CASE REPORT

Primary colorectal non-Hodgkin's lymphoma in a retropositive patient

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SUMMARY

A 43-year-old man known case of retropositive illness presented with abdomen pain of 15 days and loose stools for 10 days. Loose stools were foul smelling and blood stained. The patient was a known case of type 2 diabetes and retroviral illness on highly active antiretroviral therapy (HAART). General physical examination was normal. On examination an irregular mass was palpable in the right iliac fossa and right flank. The mass was hard in nature; irregular with restricted mobility. There was no palpable liver or spleen. CT of the abdomen and pelvis revealed a well-defined heterogeneously enhancing hypodense mass lesion measuring 16×11.7×12 cm involving the ileocaecal region and extending medially and inferiorly to sigmoid colon and rectum. A colonoscopy showed a proliferative highly vascular mass 15 cm from anal verge at the rectosigmoid junction. Histopathology revealed sheets of neoplastic lymphoid cells in rectal wall suggestive of non-Hodgkins lymphoma.

BACKGROUND

Primary colorectal non-Hodgkin's lymphoma (NHL) is a rare tumour. They are slow growing and usually present late. Symptoms are non-specific and obstruction of the large bowel is rare. HIV infection is a major risk factor for NHL and the disease is usually advanced due to immune-compromised state. Early detection and aggressive treatment in the form of surgery and chemotherapy is necessary to achieve a better survival rate.

CASE PRESENTATION

A 43-year-old man known case of retropositive illness presented with abdomen pain of 15 days and loose stools for 10 days. Pain was generalised, insidious in onset and progressively increasing in nature. Loose stools were present for 10 days, 3–4 times a day and foul smelling. It was associated with frank blood in stools. The patient neither had any discomfort of altered bowel habits in the past nor felt any mass per abdomen. No history of vomiting or yellowish discolouration of sclera. There was no history of fever, no history of travel, eating food outside or similar history in family members. The patient was a known case of type 2 diabetes and retroviral illness on HAART. General physical examination was normal. On abdomen examination an irregular mass was palpable in right iliac fossa and right flank. The mass was hard in nature; irregular with restricted mobility. There was no palpable liver or spleen. Colonoscopy, CT of the abdomen and

chest were performed. Histopathology was reported as NHL. The disease was staged as I (E).

INVESTIGATIONS

A CT of the abdomen and pelvis revealed a well-defined heterogeneously enhancing hypodense mass lesion measuring 16×11.7×12 cm involving the ileocaecal region and extending medially and inferiorly to the sigmoid colon and rectum (figure 1). No evidence of calcification or haemorrhage was seen. The mass was seen causing the narrowing of the caecum and rectum. No evidence of obstruction was observed. The rest of the colon was normal. Multiple enhanced lymph nodes were seen in the right iliac and lumbar region, largest measuring 2.7×2.2 cm. The mass lesion is seen abutting anteriorly the anterior abdominal wall with loss of fat planes at some places suggestive of adherence. Posteriorly the lesion is abutting the right psoas muscles and ureter. Posteroinferiorly, the lesion is abutting the urinary bladder with maintained fat planes. The lesion was encasing the right common and external iliac vessels all around.

The chest X-ray was normal. No mediastinal widening seen.

Figure 1 CT of the abdomen and pelvis showing a well-defined heterogeneously enhancing hypodense mass lesion measuring 16×11.7×12 cm involving the ileocaecal region and extending medially and inferiorly to sigmoid colon and rectum.

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Figure 2 Colonoscopy showing a proliferative highly vascular lesion at the rectosigmoid junction.

CT of the chest showed benign lymph nodes along both the jugular chains largest measuring 1.4×0.6 cm. Fibrosis with bronchiectatic changes seen in posterior segment of right upper lobe.

A colonoscopy was carried out and revealed proliferative highly vascular mass about 15 cm from anal verge at the rectosigmoid junction extending for about 5 cm (figure 2). Scope could be passed beyond the lesion and there was no evidence of obstruction. The descending, transverse and ascending colon were normal.

Blood investigations: haemoglobin 10.6 g/dL, total leucocyte count $12\,100/\mu\text{L}$, lymphocyte 9.7%, monocyte 5.8%, neutrophil 80.9%, platelet $394\,000/\mu\text{L}$, lactate dehydrogenase 652 IU/L. Total bilirubin 0.3 mg/dL, direct bilirubin 0.1 mg/dL, albumin 1.8 g/dL, total protein 4.8 g/dL, alkaline phosphatase 168 U/L, aspartate aminotransferase and alanine transaminase within normal limits.

Histopathology revealed sheets of neoplastic lymphoid cells in rectal wall suggestive of NHL, high grade, diffuse large B-cell lymphoma (figure 3). Immunohistochemistry showed CD138 and negative for CD20, CD3, CD30. Bone aspiration did not show any marrow infiltration.

CD4 count of absolute 42 (290–2600), CD3 of 554 (600–3700), CD8 of 494 (190–2120) and ratio of CD4/CD8 of 0.09 (0.6–2.8).

Figure 3 Histopathology showing sheet of neoplastic lymphoid cells in rectal wall suggestive of non-Hodgkins lymphoma.

DIFFERENTIAL DIAGNOSIS

Carcinoma rectum.

TREATMENT

In view of the advanced colorectal lesion involving the right ureter and the iliac vessels surgical resection was not considered. The patient was advised rituximab with CHOP (R-CHOP) regime by the multidrug therapy team.

OUTCOME AND FOLLOW-UP

The patient received a second cycle of chemotherapy and is on regular follow-up. The outcome of the lesion with the R-CHOP regime is yet to be evaluated.

DISCUSSION

Primary gastrointestinal NHL is an uncommon tumour, representing only 0.2–1.2% of all colonic malignancies. However, in NHL, the most common extranodal site is the gastrointestinal system. Most prominent sites of primary lymphoma in the gastrointestinal (GI) tract include the stomach (roughly 50%), the small bowel (30%) and the colon and rectum (20%).¹

Our case involves an elderly man with primary colorectal NHL and who was suffering from retroviral illness. Current evidence is present that lists HIV infection as a major risk factor for NHL.²

The incidence is more common in men over the age of 70 years. The patient, when symptomatic, tends to present with abdominal pain, along with constitutional symptoms such as weight loss and anorexia.

The definition of a primary GI lymphoma was given by Dawson *et al* in 1961: the five components include³

1. No palpable superficial lymph nodes on presentation;
2. No enlarged mediastinal nodes on chest X-ray;
3. Normal range of total and differential white cell count;
4. Involvement of only regional lymph nodes, discovered on surgery;
5. Liver and spleen remain without disease.

Musshoff's modified Ann Arbor staging of NHL is the one most commonly used.

Stage	Primary extra-nodal involvement
I	Involvement of extra-lymphatic organ or tissue
II1	Involvement of an extra-lymphatic organ including the regional lymph node (LN) (II1) or another adjacent extra-lymphatic organ (II1E) above or below the diaphragm
II2	Involvement of an extra-lymphatic organ and LN that extends over the regional LN and that can include another extra-lymphatic organ (II2E)
III	Involvement of an extra-lymphatic organ with LN involvement above and below the diaphragm including another extra-lymphatic organ (III E) or the spleen (IIIS) or both (IIISE)
IV	Diffuse or disseminated organ involvement with or without LN involvement.

Radiographic procedures used to diagnose and evaluate GI lymphomas include CT of the abdomen and barium enema. CT is particularly informative in telling colonic wall thickening, extent of nodal involvement and distant metastasis. Barium enema allows us to better understand the morphology of the mass as well as assess the mucosa. There may be focal or diffuse lesions. Focal lesions can be polypoid, circumferential with smooth mucosal surface, circumferential with extensive ulceration, a cavitary mass, a mucosal fold thickening and a focal mucosal nodularity. Diffuse lesions are described as ulcerated or

nodular.⁴ Despite causing mucosal abnormalities, lymphomas very rarely present as intestinal obstruction. The lesions tend to be long and annular, as opposed to the 'apple core' lesions seen in adenocarcinoma. Colonic lymphomas may rarely present dramatically as surgical emergencies. Positron emission tomography scan has very good value as a tool for detecting residual lesions after treatment of lymphoma, but it is considered to be of roughly the same value as regular CT in detecting primary lesions prior to treatment.⁵

Treatment of NHL is now considered multimodal. In addition to that, resistance to the previously used CHOP regime (cyclophosphamide, doxorubicin, vincristine, prednisolone) is now prevalent, especially among indolent lymphomas.⁶⁻⁷ In case of localised disease (stages I and II), surgical resection is carried out, along with removal of all gross tissue and anastomosis of the bowel segments. Surgery with chemotherapy is preferred in highly aggressive lymphomas. R-CHOP regime is used.⁸ For indolent lymphomas, surgery may be performed for stage I and stage II lesions, with radiotherapy used only if margins are positive for residual tumour tissue. Stages III and IV are inoperable, but surgery may be carried out to reduce bulk and alleviate symptoms.

Learning points

- ▶ Primary gastrointestinal non-Hodgkin's lymphoma (NHL) is a rare tumour, representing only 0.2–1.2% of all colonic malignancies.
- ▶ Most prominent sites of primary lymphoma in the gastrointestinal tract include the stomach (50%), the small bowel (30%) and the colon and rectum (20%).
- ▶ HIV infection as a major risk factor for NHL.
- ▶ Surgery with chemotherapy is the preferred treatment modality.

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Competing interests None.

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