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Intestinal perforation with abdominal abscess caused by extramedullary plasmacytoma of small intestine: A case report and literature review

Wang KW *et al.* Extramedullary plasmacytoma of small intestine

Abstract

BACKGROUND

Extramedullary plasmacytoma (EMP) of the gastrointestinal tract is an extremely rare disease. Clinical manifestations of EMPs are varied and depend on the location and progression of the tumor.

CASE SUMMARY

Here, we firstly report a case of intestinal perforation with abdominal abscess caused by EMP of the small intestine in a 55-year-old female patient. The patient received emergency surgery immediately after the necessary preoperative procedures. During the operation, EMP was found to have caused the perforation of the small intestine and the formation of multiple abscesses in the abdominal cavity. Partial resection of the small intestine with peritoneal irrigation and drainage was performed. EMP was finally confirmed by postoperative histopathology and laboratory tests. Additionally, we performed a literature review of gastrointestinal EMP to obtain a deeper understanding of this disease.

CONCLUSION

EMP of the small intestine may have spontaneous perforation, which requires emergency surgery. Surgical resection can obtain good therapeutic effects.

Key Words: Extramedullary plasmacytoma; Perforation; Small intestine; Gastrointestinal tract; Treatment; Case report

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Core Tip: Extramedullary plasmacytoma (EMP) of the gastrointestinal tract is an extremely rare disease, accounting for only 7% of all EMPs. ¹ Clinical manifestations of EMPs are varied and depend on the location and progression of the tumor. Here, we firstly report a case of intestinal perforation with abdominal abscess caused by EMP of the small intestine in a 55-year-old female patient. Additionally, we discussed the diagnosis and treatment of gastrointestinal EMP after a review of the literature worldwide to provide an overview of this disease.

INTRODUCTION

Plasmacytoma is a malignant tumor that originates from bone marrow hematopoietic tissue. It is characterized by an imbalance in the monoclonal proliferation of plasma cells. Extramedullary plasmacytoma (EMP) refers to a localized monoclonal plasma cell proliferation that occurs in soft tissues without bone marrow involvement. It is a rare type of malignant monoclonal plasma cell lesion, accounting for approximately 2%-3% of all plasmacytomas^[1,2]. Plasmacytoma primarily occurs in the upper respiratory tract but is rarely found in the gastrointestinal tract. Gastrointestinal EMP only accounts for approximately 7% of all EMPs^[3]. EMP is found in all parts of the gastrointestinal tract, including the small intestine^[4-7]. Clinical manifestations of gastrointestinal EMPs vary with the location and progression of the tumor and lack specificity. Common clinical manifestations include abdominal pain, abdominal discomfort, changes in bowel habits, gastrointestinal bleeding and intestinal obstruction^[8-12]. However, there are no reports of spontaneous perforation and abdominal abscess caused by EMP of the small intestine. Reports on EMP of the small intestine are mostly single case reports, and most of the patients underwent routine surgery^[7,13]. It is rare to find this disease during an emergency surgery. In this paper, we firstly present a case of intestinal perforation with abdominal abscess caused by EMP of the small intestine and review the relevant literature from PubMed.

³ CASE PRESENTATION

Chief complaints

A 55-year-old female was admitted to the Department of Emergency of our hospital with sudden abdominal pain and abdominal distension.

History of present illness

The patient's symptoms started 3 d prior and were accompanied by nausea and vomiting without gas or defecation. Since onset, the patient had a loss of appetite, limited diet, poor sleep and decreased urination. No significant change in body weight was noted.

History of past illness

The patient's previous medical history was not remarkable. She and her family had no history of multiple myeloma (MM) or other gastrointestinal diseases.

Personal and family history

The patient has no personal and family history.

Physical examination

During physical examination, the patient had a normal heart rate and mild hypotension. The patient's abdomen was slightly distended, and the abdominal tenderness was more severe in the left upper abdomen accompanied by rebound pain and muscle tension.

Laboratory examinations

Laboratory tests showed the following: White blood cells $10.5 \times 10^9/L$, (NE) $9.63 \times 10^9/L$, NE% 91.7%, hemoglobin 108 g/L, and PLT $330 \times 10^9/L$. Liver and kidney function were normal.

Imaging examinations

Enhanced computed tomography (CT) showed that the small intestinal lumen in the upper left abdomen was dilated with gas and fluid accumulation, and showed multiple fluid-gas level changes were noted. The intestinal wall was edematous and thickened, and the density of the surrounding fat interspace had increased. Small air bubbles were scattered under the left diaphragm, and multiple encapsulated effusions were observed between the small intestines. These imaging findings suggested local perforation and multiple abscesses in the abdominal cavity (Figure 1).

FINAL DIAGNOSIS

Microscopic analysis showed that the pathological specimen displayed a large number of neoplastic plasma cells with inflammatory cell infiltration (Figure 2A). These plasma cells were positive for CD38 (+), CD138 (+), kappa (+), lambda (week+), CD79a (week+), and MUM1 (+) and negative for creatine kinase (-), CD117 (-), Dog-1 (-), S-100 (-), B cell lymphoma-2 (-), beta-catenin (-), CD56 (-), immunoglobulin G4 (-) and Pax-5 (-) with a Ki-67 proliferative index of 10% (Figures 2B-F). The final pathological specimens were highly suspicious of plasmacytoma. Postoperative laboratory tests showed that the bone marrow cytology was normal and no abnormal monoclonal plasma cells were detected in the flow cytometric analysis. Urine free light chain and serum immunofixation electrophoresis were also normal. Lytic lesions were not found on X-rays. Therefore, the final diagnosis of this patient was primary EMP of the small intestine.

TREATMENT

Considering that the patient may have a perforation of the digestive tract, we performed emergency surgery. During the operation, we found that the small intestinal serosa 100 cm away from the Treitz ligament had a dark-red polyp-like protrusion with a perforation approximately 0.5 cm in diameter at the top. The local intestinal wall was hyperemic, edematous and thickened, and the surface of the surrounding small intestine and lateral peritoneum was covered with many purulent masses (Figure 3).

Several abscesses were observed between the left paracolic groove and small intestine and filled with a yellow, turbid fluid. After the abscesses were removed, the abdominal cavity was flushed with a large amount of warm normal saline. Then, a segment of the jejunum 33 cm in length was resected, and a primary side-to-end anastomosis of the small intestine was performed. The lumen of the intestinal tube 6 cm from the nearest end resection margin was narrow with a diameter of approximately 1.5 cm. The serosal surface was similar to a polypoid with a size of approximately 2 cm × 1 cm × 1 cm.

OUTCOME AND FOLLOW-UP

The patient had a good postoperative recovery with no complications, and she was discharged smoothly from the hospital one week after her surgery. As of August 1, 2021, she has been regularly followed up for 2 years at an outpatient clinic, and there have been no signs of recurrence or metastasis.

DISCUSSION

Primary plasmacytoma of the small intestine is rare in clinical practice. Here, we firstly report a case of intestinal perforation with abdominal abscess caused by EMP of the small intestine in a 55-year-old female. The diagnosis is based on a pathologically confirmed small intestinal mass with clonal growth of plasma cells, normal bone marrow histological examination, and normal serum monoclonal immunoglobulin levels^[14]. EMP can be divided into two types: Primary and secondary. EMP can also present as a secondary tumor of another plasma cell neoplasm, such as MM^[15]. MM must be excluded before the diagnosis of primary EMP^[16]. The case we reported had no positive laboratory or imaging findings of MM, which met the diagnostic criteria of primary EMP. In this paper, we performed a review of the well-documented primary gastrointestinal EMP cases in the last 20 years and presented these results in table form (Table 1)^[4-7,11,17-45]. These results show that gastrointestinal EMP is common in patients over the age of 50 years, and the incidence rate is higher in men compared with women (2:1). The clinical manifestations of gastrointestinal EMPs vary with the location of the

tumor and lack specificity. In the early stage, this disease is often asymptomatic, and patients often seek medical treatment because of pain or discomfort caused by local tumor compression. Other clinical manifestations include gastrointestinal bleeding or obstruction, changes in bowel habits, *etc.* In our case, the patient presented with sudden abdominal pain and abdominal distension, which may have been caused by intestinal perforation. CT images usually show an infiltrating mass with clear boundaries. When the mass is large, a liquefied necrotic area may appear in the center. However, until now, there has been no description of the specific imaging characteristics of EMP^[46]. Therefore, the role of imaging examinations in differentiating gastrointestinal EMP from other neoplastic diseases is limited. EMP may be occasionally misdiagnosed as cancer^[47], stromal tumors or inflammatory bowel disease^[41]. Hence, the accurate diagnosis of gastrointestinal EMP still depends on histopathological results. For gastrointestinal EMP, endoscopic biopsy is a convenient and practical diagnostic method.

Given the rarity of gastrointestinal EMP, unified treatment guidelines for this disease are not available. At present, complete surgical resection is a good choice for the treatment of gastrointestinal EMP. Several studies have reported that patients with gastrointestinal EMP can be completely cured after surgical resection of tumors^[21,24,34,40]. Most of the patients underwent routine surgery. However, the EMP patient we reported with perforation of the small intestine required emergency surgery. In addition to perforation of small intestinal EMPs, perforation of colon EMPs can also occur. Kitamura *et al*^[40] reported one case of EMP in the sigmoid colon with perforation. The patient underwent emergency surgery without postoperative adjuvant chemotherapy with no recurrence after 14 mo of regular follow-up. In recent years, endoscopic treatments, such as endoscopic mucosal resection or endoscopic submucosal dissection, have become increasingly popular in gastrointestinal EMP surgery and have obtained a good therapeutic effect^[18,20,25]. Due to the high sensitivity of primary EMP to radiotherapy, local radiotherapy is also an effective treatment method^[45,48]. At present, many hospitals use radiotherapy as an adjuvant treatment for patients with

gastrointestinal EMP after surgery to prevent local recurrence or metastasis. Moreover, radiotherapy can also represent an additional therapeutic option for cases with incomplete resection, lymph node involvement or recurrence. There are also some results suggesting that EMP is well controlled with a dose of 40 Gy or more^[49]. In cases that are small, well-defined, or postexcision with positive margins, 40 Gy is acceptable^[50]. Currently, most studies in this area are retrospective, and more prospective randomized controlled studies are needed to verify these results.

EMP is a low malignancy tumor with a good prognosis. Local recurrence or recurrence at other sites occurred in 7.5% and 10% of patients, respectively, and the 15-year survival rate was 78%^[51]. Given that EMP may recur or progress to MM in some patients, regular long-term follow-up is recommended and necessary. Detailed medical records, physical examination, laboratory tests, including complete blood cell count, beta-2 microglobulin and immunoglobulin levels, renal function, and imaging examination of the abdomen are required for patients during follow-up^[52].

CONCLUSION

In conclusion, EMP of the small intestine is extremely rare and lacks specific clinical and imaging manifestations. EMP may be associated with spontaneous perforation, which requires emergency surgery. We firstly report a case of intestinal perforation caused by EMP of the small intestine. The diagnosis of EMP still depends on the histopathological results. Surgical resection and radiotherapy can obtain good therapeutic effects. The cooperation of a multidisciplinary team, including pathologists, hematologists, radiologists and surgeons, is needed to develop the best diagnostic and therapeutic plan for gastrointestinal EMP.

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