A Clinically-occult Gastrointestinal Stromal Tumor in A Meckel's Diverticulum Presenting as Hollow Organ Perforation

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Meckel's diverticulum is the most common congenital anomaly of the gastrointestinal tract. However, a neoplasm is a rare complication of Meckel's diverticulum. We report a case of a ruptured gastrointestinal stromal tumor (GIST) in a Meckel's diverticulum, presenting as hollow organ perforation, in a 76 year-old woman. To our knowledge, the case we presented here is the 6th report describing a perforated GIST within a Meckel's diverticulum. In addition, the diverticular neoplasm in our case was clinically occult because of an unusual tumor configuration. Since the treatment of asymptomatic Meckel's diverticula remains controversial, our case raises suspicion that managing asymptomatic Meckel's diverticula by pure observation may leave some clinically occult diverticular neoplasms untreated. The role of prophylactic diverticulectomy requires further evaluation. *(Chang Gung Med J 2011;34(6 Suppl):56-61)*

Key words: Meckel's diverticulum, gastrointestinal stromal tumor (GIST), diverticular neoplasm, hollow organ perforation

Symptomatic Meckel's diverticulum manifesting with acute abdomen is not an unusual condition. In most circumstances, the symptoms come from alteration of Meckel's diverticulum per se, such as intussusception and ulceration.[1,2] We report a rare case of Meckel's diverticulum, in which the clinical symptoms came from rupture of a gastrointestinal stromal tumor (GIST) located within the diverticulum. The diverticular GIST was clinically occult because of an unusual tumor configuration. Some authors still recommend that an asymptomatic Meckel's diverticulum be left in place without further management.[1] Our case points out a potential pitfall: if an asymptomatic Meckel's diverticulum harbors a clinically undetectable neoplasm, pure observation without surgical intervention may leave the neoplasm untreated.

CASE REPORT

A 76-year-old woman without major underlying diseases presented to the emergency department because of lower abdominal cramping pain for two days. Motion aggravated the abdominal pain, which was relieved after rest. Episodes of diarrhea and vomiting were noted. The patient denied other symptoms, such as fever, tarry stool, and bloody stool. Computed tomography (CT) revealed hollow organ perforation characterized by intraperitoneal free air pockets. A distended diverticulum filled with bezoar content was detected at the distal ileum accompanied by regional fluid accumulation. The diverticular wall was mildly thickened without an obvious mass contour (Fig. 1A).

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Received: July 1, 2011; Accepted: Oct. 6, 2011
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During emergency laparoscopic examination, a perforated diverticulum was identified at the antimesenteric ileal border 100 cm above the ileocecal valve (Fig. 1B, 1C). No other active intraabdominal lesion was found. Under the impression of Meckel's diverticulitis with perforation, a segmental ileal resection followed by anastomosis was performed.

Grossly, the resected ileum revealed an antimesenteric diverticulum 3.9 cm in the maximal dimension. At the diverticular tip, a 3.2 cm crescent-shaped fusiform lesion, up to 1 cm in maximal thickness, was found extending along the curvature of the diverticular wall without an outward bulge (Fig. 1D). Since the lesion was brownish and had cystic changes, a ruptured Meckel's diverticulum covered by a hematoma was considered. However, a diverticular tumor was also suspected because of the unusual elastic texture on palpation.

Microscopically, a GIST arising from a Meckel's diverticulum was diagnosed by hematoxylin and eosin and immunohistochemical stains (Fig. 2). Tumor rupture with perforated serosa, areas of increased mitoses up to 6 per 50 high-power fields, focal prominent nuclear atypia, occasional tumor giant cells, and tumor necrosis was discernible. According to a recently-proposed classification,[3] the neoplasm belonged to the 'high malignant potential' category.

DISCUSSION

Meckel's diverticulum is the most common congenital anomaly of the gastrointestinal tract (incidence: 0.6%-4%).[1,4] A Meckel's diverticulum is clinically insignificant unless complications occur. Only 4%-16% of Meckel's diverticula produce symptoms,

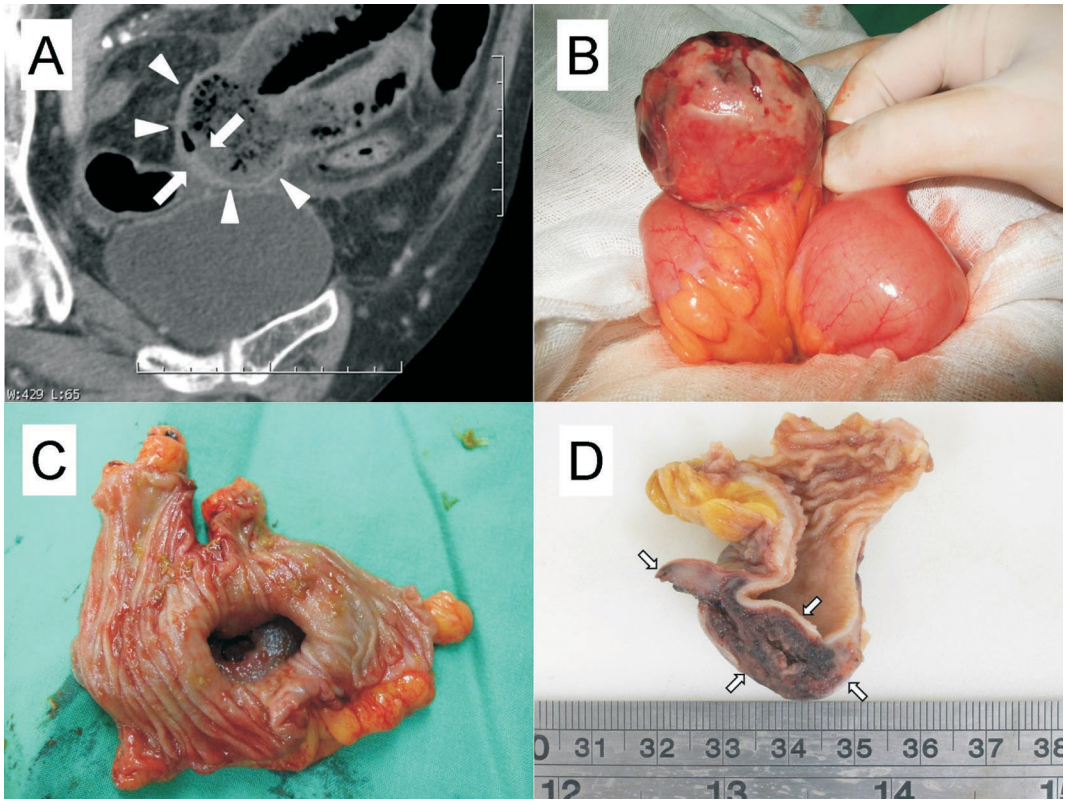

Fig. 1 CT image demonstrates a mechanical ileus with an outpouch (arrowhead) at the distal ileum. The outpouch is expanded by an intraluminal bezoar. The tip of the outpouch (arrow) is mildly thickened (A). External view of the perforated Meckel's diverticulum at the antimesenteric border of the terminal ileum (B). Internal view of the Meckel's diverticulum (C). A brownish fusiform lesion (arrow) is seen at the tip of the diverticulum after bisection. The lesion mimics a hematoma or purulent exudate covering the perforated Meckel's diverticulum (D).

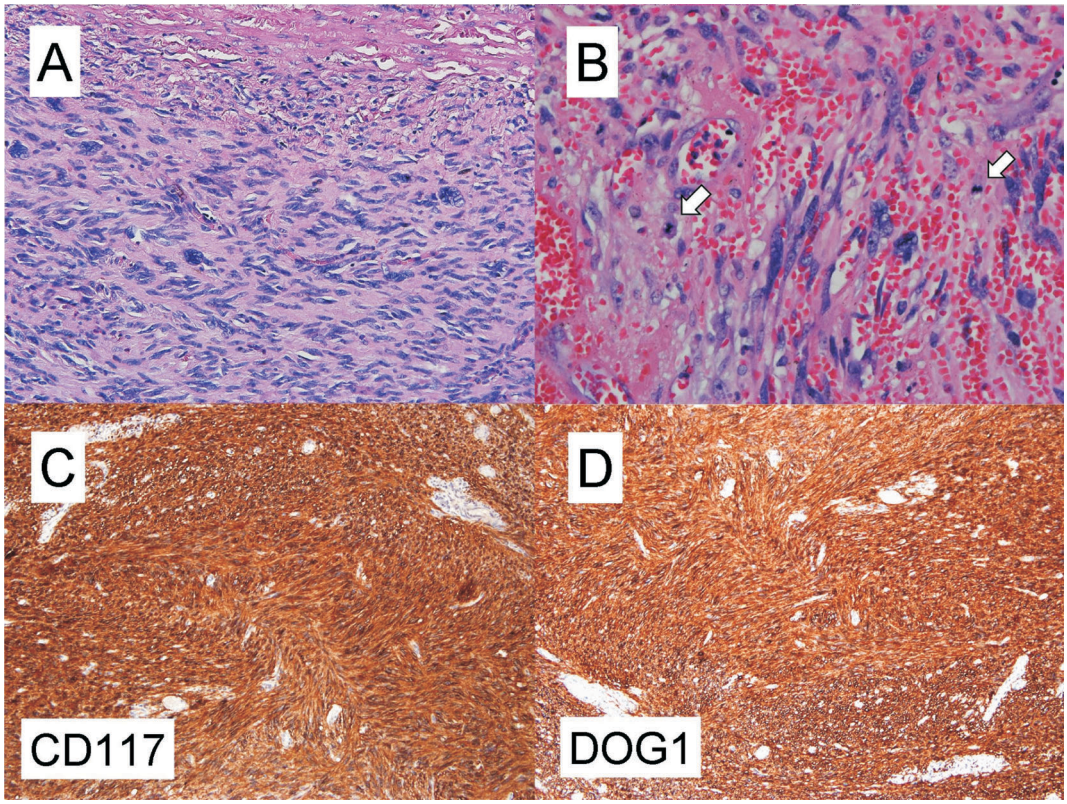

Fig. 2 The tumor cells are spindle-shaped with foci of nuclear atypia and tumor giant cell formation (A, hematoxylin and eosin (H & E), x 200). Increased mitotic activity (arrows) is seen (B, H & E, x 400). The tumor cells are diffusely positive for CD117 (C, x 100) and DOG1 (D, x 100) stains. A gastrointestinal stromal tumor of high malignant potential was diagnosed.

mainly caused by intussusception and ulceration.[1,2] A neoplasm is a rare complication observed in only 0.5%-3.2% of Meckel's diverticula.[5-8] Carcinoid tumors are the most common (33%-44%) primary diverticular malignancies, followed by leiomyosarcoma (18%-25%) and adenocarcinoma (12%-16%).[9] GISTs represent 12% of primary tumors in Meckel's diverticula.[5,10]

GISTs are the most common mesenchymal tumors of the gastrointestinal tract.[3,11] The stomach is the most frequent site (70%) for GISTs, followed by the small intestine (20%-30%). About 13% of small intestinal GISTs are incidentally detected during management of other diseases.[3,9] A few clinical settings are associated with the development of GISTs, such as neurofibromatosis type 1 (Von Recklinghausen disease) and Carney triad.[3,9,11] The clinical behavior of a GIST is strongly related to its size and mitotic activity: large size and frequent mitoses are indicative of higher malignant potential.[3] Under microscopic examination, most GISTs appear as spindle cell tumors with a storiform pattern. Epithelioid morphology, signet ring cells, and the so-called gastrointestinal autonomic nerve tumor variant may be discernible in some cases. Positive CD117 and DOG1 stains are diagnostic immunohistochemical indicators of GISTs.[3,11]

Our patient had a Meckel's diverticulum complicated by a GIST. This rare combination has been reported in the literature.[9,10,12-15] However, our case was unique in two aspects. First, unlike most diverticular neoplasms showing chronic nonspecific symptoms, the diverticular GIST in our case manifested with hollow organ perforation because of the tumor rupture. In fact, a diverticular neoplasm seldom causes acute abdomen. To our knowledge, there are only five case reports of a small intestinal GIST causing perforation of the Meckel's diverticulum in

the English literature.[12-15] Here we present the 6th case report. Second, the diverticular GIST in our case was clinically occult because of the unusual tumor configuration: since the brownish GIST grew smoothly along the diverticular wall without an outward bulge, the neoplasm grossly mimicked a hematoma covering a perforated Meckel's diverticulum. It was also difficult to recognize the fusiform diverticular GIST on the preoperative CT images, because the neoplasm was stretched to a more slender shape when the Meckel's diverticulum was distended by the intraluminal bezoar. Actually, most published cases described diverticular GISTs as an indurated, rounded mass detectable either by standard diagnostic modalities or explorative operations.[9,10,12,14,15] One other reported case showed a presentation similar to our case.[13] In that report, a 2 cm GIST could not be identified until the specimen of a ruptured Meckel's diverticulum was put under microscopic examination. Unfortunately, there was no detailed description of the macroscopic findings of that grossly invisible neoplasm.

The standard treatment for symptomatic Meckel's diverticula is surgical resection; however, the management of asymptomatic Meckel's diverticula is controversial.[1] Some authors have suggested that asymptomatic Meckel's diverticula should not be removed unless the patient is at an increased risk of developing complications. The risk factors included male gender, young age, diverticulum larger than 2 cm, and presence of heterotopic tissue.[2,16] Morbidity following prophylactic diverticulectomy has been claimed to exceed the lifetime complication risk of Meckel's diverticulum (9% versus 4.2%).[8] However, since operation-related sequelae have decreased recently, many authors accept prophylactic diverticulectomy for asymptomatic patients.[14,15,17,18] In contrast to the significant morbidity of up to 33% with complicated Meckel's diverticula, the postoperative morbidity associated with prophylactic diverticulectomy is between 0% and 6%.[1]

Our case demonstrated another uncommon advantage of prophylactic diverticulectomy. A Meckel's diverticulum may harbor a clinically undetectable neoplasm. If our patient had presented with an asymptomatic Meckel's diverticulum, pure observation without surgical intervention may have left a GIST with high malignant potential untreated.

Debate continues on the management of asymptomatic Meckel's diverticula, and the role of prophylactic diverticulectomy requires further evaluation. Based on the experience in our case, we suggest that when an asymptomatic Meckel's diverticulum is incidentally found, a prophylactic diverticulectomy should be considered if the diverticulum shows unusual mural thickening on image study or an unusual elastic texture during intraoperative palpation.

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以中空器官穿孔爲臨床表現的梅克爾憩室內胃腸道間質細胞瘤病例報告暨文獻回顧

周岳弘 塗昭江[1] 黃俊傑[2] 謝明書[3]

梅克爾憩室雖然是最常見的先天性腸道異常，併發憩室內腫瘤卻是一個罕見的情況。我們報告一例潛藏在梅克爾憩室內的胃腸道間質細胞瘤，病人因爲腫瘤的破裂、臨床上產生中空器官穿孔的症狀而求診。此外，由於該腫瘤的形狀扁平且嵌合在憩室的腸壁上，導致術前電腦斷層檢查與手術過程中，均難以察覺到憩室內潛藏有一顆腫瘤。目前在臨床實務上，對於意外發現、無症狀的梅克爾憩室，可能會採取追蹤觀察的方式處理，而非預防性的手術切除。我們的病例闡述一個可能的風險：如果我們對於無症狀的梅克爾憩室一律採取單純的追蹤觀察，有時將會延遲對於潛藏憩室腫瘤的治療。最後，我們回溯性的分析本病例，提供可能可以提早發現潛藏憩室腫瘤的方法。(長庚醫誌 2011;34(6 Suppl):56-61)

關鍵詞：梅克爾憩室，胃腸道間質細胞瘤，憩室腫瘤，中空器官穿孔

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