Biclonal Extramedullary Plasmacytoma Arising in the Peritoneal Cavity: Report of a Case

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Abstract
We report a rare case of extramedullary plasmacytoma, which arose either in the ileum or the ileal mesentery. A 70-year-old woman presented with a high fever and symptoms of bowel obstruction. Computed tomography and magnetic resonance imaging showed a large heterogeneous tumor in the peritoneal cavity. Serum immunoelectrophoresis revealed a biclonal increase of IgA-κ and IgG-κ. At surgery, we found that the parenchyma of the fragile tumor had firm communication with the ileal mesentery, and the cavity of the tumor communicated with the ileal lumen. After a temporary regression following surgery and chemotherapy, the tumor grew rapidly. Although there was no evidence of progression to multiple myeloma, the patient died of cachexia less than 4 months after surgery.

Key words Extramedullary plasmacytoma · Biclonal · Peritoneal

Introduction
Plasmacytoma is a neoplastic proliferation of plasma cells. While this tumor usually arises in bone marrow, it may originate in almost any tissue throughout the body. Only about 4% of plasma cell tumors originate outside the bone marrow and are known as extramedullary plasmacytomas (EMPs), the majority of which develop in the soft tissue of the head and neck. Only 10% of EMPs develop in the gastrointestinal tract, the stomach being the most frequent site.1 We describe an unusual case of EMP arising in the peritoneal cavity. The parenchyma of the tumor had a firm communication with the ileal mesentery, and the cavity of the tumor communicated with the lumen of the ileum.

Plasmacytomas sometimes produce M-protein; however, only 1.5% of these tumors have been reported to produce two or more M-proteins.2 The EMP in this case produced two M-proteins, namely, IgG-κ and IgA-κ. Unlike most other reported cases of EMP, the patient had a very poor prognosis.

Case Report
A 70-year-old woman presented to her family physician with a high fever, abdominal pain, and vomiting. Computed tomography (CT) showed a large tumor in the peritoneal cavity, and she was admitted to our hospital about 2 weeks later for further investigations and treatment. She had undergone a right mastectomy for breast cancer at the age of 60, and had been suffering from hypothyroidism since the age of 55. She looked very ill because of the high fever, nausea, and vomiting, but no lymph node swelling was detected in the neck, maxillary, or inguinal regions. Her abdomen was distended, and a large soft tumor with a poorly defined contour was palpable in the whole abdomen, mainly confined to the lower abdomen.

Laboratory analysis showed a decreased hemoglobin concentration (10.1 g/dl), a normal leukocyte count (6 300/mm³), and a slightly increased C-reactive protein level (total level, 3.5 mg/ml). The serum transaminase levels were slightly increased, but the tumor markers such as carcinoembryonic antigen, α-fetal protein, carbohydrate antigen 15-3, and NCC-ST439 were all within the normal range. The serum total protein level was 6.5 g/dl, with 43.8% albumin, 5.6% α1-globulin, 11.2% α2-globulin, 22.8% β-globulin, and 16.6% γ-globulin. The immunoglobulin levels were as follows: IgG, 1680 mg/dl (normal range 870–1700), IgA, 637 mg/dl (normal range 110–410), and IgM, 29 mg/dl (normal range 0.6–16).
range 46–260). Serum immunoelectrophoresis revealed a biclonally increased production of IgA-κ and IgG-κ (Fig. 1), but Bence Jones protein was not detected in the urine. A systemic X-ray examination of the whole body did not show any other abnormalities. A microscopic examination of bone marrow tissues showed no abnormal proliferation of plasma cells.

Ultrasonography showed a 76 × 51-mm heterogeneous tumor adjacent to the uterus and abdominal CT scan showed a tumor with soft tissue density, containing air. Intravenous contrast medium failed to enhance the tumor parenchyma (Fig. 2). Magnetic resonance imaging (MRI) showed that the tumor had heterogeneous signal intensity (Fig. 3). A barium meal study of the small intestine showed irregular mucosa in the ileum, and revealed a communication between the tumor cavity and the ileal lumen.

Following admission, new small tumors appeared everywhere in the subcutaneous tissues of the patient’s body and on her left nipple. Histological examination of a subcutaneous tumor showed massive cellular infiltration of atypical plasma cells. However, we decided to remove the abdominal tumor to relieve the bowel obstruction and to obtain a definitive diagnosis. At surgery, a large fragile tumor was found in the lower peritoneal cavity, and many lumps were scattered throughout the upper peritoneal cavity. The largest tumor, which was located in the lower peritoneal cavity, communicated with the ileum and ileal mesentery 40 cm proximal to the ileocecal junction. The tumor infiltrated the serosa of the intestinal wall, especially of the sigmoid colon and small intestine, as well as the parietal peritoneum. The bulk of the tumor mass was extirpated with combined resection of the terminal ileum and ascending colon.

The resected tumor was 13 × 10 × 12 cm in size and weighed 2000g. Macroscopically, the parenchyma of the tumor had firm communication with the ileal mesentery, and a cavity within the tumor communicated with the ileal lumen (Fig. 4). Histological examination revealed that it was comprised of a proliferation of immature-appearing plasma cells, each of which had an...