

Available online at [www.sciencedirect.com](http://www.sciencedirect.com)

ScienceDirect

journal homepage: [www.elsevier.com/locate/radcr](http://www.elsevier.com/locate/radcr)

## Case Report

# Extramedullary plasmacytoma involving rectum: A case report and literature review

Qi-Ting Lin, MD, Xiang-Ran Cai, MD\*

Medical Imaging Center, First Affiliated Hospital, Jinan university, No. 613 West Huangpu Avenue, Tianhe District, Guangzhou 510630, Guangdong, China

## ARTICLE INFO

## Article history:

Received 3 January 2021

Revised 10 January 2021

Accepted 11 January 2021

## Keywords:

Extramedullary plasmacytoma (EMP)

Gastrointestinal

Neoplasm

Laparoscope

## ABSTRACT

Extramedullary plasmacytoma (EMP) of the gastrointestinal tract is an extreme rarity. Clinical manifestations of EMPs are varied, depending on the position and progress of tumor. Here we report a case of an EMP involving rectum in an 80-year-old, male patient with a change of bowel habit. Computed tomography scanning confirmed a circumscribed, isoattenuating mass with the obvious heterogeneous enhancement. Patient received the surgical resection by laparoscope and the plasmacytoma was finally confirmed by the pathology. Furthermore, we made a literature review about the EMP of gastrointestinal tract to get the further study. Finally, we found out there is no specificity in imaging examination. Diagnosis of EMP still depends on the histopathology.

© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

## Introduction

Plasmacytoma is a malignant tumor originating from the hematopoietic tissue of bone marrow, which is characterized by dysregulated monoclonal proliferation of plasma cells and produces monoclonal immunoglobulin. Plasmacytoma mostly arises in the bone marrow (intramedullary), presenting as the multiple myeloma (MM) or solitary plasmacytoma of bone. In contrast, extramedullary plasmacytoma (EMP) is a rare entity, which accounting for about 3% of the plasmacytoma [1]. EMP most commonly affects the upper respiratory tract and nasopharynx, and rarely originates from the gastrointestinal tract. Gastrointestinal EMP occupies about 5%

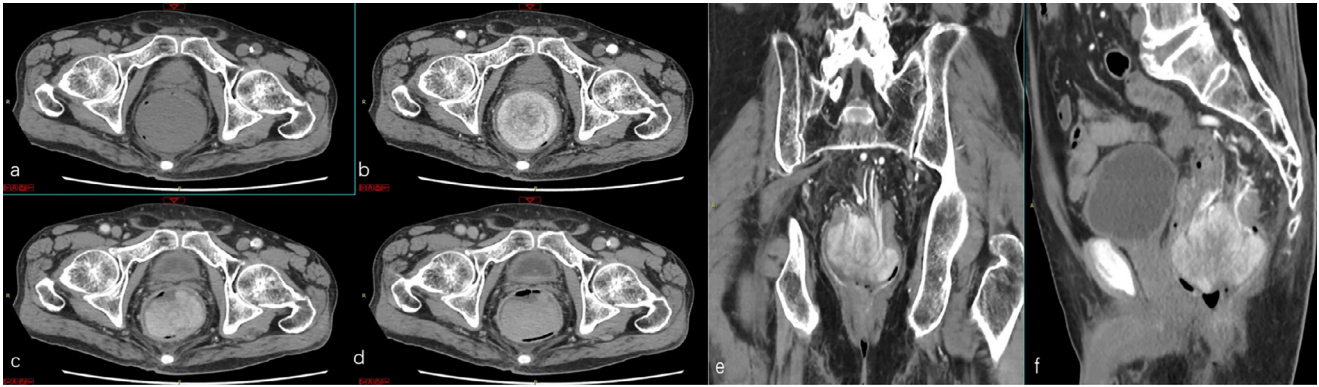
of all EMPs [2]. It often occurs in the small bowel, followed by the stomach and colon [3]. It is extremely rare to occur in the large intestine. Clinical manifestations are variable depending on the location and invasive depth of tumor [4]. The common clinical presentation of a gastrointestinal EMP is abdominal pain, change of bowel habit, and bowel obstruction. In addition, gastrointestinal bleeding has also been reported [4,5]. The average age of onset is 55 years old, with a male predilection [6]. Diagnostic and treatment plan of gastrointestinal EMP requires the multidisciplinary cooperation including the hematologist, surgeon, radiologist, and pathologist [6]. In this paper, we are going to present a case of rectal EMP and review the relevant reports about the EMP in the PubMed.

Abbreviations: EMP, extramedullary plasmacytoma; MM, multiple myeloma; TSGF, tumor-specific growth factor; PET-CT, positron emission tomography-computed tomography; ESD, endoscopic submucosal dissection; EMR, endoscopic mucosal resection.

\* Corresponding author.

E-mail address: [caixran@jnu.edu.cn](mailto:caixran@jnu.edu.cn) (X.-R. Cai).<https://doi.org/10.1016/j.radcr.2021.01.023>

1930-0433/© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)



**Fig. 1 – (a-f) (a)-(d) Axial CT. (a) Noncontrast CT scan shows a round intraluminal mass in the rectum. (b-d) After the administration of contrast medium, the mass appears as the obviously heterogeneous enhancement on arterial and venous phases. Feeding arteries are noted in coronal (e) and sagittal (f) images on arterial phase.**

## Case presentation

An 80-year-old male patient complained of the change of his bowel habit and inhibited defecation in the past 1 month. The symptoms included the increased frequency of defecation, such as more than once a day and even up to 6 times a day, or constipation. Besides, the stools became more loose and even watery occasionally. He was admitted to a local hospital and performed the colonoscopy, which found the mass in rectum. In order to receive the further diagnosis and treatment, he referred to the gastrointestinal surgery department of our hospital. Digital rectal examination revealed a palpable solid mass approximately 8 cm from the anal verge with the poor activity. No blood was found on the finger sleeve. Routine blood test suggested the identified anemia (RBC  $3.38 \times 10^{12}/L$ , hemoglobin 94 g/L) and elevated malignant tumor-specific growth factor (77 U/mL). However, the tumor markers including CA153, CA125, CA199, CEA, and AFP were all within the normal limits. Bone marrow biopsy demonstrated the active proliferation in granulocyte system and nucleated red cell, as well as the appearance of abnormal plasma cells. He had no history of MM.

The patient underwent the enhanced computed tomography (CT) scan of the whole abdomen. CT images (Fig. 1a-f) confirmed a 6.3 cm  $\times$  5.0 cm  $\times$  4.2 cm circumscribed mass protruding into the lumen, which was approximately 5.1 cm from the anal verge. On unenhanced CT scanning, the mass was iso-attenuating and the CT value was about 36 HU. After the administration of contrast medium, the mass showed the obviously heterogeneous enhancement on arterial phase, the persistent enhancement on venous and parenchymal phases. The boundary between the mass and the normal rectal wall was clear and the adjacent fat was intact with absence of swollen lymph node. Besides, the tumoral feeding arteries were found on arterial phase and some feeding vessels originated from the abdominal aorta.

Trans-anal biopsy suggested a malignant tumor and so the surgeons decided to perform a radical resection of the mass by laparoscope. The final pathological result displayed a large number of neoplastic plasma cells in the microscope,

which were closely arranged. Immunohistochemistry (Fig. 2b-d) of the specimen manifested CD138(+), CD38(+), kappa (+), LCA(+), Ki 67 (about 60%), lambda(-), CD56(-), AE1/AE3(-), and CD34(-), which was consistent with a moderately differentiated plasma cell tumor. The further bone marrow aspiration excluded MM. The serum protein electrophoresis and routine urine test were normal. Therefore, this case was finally diagnosed with primary plasmacytoma of rectum.

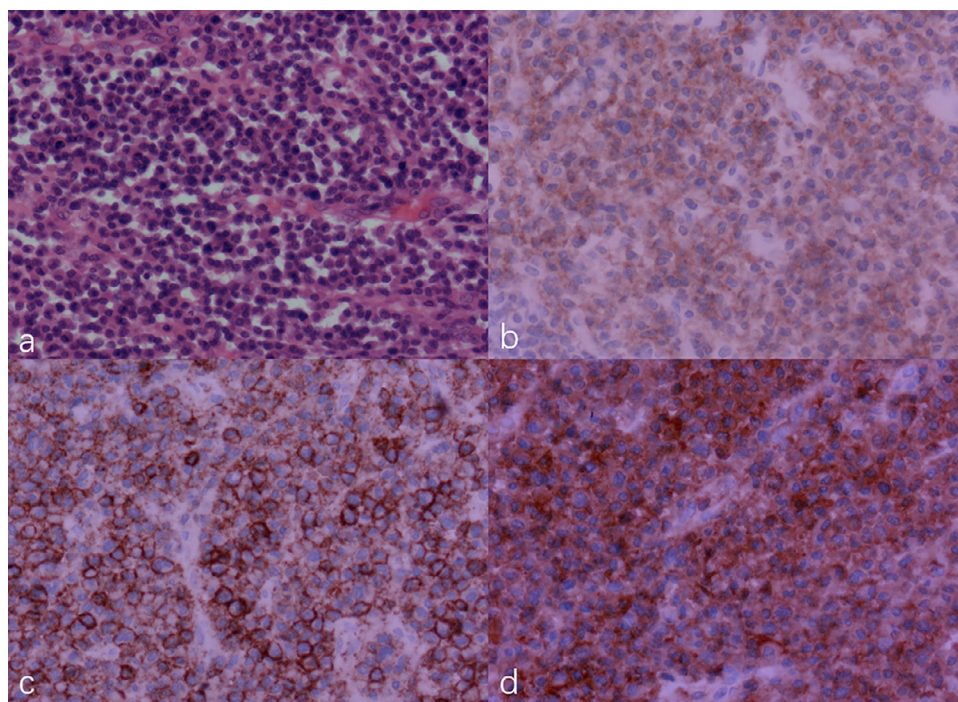
## Discussion

The majority of EMP are accompanied with MM and the primary gastrointestinal EMP without MM is extremely rare. Gastrointestinal EMP can be solitary or accompany, precede, or occur secondary to the onset of MM [7]. Making a diagnosis of primary EMP must rule out the MM, which is decided by the absence of Bence-Jones protein in the urine, normal serum electrophoresis, and normal bone marrow biopsy [8]. The case we presented has no indications of clinical, laboratory or imaging findings of MM, which satisfies the criteria of primary EMP.

In this paper, we not only present a case, but also conduct a case review of EMPs in the gastrointestinal tract. In recent years, EMP located in the gastrointestinal tract has been reported occasionally. After making systematic inquiries on PubMed from 2010 to now, about 22 case reports of primary gastrointestinal EMPs were reported, including 7 in stomach, 2 in duodenum, 1 in small bowel, 3 in ileocecum, 1 in appendix, 6 in colon, and 2 in rectum.

The clinical symptoms of gastrointestinal EMP appear as nonspecific. In our case, the patient presented as the change of bowel habit. EMPs of the gastrointestinal tract usually manifest with nonspecific abdominal pain, abdominal mass, gastrointestinal bleeding, vomiting, changes in bowel habits, bowel obstruction, and intussusception, which have all been previously reported [5,9,10].

Bone marrow and pathological biopsies are the indispensable examinations for confirmation of EMP. Bone marrow biopsy has no evidence of peripheral plasmacytosis or clonal marrow. Microscopic findings of pathological biopsies



**Fig. 2 – (a, x 200) H&E examination revealed a dense of neoplastic plasma cells. (b-d, x 200) Histological findings: CD38(+), CD138(+), kappa (+).**

show the dense of neoplastic plasma cells and intranuclear immunoglobulin inclusion [11]. Normal serum protein electrophoresis and absence of Bence-Jones protein in urine exclude the MM. Besides, the imaging modalities, such as CT, magnetic resonance imaging (MRI) and positron emission tomography-computed tomography (PET-CT) are usually also used to detect the EMP before the surgery. CT and MRI scan can confirm the position and size of the tumor, the relationship with the adjacent organs, and whether there is lymph node metastasis [2]. PET-CT can show underlying bone lesions and find out the area of hypermetabolism, which reveals existence of malignant lesions [2]. As a matter of fact, endoscopy and biopsy play an essential role in diagnosis of gastrointestinal lesions.

CT images generally demonstrate an infiltrative tumor or mass with the clear boundary. When the tumor is large enough, it may appear as the necrotic area. From now on, there is no specific imaging feature about EMP [12]. In the cases we reviewed, CT imaging findings are varied. First, it presents as a well-demarcated mass inside or outside the lumen of the gastrointestinal tract with homogeneous enhancement. In one previously published case, an extraluminal mass with well-defined margin was seen in the stomach, appearing as the homogeneous density with gradual enhancement [13]. Besides, EMP in the gastrointestinal tract could show focal wall thickening and luminal narrowing with obvious enhancement [9,14–16]. Sometimes, due to the thickening of the intestinal wall, it can cause the obstruction in the upper segment of the intestine [17]. Occasionally, peripheral lymph node swelling, the serosal layer of the intestine involved and infiltration of the surrounding fatty tissue can be observed. In another case,

a small intestinal tumor was accompanied by a psoas mass [6]. What's more, in one case with ileocecal plasmacytoma due to perforation, the CT scanning showed pneumoperitoneum and inflammatory exudation around the terminal ileum and cecum [18].

On MRI scanning, Ryu and Cohen-Hallaleh [12] reviewed 21 patients who were diagnosed as EMP, which appear as iso-to-hypointensity to the muscle on T1-weighted image, iso/hyperintensity on T2-weighted image and variable enhancement. Diffusion-weighted imaging revealed the restricted diffusion within the tumor. Actually, due to artifacts of the bowel peristalsis, CT and endoscopy will be the first choice for tumors of the gastrointestinal tract rather than MRI.

PET-CT could locate the area of hypermetabolism, which is used to identify the malignant lesions, but not for the qualitative diagnosis. Because of hypermetabolism in EMPs, we can observe the accumulation of fludeoxyglucose uptake in PET-CT [6,15].

From now, there is no report to describe the imaging feature of EMP optimally [19]. Hence, imaging examinations have a limited role in identifying EMP of gastrointestinal tract and may be misdiagnosed as other diseases, such as carcinoma, stromal tumors, lymphoma, metastases, or inflammatory bowel disease [20]. Therefore, the accurate diagnosis of gastrointestinal EMP requires the histopathological analysis. What's more, bone marrow biopsy can help us to rule out MM.

There is still no unified guideline for treatment of gastrointestinal EMP because of its rarity. Though plasma cell tumors are highly radiosensitive, surgical resection has been proposed as the first choice for gastrointestinal EMP. Radiotherapy is more commonly used for EMP of head and neck.

Besides, radiation therapy can be the additional therapy for incomplete resection, lymph node metastasis or recurrence [21]. For the past few years, endoscopic therapy such as endoscopic submucosal dissection or endoscopic mucosal resection has increasingly become a substitute of surgery for gastrointestinal EMP [11,22].

Generally, EMP has an indolent process and better prognosis than MM and solitary intramedullary plasmacytoma, with a 70% of 10-year survival rate [21]. Since EMP may recur or progress to MM in a few cases, long-term follow-up is recommended and necessary.

In conclusion, EMP of the gastrointestinal tract is extremely rare, and its clinical and imaging manifestations are not specific. Therefore, bone marrow examination and histopathological analysis are the golden standard for diagnosis. EMP in the gastrointestinal tract prefers the surgical resection. Postoperative follow-up is indispensable.

#### REFERENCES

- [1] Strojanc P, Soba E, Lamovec J, Munda A. Extramedullary plasmacytoma: clinical and histopathologic study. *Int J Radiat Oncol Biol Phys* 2002;53(3):692–701.
- [2] Zhang D, Cao D, Shen D, Mulmi Shrestha S, Yin Y. Extramedullary plasmacytoma occurring in ileocecum: a case report and literature review. *Medicine (Baltimore)* 2017;96(51):e9313.
- [3] Ding W, Tan Y, Qian Y, Xue W, Wang Y, Xi C, et al. Primary plasmablastic plasmacytoma in the stomach of an immunocompetent adult: a case report. *Medicine (Baltimore)* 2019;98(4):e14235.
- [4] Comba IY, Torres Luna NE, Cooper C, Crespo M W, Carilli A. A rare case of extramedullary plasmacytoma presenting as massive upper gastrointestinal bleeding. *Cureus* 2019;11(1):e3993.
- [5] Iosif E, Rees C, Beeslaar S, Shamali A, Lauro R, Kyriakides C. Gastrointestinal bleeding as initial presentation of extramedullary plasma cell neoplasms: a case report and review of the literature. *World J Gastrointest Endosc* 2019;11(4):308–21.
- [6] Glasbey JC, Arshad F, Almond LM, Vydianath B, Desai A, Gourevitch D, et al. Gastrointestinal manifestations of extramedullary plasmacytoma: a narrative review and illustrative case reports. *Ann R Coll Surg Engl* 2018;100(5):371–6.
- [7] Lu T, Pu H, Zhao G. Primary pancreatic plasmacytoma: a rare case report. *BMC Gastroenterol* 2017;17(1):167.
- [8] Wallace E, Stewart Z, Theriot D, Shaffer W, Guillory S, Hanemann M. Atypical presentation of extramedullary plasmacytoma. *Ochsner J* 2018;18(1):101–3.
- [9] Lee SH, Ahn BK, Baek SU, Chang HK. Primary isolated extramedullary plasmacytoma in the colon. *Gastroenterol Res* 2013;6(4):152–5.
- [10] Mjoli M, Vorajee N, Naidoo Y, Madiba T. Solitary extramedullary plasmacytoma of the colon, rectum and anus. *S Afr J Surg* 2016;54(2):45–7.
- [11] Park CH, Lee SM, Kim TO, Kim DU, Jung WJ, Kim GH. Treatment of solitary extramedullary plasmacytoma of the stomach with endoscopic submucosal dissection. *Gut Liver* 2009;3(4):334–7.
- [12] Ryu SW, Cohen-Hallaleh V. Imaging features of extramedullary plasmacytoma. *J Med Imaging Radiat Oncol* 2020;64(1):44–51.
- [13] Zhao ZH, Yang JF, Wang JD, Wei JG, Liu F, Wang BY. Imaging findings of primary gastric plasmacytoma: a case report. *World J Gastroenterol* 2014;20(29):10202–7.
- [14] Fukuhara S, Tazawa H, Okanobu H, Kida M, Kido M, Takafuta T. Successful treatment of primary advanced gastric plasmacytoma using a combination of surgical resection and chemotherapy with bortezomib: a case report. *Int J Surg Case Rep* 2016;27:133–6.
- [15] Han YJ, Park SJ, Park MI, Moon W, Kim SE, Ku KH. Solitary extramedullary plasmacytoma in the gastrointestinal tract: report of two cases and review of literature. *Korean J Gastroenterol* 2014;63(5):316–20.
- [16] Zihni İ, Dinç R, Canpolat S, Cengiz F, Uslu A. Extramedullary plasmacytoma of the colon: a case report. *Ulus Cerrahi Derg* 2014;30(4):231–3.
- [17] Zhu Z, Cao H, Chen L. Incomplete colonic obstruction caused by extramedullary plasmacytoma. *Clin Gastroenterol Hepatol* 2017;15(3):e69–70.
- [18] Gabriel EM, Savu M. Discovery of a rare ileocecal plasmacytoma. *J Surg Case Rep* 2014;2014(3):1–4.
- [19] Çelebi İ, Bozkurt G, Polat N. Tonsillar plasmacytoma: clues on magnetic resonance imaging. *BMC Med Imaging* 2018;18(1):19.
- [20] Gupta V, Nahak B, Sakhuja P, Agarwal AK, Kumar N, Mishra PK. Primary isolated extramedullary plasmacytoma of colon. *World J Surg Oncol* 2007;5:47.
- [21] Miwa W, Hiratsuka T, Tei S, Sato K, Yo K. Solitary extramedullary plasmacytoma of the rectum complicating ulcerative colitis. *Clin J Gastroenterol* 2019;12(2):160–5.
- [22] Kodani T, Osada T, Terai T, Ohkusa T, Shibuya T, Sakamoto N. Successful endoscopic mucosal resection of a solitary extramedullary plasmacytoma in the sigmoid colon. *Endoscopy* 2011;43(Suppl 2 UCTN):E298–9.