CASE REPORT

High-grade GI lymphoma presenting with diarrhoea

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SUMMARY

A 63-year-old male carpenter presented with diarrhoea after returning from a holiday in Egypt with no other abdominal features. Physical examination showed generalised lymphadenopathy and splenomegaly. Blood tests including inflammatory markers were within normal limits. Microbiological stool sample analysis was unremarkable. Endoscopies of the large bowel showed patchy inflammation from the rectum to the hepatic flexure with multiple tiny polyps. Histology revealed a lymphocytic infiltrate suggestive of multiple lymphomatous polyposis. Some time later, he presented with small bowel obstruction requiring emergency laparotomy and ileostomy. After recovery from surgery he started definite treatment with chemotherapy resulting in complete remission.

BACKGROUND

Lymphomatous polyposis should be included in the differential diagnoses of patients presenting with lower gastrointestinal symptoms. Interestingly this patient presented with symptoms predominantly related to gastrointestinal tract with no systemic features of lymphoma.

CASE PRESENTATION

A 63-year-old male carpenter presented with diarrhoea after returning from a holiday in Egypt to the gastroenterology clinic. He had 4–5 loose motions a day with no blood or mucus. There were no other abdominal symptoms and he did not have night sweats or weight loss.

Apart from essential hypertension and a previous inguinal hernia repair his medical history was unremarkable. There was no relevant family history and was taking atenolol and aspirin, long term. He was a smoker with a moderate alcohol intake. Physical examination showed generalised lymphadenopathy and splenomegaly.

INVESTIGATIONS

Routine blood tests (haemoglobin 15.2 g/dl, white cell count $6.5 \times 10^9/l$, platelet $180 \times 10^9/l$, lymphocyte $2.2 \times 10^9/l$, creatine 86 $\mu\text{mol/l}$, albumin 45 g/l, bilirubin 7 $\mu\text{mol/l}$, alkaline phosphatase 85 U/l, alanine transaminase 13 U/l, C reactive protein 3 mg/l, erythrocyte sedimentation rate 6 mm/h) were within normal limits. The lactate dehydrogenase was also normal at 307 U/l. Microbiological stool sample analysis showed no evidence of bacterial or parasitic infection.

A flexible sigmoidoscopy followed by a full colonoscopy showed a prominent ileocecal valve and mild patchy inflammation from the rectum to the hepatic flexure with multiple tiny polyps, suggestive

of pseudopolyps (figure 1), from which biopsies were taken.

The histological analysis of the polyp biopsy revealed a nodular large bowel infiltration by a uniform population of lymphocytic cells with irregular nuclei.

Immunohistochemistry staining showed the lymphocyte population positive for CD5, 20, 43 ($\pm$), cyclin D1 and negative for CD3, 10 and 23. Interestingly, the Ki67 proliferation fraction was less than 10%. These findings confirmed the diagnosis of non-Hodgkin's mantle cell lymphoma as a result of the t(11;14)(q13;q32) chromosomal translocation (figure 2).

This with the endoscopic appearance of multiple polyps suggested multiple lymphomatous polyposis.

A staging CT scan showed widespread infra and supra diaphragmatic lymphadenopathy with moderate hepatosplenomegaly. In addition there was diffuse thickening of the colon up to the hepatic flexure with a scattered nodular filling defect in the small bowel. Radiological features confirmed stage IVe disease. There was also bone marrow involvement with a trephine biopsy showing a marked patchy inflammation of small- to medium-sized lymphoid cells with immunohistochemistry identical to the large bowel biopsy above.

DIFFERENTIAL DIAGNOSIS

1. Inflammatory bowel disease
2. Chronic ischaemic colitis
3. Travellers diarrhoea
4. Schistosoma infection

TREATMENT

A combination of the monoclonal anti-CD20 antibody rituximab, conventional chemotherapy

Figure 1 Colonoscopy image shows multiple polyps.

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Figure 2 (A) Histology image of the biopsy taken from the colonic polyps. (B) Immunohistochemistry staining of the colonic biopsy shows the lymphocyte population positive for CYCLIN D1. (C) Immunohistochemistry staining of the colonic polyp biopsy shows the lymphocyte population positive for CD5. (D) Immunohistochemistry staining of the colonic polyp biopsy shows the lymphocyte population positive for CD20.

(cyclophosphamide, vincristine, doxorubicin, prednisolone) and high-dose cytarabine was used. The Nordic protocol, used in this patient, incorporates such a regime.

OUTCOME AND FOLLOW-UP

After a short period and prior to starting chemotherapy, he presented with acute abdominal pain and distension.

Physical examination showed signs of intestinal obstruction, confirmed on a CT scan revealing an intussuscepting mass in the large bowel (figure 3). He underwent emergency laparotomy with ileostomy.

Chemotherapy was tolerated well resulting in a complete response with a resolution of his lymphadenopathy and hepatosplenomegaly.

DISCUSSION

Only a few cases have been published in the literature regarding multiple lymphomatous polyposis due to its rarity.

Although an uncommon presentation of a common complaint, endoscopic and histological examination of the gastrointestinal tract is an important component in the workup. To differentiate lymphomatous polyposis from other conditions in the differential diagnosis such as inflammatory, infective and ischaemic colitis this approach is essential.

It is established that gastrointestinal tract involvement in mantle cell lymphoma confers a poorer prognosis.¹

The recent British Committee for Standards in Haematology guidelines (2012)² recommend an intensive approach in managing younger patients with a good performance status using a

Figure 3 CT scan of the abdomen shows small bowel distension secondary to intussuscepting mass in the large bowel.

combination of rituximab, conventional chemotherapy and high-dose cytarabine augmented by a autologous stem cell transplant.

Learning points

- ▶ Lymphoma of the gastrointestinal tract is a rare condition but well recognised.³
- ▶ Gastrointestinal tract involvement of mantle cell lymphoma confers a poorer prognosis.¹
- ▶ Thirty-five per cent of mantle cell lymphoma may present with diarrhoea and an obstructive tumour can lead to the need for surgery in 10% of cases.^{1 4}
- ▶ Surgical intervention in this case was important in treating the acute emergency allowing chemotherapy to be delivered safely after a period of recovery.

Contributors SA contributed in collecting the notes, images, followed up the case and writing the manuscript and finding the references. AM reviewed the manuscript particularly from language and grammar point of view, helped in finding the appropriate references and obtained patient consent. SH performed the colonoscopy and suggested writing up this case and publish it.

Competing interests None.

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