CASE REPORT

Lymphoma in an ileostomy

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Lymphoma developing in an ileostomy is an extremely rare complication. The presentation is similar to the commoner, yet still rare, adenocarcinoma but the staging and management of the condition differs.

78 year old man was referred to the surgical outpatient clinic by the stoma care clinical nurse specialist. He was having increasing difficulties with his long standing right iliac fossa end ileostomy that had been created 20 years earlier for ulcerative colitis. He noticed that it was functioning but prolapsed, extending to approximately 10 cm from the skin surface. It was very hard, swollen with a diameter of 6.4 cm, with the tip appearing bruised and black, stenosed, and bleeding to the touch. These changes had occurred over a six month period and been associated with several episodes of bleeding. It was painless but he was having great difficulty in obtaining a satisfactory seal with the stoma appliances. His weight was steady and his appetite good. He was a hypertensive on treatment and was otherwise well.

Examination confirmed the above findings of a swollen, necrotic, and stenosed stoma (see fig 1) and also revealed two skin deposits adjacent to the stoma and a large parastomal hernia. A clinical diagnosis of a malignant change in an ileostomy was made and after discussion with the patient, a decision to undertake an en-bloc excision and refashioning of the stoma was made. A preoperative biopsy was not carried out as it was felt that it would not change the above management plan.

A midline laparotomy was carried out to clinically stage the disease. No obvious mesenteric or para-aortic nodal enlargement was detected. The liver and spleen were impalpable due to adhesions. The ileostomy was excised en-bloc with the abdominal wall through a transverse elliptical incision. A new end ileostomy was fashioned in the left iliac fossa.

Histology revealed a B-cell non-Hodgkins lymphoma of the ileum. The tumour appeared to have been completely excised.



Figure 1 Stoma preoperatively showing infiltration of the skin and necrosis.

Standard staging tests showed that the tumour appeared to be localised. In view of its large size and grade II histology and hence a high probability of occult disease, chemotherapy was offered with PACEBO (prednisolone, Adriamycin, cyclophosphamide, etoposide, bleomycin, and oncovin), an eight week protocol discussed by the British National Lymphoma Investigation for elderly patients with non-Hodgkin's lymphoma.¹

The patient could complete only four weeks out of the recommended eight of adjuvant chemotherapy due to toxicity as he developed overt congestive cardiac failure. Follow up computed tomography six months postoperatively has shown no signs of relapse/recurrence.

DISCUSSION

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This association has been reported on only once before and this was in a patient with transfusion related AIDS, two years after undergoing a total colectomy and ileostomy for indeterminate colitis.²

Adenocarcinoma of the small intestine is rare compared with other sites in the gastrointestinal tract. Of the four major primary small bowel tumours (adenocarcinomas, lymphomas, carcinoid, and leiomyosarcomas), the former two are associated with diseases that seem to increase the risk of developing these malignancies. Both immunoproliferative small bowel disease and coeliac disease predispose patients to the development of primary lymphoma.³ The association of coeliac disease with T-cell lymphoma is well recognised.

Ileostomies are associated with a number of complications such as skin excoriation, stenosis, parastomal herniation, intestinal obstruction, retraction or prolapse of the stoma, abscess and fistula formation, and ileitis.4 Adenocarcinoma arising in the abnormally placed small intestinal mucosa (that is, ileostomy, ileoanal pouch) several years after the initial operation is being increasingly recognised and reported.^{4 5} The intestinal flora in an ileostomy come to resemble colonic flora6 and this has been proposed as a potentially carcinogenic factor.7 Further, colonic metaplasia and dysplasia has been found in the mucosa adjacent to an adenocarcinoma. It has been postulated that chronic physical or chemical irritation of an ileostomy predisposes the ileal mucosa to colonic metaplasia, dysplasia, and frank malignant change.⁷⁻⁹ To prevent ileostomy carcinoma it is recommended that a biopsy of all polyps at the mucocutaneous junction and of any non-prolapse associated polyps elsewhere on the stoma occurring >15 years after ileostomy construction is done.¹⁰ En-bloc resection of the ileostomy, wide resection of the anterior abdominal wall, and transposition of the stoma to a new site have been shown to provide the best prognosis for adenocarcinoma arising in an ileostomy.9 11-13

A preoperative biopsy, particularly at an early stage in the above case, might have changed management. It would have enabled a discussion of the case with haematology and oncology teams with regards to primary chemotherapy and radiotherapy as alternatives to surgery. Their advantages and disadvantages, however, have to be considered in each individual case, taking into account age, past medical history, presentation, and staging of the disease. The advantage of avoidance of surgery has to be balanced with the risk of toxicity as in our case, and sepsis after chemotherapy due to the presence of tissue necrosis. We are doubtful whether radiotherapy would have been helpful in the presence of an established stenosis: we think it would have been more likely to make matters worse. Radiotherapy may have succeeded in down-staging or even curing the local disease at the risk of radiation enteritis especially in the presence of a parastomal hernia.

We therefore think that we did perform the right operation in our case, although we concede that we should have secured a firm preoperative diagnosis.

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