Clinical Case Report

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Primary plasmablastic plasmacytoma in the stomach of an immunocompetent adult

A case report

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Abstract

Rationale: Extramedullary plasmacytomas (EMP) are tumors composed by a monoclonal population of plasma cells that arise in extraosseus tissues, occupying <5% of all plasma cell neoplasms. Gastrointestinal solitary extramedullary plasmacytoma is rare, just comprises about 5% of all EMPs. The most common site is small intestine. The gastric incidence is much rare and especially the plasmablastic plasmacytoma in the stomach.

Patient concerns: A 65-year-old man had an epigastric discomfort and mass for about 2 months. Gastroscopy revealed a malignant tumor in the gastric body. Abdomen computed tomography (CT) showed that the gastric cavity was filled, and the irregular soft tissue shadow was seen in the greater curvature, and the enhancement was obvious. To get more tissue, we conducted stomach puncture biopsy. Pathology showed small-round cell malignant tumors. And immunohistochemical examinations revealed that the tumor tend to be a plasma cell tumor.

Diagnosis: Gastric plasma cell tumor.

Interventions: Distal gastrectomy was performed to treat the tumor.

Outcomes: In addition to ascites caused by hypoproteinemia, there were no postoperative complications. Postoperative pathologic report showed plasmablastic plasmacytoma. Histopathologic examination of the specimen revealed plasmablastic plasmacytoma originating in the stomach with transmural extension, but without lymph node metastasis. The patient is regularly followed up at a postoperative clinic and is doing well, and at present there is no plan for adjuvant treatment.

Lessons: Surgical resection is good option for gastrointestinal EMP.

Abbreviations: CT = computed tomography, EBV = Epstein-Barr virus, EMP = extramedullary plasmacytoma, MALT = mucosa-associated lymphoid tissue, MM = multiple myeloma, MRI = magnetic resonance imaging, PET-CT = positron emission tomography-computed tomography, SPB = solitary plasmacytoma of bone.

Keywords: extramedullary plasmacytoma, gastric, plasmablastic

1. Introduction

Plasmacytoma is a plasma cell dyscrasia in which a plasma cell tumor grows within the axial skeleton or within soft tissue. In 2003, The International Myeloma Working Group divided it into 3 categories: solitary plasmacytoma of bone (SPB); extramedullary plasmacytoma (EMP), and multiple plasmacytomas that are either primary or recurrent.[1] Extramedullary plasmacytomas are tumors composed by a monoclonal population of plasma cells that arise in extraosseus tissues, occupying <5% of all plasma cell neoplasms, normally presenting as localized lesions, usually in the head and neck region.[2] Gastrointestinal solitary extramedullary plasmacytoma is rare, just comprises about 5% of all EMPs.[3] The most common site is small intestine. The gastric incidence is much rare and especially the plasmablastic plasmacytoma in the stomach. In this study, we report a case of primary plasmablastic plasmacytoma in the stomach of an immunocompetent adult.

2. Patient consent

Patient has provided informed consent for publication of the case. This case report was approved by the ethical committee of Wujin Hospital affiliated to Jiangsu University.

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DW and YT have contributed equally to this manuscript as co-first authors.

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Ethics approval and consent to participate: This case report was approved by the ethical committee of Wujin Hospital affiliated to Jiangsu University.

Consent for publication: Not applicable.

The authors declare no conflicts of interest.

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3. Case

In June 2018, a 65-year-old man presented at an outside facility with an epigastric discomfort and mass for about 2 months. The movable mass started as a small lump and gradually increased in size. Laboratory values were unremarkable. Gastroscopy revealed a malignant tumor in the gastric body. Pathology showed small cell malignant tumors. Histomorphometry showed that the tumor originated in the lymphoid and hematological system. There was too little biopsy tissue to determine the differentiation. In July 2018, the patient presented to our hospital for further diagnosis and treatment. We performed upper gastrointestinal contrast to identify the tumor site. Barium x-ray revealed that the tumor was located in the gastric body and about 102 mm size (Fig. 1). Abdomen computed tomography (CT) showed that the gastric cavity was full of chyme, and the irregular soft tissue shadow was seen in the greater curvature, and the enhancement was obvious (Fig. 2). It was considered as a malignant tumor of gastric body. To get more tissue, we conducted stomach puncture biopsy. Pathology showed small-round cell malignant tumors. And immunohistochemical examinations revealed that the tumor tend to be a plasma cell tumor. To exclude associated multiple myeloma (MM), patient underwent bone marrow biopsy. His bone marrow morphology test, peripheral blood smear, and serum protein electrophoresis were found to be normal. In September 2018, the patient underwent distal gastrectomy. The tumor located in the anterior wall of the stomach body near the greater curvature (Fig. 3). The tumor was a centrally ulcerated, relatively well-demarcated bulky mass, measuring 90×90 mm in diameter (Fig. 4). Histopathologic examination of the specimen revealed plasmablastic plasmacytoma originating in the stomach with transmural extension (Fig. 5), but without lymph node metastasis. These cells were positive for CD38, CD138, MUM-1, bcl-2, and CD23 and negative for CD3, CD20, CD79a, CyclinD1, CD10, bcl-6, CD5, CK, EMA, and CD30, with a Ki-67 proliferative index of about 70% (Fig. 6). The patient had a good postoperative recovery and other than ascites caused by hypoproteinemia, there was no complications and the patient was successfully discharged. To October 1, he is regularly followed up at a postoperative clinic and is doing well, and there is no plan for adjuvant treatment.

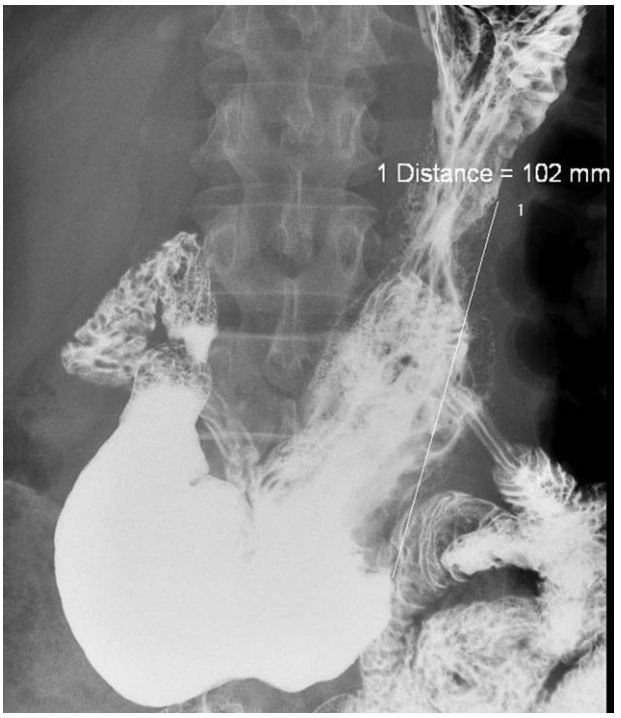

Figure 1. Barium x-ray revealed that the tumor was located in the gastric body and about 102 mm size.

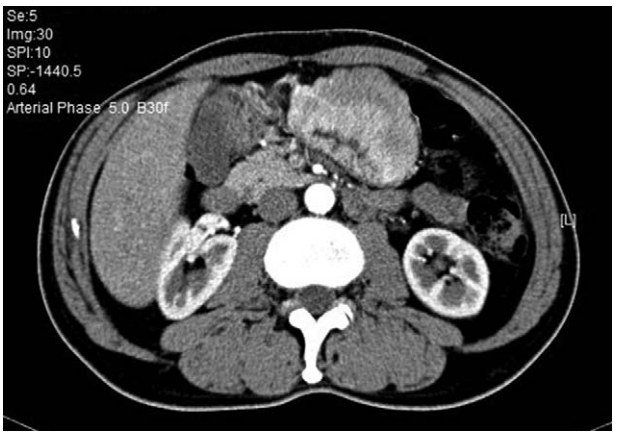

Figure 2. Abdomen computed tomography showed that the gastric cavity was filled, and the irregular soft tissue shadow was seen in the greater curvature, and the enhancement was obvious.

4. Discussion

EMP comes from soft tissue, which is solitary and has no evidence of MM. The incidence of EMP is lower than SBP, accounts for only 3% to 5% of all plasma cell disease.[4] 80% of EMPs occur in the upper aerodigestive tract, including the oropharynx and nasopharynx, nasal cavities, sinuses, and larynx. Some gastrointestinal plasmacytomas actually may represent mucosa-associated lymphoid tissue (MALT) lymphomas with extreme plasmacytic differentiation.[5] Gastrointestinal involvement occurs in only 5% of patients with extramedullary involvement and most commonly affects the small bowel, followed by the stomach, colon, and esophagus, in order of frequency of

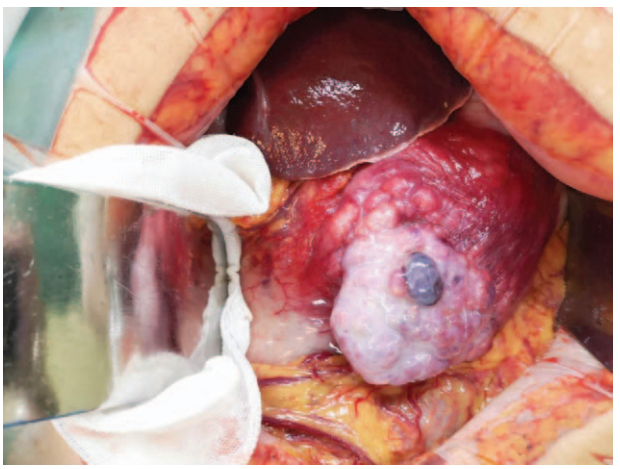

Figure 3. The tumor located in the anterior wall of the stomach body near the greater curvature.

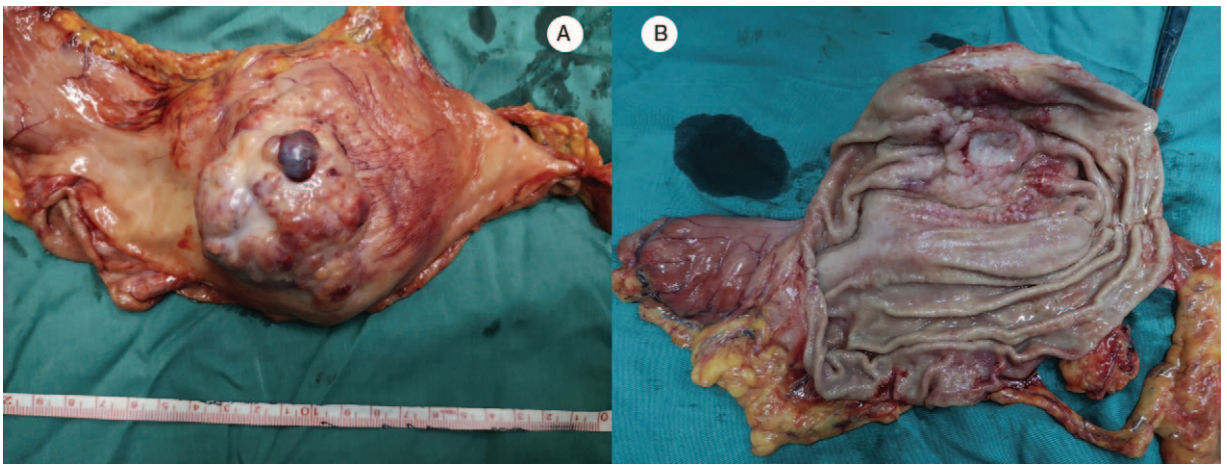

Figure 4. The tumor was a centrally ulcerated, relatively well-demarcated bulky mass, measuring 90 × 90 mm in diameter. A. outside view, B. inside view.

involvement.[1,6,7] EMP ratio of male to female is 4:1. And the disease is not common in children or adolescent.[8]

Plasmablastic plasmacytoma is a quintessential undifferentiated round cell tumor.[9] Highly aggressive plasmablastic plasmacytomas contain mainly plasmablasts and resemble plasmablastic lymphoma. It is possible that there could be a genetic overlap between plasmablastic lymphoma and plasmablastic plasmacytoma. These 2 tumors may be different manifestations of malignancies with a common derivation from B-cells at a later stage of B-cell maturation, and a common genetic feature that imparts a plasmablastic morphology and an aggressive clinical course.[10] The high grade anaplastic cell morphology brings into the differential diagnosis, various malignancies including—undifferentiated carcinoma, lymphoma, rhabdomyosarcoma, melanoma, neuroendocrine tumor, and neuroblastoma, all having different biological characteristics, predicators, treatment protocols, and prognoses.[11]

Increasing evidence showed that *Helicobacter pylori* (*H pylori*) infection was involved in the pathogenesis of primary gastric plasmacytoma.[12–14] According to current research of the multistep pathogenesis of plasma cell disorders, it is possible that antigenic material originating from the *H pylori* may stimulate lymphoplasmacytic clones to produce initially a polyclonal and subsequently a monoclonal immune response.[15,16] Papadaki et al[17] reported a patient having a stage I primary gastric plasmacytoma completely regressed after *H pylori* eradication. Evidence of Epstein-Barr virus (EBV) infection is common in plasmablastic lymphoma, but uncommon in plasmacytoma and therefore helpful in differential diagnosis.[18] Although most EMPs occur in head and neck area, a region rich in EBV-associated tumors, these tumors are rarely positive for EBV, especially in immunocompetent hosts.[19] However, EBV-associated plasmacytomas are also reported.[20–22] Unfortunately, We did not know whether this patient had *H pylori* or EBV infection.

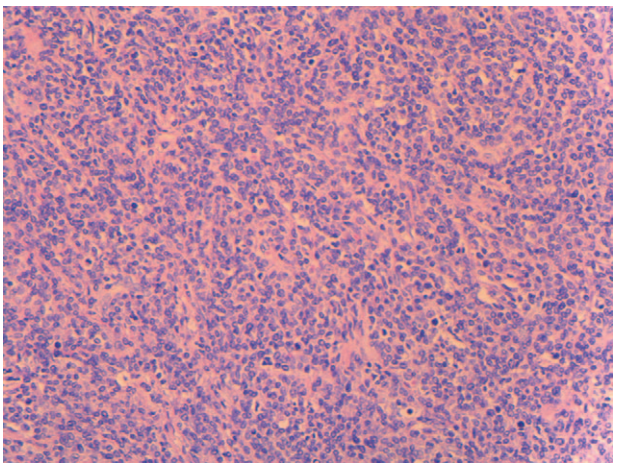

Figure 5. Histopathologic examination revealed plasmablastic plasmacytoma (HE, ×200).

Gastrointestinal EMPs could present as anorexia, weight loss, nausea, abdominal pain, vomiting, occult blood loss, abdominal mass, and rarely with overt gastrointestinal bleeding. So we should take more examinations for further diagnosis. It tends to be identified at a late stage if an endoscopic examination is not performed.[7] The endoscopic appearance of gastric EMP varies—thickened folds, polyposis, ulcers, ulcerated masses (as in the present case), plaque-like involvement, and even linitis plastica, which needs to be differentiated from gastric cancer.[5] There are some accessory examinations to help us diagnose EMP, such as CT scan, magnetic resonance imaging (MRI), or positron emission tomography-computed tomography (PET-CT), especially gastric puncture cytology and bone marrow biopsy. Gastric puncture cytology could make up for the deficiency of gastroscope biopsy.[23] Bone marrow biopsy could help to identify MM. CT scan and MRI can help to find parenchymal lesions and the size and location of the lesion and the relationships with surrounding organizations. PET-CT can find out the position of hypermetabolism which can identify probability of malignancy. For gastrointestinal EMP, endoscopy and biopsy have practical significances of diagnosis. However, Barium x-ray is not sensitive to EMPs, because EMPs don't derive from the mucosa.

For most EMPs occur in upper aerodigestive tract, which is difficult to have an operation, local radiotherapy is the preferred therapeutic modality.[24] However, the instant surgical resection of the mass is another good option for gastrointestinal EMP, having the lowest recurrence rate. The previous investigation provides evidence that surgery alone gives the best results in cases

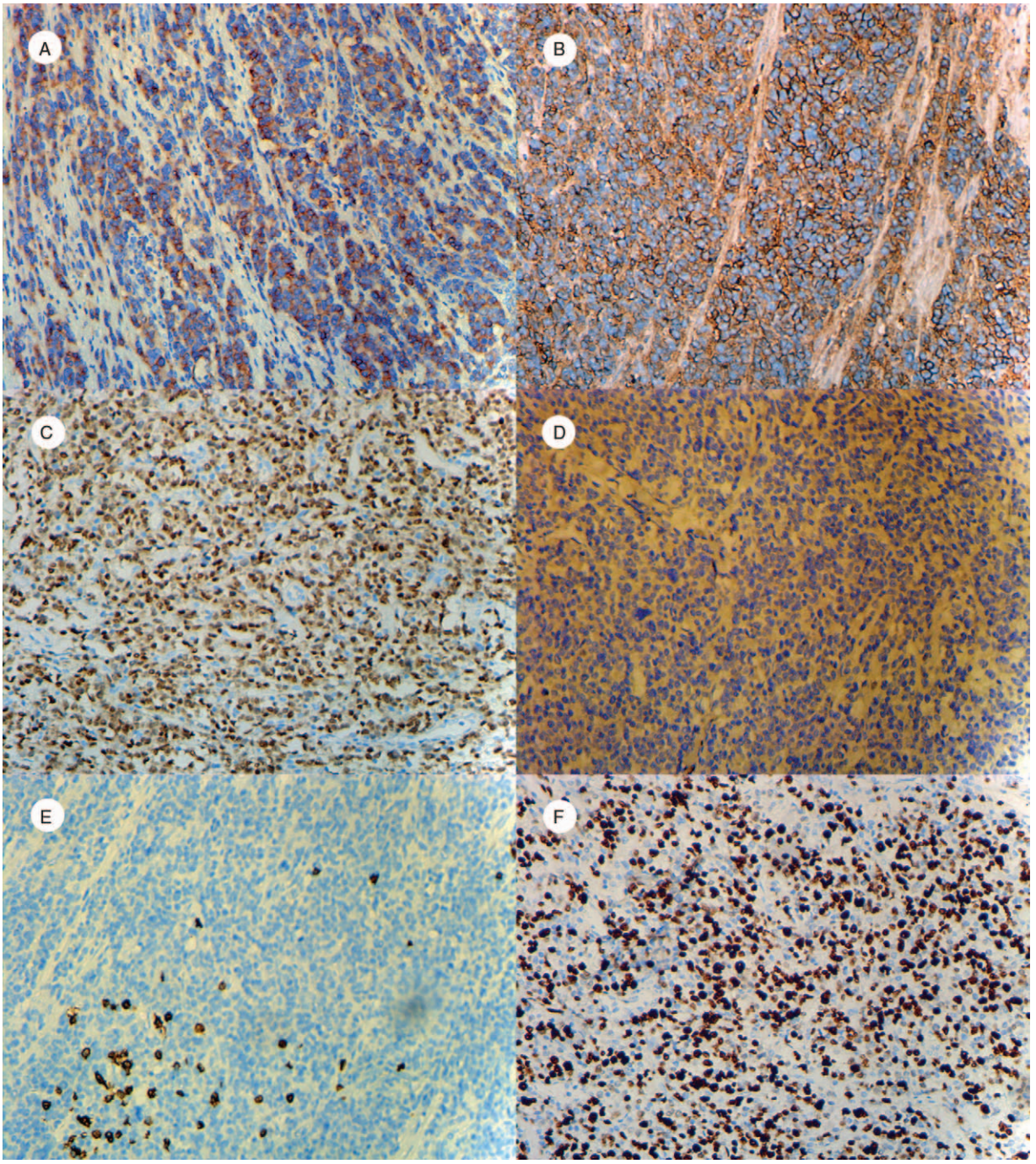

Figure 6. Immunohistochemistry for the specimens presented CD38(+), CD138(+), MUM-1(+), bcl-2(+), CD23(+), CD3(–), CD20(–), CD79a(–), CyclinD1(–), CD10 (–), bcl-6(–), CD5(–), CK(–), EMA(–), CD30(–), Ki67(about 70%) (×200). A. CD38(+), B. CD138(+), C. Mum-1(+), D. bcl-2(+), E. CD20(–), F. Ki67(about 70%).

of EMP when it is resectable. However, if complete surgical tumor resection is doubtful or impossible and/or if lymph node areas are affected, then combined therapy (surgery and radiation) is recommended.[25] In this case, the tumor was completely removed and there were no metastatic lymph node, so no further treatment was required. In certain patients for whom surgery is contraindicated, radiotherapy is applied.

EMP had better prognosis than MM and SBP.[26] After treatment for EMP in non-upper aerodigestive regions, 64.7% of all patients had no recurrence or MM, 21.2% had recurrence, and 14.1% had converted to MM.[25] Despite the typical presentation as a locally destructive tumor, plasmacytoma is highly radiosensitive, and 70% to 80% survival may be obtained with the use of radiotherapy.[27] Most patients died of non-EMP related disorders and more than two-thirds of patients survived for >10 years.[28,29] We were fortunate to have the neoplasm complete resection, and prevented him from radiotherapy-related complications.

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Author contributions

Xuezhong Xu performed this operation. Yulin Tan, Wenbo Xue, and Wei Ding participated in this operation. Kefeng Gu carried on the pathology analysis. Yibo Wang, Cheng Xi, and Yixin Xu participated in perioperative management. Wei Ding and Yan Qian designed the study and wrote the manuscript. Xuezhong Xu edited the manuscript as the corresponding author.
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Methodology: Yulin Tan, Wenbo Xue, Kefeng Gu.
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Resources: Yibo Wang, Cheng Xi, Yixin Xu.
Writing – original draft: Wei Ding, Yan Qian.
Writing – review & editing: Xuezhong Xu.
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