Mixed adenocarcinoma/carcinoid tumour of large bowel in a patient with Crohn’s disease

Y L Hock, K W M Scott, R H Grace

Abstract
A 50 year old woman with a 20 year history of Crohn’s disease underwent laparotomy which revealed extensive disease in the small and large bowel, and this was resected. Gross examination of the resected bowel showed features of Crohn’s disease as well as a polypoid tumour in the caecum. Histopathological examination of the tumour showed it to be an infiltrating mixed adenocarcinoma/carcinoid tumour arising in a tubulovillous adenoma.

Random sampling of the rest of the bowel affected by Crohn’s disease also showed a focus of dysplasia and adenomatous change. It is suggested that Crohn’s disease may have played a part in the pathogenesis of the tumour.

Case report
A 50 year old woman with a 20 year history of small and large bowel Crohn’s disease was admitted to hospital for laparotomy following the failure of medical treatment for increasingly severe right iliac fossa pain. There were no clinical features suggestive of a carcinoid syndrome and there was no clinical suspicion, biochemical tests for carcinoid syndrome were not performed.

Laparotomy showed Crohn’s disease from 15 cm proximal to the ileo-caecal valve to the junction of the descending and sigmoid colon. The region of the appendix was adherent to the anterior abdominal wall. Resection of the affected bowel together with ileo-sigmoid anastomosis was performed.

Pathological examination
The specimen included 15 cm of terminal ileum and 45 cm of large bowel. A paracolonic abscess encasing the appendix was noted. On opening the bowel, there was a continuous segment of mucosa showing the typical “coble stone” appearance of Crohn’s disease, measuring 32 cm in length and involving the

mucosa. Histological analysis showed the polypoid lesion to be a tubulovillous adenomatous polyp, which showed progressive dysplasia and became continuous with an area of infiltrating malignant tumour composed of an intimate admixture of two components: one component showing features of moderately differentiated colonic adenocarcinoma (fig 1), and the other showing solid nests and trabeculae of uniform cells, surrounded by peripheral rims of smaller cells with hyperchromatic nuclei (fig 2), typical of carcinoid tumour.

Occasional rosette formations were also seen. The foci of carcinoid areas showed negative diazo and Grimelius reactions, but strong positivity with neuron specific enolase on immunocytochemistry. The tumour had infiltrated through the full thickness of the bowel wall and into the pericolic fat.

Sections from the rest of the large bowel, as well as from the ileum, showed transmural chronic inflammation with numerous lymphoid aggregates. Giant cell granuloma were also seen in the muscularis propria and in the submucosa.

About 12 cm distal to the malignant tumour, the inflamed colonic mucosa also showed a separate small focus of villous adenomatous change with low grade epithelial dysplasia (fig 3A). There were no metastases or granulomata in the lymph nodes recovered.

The pathological diagnosis was mixed adenocarcinoma/carcinoid tumour (composite carcinoma carcinoid tumour) occurring in a colon affected by Crohn's disease.

Discussion

There is an accepted association between Crohn's disease and cancer of the gastrointestinal tract, and the relative risk compared with the normal population ranges from fourfold to 20-fold according to various epidemiological studies. In real terms, roughly 2% of patients with Crohn's disease will develop cancer in the course of the disease.

Although the risk of colorectal cancer in patients with ulcerative colitis is related to the duration of disease, patients with Crohn's disease are at risk even in the first 10 years of their disease, and the disease activity does not appear to affect the risk of cancer.

It has been established that malignant change in Crohn's disease is often a chance discovery by the pathologist, and 59% of all cancers complicating Crohn's disease have only been discovered during pathological examination of the surgical specimens. As in ulcerative colitis, adenocarcinoma of the large bowel in Crohn's disease occurs characteristically in association with foci of dysplasia and villous adenomatous change.

The concurrence of typical carcinoid admixed with adenocarcinoma of the gastrointestinal tract is a well known although an unusual phenomenon that has been reported in the oesophagus, stomach, gall bladder, colon and appendix, and has been termed composite carcinoma carcinoid tumour.

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**Figure 2** Another focus shows solid nests of uniform cells surrounded by peripheral rims of smaller cells with hyperchromatic nuclei typical of carcinoid tumour (haematoxylin and eosin).

**Figure 3** Villous adenomatous change showing epithelial dysplasia in: (A) mucosa further away from the malignant tumour; (B) the adjacent mucosa. The infiltrating tumour is also apparent in the left hand corner (haematoxylin and eosin).
Epithelial myoepithelial tumour of the tracheal gland

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Abstract

A case of epithelial myoepithelial tumour originating from the tracheal gland in a 57 year old woman is described. The tumour was removed by segmental tracheal resection and end-to-end anastomosis. Histologically, the tumour comprised clear cells and presented a monophasic pattern. Immunohistochemical analysis showed that the tumour cells were positive for both S-100 protein and smooth muscle actin, suggesting that this tumour resembles a subtype of epithelial-myoid epithelial carcinoma described in the 1990 WHO international classification of salivary glands.

Although some reports describe a clear cell dominant epithelial myoepithelial carcinoma, in this case local invasiveness or regional lymph node metastasis was not proved through investigation. It is therefore concluded that this was an epithelial myoepithelial tumour rather than a carcinoma.


Among the cases difficult to classify histologically, the rare case presented here, that of a tracheal tumour, is thought to be a clear cell dominant epithelial myoepithelial tumour.

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