CASE REPORTS

Adenocarcinoma of the gastro-oesophageal junction with a synchronous carcinoid of the duodenum

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Abstract

Duodenal carcinoids are rare tumours. There is an increased incidence of primary carcinomas, especially in the gastrointestinal tract, which occur synchronously with gastrointestinal tract carcinoids. However, the synchronous occurrence of adenocarcinoma of the gastro-oesophageal junction with a duodenal carcinoid has not been previously described. A case report is presented, with discussion of carcinoid tumours and management when occurring synchronously with non-carcinoid tumours.

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A man was referred for an oncological opinion after a curative resection of a gastro-oesophageal junction adenocarcinoma, because of the discovery of a synchronous duodenal carcinoid. This is the first time a duodenal carcinoid synchronously occurring with an adenocarcinoma of the gastro-oesophageal junction has been described in the literature.

Case report

The patient, a 68 year old man, initially presented with an upper gastrointestinal bleed. Endoscopy showed a polyp in the lower oesophagus, histology of which showed “low grade dysplasia”. Subsequent endoscopy showed unequivocal high grade dysplasia with significant risk of carcinoma *in situ*. He opted to undergo oesophagogastrrectomy and histology showed a fully resected adenocarcinoma of the gastro-oesophageal junction (graded T1N0M0).

A duodenal specimen was found to have a 0.4 cm tumour with one lymph node positive. The cells of this tumour had granular eosinophilic cytoplasm and immunohistochemistry showed expression of synaptophysin, features consistent with a diagnosis of carcinoid. Due to this finding he was referred for an oncology opinion. He was found to be fit and well and recovering from his operation. Before surgery he had had no symptoms pertinent to his carcinoid tumour. Examination was normal and 24 hour urinary 5-hydroxyindole-acetic acid (5-HIAA) was within the normal range at

22 µmol/24 hours (normal range 0–50 µmol/24 hours). His gastro-oesophageal tumour had been fully resected with no evidence of metastatic disease and therefore required no further treatment other than observation—there is no role for adjuvant chemotherapy in this scenario. The carcinoid tumour, despite the presence of one local nodal metastasis, also requires no active treatment other than observation.

Discussion

Carcinoid tumours of the gastrointestinal tract arise from neuroendocrine cells that line the tract. They occur most frequently in the appendix (35%) and ileum (20%). These are the “midgut” carcinoids. Ten per cent occur in the rectum (“hindgut” carcinoid), 10% in the stomach, and 1.5%–5% in the duodenum (“foregut” carcinoids). Less commonly they can occur elsewhere in the gastrointestinal tract. The annual incidence of small intestine carcinoids is reported as 2.8 per million population.¹ The incidental finding at necropsy is much higher: overall annual incidence of malignant carcinoid was 21 per million population per year in one study.²

Histologically they are usually easy to recognise, but in uncertain cases immunohistochemistry detects neuroendocrine markers, such as neurone specific enolase, chromogranins, synaptophysin, serotonin, and neurohormonal peptides.³ Carcinoid cells typically synthesise serotonin (5-HT) from dietary tryptophan in greater proportion than in healthy individuals—where 99% of dietary tryptophan is converted to nicotinic acid and only 1% is converted to 5-HT. In patients with carcinoid tumours the production of 5-HT and therefore its metabolite 5-HIAA is increased. This does not usually cause any problems unless serotonin and other secretory products are secreted directly into the systemic circulation, for example, when liver metastases are present. In the carcinoid syndrome, usually secondary to a metastatic midgut carcinoid, serotonin and other gastrointestinal tract peptides (for example, pancreatic polypeptide, glucagon, vasoactive intestinal peptide, calcitonin, glucagon), are released and the well recognised features of flushing, diarrhoea, wheezing, abdominal pain, and carcinoid heart disease can occur. It is not known exactly how all the hormones contribute to the various symptoms and signs, however.

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Carcinoid tumours are associated with an increased incidence of secondary primary malignancies, especially adenocarcinoma. A retrospective review by Gerstle *et al* of 69 patients with carcinoids in their gastrointestinal tracts found that 29 (42%) had synchronous tumours and three (4%) had metachronous tumours.⁴ Carcinoma of the colon was the second primary in 7/32 patients (21.9%), with the gastrointestinal tract accounting for 42.9% of the second primary site of tumour overall. Of the patients with synchronous tumours, similar to our patient, none had symptoms attributable to the carcinoid tumour. Another series from France reviewed 270 cases of gastrointestinal tract carcinoid and 21 were associated with synchronous tumours⁵; two thirds were associated with adenocarcinoma of the colon, confirming the observation that the large intestine is the most common site for a synchronous primary to occur with a gastrointestinal tract carcinoid. Less common combinations include: appendix carcinoid occurring with a T cell lymphoma of the ileum,⁶ oesophageal carcinoid occurring with oesophageal adenocarcinoma in Barrett's oesophagitis,⁷ and uterine carcinoid occurring with adenocarcinoma also in the uterus.⁸ Duodenal carcinoid is rare and associated synchronous primary tumours have been rarely reported in the literature. It has been described in association with gastric leiomyoblastoma.⁹ The synchronous occurrence of adenocarcinoma of the gastro-oesophageal junction with duodenal carcinoid has not been previously described in the literature, though the coincidence may partly be explained by the increasing incidence of gastro-oesophageal junction adenocarcinomas,¹⁰ and to a lesser extent the previously described increased incidence of foregut carcinoids.¹¹

How is the patient managed once a diagnosis of synchronous carcinoma with carcinoid is made? As nearly all are discovered incidentally they are invariably at an early stage and usually fully resected. Management is directed towards the carcinoma. This patient had a potentially curative resection for a small gastro-oesophageal junction carcinoma and further treatment in the form of adjuvant chemotherapy is not indicated. Prognosis is also determined usually by the non-carcinoid

tumour. The five year survival rate for fully resected carcinoid depends upon site, with a worse prognosis for small intestine tumours (~ 75%) than for the appendix (~99%). The prognosis for this patient is however quite good, as the five year survival rate for a T1N0M0 oesophageal carcinoma is approximately 66%.¹² His carcinoid was fully resected also, and for both tumours observation is all that is required at present.

In conclusion, this is the first time a duodenal carcinoid in association with an adenocarcinoma of the gastro-oesophageal junction has been described. It is important to be aware of the possibility of synchronous cancers, especially in those patients presenting with colonic carcinomas. It is advised that patients presenting with any neoplasm in the gastrointestinal tract should undergo thorough exploration of the peritoneal cavity and its organs at initial surgery to exclude this,¹³ but in general the management and prognosis depend on the non-carcinoid malignancy.

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