

Adenocarcinoid Of The Midgut: An Unusual Presentation Of A Rare Tumour

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Abstract

Adenocarcinoids or Goblet cell tumours are rare tumours of the gastro-intestinal tract commonly seen in the appendix. There is no definite investigation to prove the presence of these tumours. Only histology confirms presence of adenocarcinoids as, tumours with microscopic features of an infiltrating tumour with distinct glandular differentiation and focal mucin production and are a variant of carcinoids showing features of both carcinoids and colonic adenocarcinomas.

These tumours appear to be more aggressive than carcinoids but not as aggressive as adenocarcinomas. Radical surgical resection is curative and palliative chemotherapy appears to improve the 5-year survival. Here we present an unusual case report of such a rare tumour exclusively in the small bowel.

INTRODUCTION

Adenocarcinoids are rare tumours of the gastrointestinal tract commonly seen in the appendix, showing features of carcinoid as well as adenocarcinoma (1). Hereby, we report an unusual presentation of one such rare tumour in the small intestine.

CASE REPORT

A 63-year-old gentleman was referred with upper abdominal pain, vomiting and progressive weight loss. Examination revealed a tender mass in the left hypochondrium. X-ray as well as ultrasound scan of the abdomen were normal. Oesophago-gastro-duodenoscopy and barium enema were also normal. Computed tomography revealed a mass around the duodeno-jejunal flexure.

The case was discussed at a multi-disciplinary meeting on numerous occasions and eventually a decision to perform small bowel contrast study was taken, which showed an area of irregularity in the upper jejunum.

Exploratory laparotomy, performed with a tentative pre-operative diagnosis of small bowel lymphoma showed a mass in the left hypochondrium adherent to the anterior abdominal wall. It involved the proximal jejunum 2cm beyond the duodeno-jejunal flexure and infiltrated into the distal transverse colon. There were two other distinct lesions in the omentum and a separate stricturing lesion in the

terminal ileum with no distant metastases.

A wide en-bloc resection of the entire lesion including the loop of jejunum and the left hemi colon along with excision of the adjacent abdominal muscle was performed. Continuity of the gut was restored with an end-to-end duodeno-jejunal and anastomosing the proximal transverse colon to the distal sigmoid colon. Omentectomy was performed and the ileal stricture was also resected. The patient made an uneventful recovery.

Gross pathological examination revealed tumour primarily arising from the jejunal mucosa and infiltrating into the colon from the serosal aspect with intact colonic mucosa with synchronous tumour in the terminal ileum (2). Both tumours had a distinct pale yellow appearance and showed microscopic features of an infiltrating tumour with distinct glandular differentiation and focal mucin production. There was both perineural and perivascular invasion with marked desmoplastic response. The morphological features were highly suggestive of tumours showing both endocrine and glandular differentiation. Neurone specific enolase and synaptophysin were both focally positive in the tumour cells confirming the endocrine component, fulfilling the criteria for adenocarcinoid tumours.

Figure 1

Figure 1: Cut section of the jejunum showing mucosal ulceration due to tumour with infiltration of tumour into adjacent mesenteric fat.

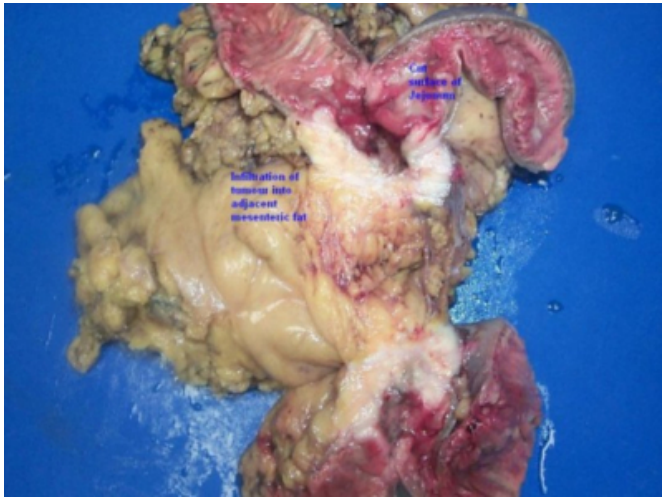
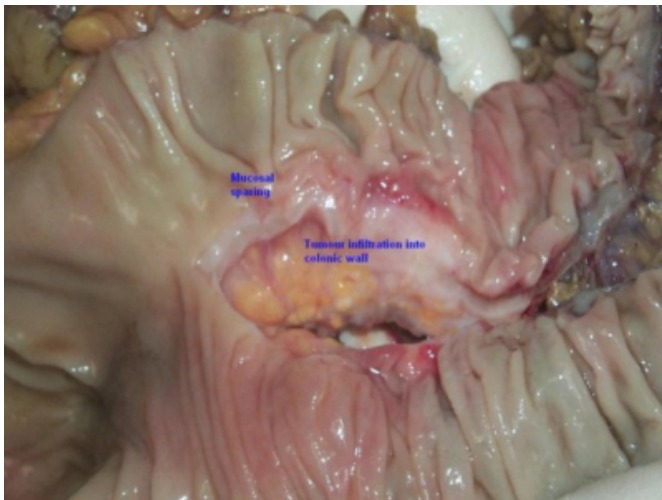


Figure 2

Figure 2: Cut section of the transverse colon showing tumour infiltrating into the wall with mucosal sparing.



DISCUSSION

Adenocarcinoids, also called “goblet cell carcinoids”, are rare tumours of the gastrointestinal tract. They are variants of carcinoid tumours arising in the small bowel (commonly in the appendix) with a low propensity to metastasize (^{1,3}). These tumours classically show histological features of both carcinoids and well-differentiated colonic adenocarcinomas (⁴). There is some evidence to suggest the role of mutation of p53 tumour suppression gene in the aetiology of this tumour (⁵).

Extensive literature search have shown adenocarcinoids to be unusual in the small intestine and occurring more

commonly in the appendix (^{1,3}).

Electron microscopy studies have shown both mucin droplets and neurosecretory-type granules in adenocarcinoids. However, there is some controversy as to whether they are located in the same cell (⁴).

Collision or composite tumours are different from adenocarcinoids in that they represent different tumours occurring at the same site whereas adenocarcinoids represent a single tumour showing neurosecretory and mucinous differentiation (^{6,7}). These tumours appear to be more aggressive than classical carcinoids but not as aggressive as adenocarcinomas (^{1,3}).

Curative treatments appear to be radical surgical resection. Palliative chemotherapy appears to improve the 5-year survival rate in patients with advanced and metastatic disease (⁸).

CONCLUSION

This case had an atypical mode of presentation and represents multifocal adenocarcinoids arising in the small intestine, an unusual site. It highlights the value of a multidisciplinary approach to such unusual cases whereby pathologists, radiologists, surgeons and oncologists can interact together in reaching the diagnosis and planning appropriate management.

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