

CASE REPORT

위장관에 발현한 고립 골수외 형질세포종: 두 증례 보고 및 문헌 고찰

한유진, 박선자, 박무인, 문 원, 김성은, 구기환, 옥소영

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Solitary Extramedullary Plasmacytoma in the Gastrointestinal Tract: Report of Two Cases and Review of Literature

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Solitary extramedullary plasmacytoma (EMP) is a plasma cell neoplasm without bone marrow involvement. EMPs are rare in the gastrointestinal (GI) tract. We report two cases of primary EMP, one in the transverse colon and the other in the stomach. In the first case, a mass on the transverse colon was found on colonoscopy. The patient underwent left hemicolectomy and has been followed-up for 3 years without recurrence postoperatively. The latter case had several masses in the stomach. The patient underwent bypass surgery and has received supportive care for 1 month. Histopathologic specimens of both the cases showed a monoclonal lambda chain EMP. Subsequent investigations included a bone marrow biopsy, serum IgA, IgG, IgM and serum protein electrophoresis, and the results were negative for multiple myeloma in both the cases. Solitary EMP in the GI tract can be mistaken for colon cancer or stomach cancer on endoscopy; therefore, a sufficient number of biopsy specimens can help diagnose solitary EMPs. Surgical resection alone or with radiation therapy in cases with positive surgical margin is currently the only treatment for solitary EMP in the GI tract. Further study is necessary to determine disease prognosis and to investigate other treatment methods. (*Korean J Gastroenterol* 2014;63:316-320)

Key Words: Solitary extramedullary plasmacytoma; Stomach; Transverse colon

INTRODUCTION

Solitary extramedullary plasmacytoma (EMP) is a plasma cell neoplasm that does not include bone marrow involvement. Compared to multiple myeloma, the most common primary bone cancer in adults,¹ EMPs are uncommon tumors constituting 3-5% of all plasma cell neoplasms.² EMPs usually present in the upper respiratory tract or oropharyngeal area; however, a few cases of EMPs occurring in the gastro-

intestinal (GI) tract have been reported.^{3,4} This study reports on two cases of EMP in the GI tract: one in the transverse colon and the other in the stomach.

CASE REPORT

1. Case 1

A 49-year-old man was admitted with complaints of periumbilical abdominal pain that had continued for one year pri-

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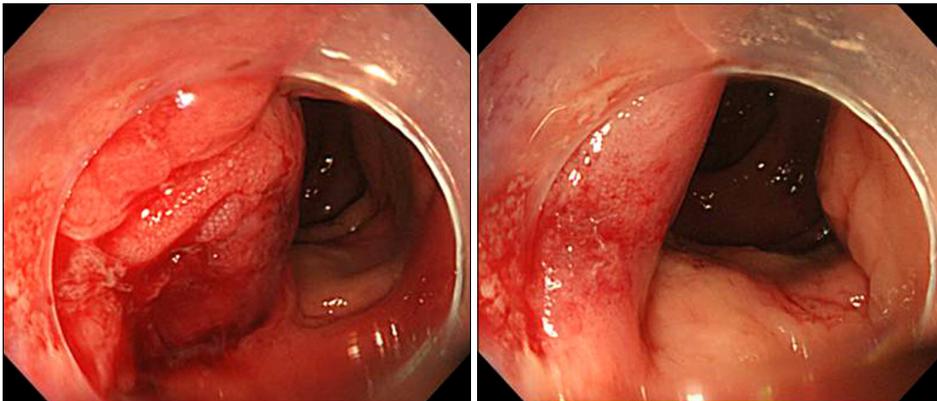


Fig. 1. Colonoscopy showed edematous and nodular mucosal changes 45-55 cm from the anal verge, and a mass-like lesion that protruded into the lumen of the transverse colon 50 cm from the anal verge.

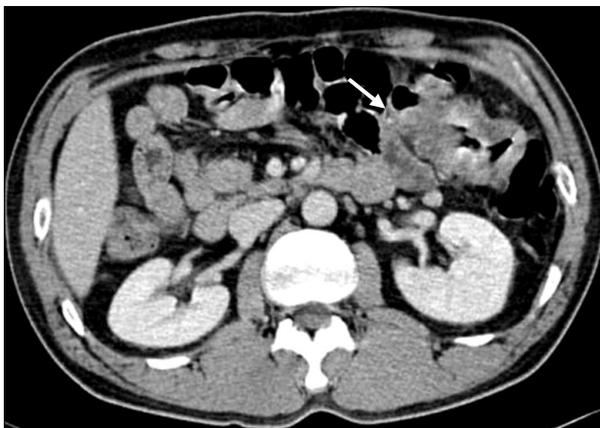


Fig. 2. CT showed enhancing circumferential wall thickening and luminal narrowing (arrow) in the transverse colon (portal phase).



Fig. 3. Hypermetabolism in the transverse colon (arrow) (max SUV=7.7) without distant metastasis was observed on PET-CT.

or to presentation. Before admission to our hospital, he visited a local medical center and underwent a colonoscopy. The colonoscopy showed luminal narrowing with a mass 50 cm from the anal verge (AV), and he was transferred to our hospital in order to undergo further evaluation. The findings of colonoscopy performed at our hospital also showed edematous and nodular mucosal changes 45-55 cm from the AV and a mass protruded into the lumen of the transverse colon 50 cm from the AV (Fig. 1). Biopsy obtained during colonoscopy suggested malignancy, however, we were unable to confirm the type of malignancy since immunohistochemical test results, including cytokeratin (cocktail), cytokeratin 20, myeloperoxidase, CD3, and CD 20, were negative. Complete blood count and routine biochemistry laboratory studies were within normal limits. Serum CEA was 2.27 ng/mL and CA 19-9 was below 0.60 U/mL. CT scan showed an enhanced circumferential wall thickening and luminal narrowing in the trans-

verse colon (Fig. 2), which infiltrated into the pericolic fat plane, with enlargement of the pericolic lymph nodes. Hypermetabolism in the transverse colon (max SUV=7.7) without distant metastasis was observed on PET-CT (Fig. 3) and an extended laparoscopic left hemicolectomy was subsequently performed. There were no significant complications related to the operation, and gross findings showed a relatively well-demarcated mass measuring 1.7×1.5 cm in size involving the muscularis propria (Fig. 4) and synchronous involvement of eight of 50 regional lymph nodes. Abundant plasma cells were observed in H&E staining light microscopy of the lymph nodes and the mass, and immunohistochemistry indicated that the specimen was positive for lambda light chain, which was indicative of tumor monoclonality, but negative for kappa light chain, CD20,

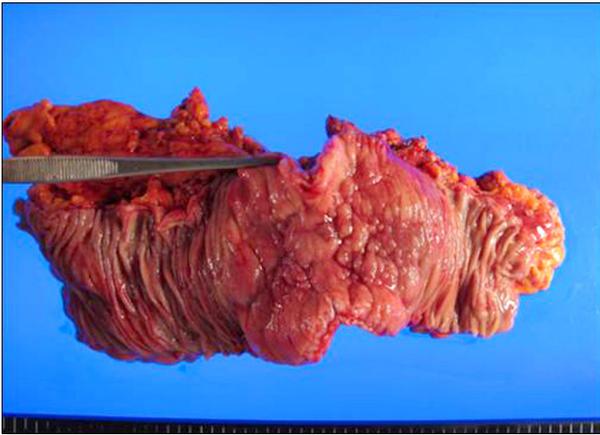


Fig. 4. Gross findings showed a relatively well-demarcated mass measuring 1.7×1.5 cm in size.

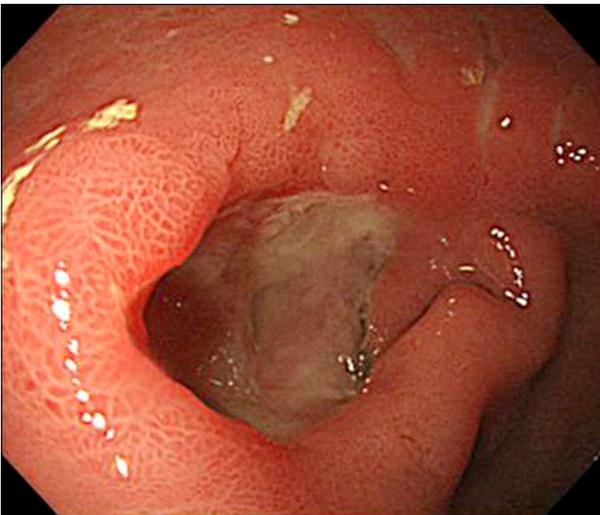


Fig. 5. A mass measuring approximately 3-4 cm in size with sharp margins and central ulceration was observed on the anterior wall near the deformed pylorus ring, which was thought to be Bormann type II gastric cancer.

CD3, and CD45RO. Normocellular marrow was found on bone marrow examination and serum IgG, IgA, and IgM levels were within the normal ranges. In addition, serum protein electrophoresis showed no definite M protein, and Bence-Jones protein was not detected in the urine. No gross bony abnormalities were seen on the skull X-ray and MRI of the whole spine showed no evidence of bone plasmacytoma. The patient has been followed-up for three years without recurrence postoperatively and chemotherapy or radiotherapy was not used as a treatment modality.

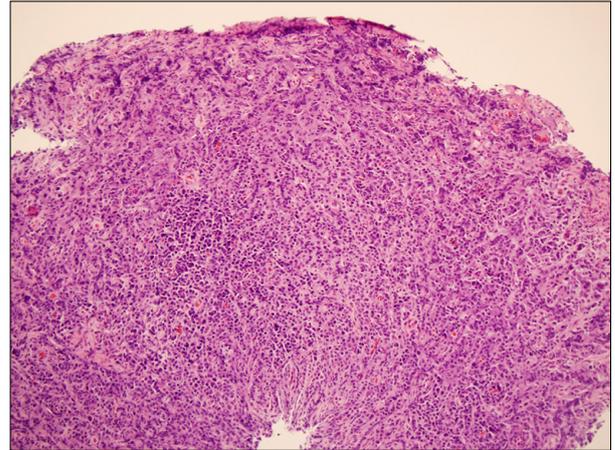


Fig. 6. Repeated endoscopic biopsy indicated plasmacytic infiltration, seen with H&E (×100).

2. Case 2

A 62-year-old man visited our hospital complaining of nasal congestion and rhinorrhea for one month. MRI and endoscopy of the sinuses showed nasal polyps and a nasal polypectomy with septoplasty was performed. Plasmacytoma was found on a biopsy and additional diagnostic tests were performed. No evidence of plasma cell infiltration was found on bone marrow aspiration and serum IgG, IgA, and IgM levels were within the normal ranges. Serum protein electrophoresis identified no definite M protein and no Bence-Jones protein was detected in the urine. Skull X-ray showed that the mass had destroyed a part of the nasal cavity and the left ethmoid and maxillary sinuses, however, no other bony destruction was seen. PET-CT showed hypermetabolism in the antrum of the stomach. Endoscopy found a mass measuring approximately 3-4 cm in size with central ulceration with sharp margins in the anterior wall near the deformed pylorus ring, which was thought to be Bormann type II gastric cancer (Fig. 5). However, the biopsy showed no evidence of malignancy. A biopsy was repeated in order to obtain a confirmative specimen and plasmacytoma was confirmed with additional biopsies, in which plasmacytic infiltration was observed on H&E staining (Fig. 6). On immunohistochemical staining, the neoplastic cells were positive for lambda light chain and negative for kappa light chain and cytokeratin (Fig. 7). The mass was diagnosed as a solitary EMP in the stomach and its treatment of choice was a curative surgery. However, due to invasion of the mass to adjacent tissues, a curative surgery was not feasible and a palliative gastric bypass surgery was performed

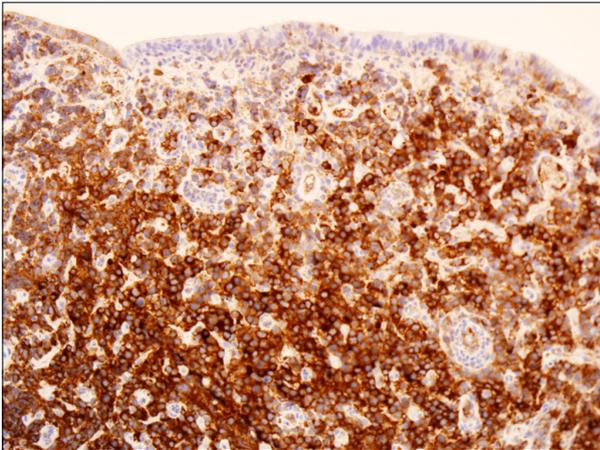


Fig. 7. Immunohistochemical staining revealed positive lambda light chains, indicating that the tumor was monoclonal ($\times 200$).

instead. Currently, the patient has received one month of follow-up care after the bypass surgery, and no additional treatment, such as adjuvant chemotherapy or radiotherapy, has been administered due to general weakness.

DISCUSSION

Solitary plasmacytoma is defined as a localized accumulation of neoplastic monoclonal plasma cells without evidence of a systemic plasma cell proliferative disorder. Solitary plasmacytoma can be classified based on location in solitary plasmacytoma of the bone or solitary EMP.⁵ EMP is a plasma cell neoplasm without bone marrow involvement. It is less common than solitary bone plasmacytoma and constitutes 3-5% of all plasma cell neoplasms.^{2,6} Patients diagnosed with solitary EMP usually present with non-specific symptoms and have no significant abnormal findings in their urine or serum. Often, there are no other diagnostic findings on endoscopy or radiologic studies. Therefore, a biopsy is necessary in order to confirm EMP.⁷ Based on endoscopic findings, we initially suspected that the masses in both cases were colon cancer and stomach cancer, respectively; however, the biopsy results were consistent with plasmacytoma. In the first case, the colonoscopic biopsy specimens suggested malignancy; however, the exact nature of the mass was unclear, and a surgical excision was required for diagnosis. In the latter case, EMP in the stomach was diagnosed with repeated endoscopic biopsies. Therefore, obtaining a sufficient number of biopsy specimens is critical for ac-

curate diagnosis of EMP.

EMPs comprise 3-4% of all plasma cell tumors and occur mainly in the upper respiratory tract (80-90%); the GI tract is involved in only approximately 7% of cases.²⁻⁴ All segments of the GI tract may be involved, and the small bowel is the most common site of occurrence, followed by the stomach, colon, and esophagus.⁸⁻¹⁰ The first case was EMP involving the transverse colon with regional metastasis to pericolic lymph nodes. An extended laparoscopic left hemicolectomy was performed due to concern about the possibility of colon cancer; however, the final biopsy showed that the mass was colonic EMP. The second case was EMP involving the stomach in a patient with maxillary plasmacytoma. We verified that there were no multiple myeloma manifestations such as monoclonal gammopathy or bone marrow plasmacytosis. There was no evidence of solitary plasmacytoma of the bone in either case.

The treatment of choice for EMP in the head and neck area is radiation therapy (RT), while surgical removal should be performed first for patients with EMP that presents in other anatomic regions. RT should be performed after primary surgery if the tumors have an inadequate surgical margin. Endoscopic submucosal dissection of EMP involving only the submucosa of the stomach has also been reported.¹¹ EMP has a better prognosis than other lymphoproliferative diseases, with a 10-year overall survival rate of 70%; when surgical treatment is performed along with RT, the reported median overall survival is 7-9 years.^{5,12} Close follow up of patients with EMP is important because 10-20% of patients progress to multiple myeloma.⁷ Our first patient underwent surgical resection and has been followed-up for three years without recurrence. Gastric bypass surgery was required in the latter case because an adequate resection margin was not obtained. The patient underwent bypass surgery and has received supportive follow-up care for one month.

Solitary EMPs in the GI tract are rare and can easily be mistaken for adenocarcinoma on endoscopy. Consequently, a sufficient number of biopsy specimens are necessary for accurate diagnosis of EMP. Currently, surgical resection alone or with RT in cases of inadequate surgical margins is the only treatment for solitary EMP in the GI tract. Conduct of further study will be necessary in order to determine prognosis and to investigate other treatment methods.

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