CASE REPORT

Primary ileocaecal B-cell non-Hodgkin lymphoma (NHL): a rare underlying cause of right iliac fossa pain

Paschalis Gavriilidis, Barbara Christoforidou, Ilektra Michalopoulou, Anastasia Nikolaidou

Department of Pathology,
Theageneio Anticancer
Hospital, Thessaloniki, Greece

Correspondence to
Paschalis Gavriilidis,
pgavriilidis@yahoo.com

SUMMARY

Primary gastrointestinal non-Hodgkin lymphomas (NHL) account for about 0.9% of all gastrointestinal tract tumours. They are usually associated with ulcerative colitis, Crohn's disease, previous radiotherapy, renal transplantation and AIDS. We present a 36-year-old immunocompetent man, previously well with abdominal pain, weight loss and palpable mass in the right iliac fossa. A CT scan of the abdomen showed a thickening of the terminal ileum and the caecum with characteristic aneurysmal luminal dilation. Given the patient's presentation and radiology findings, a right hemicolectomy was performed. The suspected diagnosis of NHL, which was based on the imaging findings, was confirmed at histological analysis.

BACKGROUND

Primary ileocaecal lymphoma is defined as occurring if the patient has a predominant ileocaecal lesion which has been the cause of his/her presentation. Patients in whom neither ileocaecal involvement occurred after the diagnosis of lymphoma nor in whom ileocaecal disease is not the cause of their presentation are included.¹

CASE PRESENTATION

A 36-year-old immunocompetent Caucasian man presented with right iliac fossa pain, fever, constipation and weight loss of 6 kg in 7 months. He looked cachectic with temperature 38.7, pulse 90/min with normal blood pressure and respiratory rate. Clinical examination revealed right-sided abdominal peritonism and palpable mass. No lymphadenopathy. His laboratory investigations revealed anaemia, leucocytosis and high C reactive protein. High-serum levels of lactic dehydrogenase were detected. A CT scan of the abdomen revealed a wall thickening of the terminal ileum and the caecum with characteristic aneurysmal luminal dilation (figure 1). Thickened terminal ileum and caecum with enlarged mesenteric lymph nodes were identified on laparotomy. After careful examination of the rest of the abdomen, no pathology was detected elsewhere. Right hemicolectomy was performed. The histopathological report confirmed the diagnosis of diffuse large B-cell non-Hodgkin lymphoma (NHL) (figures 2–4).

Figure 1 Mural thickening of the terminal ileum and the caecum with characteristic aneurysmal luminal dilation.

INVESTIGATIONS

HIV, Widal test, Venereal Disease Research Laboratory and brucella titres were all negative. Skin tuberculin test and tumour markers were normal. Chest x-ray was normal. Bone marrow biopsy was normal.

DIFFERENTIAL DIAGNOSIS

The latest WHO classification recognises five histological subtypes of B-cell lineage: extranodal marginal lymphomas (MALT lymphoma), follicular lymphoma, mantle cell lymphoma, diffuse large B-cell lymphoma and Burkitt's lymphoma.

Figure 2 Infiltration of the lamina propria of the caecum with numerous neoplastic large B-cells embedded in a background of small lymphocytes that represent T-cells (HE x400).

To cite: Gavriilidis P, Christoforidou B, Michalopoulou I, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009052

Figure 3 Infiltration of the lamina propria of the terminal ileum with numerous neoplastic large B-cells embedded in a background of small lymphocytes that represent T-cells (HE $\times 200$).

TREATMENT

He underwent right hemicolectomy and referred to haematologists for adjuvant therapy.

OUTCOME AND FOLLOW-UP

The patient recovered uneventfully, was discharged on the eighth postoperative day and was referred to haematologists for adjuvant treatment.

DISCUSSION

Gastrointestinal lymphoma is an uncommon disease, but is the most common occurring extranodal lymphoma and is almost exclusively of non-Hodgkin type.² Gastrointestinal NHL accounts for 4–12% of all NHL.^{1,2}

The latest WHO classification³ recognises five histological subtypes of B-cell lineage: extranodal marginal lymphomas, follicular lymphoma, mantle cell lymphoma, diffuse large B-cell lymphoma and Burkitt's lymphoma.

In 1961, Dawson *et al*⁴ established the criteria for the diagnosis of primary colorectal lymphomas. These are (1) no enlarged superficial lymph nodes when the patient is first seen; (2) chest radiographs were without obvious enlargement of the mediastinal nodes; (3) white cell counts, both total and differential, are

Figure 4 Immunohistochemistry for the leucocyte common antigen (LCA) shows strong reactivity of the neoplastic cells (LCA $\times 200$).

within normal range and bone marrow biopsy is also normal; (4) at laparotomy, only regional nodes are affected by the disease; and (5) the liver and the spleen are free of tumour.

Primary ileocecal NHL is uncommon.^{1,2} Its clinical presentation is that of abdominal pain, altered bowel habits and weight loss, together with the finding of the abdominal mass. Primary ileocaecal NHL is commoner than colorectal, but rarer than gastric lymphoma.^{5,6}

Lower gastrointestinal bleeding can be found in 13–82% of patients.^{7–9} Infrequency of these complaints suggests that, despite the large size of the lymphoma, mucosal ulceration does not usually occur.⁷

CT shows uniform thickening of the intestinal wall, without any associated desmoplastic reaction. Infiltration of the muscularis propria and the autonomic plexus may result in atonic, aneurysmal dilation of the lumen.¹⁰ Sometimes lymphoma may form an annular napkin-ring lesion that mimics carcinoma.¹⁰ When CT reveals the presence of a bulky mass or diffuse infiltration with the preservation of fat planes and no obstruction, multiple site involvement and associated bulky lymphadenopathy, lymphoma should be the primary consideration in the differential diagnosis and must be excluded by biopsy.

The small number of patients with various histological subtypes and different stage at presentation results in unclear treatment protocol.

It is reported that patients receiving chemotherapy generally survived longer than those did not (median 34 months vs 14 months).^{2,8}

Learning points

Surgery is the main treatment for primary colorectal lymphomas:

- ▶ It provides data for prognosis, including histology, extent and stage of the lesion.
- ▶ It offers the opportunity for cure either in association with adjuvant chemotherapy or not.
- ▶ It prevents complications such as haemorrhage, obstruction or perforation from occurring spontaneously or during chemotherapy.

Contributors PG managed and operated the patient. BC, IM and AN studied the surgical specimen. All the doctors reviewed the literature.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Auger MJ, Allan NC. Primary ileocecal lymphoma. A study of 22 patients. *Cancer* 1990;65:358–61.
- 2 Bush RS. Primary lymphomas of the gastrointestinal tract. *JAMA* 1974;228:1291–4.
- 3 Harris BL, Jaffe ES, Diebold J, *et al*. Lymphoma classification: from controversy to consensus: the R.E.A.L and WHO classification in lymphoid neoplasms. *Ann Oncol* 200;11(Suppl 1):3–10.
- 4 Dawson IM, Cornes JS, Morson BC. Primary malignant lymphoma tumours of the intestinal tract. Report of 37 cases with a study of factors influencing prognosis. *Br J Surg* 1961;49:80–9.
- 5 Fleming ID, Mitchell S, Dilawari RA. The role of surgery in the management of gastric lymphoma. *Cancer* 1982;49:1135–41.
- 6 Rosenfelt F, Rosenberg SA. Diffuse histiocytic lymphoma presenting with gastrointestinal tract lesions: the Stanford experience. *Cancer* 1980;45:2188–93.

- 7 Cai S, Cannizzo F Jr, Bullard Dunn KM, *et al.* The role of surgical intervention in non-Hodgkin's lymphoma of the colon and rectum. *Am J Surg* 2007;193:409–12; discussion 412.
- 8 Fan CW, Changchien CR, Wang JY, *et al.* Primary colorectal lymphoma. *Dis Colon Rectum* 200;43:1277–82.
- 9 Cho MJ, Ha CS, Allen PK, *et al.* Primary non-Hodgkin lymphoma of the large bowel. *Radiology* 1997;205:535–9.
- 10 Montgomery M, Chew FS. Primary lymphoma of the colon. *Am J Roentgenol* 1997;168:688.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow