CASE REPORT

Extramedullary plasmacytoma mimicking colon carcinoma: an unusual presentation and review of the literature

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SUMMARY

A 72-year-old woman presented to outpatient clinic with fatigue, light-headedness, dyspnoea and dark stool suggestive of lower gastrointestinal bleeding. She was previously diagnosed with multiple myeloma and completed 9 cycles of chemotherapy with bortezomib, lenalidomide and dexamethasone. She had very good partial response. A CT scan of the abdomen revealed a 9 cm mass at the hepatic flexure of the large intestine with an apple core deformity causing a marked narrowing of the lumen. Colonoscopy confirmed a large, nearly obstructing ulcerative mass in the distal right colon. The patient underwent a right hemicolectomy, distal ileal resection and lymph node dissection. Histopathology confirmed the mass as a plasmacytoma. Postoperatively, the patient was started with bortezomib and liposomal doxorubicin followed by carfilzomib. She showed excellent response to the chemotherapy.

BACKGROUND

Extraskeletal manifestations of multiple myeloma (MM) are rare. MM can directly extend from bone into paraspinal soft tissue or it can present as a secondary plasmacytoma at a distant site. Extramedullary plasmacytoma (EMP) occurs secondary to MM and most commonly involves the soft tissue, skin, lymph nodes, liver, spleen and kidneys. MM involving the colon is rare with less than 25 reported cases. We present a case of secondary EMP affecting the ascending colon that occurred despite a very good partial response (VGPR) to therapy. Our patient's presentation and clinical findings looked highly suspicious for a typical colonic carcinoma; hence, it was surprising when the pathological diagnosis was EMP. Additionally, gastrointestinal (GI) EMP may histologically show poorly differentiated cells, as seen in our case, or resemble a lymphoma. This case presents a rare presentation of MM involving the colon, as well as highlights the mimicking ability and aggressive nature of MM to localise at a distant site despite responding well to therapy.

CASE PRESENTATION

A 72-year-old woman presented to clinic with fatigue, light-headedness and dyspnoea along with occasional abdominal pain relieved by bowel movements. She also mentioned dark stools of 1-week duration. She had previously been diagnosed with IgM κ multiple myeloma and exhibited VGPR after nine cycles of induction chemotherapy with

bortezomib, lenalidomide and dexamethasone. Her diagnosis of MM was made after sustaining a pathological fracture of the femur 11 months prior. Four weeks after the fracture, she developed a pulmonary embolism consequent to a deep vein thrombosis of the left lower extremity. Subsequently, she was started on warfarin. Other medical history included hypertension, hypothyroidism, gastro-oesophageal reflux and a history of gastric ulcers. She had no family history of colorectal cancer and her colonoscopy 3 years prior was normal. On physical examination, she appeared ill and anaemic. Her vital signs at presentation were as follows: temperature 97.4°F, pulse 75 bpm, respiratory rate 18 breaths/min, blood pressure 101/61 mm Hg and oxygen saturation 98% on room air. Her abdomen was soft, non-tender and non-distended with no palpable masses. Stool was positive for occult blood. The rest of the physical examination was unremarkable.

INVESTIGATIONS

Laboratory work up revealed haemoglobin level at 8.1 g/dL. Further, it was noted that the patient's M-spike had risen over a period of 1 month from 0.6 to 2.3 g/dL and her IgM level from 1117 to 3069 mg/dL. CT scan of the abdomen revealed a 9 cm mass at the hepatic flexure of the large intestine with an apple core deformity causing a marked narrowing of the lumen (figures 1 and 2). Colonoscopy confirmed a large, nearly obstructing ulcerative mass in the descending colon near the hepatic flexure (figure 3). A laparotomy with a right hemicolectomy, distal ileal resection and lymph node dissection was performed. Histopathology of

Figure 1 Abdominal CT scan revealing a 9 cm mass at the hepatic flexure of the large intestine with an apple core deformity causing marked narrowing of the lumen.

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Figure 2 Abdominal CT scan revealing a 9 cm mass at the hepatic flexure of the large intestine with an apple core deformity causing marked narrowing of the lumen.

the mass showed ulcerated colonic mucosa infiltrated by sheets of poorly differentiated tumour cells (figure 4) with characteristic staining (figure 5) and markers confirming a plasmacytoma (positive for CD138; negative for CK7, CK20 and CDX2; κ restricted by in-situ hybridisation). In addition, 7 of the 44 lymph nodes showed plasma cells in the subcapsular sinus. The postoperative course was uneventful.

TREATMENT

Following laparotomy, the patient's treatment regimen was modified to bortezomib and liposomal doxorubicin every 3 weeks for 3–4 cycles followed by carfilzomib to decrease the tumour burden until complete remission. Currently at the seventh week of her postoperative period, she is clinically doing well with a decrease in her M-spike to 1.5 g/dL and IgM level to 2435 mg/dL.

DISCUSSION

Plasma cell neoplasms are generally characterised into the following four groups: MM, plasma cell leukaemia, solitary plasmacytoma of the bone and EMP.¹ EMP is rare, constituting less than 5% of all plasma cell neoplasms.^{1–5} It is classified as primary (when it develops as a solitary, sporadic lesion) or secondary, on the basis of a preceding plasma cell neoplasm.^{1,6} Primary EMP is more common than secondary.⁷ The reported prevalence of

Figure 4 Histopathology of the colonic mass showing ulcerated colonic mucosa infiltrated by sheets of poorly differentiated plasma cells.

EMP ranges from 2% to 20% in patients with newly diagnosed MM and 2% to 17% in patients later in the course of MM.^{8–13} EMP secondary to MM most commonly involves the soft tissue, skin, lymph nodes, liver, spleen and kidneys.^{11,14–16} GI manifestations of MM are very rare.^{7,14,17–21} When GI involvement occurs, the stomach and small intestine are the most commonly involved sites,^{14,19–21} while the colon and oesophagus are the least.^{19–21} We reviewed the literature and found less than 25 reported cases of MM manifesting as a colonic plasmacytoma. Some cases presented with complication of amyloidosis and some presented with mechanical obstruction with plasmacytoma. There are multiple reported cases of EMP without multiple myeloma involving the GI tract. In table 1, we mention a few

Figure 3 Colonoscopy showing a large, nearly obstructing ulcerative mass in the descending colon.

Figure 5 Immunohistochemistry of the colonic mass showing positive staining for CD138.

Table 1 Previously reported cases of extramedullary plasmacytoma involving gastro-intestinal tract

Case report	Age (years)	Sex	Symptoms	Treatment modalities	Outcome
Venizelos <i>et al</i> ¹⁴	61	F	Acute large bowel pseudo-obstruction	Combined chemotherapy and radiotherapy	Recovered
Herbst <i>et al</i> ²¹	79	M	Back pain and constipation	Surgery	Died
Göral <i>et al</i> ²²	55	M	Thoracic pain, abdomen distention, pseudo-obstruction	Conservative management for obstruction, chemotherapy for multiple myeloma	Recovered
Islam <i>et al</i> ²³	52	M	Haematochezia	Surgery and chemotherapy	Recovered
Triantafyllou <i>et al</i> ²⁴	43	M	Colicky abdominal pain, microcytic hypochromic anaemia	Died before treatment	Died
Kakati <i>et al</i> ²⁵	57	M	Incidental finding in CT scan with extensive infiltrating soft tissue thickening in the ascending, transverse and descending colon	Stem cell transplant and melphalan	Recovered
Dawson <i>et al</i> ²⁶	60	F	Hematemesis and melena	Conservative	Died
Rogulj <i>et al</i> ²⁷	66		Article in Croatian		
Rao and Yaghmai ²⁸	45	M	Malaise, nausea and vomiting	Conservative	Died
Abouna ²⁹	75	M	Intestinal obstruction	Surgery	Recovered
Weintraub <i>et al</i> ³⁰	74	M	Diffuse abdominal pain, diarrhoea and weight loss	Surgery	Died

F, female; M, male.

cases of multiple myeloma with plasmacytoma involving the GI tract.

Our patient is one such case, presenting with an EMP of the ascending colon 11 months into the treatment of her IgM κ MM. She had received nine cycles of induction chemotherapy with bortezomib, lenalidomide and dexamethasone, and was exhibiting a VGPR with an M-spike at 0.6 g/dL and IgM level at 1117 mg/dL. It is presumed that her sudden increase in M-spike to 2.3 g/dL and IgM level to 3069 mg/dL was associated with the formation of her colonic EMP. At this point, she developed symptoms of abdominal pain and rectal bleeding while on warfarin therapy. The EMP was 9 cm in diameter at the time of resection. This suggests the need for suspicion of a secondary EMP when a previously responding MM exhibits an increasing serum M-spike and immunoglobulin level. In the setting of new GI symptoms, evaluation for a GI EMP should be considered. The diagnosis of a GI EMP can be clinically^{5 20 21} and histologically^{1 5 7} difficult because it may mimic a carcinoma or lymphoma of the colon. Our case was no different. She presented with a lower GI bleed and an apple core lesion on imaging—classic findings of adenocarcinoma of the colon. A typical EMP displays sheets of plasma cells on histological evaluation. However, GI EMP may show poorly differentiated cells with loss of recognisable features or a differentiation similar to that seen in some types of lymphoma.^{1 5 7 20} Our patient's mass formed sheets of poorly differentiated tumour cells without signs of plasmacytoid differentiation. The immunophenotypic diagnostic requirements of MM—positive CD138 and κ restriction—were the only typical features in our patient. When a history of MM is present, clinicians and pathologists must keep in mind the mimicking potential of plasmacytomas when diagnosing a GI neoplasm. The management of EMP includes surgery, radiotherapy and chemotherapy. In cases of secondary EMP, systemic therapy is required. Although thalidomide produces good medullary and serological responses of myeloma, it is not effective against extramedullary (EM) disease.³¹ Bortezomib has shown good efficacy and is currently the preferred choice of treatment for MM with EM manifestations.^{31 32} Additionally, there is evidence suggesting that lenalidomide could be useful in secondary EMP refractory to bortezomib.^{32–34} However, our patient had received nine cycles of bortezomib and lenalidomide. She was showing a VGPR with M-spike and IgM levels trending down. Unfortunately, GI

localisation still occurred, which demonstrates the aggressive nature of this disease.⁵ The literature describes myeloma involving the GI tract as a late event associated with a poor prognosis.^{7 17 20 21} For that reason, our patient's therapy was modified to bortezomib and liposomal doxorubicin followed by carfilzomib. She is under close monitoring for signs of recurrence.

Learning points

- ▶ Manifestations of multiple myeloma (MM) in the gastrointestinal (GI) system are rare.
- ▶ The colon, in particular, is one of the least commonly involved sites for MM to localise in.
- ▶ MM can manifest as a plasmacytoma in distant sites despite responding well to therapy.
- ▶ An extramedullary plasmacytoma (EMP) in the large bowel may clinically mimic a carcinoma of the colon. Histological diagnosis of a GI EMP may also be difficult, because tumour cells can be poorly differentiated or show features that resemble a lymphoma.

Competing interests None declared.

Patient consent Obtained.

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