Primary squamous cell carcinoma of the terminal ileum

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Abstract
A case of squamous cell carcinoma of the terminal ileum with no underlying duplication or inflammatory disorder is described. The neoplasm seemed to have originated from the surface epithelium, invading the wall and metastasising to the regional lymph nodes. The 65 year old patient was free of disease three years after having had the tumour removed.

Previous reports of squamous carcinoma of the small intestine have been associated with intestinal duplication or metastatic disease from distant sites.

Case report
A 62 year old man was admitted to hospital with a 12 month history of intermittent mid-abdominal colic and vomiting. A barium enema performed as an outpatient before admission showed an extrinsic filling defect in the caecum. No reflux of barium into the terminal ileum was shown. A computed tomogram of the abdomen showed an homogeneous lobulated mass in the right iliac fossa which seemed to affect small bowel loops. There was no evidence of metastatic deposits in the liver. A chest x-ray picture, routine biochemical tests, and blood count were within normal limits.

At laparotomy, a tumour was found arising from the terminal ileum, with enlarged nodes in the ileocaecal region. There was no other evidence of intra-abdominal disease. A resection of the terminal ileum with a limited right colectomy was carried out to remove all macroscopic disease. Intestinal continuity was restored by end to end anastomosis. The patient made an uneventful recovery and was alive and well three years later with no evidence of recurrence.

Pathology
The resected specimen consisted of 40 cm of small intestine, the appendix, and 10 cm of large bowel with the associated mesentry. There was a fungating tumour sparing the terminal 5 cm of ileum but affecting the mucosa of the next 5 cm and spreading through the thickness of the wall into the mesentery. Focal areas of calcification were identified on cut section.

Paraffin wax sections showed the tumour to be a moderately differentiated keratinising squamous cell carcinoma arising from the surface epithelium. Intercellular bridges and keratinous cysts and pearls were prominent (figure). The tumour had infiltrated the wall and extended into the mesentery and paraintestinal lymph nodes. There was evidence of vascular invasion, but none of glandular differentiation or adenomatous change in any part of the tumour.

Mucin stains and Gremelius argyrophil stains were negative. Electron microscopical examination showed features typical of squamous cell differentiation. The neoplastic cells contained well formed tonofilaments, while microvilli, glyocalceal bodies; mucinous granules were absent.

Discussion
The spread of tumour into the small bowel as a result of secondary squamous carcinoma has been well documented.1-3 Primary malignant tumours, however, are rare and the squamous carcinomas have so far been reported in cases of duplication.4

In the colon squamous cell differentiation is seen in about 0·05% of adenocarcinoma5 and in 0·4% of adenoma.6 The pathogenesis of squamous cell carcinoma of the colon is unknown but in some cases it has been associated with carcinoma
complicating ulcerative colitis,7 schistosomiasis,4 pelvic irradiation5 and villous adenoma.

The possibilities for squamous differentiation in the intestine may result from any of the following mechanisms:

1. malignant transformation of heterotopic rests of squamous epithelium in the submucosa;
2. aberrant differentiation of stem cells to squamous cells with subsequent malignant change;
3. squamous metaplasia of glandular cells with subsequent malignant change;
4. transformation of an adenocarcinoma into an epidermoid carcinoma.

We believe that our patient had a primary squamous cell carcinoma of the terminal ileum without underlying adenoma, inflammatory disease, or duplication. As far as pathogenesis is concerned, we cannot exclude any of the four basic mechanisms proposed above.