CASE REPORT

Anorectal involvement in a patient with multiple myeloma

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SUMMARY

Multiple myeloma is a neoplastic proliferation of monoclonal plasma cells. Symptomatic gastrointestinal involvement is uncommon. We report the case of a 45-year-old patient admitted with an anorectal polypoid lesion, which progressed to colonic obstruction. Investigation revealed a secondary plasmacytoma associated with multiple myeloma. We discuss the characteristics of this rare entity with poor prognosis, its clinical implications and treatment options.

BACKGROUND

Anorectal plasmacytoma is a very rare entity and its association with multiple myeloma has never been reported to date. We discuss the clinical implications and the therapeutic options of this condition with poor prognosis.

CASE PRESENTATION

A 45-year-old man was admitted to our gastroenterology department with a 4-month history of diarrhoea, rectal bleeding, tenesmus, abdominal pain and weight loss (12% of total body weight). His medical history was unremarkable. There was neither family history of inflammatory bowel disease nor colorectal cancer.

INVESTIGATIONS

Flexible sigmoidoscopy revealed a large polypoid lesion that extended from the proximal anal canal to the distal rectum, covering about one-third of the rectal lumen (figure 1). Proximal colonic mucosa was normal. Previous total colonoscopy, performed 3 months earlier, showed no remarkable pathological features.

Figure 1 Flexible sigmoidoscopy revealing a large polypoid lesion with proximal anal and distal rectal involvement covering about one-third of the rectal lumen.

Figure 2 Histopathological examination of the anorectal lesion showing subepithelial plasmacytic cell infiltration (A). Immunohistochemical staining positive for CD138 (B) and CD38 (C), with restraint expression of lambda chains (D).

Laboratory evaluation revealed the following: haemoglobin 8.7 g/dL, white blood cell count 6.5×10^9 cells/L, platelet count 158×10^9 cells/L, urea 47 mg/dL, serum creatinine 2.43 mg/dL, lactate dehydrogenase 166 U/L, serum calcium 8.7 mg/dL, C reactive protein 14.7 mg/L, serum $\beta 2$ microglobulin 10.17 mg/L and hypogammaglobulinaemia with monoclonal peak (undetectable serum IgM and IgA, serum IgG 4.4 g/L). Serum immunofixation revealed monoclonal lambda free light chain. A 24-hour urine collection was positive for Bence-Jones protein. Bone marrow aspiration and biopsy showed 73% of monoclonal plasma cells, confirming the diagnosis of lambda light chain multiple myeloma.

Histopathological examination of the anorectal lesion showed subepithelial plasmacytic cell infiltration, and immunohistochemical staining positive for CD138 and CD38, and Ki-67 of almost 100%, compatible with plasma cell neoplasm (figure 2).

The patient developed intestinal obstruction with important colonic distension and air-liquid levels in the abdomen radiograph.

Abdominopelvic CT revealed colonic distension induced by anorectal polypoid lesion measuring $11 \times 7 \times 8$ cm, two perirectal tissue lesions adjacent to rectosigmoid transition, and multiple lytic lesions in the lumbar column and pelvis (figure 3).

DIFFERENTIAL DIAGNOSIS

- ▶ Solitary anal plasmacytoma
- ▶ Squamous cell carcinoma
- ▶ Adenocarcinoma
- ▶ Small cell carcinoma

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Figure 3 Abdominopelvic CT showing anorectal polypoid lesion measuring 11×7×8 cm (A), perirectal tissue lesion adjacent to rectosigmoid transition (B) and lytic lesion in the pelvis (C).

- ▶ Lymphoma of the anal canal
- ▶ Giant condyloma

TREATMENT

After discussion with haematology and surgery, the patient started high-dose dexamethasone (40 mg per day for 2 days) with clinical resolution of the obstruction. Treatment with VCD (bortezomib, cyclophosphamide and dexamethasone) was started.

OUTCOME AND FOLLOW-UP

After two VCD cycles, the disease progressed to secondary plasma cell leukaemia, showing leucocytosis of $100\,000 \times 10^9$ with 69% of circulating plasma cells, thrombocytopenia (platelet count 46×10^9 cells/L) and anaemia (haemoglobin 6.3 g/dL). There was recurrence of anal obstruction requiring derivative colostomy. Despite poor prognosis, he started treatment with VDT-PACE (bortezomib, dexamethasone, thalidomide, cisplatin, doxorubicin, cyclophosphamide and etoposide), but died 2 months after diagnosis from alveolar haemorrhage.

DISCUSSION

Gastrointestinal involvement at the time of initial diagnosis of multiple myeloma is extremely rare and, when present, is usually associated with rapid progression of the disease.¹ The median overall survival in a large retrospective study of patients with multiple myeloma involving the gastrointestinal system was 7 months. Gastrointestinal plasmacytomas may affect any part of the gastrointestinal tract, but the small bowel is the most common site of infiltration, followed by the stomach, colon and oesophagus.¹ They account for <5% of all extramedullary plasmacytoma.¹

Clinical manifestations depend on the site and extension of involvement and may range from nausea, vomiting, weight loss obstruction and bleeding.²

Few cases of anal solitary plasmacytoma have been reported in the literature, and the patients were successfully treated with

radiotherapy.^{3 4} To our knowledge this is the first case of anal plasmacytoma associated with multiple myeloma reported to date.

Treatment with high-dose dexamethasone was transiently effective in our patient in reducing tumour volume and allowed colonic decompression. However, due to disease progression under treatment, he ultimately required derivative colostomy. The disease eventually progressed with a fatal outcome, as expected in cases refractory to bortezomib and immunomodulatory drugs.

Learning points

- ▶ Unlike solitary lesions, which are preferably treated with surgical resection and/or radiotherapy, gastrointestinal plasmacytomas associated with multiple myeloma require a more aggressive approach, reserving surgery and radiotherapy for cases of uncontrolled bleeding and obstruction.⁵
- ▶ Intensive chemotherapy bortezomib-based regimens are recommended, followed by autologous stem cell transplantation.⁶ The regimen VDT-PACE (bortezomib, dexamethasone, thalidomide, cisplatin, doxorubicin, cyclophosphamide and etoposide) is of particular value in these patients who present with aggressive disease on diagnosis, as in extramedullary gastrointestinal plasmacytomas or plasma cell leukaemia.
- ▶ Allogeneic transplantation can be considered in younger patients.⁷ However, new therapeutic approaches are necessary in this limited prognosis scenario, and the patients should always be considered for clinical trials.

Contributors PR has produced the report and literature review. TC and ALT assisted in the production of the report and were also involved in the care of the patient. MJB oversaw the creation of the report.

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