Extramedullary Plasmacytoma of the Small Intestine:
A Case Report

Naoyuki Kobayashi,¹ Masatoshi Kawamura,¹ Yoshihumi Niitsu,²
Masako Suzuki,³ Yukiyoshi Esaki,³ Shinji Takahashi,¹
Taiki Masuda,¹ Junjiro Katsurada,¹ Takeo Ogino,¹
Takeharu Kawamura,¹ Takeshi Machida,¹ Nobuyuki Sakamoto,¹
and Sotaro Kanno ¹

Extramedullary plasmacytoma (EMP) of the gastrointestinal tract is an uncommon-type of neoplasms. This report describes a case of EMP in the small intestine. A 78-year-old male, with abdominal discomfort and vomiting, was found to have a 7-cm sized tumor of the small intestine and swollen lymph nodes. He underwent resection of the bowel and lymph nodes. Histological examination of the resected specimens revealed compact tumor cells from mucosa to suberosa and infiltrate into the fat tissue of the mesentery and the lymph nodes. Pathological examination of the resected specimen revealed plasma cell-like round cells, Dutcher bodies, and Russell bodies. Immunohistochemical examination showed that the cytoplasm of the tumor cells were positive for CD138 and the nuclei were positive for cyclin D1. Four months after surgical resection, the patient had recurrence of tumor cells in the para-aortic lymph nodes and lumbar vertebrae. Chemotherapy treatment was ineffective. Surgical resection followed by chemotherapy is thought to be effective in the case of intestinal obstruction, although there is a possibility of devastation which is resistant to chemotherapy. We propose that tumor cells positive for cyclin D1 may play a key role in devastation and could be a useful indicator of prognosis. (Kitakanto Med J 2007; 57: 251~254)

Key Words: extramedullary plasmacytoma, small intestine, cyclin D1

Introduction

Extramedullary plasmacytoma (EMP) of the gastrointestinal tract is an uncommon neoplasm. Since 1970, 62 cases of EMP of the gastrointestinal tract have been reported.¹ We present a case of EMP of the small intestine which was resected for the treatment of obstruction but thereafter developed plasma cell leukemia with multiple bone metastases, and review the literature related to this rare tumor.

Case

A 78-year-old male was admitted to our hospital because of abdominal discomfort and vomiting. His body temperature was up to 37.0°C and a maximum white blood cell count of 11,640/mm³ with epigastralgia which was controllable with analgesics. The abdominal X-ray showed dilatation of the stomach and the small intestine (Fig. 1). Computed tomography revealed a 7-cm sized tumor of the small intestine and swollen lymph nodes (Fig. 2). The chest X-ray was normal and a flat bones roentgenogram was negative for osteolytic lesions. The serum β2-microglobulin level was within normal ranges and the Bence Jones' protein was not detectable in the concentrated urine sample. To treat the bowel obstruction, the patient underwent resection of the bowel and swollen lymph nodes. The resected specimen showed a 7-cm sized circumferential yellowish solid tumor with ileal ulceration and swollen lymph nodes (Fig. 3). A slice section of the resected specimen showed yellowish solid tumor tissue spread from mucosa to suberosa of the ileum (Fig. 4). Cytological findings from a touch smear of the resected tumor slice revealed tumor

¹ Department of Surgery, Sayama Hospital ² Department of Internal Medicine, Sayama Hospital ³ Department of Pathology, Sayama Hospital
Received: June 31, 2007
Address: NAOYUKI KOBAYASHI Department of Surgery, Sayama Hospital 1-33 Unoki, Sayama, Saitama 350-1323 Japan
cells which were well differentiated to plasma cells. These cells were composed of plasma cell-like round cells which have cart-wheel-shaped nuclear chromatin and a perinuclear halo. The nuclei were non-uniform in size. A grape cell with orange G positive immunoglobulin granules was found in the center (Fig. 5). Pathological examination of the resected specimen also showed plasma cell-like round cells which had an amphophilic cytoplasm and binucleated cells. Dutcher bodies and eosinophilic microspheres (Russell bodies) were also revealed (Fig. 6). Immunohistochemical analysis demonstrated that the tumor cells were LCA®, CD20®, CD79®, CD3®, CD5®, CD10®, BCL-2®, CD38®, CD56®, CD138®, vimentin®, cyclin D1®, and Ki–67® (Fig. 7). Ig lambda was monotypic and Ig kappa was negative. IgG and IgA were difficult to determine but the tumor cells were negative for IgM. These findings did not support the diagnosis of lymphoplasmacytic lymphoma. At this point serum and urinary monoclonal protein were not detected. From these findings, our patient was diagnosed as having the IgA-λ type of plasmacytoma. Four months after the procedure, computed tomography revealed the para-aortic lymph nodes were swollen. The patient had lumbar and a MRI showed a 2-cm sized lesion in the lumbar vertebrae. 13 months after the procedure, the serum IgA level gradually increased to 1438 mg/dl. Immunodiffusion of the serum globulin fraction identified the IgA-λ monoclonal protein. However, a bone marrow aspirate biopsy revealed a relatively small number of plasma cells (1.4% of all cells). Due to the recurrence of plasmacytoma, the patient underwent seven cycles of melphalan/prednisone (MP) therapy which was ineffective. Thereafter he was treated by radiotherapy (39 Gy) to his lumbar region and was administered morphine sulfate to relieve invertebrate pain. 16 months after the resection, he was diagnosed with plasma cell leukemia as the number of absolute plasma cells increased up to 51% of the blood leukocytes. An additional two cycles of vincristine/doxorubicin/dexamethasone (VAD) therapy achieved no remission.

**Discussion**

Plasmacytoma is a neoplastic proliferation of plasma cells which can occur in almost any tissue of the body. Most EMPs occur in the upper aerodigestive tract (UAD). However, forty percent of EMPs in non-UADs were found in the gastrointestinal tract, occurring mostly in the colon and rectum, followed by the stomach and the small bowel. EMPs occur most commonly in the 40–70 age group and the male to female ratio is 3:1. Little has been reported about EMP and ethnicity. EMP usually originates in the bone marrow and can manifest into multiple tumors.

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**Fig. 1** The abdominal X-ray showed dilatation of stomach (arrow heads) and small intestine (arrows).

**Fig. 2** Computed Tomography revealed a 7-cm sized tumor of the small intestine (arrow) and lymph nodes swelling (arrow heads).

**Fig. 3** The resected specimen showed a 7-cm sized circumferential elastic hard tumor with ileal ulceration (black arrow) and lymph nodes swelling (white arrows).
Extensive research of the literature revealed only 11 cases of EMP in the small intestine including our case.\textsuperscript{2–11} The sites of the lesions reported include the duodenum in 4 cases, the jejunum in 6 cases, and the ileum in one case. In early 1980s, an immunoperoxidase technique enabled the detection of monoclonal plasma cell populations. Among the reported EMP cases in the jejunum and the ileum including ours, the IgA type was found to be predominant (56\%, 5/9 cases). Using immunohistology, it is still unclear whether each immunoglobulin class attributes to a different prognosis.

Pathological classification of a tumor, which is located extramedullary and is microscopically well differentiated to plasma cells, can be problematic. In this case, the lesion was formed extramedullary and it was unclear whether it was a mucosa-associated lymphoid tissue (MALT) lymphoma or a plasmacytoma. The diagnosis of a plasmacytoma was eventually made as follicular structures and lymphoepithelial lesions (LEL) were not seen, and the tumor cells were in a homogenous pattern consisting of CD20-negative plasma-like cells. The metastasized tumor of multiple myeloma was ruled out because a bone marrow aspirate biopsy revealed relatively small number of plasma cells, the results of radiologic skeletal survey were normal, and the urinary Bence Jones’ protein was not detectable.

Surgical resection alone can give the best results with cases of EMP of the UAD.\textsuperscript{1} Furthermore, EMP is highly curable when radiotherapy is used with or without previous surgery with a reported 10-year disease-free survival of 87\% and overall survival rate of 61\%.\textsuperscript{12,13} However, Wiltshaw et al reported on the effectiveness of additional chemotherapy with mel-
phalan, prednisone, cyclophosphamide or vincristine alone or in combination with complete remission rates of 50%-88%.14,15 These results suggested that surgery followed by chemotherapy is an effective treatment. However, MP therapy was ultimately not an effective treatment for our patient. A larger case-controlled study is needed to confirm the efficacy of this approach.

Some EMP cases are known to develop multiple myeloma. After treatment of EMP in non-UAD regions, 21.2% of patients had recurrence and 14.1% of them converted to multiple myeloma.1 Hoehctlen-Vollmar et al state that the cyclin D1 gene is immunohistochemically detectable in up to one third of all cases of multiple myeloma. They demonstrated that amplification of this gene correlated significantly with the bone marrow infiltration and concluded that the detection of cyclin D1 amplification seems to be of promising prognostic value in multiple myeloma.16 However, cyclin D1 detection has not been reported previously with EMP of the small intestine. Using immunohistochemistry, we found the tumor cells of the EMP from our case were positive for cyclin D1.

In conclusion, among reported EMP cases including our patients, the IgA type is the most predominant. Surgical resection followed by chemotherapy is thought to be effective in cases of intestinal obstruction. However, clinicians should be cautious because there is a possibility of devastation, like our case, which is resistant to chemotherapy. Therefore, we propose that tumor cells immunohistochemically positive for cyclin D1 may play a key role for devastation and could be a useful indicator of prognosis.

References