

Extramedullary plasmacytoma occurring in ileocecum

A case report and literature review

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Abstract

Rationale: Gastrointestinal solitary extramedullary plasmacytoma (EMP) is rare, just occupies about 5% of all EMPs. The most common site is small intestine followed by stomach. The colorectal incidence is much rare.

Patient concerns: A 63-year-old female had an episodic pain around the umbilicus for about one week. The hyperemia and edema in the ileocecal mucosa were found in colonoscopy, and the endoscopy could not cross the ileocecal valve. The pathology specimens showed a high index suspicion of plasmacytoma.

Diagnoses: The patient was diagnosed with extramedullary plasmacytoma.

Interventions: A right hemicolectomy surgery was performed to treat the disease.

Outcomes: Post surgery pathologic report showed low grade malignant mucosa associated marginal zone B cell lymphoma.

Lessons: We report a case of an extramedullary plasmacytoma in ileocecum with abdominal pain and a review of extramedullary plasmacytoma.

Abbreviations: EMP = extramedullary plasmacytoma, EMR = endoscopic mucosal resection, ESD = endoscopic submucosal dissection, HP = *Helicobacter pylori*, MM = multiple myeloma, SBP = solitary bone plasmacytoma, UKMF = United Kingdom Myeloma Forum.

Keywords: extramedullary plasmacytoma, gastrointestinal, ileocecum

1. Introduction

Plasma cell neoplasms can be divided into four groups: multiple myeloma (MM), plasma cell leukemias, solitary bone plasmacytoma (SBP), and solitary extramedullary plasmacytoma (EMP).^[1] Gastrointestinal solitary extramedullary plasmacytoma (EMP) is rare, just occupies about 5% of all EMPs. The most common site is small intestine followed by stomach. The colorectal incidence is much rare.

2. Case

A 63-year-old female was presented to our hospital with an episodic pain around the umbilicus for about 1 week. There were no nausea, vomit, haematemesis, melena, hematochezia, or the

absence of anal venting of faeces or flatus. She had no history of multiple myeloma and other disease. The patient underwent gastric endoscopy and colonoscopy. She was found to have *Helicobacter pylori* (HP) positive in gastric endoscopy.

The hyperemia and edema in the ileocecal mucosa were found in colonoscopy, and the endoscope could not cross the ileocecal valve (Fig. 1). Biopsies were obtained from the ileocecal valve and the opening of appendix. The pathology specimens showed a high index suspicion of plasmacytoma (Fig. 2). Immunohistochemistry for the specimens manifested CD38(+), CD138(+), Ki 67 (about 15%), Lambda(±), Kappa(3+), Pan CK(-), Mum-1 (+) (Fig. 3). The laboratory data indicated her WBC was $12.42 \times 10^9/L$, a positive vomit and stool occult blood, and normal amylase level. The serum immunofixation electrophoresis, urine immunoglobulins (κ, λ) light chains serum, β_2 microglobulin, and bone marrow aspirate index were in normal level. Serum free light chains showed a high level of kappa light chains. The level of immunoglobulin IgM was 0.359 g/L (NR:0.4 – 2.3 g/L). Lymphocyte subsets flow analysis showed a low level of CD3+, CD19+, CD45+ brightSS Clow, CD3+ CD8+, CD3+ CD4+. CT scan showed a mass in ileocecal region (Fig. 4). Finally, the patient was diagnosed with extramedullary plasmacytoma (EMP) and a right hemicolectomy surgery was performed to treat the disease. Postsurgery pathologic report showed low grade malignant mucosa associated marginal zone B cell lymphoma with plasma cell differentiation and tumor cell metastasis to perienteric lymph nodes.

3. Discussion

Extramedullary plasmacytomas come from soft tissue, which is solitary and have no evidence of multiple myeloma. The incidence of EMP is lower than SBP, accounts for only about 3% of all the

Editor: N/A.

Grant support: This work was supported by grants from National Natural Science Foundation of China (No. 81602432), Program for Jiangsu Provincial Natural Science Foundation of China (BK20140652).

The authors have no conflicts of interest to disclose.

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Medicine (2017) 96:51(e9313)

Received: 1 November 2017 / Received in final form: 24 November 2017 /

Accepted: 27 November 2017

<http://dx.doi.org/10.1097/MD.00000000000009313>

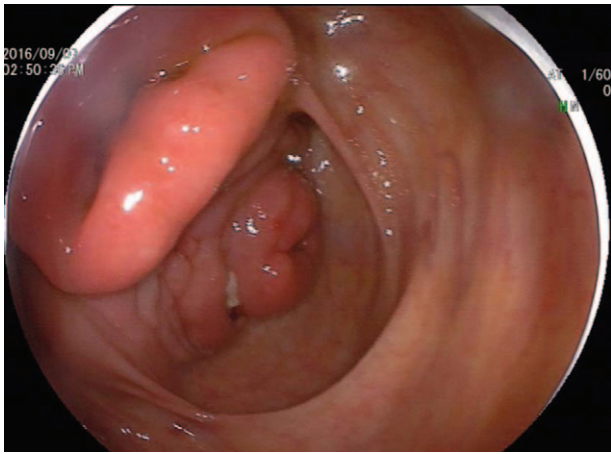


Figure 1. The colonoscopy showed hyperemia and edema of the ileocecal mucosa.

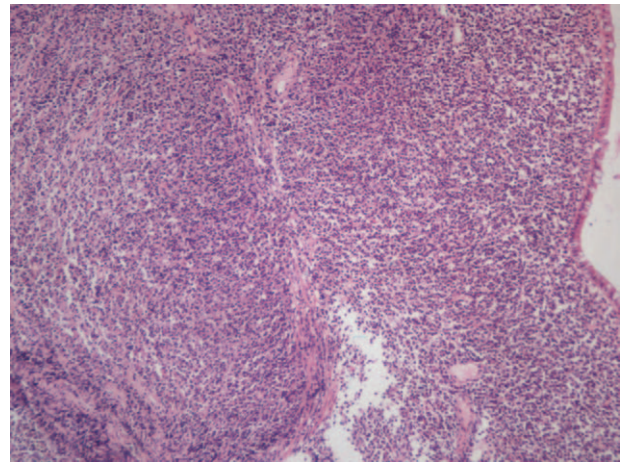


Figure 2. The pathology specimens showed a high index suspicion of plasmacytoma (HE, $\times 100$).

plasma cell neoplasia.^[2] However, the relationship of EMP and MM is not clear nowadays. About 1/3 of the EMP patients are diagnosed as multiple myeloma within two years after the symptoms.^[3] Some studies showed EMP may occur in the early MM and the patient could be easily misdiagnosed in this phase. Extramedullary plasmacytoma regularly occurs in the areas that are full of plasmacyte cells such as in gastrointestinal tract, lymph nodes, spleen, urogenital region and so on.^[4] Some articles reported extramedullary plasmacytoma could be divided into 3 stages. The first and second stage refer to the localized disease or extend to the lymph nodes nearby.^[5,6] Therefore, the early stage of EMP could be hardly distinguished with other plasma cell neoplasms and it could extend to lymph nodes or adjacent tissue.

About 80% of EMPs occur in the head and neck region and only about 5% of all EMPs occur as gastrointestinal solitary

EMP. The most common site is small intestine followed by stomach.^[7,8] The incidence in colon is very rare,^[9,10] especially in ileocecal region is significantly low. Gabriel et al showed an extramedullary ileocecal plasmacytoma with a large ileocecal perforation.^[11,12] In recent years, extramedullary ileocecal plasmacytoma in ileocaecal region has been reported very rarely. The EMP ratio of male to female is 4:1. And the disease is not common in children or adolescent.^[13]

Symptoms of most EMP are not typical. Nasal obstruction, epistaxis, facial pain, or hoarse voice commonly occur in head and neck EMPs. Gastrointestinal EMPs could present as loss of appetite, bleeding, abdominal discomfort or obstruction. So we should take further examination to give a diagnosis of the EMP. There are some accessory examinations to help us diagnose EMP, such as fine-needle aspiration cytology, CT scan, MRI, or PET-CT.

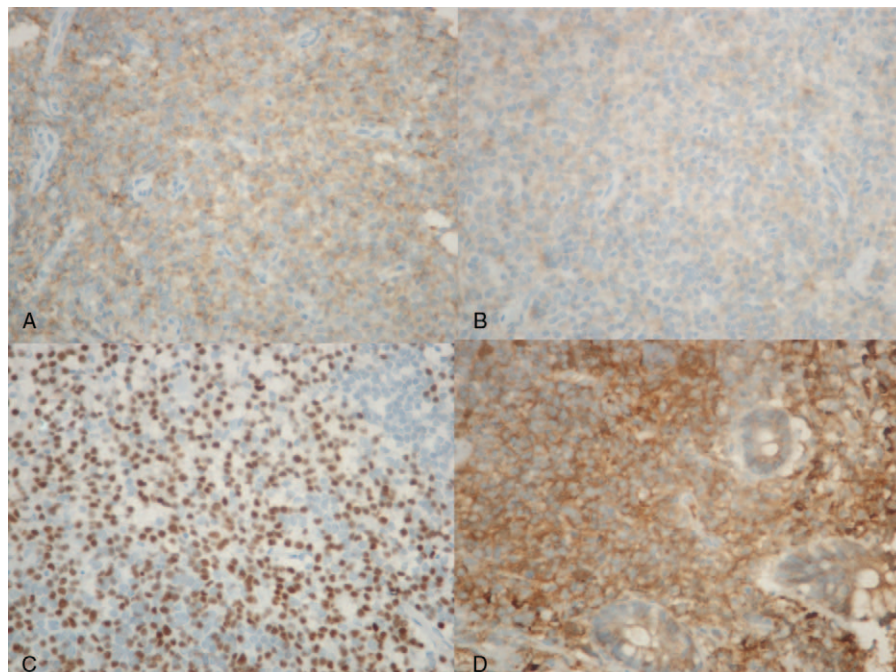


Figure 3. Immunohistochemistry for the specimens presented CD38(+), CD138(+), Ki 67 (about 15%), Lambda(\pm), Kappa(3+), Pan CK(-), Mum-1(+)($\times 400$). A. CD138 (+), B. CD38 (+), C. Mum-1 (+), D. Kappa (3+).

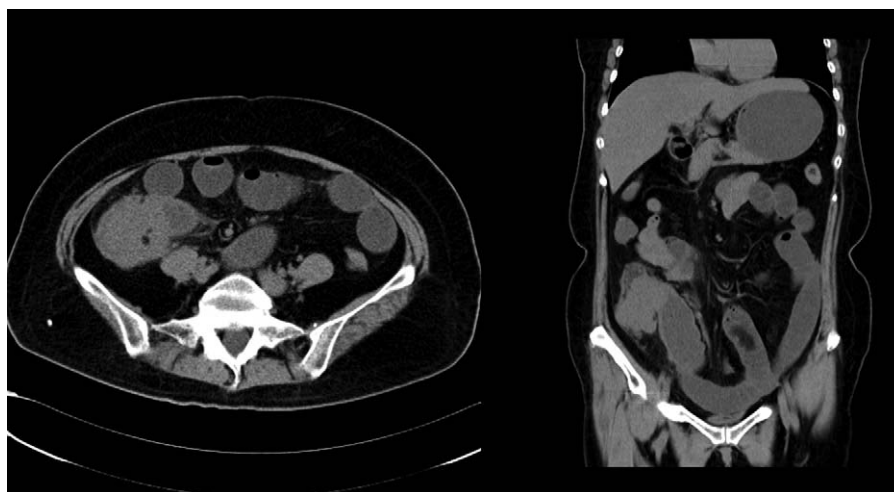


Figure 4. The CT scan showed a mass in ileocecal region.

And pathology is most important when making a definite diagnosis. Fine-needle aspiration cytology (FNAC) cannot find the plasma cells so it is difficult to make distinction between inflammatory and tumor cells.^[14] CT scan and PET-CT can help to find parenchymal lesions and the size and location of the lesion. PET-CT can find out the position of hypermetabolism which can indicate probability of malignancy. For the gastrointestinal lesion of EMP, endoscopy and biopsy are practical modes of diagnosis.

Some studies showed that *H pylori* may have relationship with infection in gastric EMP.^[15,16] But in cases of gastric EMP that we reviewed, all had negative *H pylori* infection. Although standard treatment of EMP is absent, there are some methods like radiotherapy, chemotherapy and operation. Plasmacytomas are highly sensitive to radiotherapy, with the suggesting dose of 5000 cGy.^[17] The United Kingdom Myeloma Forum (UKMF) recommended that in the head and neck EMP, radiation treatment is the better option while in the rest EMPs in other anatomic sites surgical operation should be considered first.^[18] For gastrointestinal solitary EMP, endoscopic mucosal resection (EMR) or endoscopic submucosal dissection (ESD) for the mucosal or submucosal tissue could be considered.^[19] There is a report which showed that a high-dose dexamethasone (40mg daily for 4 days every 3 weeks) treatment had an effect on ulcer type gastric EMP.^[20]

EMP had better prognosis than MM and SBP.^[21] As we have mentioned before, in the early MM, it may be misdiagnosed to EMP. So we should take a follow-up for the patients of EMP for a long time. In the aspect of prognosis, EMP may have an optimistic result of survival as the 5-year survival rate is about 82%^[22] and 10-year disease-specific survival rate is higher than 50%.^[23] Written informed consent was obtained before writing this report and the study protocol was approved by the Clinical Research Ethics Committee of Zhongda Hospital.

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