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Perforated Leiomyosarcoma of Meckel's Diverticulum

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ABSTRACT

Two cases of perforated leiomyosarcoma of Meckel's diverticulum are presented. There are only 59 cases reported in current literature, including 4 perforations. Although the condition is rare, leiomyosarcoma is the commonest tumour of Meckel's diverticulum. Its clinical presentation include abdominal pain, intestinal bleeding, abdominal mass, intestinal obstruction and less commonly, acute perforations. Both our cases presented with perforations which is unusual. Despite this late presentations both were resectable and both had no distant or local metastasis. One of our patients was 89 years old at presentation and has been disease-free 3 years after resection. The other patient was 69 years old and has also been disease-free.

Keywords: small bowel, smooth muscle tumour, sarcoma, perforation, Meckel

INTRODUCTION

Primary neoplasms originating from Meckel's diverticulum, whether benign or malignant, are uncommon. In Lie's review, he found an incidence of only 0.9% among 1,135 cases of Meckel's diverticulum that were surgically removed[1]. Malignant tumours occur 3 times more frequently than benign ones. In descending order of frequency, the commonest malignant tumours of Meckel's diverticulum are sarcomas, carcinoids and adenocarcinomas[1,2]. The commonest sarcoma arising in Meckel's diverticulum is leiomyosarcoma. The total number of reported cases of leiomyosarcoma of Meckel's diverticulum was brought up to 59 in 1991, with 4 perforations[3].

We report 2 cases of perforated leiomyosarcoma of Meckel's diverticulum.

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Case 1

An 89-year-old woman was admitted in June 1990, for generalised abdominal pain of sudden onset associated with vomiting. There were no accompanying fever, bowel or urinary symptoms. On examination, she was septic, breathless and tachycardic (pulse rate at 120/min). Her blood pressure was 180/100 mmHg. She had generalised abdominal distension, guarding and rebound tenderness. No obvious mass could be felt in the abdomen or per rectum.

Laboratory tests showed a haemoglobin level of 167 g/L and a white cell count of 6.3 x 10^9/L. Serum urea, creatinine and electrolytes were normal. Chest and abdominal X-rays did not show any pneumoperitoneum.

An emergency laparotomy was performed and we discovered extensive soilage of the peritoneal cavity. A Meckel's diverticulum was found at 25 cm from the ileocecal valve. A perforation was present and the diverticulum was wrapped around by omentum. A healthy length of bowel separates the mass from the small bowel. The diverticulum was excised with an ellipse of small bowel wall and the defect was closed primarily in two layers. Incidentally, there were multiple gallstones in the gallbladder for which a cholecystectomy was done. Appendicectomy was also performed. The liver was normal and there were no peritoneal nodules.

Her post-operative recovery was stormy. The patient required prolonged ventilation and had tracheostomy on the seventh post-operative day. She was subsequently weaned off the ventilator 2 weeks after laparotomy.

Macroscopically, the specimen was a pouch-like structure measuring 6 cm in diameter. Sections showed the presence of a lumen with a solid white tumour mass. An area of perforation was noted.

Microscopic examination revealed a tumour occupying the submucosa and muscularis mucosae composing of spindle-shaped cells with rounded to fusiform nuclei and eosinophilic cytoplasm. The tumour was cellular and pleomorphic with areas of degeneration and numerous lymphoid collections. The mitotic rate was 2 per 10 hpf. There was inflammation along the external surface consistent with perforation.

In view of the patient's age and poor respiratory status, a repeat laparotomy was not advisable. The patient was discharged well.

Two months later, the patient was seen for intermittent upper abdominal pain. A barium meal and follow-through performed was normal. The patient was last seen in June 1993. A liver ultrasound did not show any metastasis.

Case 2

A 69-year-old Malay lady was admitted in September 1992 for generalised abdominal pain of one day's duration associated with fever, vomiting and diarrhoea. The patient had intermittent episodes of

epigastric pain prior to this admission.

On examination, the patient was febrile with a temperature of 38.9°C. Her vital signs were stable and she was not jaundiced. Abdominal examination revealed generalised tenderness with guarding and rebound tenderness. There were no palpable masses and per rectal examination was normal.

Laboratory tests showed a haemoglobin level of 109 g/L, a white cell count of 15.3 x 10^9/L and the serum amylase at 58. Serum urea, creatinine and electrolytes were normal. Erect chest and abdominal X-rays did not show any pneumoperitoneum.

An emergency laparotomy was performed. A 10 x 7 cm tumour mass was found arising from the tip of a Meckel's diverticulum. The tumour had perforated and was walled off by omentum and small bowel. There was moderate peritoneal soilage. The tumour was excised with a segment of 5 cm of ileum on either side.

Post-operatively, the patient recovered uneventfully and was discharged on the ninth post-operative day.

The specimen was reported as a 10.5 x 7 x 6 cm tumour that appeared to have arisen from the wall of the mid-segment of the diverticulum. The mucosa overlying the tumour did not show any ulceration. Cut section through the tumour mass showed large areas of necrosis and haemorrhage. The rest of the mucosa appeared normal.

Microscopic examination of the tumour showed proliferation of spindle cells with moderate degrees of nuclear atypia and ample fine to granular eosinophilic cytoplasm. Some of these cells formed interlacing fascicles with features resembling a smooth muscle tumour. A thin fibrous capsule covered the outer aspect of the tumour and no perforation of the capsule was seen. Mitoses were infrequent. Masson's trichome showed cytoplasmic granular trichome red material. A diagnosis of leiomyosarcoma was made.

The patient was last seen in June 1993. An ultrasound of the hepatobiliary system did not show any metastasis.

DISCUSSION

Leiomyosarcoma is the commonest malignant smooth muscle tumour of Meckel's diverticulum. It appears to develop in patients above the age of 20 years. In a series reported by Weinstein et al, the age ranged from 22 to 76 years[4]. The patient in case 1 presented at the age of 89 years: the oldest reported to date. There is no sex predilection.

The commonest symptom of leiomyosarcoma of Meckel's diverticulum is abdominal pain followed by intestinal bleeding[3,5]. It can also present with a palpable abdominal mass, intestinal obstruction and acute perforation. Both cases presented with acute abdominal pain suggestive of bowel perforation. In both cases, no mass could be felt because of severe tenderness and guarding thus making pre-operative diagnosis difficult.

The diagnosis is usually made intra-operatively. Pre-operative investigations such as plain abdominal X-rays and barium studies are uninformative. In certain series, mesenteric angiography may suggest the diagnosis by the presence of hypervascularisation and feeding vessels[5].

The histological differentiation between leiomyoma and leiomyosarcoma is difficult. Golden and Stout suggested that if there are 2 or more mitoses per high power field, the lesion is probably malignant[6]. On the other hand, certain reviews have shown that the absence of mitoses does not exclude the possibility of the lesion being malignant. The size of the tumour has also been used to predict malignancy. In a series by Starr, he found no benign tumour larger than 7 x 5 x 5 cm and no malignant lesion smaller than 2 x 2 x 1 cm[7].

The commonest mode of spread of intestinal leiomyosarcoma is by vascular embolisation, the liver being the most frequently involved organ[3,5]. Local invasion and peritoneal seeding have also been described. Lymphatic spread is rare.

There is no unanimous approach to the management of such tumours. Lee proposes that the tumour should be excised with at least 10 cm of normal bowel on either side including the adjacent mesentery[8]. This approach is justified because the precise histological nature of the lesion (benign vs malignant) is uncertain, the possibility of involvement of regional lymph node involvement exists and there is little or no increase in morbidity between wide segmental and limited resection of the small bowel. Neither radiation therapy nor chemotherapy confers any benefit.

With regards to the long-term survival, little is known although Starr and Dockerty reported a 50% 5-year survival after curative resection[7]. In a recent series from the Cleveland Clinic, 3 significant favourable factors were identified: a long duration of symptoms, a tumour of less than 9 cm in diameter, and the absence of lymphatic or distant metastases[5]. The diagnostic difficulties faced with such tumours probably worsened the prognosis. In some reports, however, the tumour is indolent and slow-growing and the patient may survive for many years.

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