Primary extramedullary plasmacytoma of the sigmoid colon with perforation: a case report

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Abstract

Background: Extramedullary plasmacytomas account for 4% of all plasma cell tumors and occur mainly in the upper respiratory tract; gastrointestinal system involvement is rare. Extramedullary plasmacytoma of the colon with perforation has not been reported.

Case presentation: A 77-year-old woman with a 1-year history of lower abdominal pain and nausea was admitted to our hospital. An abdominal computed tomography scan revealed a sigmoid tumor with perforation. The patient underwent emergency surgery. Pathological examination led to a diagnosis of plasmacytoma of the colon. The patient did not undergo postoperative adjuvant chemotherapy. She has had no recurrence in 14 months of regular follow-up.

Conclusions: We have herein described a rare case of extramedullary plasmacytoma of the gastrointestinal tract with perforation involving the sigmoid colon.

Keywords: Extramedullary plasmacytoma, Colon neoplasms, Perforation

Background

A plasma cell tumor is an immunoproliferative monoclonal disease of the B cell line that originates from malignant transformed plasma cells. Plasmacytoma includes solitary plasmacytoma of bone and solitary extramedullary plasmacytoma.

Solitary extramedullary plasmacytoma has been rarely reported, and its natural history and diagnosis are unclear. Most such plasmacytomas occur in the nasopharynx or upper respiratory tract; only 10% of reported cases have involved the gastrointestinal tract. The stomach and small intestine are the most commonly involved sites in the gastrointestinal tract [1–3]. Primary isolated extramedullary plasmacytoma of the colon is extremely rare. No previous reports have described plasmacytoma of the colon with perforation. We herein report a rare case of primary isolated extramedullary plasmacytoma of the colon with perforation and describe the patient’s postoperative clinical course.

Case presentation

A 77-year-old woman with a 1-year history of lower abdominal pain and nausea was admitted to our hospital. Blood examination showed evidence of an inflammatory response (Table 1), and abdominal computed tomography revealed a sigmoid tumor with perforation (Fig. 1). We suspected sigmoid cancer with perforation, and the patient underwent emergency surgery. Open laparotomy revealed an extensive mass involving the sigmoid colon with surrounding contamination (Fig. 2). The abdominal mass was removed en bloc, including resection of the sigmoid colon. The abdomen was flushed to remove contamination. An artificial anus was made.

Histopathologic examination showed that the oval mass was composed of a diffuse proliferation of plasma cells (Fig. 3a). At the concavity of the site of the perforation showed the tumor cell infiltrated into the subserosa and necrosis of tissue. But we were unable to identify the site of the perforation pathologically. The surgical margins were free from tumor cells. Immunohistochemical examination revealed positivity for CD79a (Fig. 3b), immunoglobulin G, and lambda light chain (Fig. 4a, b). Other markers (CD10, CD20, and kappa light
chain) were negative (Fig. 4c). Pathological examination led to a diagnosis of plasmacytoma of the colon. The patient underwent bone marrow biopsy and bone imaging to exclude associated multiple myeloma. Her peripheral blood smear, serum protein electrophoresis, and urine immunoelectrophoresis for Bence-Jones protein were normal. Postoperatively, the patient was discharged without any complications. She did not undergo postoperative adjuvant chemotherapy and has had no recurrence in 14 months of regular follow-up.

**Table 1** Blood examination

| Blood count | Biochemical parameters | Coagulation parameters |
|-------------|------------------------|------------------------|
| WBC 8090/μL | TP 3.5 g/dL             | PT (s) 14.3            |
| RBC 360 × 10^9/μL | Alb 1.59 g/dL | PT (%) 63.3           |
| Hb 11.5 g/dL   | T-bil 1.19 mg/dL       | PT-INR 1.21            |
| Plt 21.4 × 10^9/μL | AST 28 IU/L      | APTT (s) 45.4         |
|               | ALT 18 IU/L           | ATIII 49%              |
|               | ALP 127 IU/L          | FDP 40.0 μg/mL         |
|               | LDH 219 IU/L          | D-dimers 18.20 ng/mL   |
|               | γ-GTP 14 IU/L         |                        |
| Tumor markers |                        |                        |
| CEA 1.6 ng/mL |                        |                        |
| CA19-9 5.2 U/mL |                        |                        |
| BUN 15.6 mg/dL |                        |                        |
| Cr 0.4 mg/dL  |                        |                        |
| Na 13.4 mEq/L |                        |                        |
| Cl 3.6 mEq/L  |                        |                        |
| CRP 29.49 mg/dL |                        |                        |

WBC white blood cells, RBC red blood cells, Hb hemoglobin, Plt platelets, CEA carcinoembryonic antigen, CA19-9 cancer antigen 19-9, TP total protein, Alb albumin, T-bil total bilirubin, AST aspartate transaminase, ALT alanine transaminase, ALP alkaline phosphatase, LDH lactate dehydrogenase, γ-GTP gamma glutamyl transferase, BUN blood urea nitrogen, Cr creatinine, Na sodium, Cl chloride, CRP C-reactive protein, PT prothrombin time, PT-INR prothrombin time–international normalized ratio, APTT activated partial thromboplastin time, ATIII antithrombin III, FDP fibrin degradation products

**Discussion**

Extramedullary plasmacytoma accounts for only 3 to 5% of all plasma cell diseases. These tumors may be solitary or may precede, accompany, or follow the onset of multiple myeloma. Solitary extramedullary plasmacytoma has rarely been reported, and its natural history and diagnosis are unclear. Diagnosis of solitary extramedullary plasmacytoma requires the exclusion of associated multiple myeloma, which is determined by the absence of Bence-Jones protein in the urine, normal serum

![Fig. 1 Plain abdominal computed tomography. Huge tumor is present at the sigmoid colon, and free air (arrows) is seen around the tumor](image1)

![Fig. 2 Macroscopic examination. A A type 1 tumor is present in the sigmoid colon. B The sigmoid colon is surrounded by contamination. Arrows indicate concavity suspected the site of perforation of the tumor](image2)
electrophoresis, and normal bone marrow biopsy [4]. Our present case met these criteria.

Alexiou et al. [5] reported that extramedullary plasmacytoma most often occurs in the nasopharynx or upper respiratory tract (82.2%). Only 17.8% of cases involve the gastrointestinal tract. The stomach and small intestine are the most commonly involved sites in the gastrointestinal tract. Primary isolated extramedullary plasmacytoma of the colon is extremely rare, occurring in only 0.028% of cases [5]. Therefore, its clinical features and prognosis are not well known.

The clinical presentation of extramedullary plasmacytoma of the colon is variable and may include abdominal pain, intestinal bleeding, and diarrhea. Gabriel and Savu [6] reported a rare case in which an extramedullary plasmacytoma was found with ileocecal junction perforation secondary to colonoscopic injury. This is the only previous report to describe extramedullary plasmacytoma with gastrointestinal perforation (Table 2).

In the present case, we were unable to determine the cause of the perforation by pathologic examination. We consider that the tumor was necrosed and perforated; otherwise, as the tumor grew, the intestinal internal pressure increased, resulting in perforation of the sigmoid colon.

Postoperative chemotherapy has no effect on the course of extramedullary plasmacytoma. Our patient did not undergo postoperative adjuvant chemotherapy, and
### Table 2 Well-documented cases of plasmacytoma of the colon

| Author/year        | Sex | Age (years) | Location             | Clinical features                                    | Therapy                                                                 |
|--------------------|-----|-------------|----------------------|------------------------------------------------------|------------------------------------------------------------------------|
| Vasiliu and Popa/1928 | F   | 47          | Sigmoid              | Anorexia, epigastric pain, glandular enlargement     | ?                                                                      |
| Brown and Liber/1939 | M   | 57          | Colon, rectum        | Rectal discomfort                                     | ?                                                                      |
| Hampton and Gandy/1957 | F   | 43          | Rectum               | Rectal pain and bleeding                              | Rectosigmoid resection                                                 |
| Miller/1970         | M   | 35          | Cecum                | Anemia                                               | Right hemicolecotomy                                                   |
| William/1970        | M   | 84          | Cecum                | Anemia                                               | Right hemicolecotomy                                                   |
| Neilson/1972        | F   | 82          | Sigmoid              | Pain                                                 | Resection                                                             |
| Wing/1975           | F   | 82          | Ascending colon      | Pain                                                 | Right hemicolecotomy                                                   |
| Shaw/1976           | F   | 47          | Cecum                | Diarrhea                                             | Resection                                                             |
| Staples/1977        | M   | 61          | Sigmoid              | Incidental operative finding                        | Resection                                                             |
| Daniel/1977         | M   | 21          | Descending colon     | Pain, nausea, vomiting                               | Left hemicolecotomy                                                    |
| Allion/1977         | M   | 61          | Sigmoid              | None                                                 | Sigmoid colectomy                                                      |
| Adekunle/1978       | M   | 35          | Cecum                | Pain                                                 | Right hemicolecotomy                                                   |
| Terrence/1982       | F   | 20          | Transverse colon     | Pain, rectal bleeding                                | Transverse colon resection                                             |
| Sidani/1985         | M   | 52          | Sigmoid              | Pain, rectal bleeding                                | Resection                                                             |
| Rechard/1987        | M   | 77          | Cecum                | Weight loss, anemia, pain, fecal occult blood        | Right hemicolecotomy                                                   |
| Saverio Ligato/1996 | M   | 45          | Hepatic flexure of the colon | Anemia                                               | Extended right hemicolecotomy                                           |
| Holland/1997        | M   | 62          | Sigmoid colon        | Pain                                                 | Sigmoid colectomy                                                      |
| Lattuneddu/2004     | M   | 86          | Sigmoid colon        | Pain, rectal bleeding, asthenia                      | Segmental resection of the left colon                                  |
| Gupta/2007          | M   | 42          | Diffuse colon        | Diarrhea                                             | Subtotal colectomy                                                     |
| Jones/2008          | M   | 65          | Sigmoid colon        | Dysuria, abdominal pain                              | Sigmoid colon resection                                                |
| Jone/2008           | M   | 57          | Sigmoid colon        | Fatigue, melena                                      | Hartmann resection                                                     |
| Doki/2008           | M   | 64          | Ascending colon      | Pain                                                 | Right hemicolecotomy, lymph node dissection, excision of Gerota's fascia, partial resection of the posterior portion of the liver |
| Collado Pacheco/2009 | M   | 74          | Right colon          | Diarrhea, pain, rectal bleeding                      | ?                                                                      |
| Kodani/2011         | M   | 42          | Sigmoid              | Fecal occult blood                                  | Endoscopic submucosal resection                                        |
| Nakagawa/2011       | F   | 84          | Cecum and rectum     | Medical examination                                  | Endoscopic submucosal resection                                        |
| Lee/2013            | M   | 45          | Transverse colon     | Pain                                                 | Extended left hemicolecotomy                                           |
| Zihni/2013          | M   | 54          | Descending colon     | Pain and weakness                                    | Left hemicolecotomy and small intestinal resection                     |
| Han/2014            | M   | 49          | Transverse colon     | Pain                                                 | Left hemicolecotomy                                                    |
| Emmanuel/2014       | M   | 62          | Cecum                | Perforation during diagnostic colonoscopy           | Right hemicolecotomy                                                   |
| Parnel/2015         | F   | 72          | Right colon          | Fatigue, light-headedness, dyspnea, dark stool       | Right hemicolecotomy Distal ileal resection                            |

F female, M male
she has had no relapse to date. However, careful follow-up is required.

Because primary isolated extramedullary plasmacytoma in the colon is very rare, the clinical course, treatment guidelines, and prognosis remain unclear. Further study of the clinical features of primary isolated extramedullary plasmacytoma of the colon is necessary to ensure that adequate treatment is administered.

Conclusions
We have described a rare case of extramedullary plasmacytoma of the gastrointestinal tract with perforation of the sigmoid colon. In this case, the prognosis was good because of appropriate treatment involving early surgery.

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Authors’ contributions
FK wrote the final manuscript and performed the literature search. KD supervised the writing of the manuscript. HI and TO performed the surgery. All authors read and approved the final manuscript.

Consent for publication
Written informed consent was obtained from the patient for the publication of this report and any accompanying images.

Competing interests
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