa and a tumor at the fundus of Meckel's diverticulum, which had perforated. Appendix was found to be normal. The histopathology examination showed it to be gastrointestinal stromal tumor of Meckel's diverticulum. The gastrointestinal stromal tumor causing Meckel's diverticulum perforation and peritonitis is very rare, and prior to this indexed case, only two such cases have been reported in the literature.

Keywords Meckel's diverticulum · Diverticulum · Gastrointestinal stromal tumor

Introduction

Meckel's diverticulum is the most commonly encountered congenital anomaly of the small intestine. The vast majority of Meckel's diverticula are incidentally discovered during autopsy, laparotomy, or barium studies. Tumors are observed in only 0.5–3.2 % of Meckel's diverticula, but gastrointestinal stromal tumors are rare in this location. Still rare is gastrointestinal stromal tumor causing Meckel's

diverticulum perforation and peritonitis. Here we present one such case.

Case History

A 23-year-old man presented with history of fever and pain in the right lower abdomen for the past 2 days. On examination, we found the abdomen mildly distended with tenderness and guarding. There was a vague palpable mass in the right lower abdomen. His general physical examination revealed tachycardia (pulse rate 130/min). The rest of systemic examination did not reveal any abnormality. Clinical diagnosis of acute appendicitis with local peritonitis was made.

He was taken up for emergency laparotomy. The operative findings were pus in the right iliac fossa and a large tumor at the fundus of Meckel's diverticulum, which had perforated. He underwent segmental ileal resection with Meckel's diverticulectomy and end-to-end ileoileal anastomosis. Grossly, the specimen consisted of 6.2 cm long ileal segment with attached Meckel's diverticulum on the anti-mesenteric border, measuring 4.8 cm×4.2 cm×4.2 cm. The serosal aspect of the diverticulum showed hemorrhages and exudate (Fig. 1). On slicing, almost whole of the diverticulum was replaced by the tumor except for partly ulcerated mucosa. The tumor was solid gray white with foci of hemorrhages. The adjoining ileum was unremarkable except for serosal congestion.

Microscopic examination showed partly ulcerated small bowel mucosa in the diverticulum with an underlying tumor. The tumor was composed of fascicles and focal whorling arrangement of spindle cells (Fig. 2). The cells showed minimal pleomorphism with only occasional mitotic figures (<5/50 hpf). There was evidence of perforation with acute

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serositis. Immunohistochemistry was carried out. The tumor cells showed strong positivity for CD117 (c-kit), CD 34, smooth muscle actin, and focally for S-100. Based on the morphological and immunohistochemistry findings, the diagnosis made was gastrointestinal stromal tumor arising in Meckel's diverticulum and causing perforation and peritonitis.

Discussion

Meckel's diverticulum (MD) is the most commonly encountered congenital anomaly of the small intestine, occurring in approximately 2 % of the population [1]. The lining of MD is predominantly of the small intestinal type, identical to that of adjacent bowel; however, gastric, duodenal, or colonic mucosa may also be encountered, usually near the tip [2].

The vast majority of Meckel's diverticula are incidentally discovered during autopsy, laparotomy, or barium studies. The most common clinical presentations of the MD are gastrointestinal bleeding, intestinal obstruction, and diverticulitis [1]. Incidence of tumors within MD is 0.5–3.2 % [3]. Among these, the carcinoid tumor predominates, either alone or rarely associated with carcinoid tumors of the similar histologic type in the small bowel. Other tumors include villous adenoma, adenocarcinoma, gastrointestinal stromal tumors (GIST), leiomyoma, leiomyosarcoma, and malignant melanoma [2]. Only a few definite cases of GIST occurring in MD are reported [1, 4–7].

GISTs account for approximately 0.1–3 % of all gastrointestinal neoplasms. The majority of GISTs (60–70 %) have been reported to arise in the stomach, whereas 20–

Fig. 1 Gross photograph showing ileal segment with Meckel's diverticulum

Fig. 2 Photomicrograph of the diverticulum showing small bowel mucosa and the underlying tumor consisting of fascicles of cells (×200)

30 % originate in the small intestine, and less than 10 % in the esophagus, colon, and rectum. GISTs also occur in the extraintestinal abdominopelvic sites such as omentum, mesentery, or retroperitoneum [1].

Located in the small intestine, a GIST becomes symptomatic due to abdominal pain (74 %), abdominal mass (72 %), gastrointestinal bleeding (44 %), small bowel obstruction (44 %), weight loss (16 %), fever or abscess (14 %), or urinary symptoms (12 %). Perforation of the tumor with subsequent peritonitis has been reported in 8 % of GISTs of the small intestine. However, GIST causing MD perforation and peritonitis is very rare, and prior to this indexed case, only two such cases have been reported in the literature [4, 7].

The preoperative diagnosis of tumors within MD is rare because of the few characteristic clinical and radiologic features. As with other tumors of the small bowel, GIST in MD is difficult to visualize with standard diagnostic modalities. A range of diagnostic tests can be performed in symptomatic patients, including barium studies, endoscopy, ultrasound, CT scanning, and mesenteric arteriography, but correct preoperative diagnosis is infrequent. Therefore, most of the cases are diagnosed at the time of laparotomy and microscopic examination of the specimen.

Most reliable prognostic factors are the size of the primary tumor and the mitotic index which measures proliferative activity of the cells. According to the suggested guidelines for assessing the risk of malignant potential of jejunal and ileal GISTs [8], the current case falls into low malignant potential GIST. Other prognostic factors are specific histologic subtypes (epithelioid versus spindle cell), the degree of cellular pleomorphism, and age of the patient. The indexed case was put on tyrosine kinase inhibitor imatinib mesylate (400 mg daily) and was clinically well after 1 year of follow-up.

Conflicts of interest The authors do not have any disclosable interest.

References

1. Chandramohan K, Agarwal M, Gurjar G, Gatti RC, Patel MH, Trivedi P et al (2007) Gastrointestinal stromal tumour in Meckel's diverticulum. *World J Surg Oncol* 5:50
2. Rosai J (2004) Rosai and Ackerman's surgical pathology. Missouri, Mosby Publishers, p 714
3. Yahchouchy EK, Marano AF, Etienne JC, Fingerhut AL (2001) Meckel's diverticulum. *J Am Coll Surg* 192(5):658–662
4. Hager M, Maier H, Eberwein M, Klingler P, Kolbitsch C, Tiefenthaler W et al (2005) Perforated Meckel's diverticulum presenting as a gastrointestinal stromal tumour: a case report. *J Gastrointest Surg* 9 (6):809–811
5. Johnston AT, Khan AL, Bleakney R, Keenan RA (2001) Stromal tumour within a Meckel's diverticulum: CT and ultrasound findings. *Br J Radiol* 74(888):1142–1144
6. Lorusso R, Forte A, Urbano V, Soda G, D'Urso A, Bosco MR et al (2003) Small bowel stromal tumors in a "meckelian" location. About a clinical observation. *Ann Ital Chir* 74(6):707–711
7. de la Morena LF, Fernández-Salazar L, Velayos B, Aller R, Juárez M, González JM (2007) Meckel's diverticulum and gastrointestinal stromal tumor: an unusual association. *Gastroenterol Hepatol* 30 (9):534–537
8. Miettinen M, Makhoul H, Sobin LH, Lasota J (2006) Gastrointestinal stromal tumors of the jejunum and ileum: a clinicopathologic, immunohistochemical, and molecular genetic study of 906 cases before imatinib with long-term follow-up. *Am J Surg Pathol* 30 (4):477–489