Gastrointestinal Carcinoids

N RANGABASHYAM, D SRIKUMARI, S VISWANATH

Department of Surgical Gastroenterology and Proctology, Madras Medical College, Madras 600 003

Abstract

Eleven cases of carcinoid tumour occurring in the gastrointestinal tract have been reviewed over a period of fifteen years, with their mode of presentation and management. None of our patients had the classical carcinoid syndrome or association with other endocrine anomalies or peptic ulcer. All our patients underwent surgical treatment and the results were good.

Key words: Carcinoid tumour of gastrointestinal tract; carcinoid syndrome; multiple endocrine neoplasia; surgical resection.

Introduction

The term carcinoid was originally given to small tumours in the submucosa of the ileum and appendix in the erroneous belief that they were benign. Although now recognised as malignant, the name is still appropriate because of the long survival even in the presence of metastasis. Carcinoïds, being descendants of the amine precursor uptake and decarboxylase (APUD) cell system, occur anywhere in the gastrointestinal system where Kulitschinsky cells are present—from the gastric cardia to the anus.

Material and Methods

Eleven cases (9 males, 2 females) of gastrointestinal carcinoids were evaluated by us over a period of fifteen years. The distribution of the tumours was as follows: appendix (5), stomach (3), and jejunum, ileum and rectum one each. One patient each with appendicular and jejunal carcinoids were females.

Symptoms

The symptoms depended on the site of the tumour. Carcinoid appendix presented with features of appendicitis, ie pain in the right iliac fossa. Of the three cases with gastric carcinoids, two had pain in the epigastric region associated with melena in one and loss of appetite and of weight in the other. The third patient came with abdominal distension and loss of appetite and weight. The jejunal and ileal carcinoids presented with features of intestinal obstruction in the form of vomiting, ball rolling movements, loss of appetite and weight. The case with rectal carcinoid came with bleeding per rectum and alternating constipation and diarrhoea. All these were isolated features and none of our patients had features of carcinoid syndrome or of multiple endocrine neoplasia (MEN).

Clinical Findings

All five patients with appendicular carcinoids had tenderness in the right iliac fossa. One patient with gastric carcinoid had a large, nodular mass in the epigastrium. Another had ascites with tenderness in the left hypochondrium. The third had no physical finding except for anemia and malnourishment, which were present in the other two also. The ileal and jejunal carcinoids presented with occasional visible intestinal peristalsis, whereas the case with rectal carcinoid presented with an ulcer, 3 cm in diameter, 5 cm from the anal verge.

Investigations

The basic haematological values were within normal limits, except for erythrocyte sedimentation rate which was raised in eight cases, and haemoglobin which was reduced in five cases. One patient had pulmonary tuberculosis and one was a diabetic. Barium meal study revealed filling defects in the antral region in cases with gastric carcinoids, and multiple strictures and stasis in the cases with jejunal and ileal carcinoids. A preoperative diagnosis was made based on biopsy of the ulcer in the case with rectal carcinoid.

There were no clinical or investigatory findings suggestive of pleuriglandular (MEN) syndrome. Urinary 5-HIAA estimation could not be done. Since all our cases required some form of resection, diagnosis was established only after post operative histopathological examination, except in the case with rectal carcinoid, as stated above.

Treatment

All our cases required surgery as medical management did not give symptomatic relief. The procedure used in each case and the findings on laparotomy are shown in the Table.

Table: Findings at laparotomy and surgical procedure

<table>
<thead>
<tr>
<th>Site</th>
<th>Findings at Laparotomy</th>
<th>Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stomach</td>
<td>1. Polypoid growth in the antrum with nodes in lesser and greater curvature</td>
<td>Polypectomy</td>
</tr>
<tr>
<td></td>
<td>2. Firm mass in pyloric region with eosinotic nodes</td>
<td>Gastroscopy</td>
</tr>
<tr>
<td></td>
<td>3. Ulcer in lesser curvature with subpyloric glands</td>
<td></td>
</tr>
<tr>
<td>Jejunum</td>
<td>Stricture about 25 cm from duodenal/jejunal flexure with mesenteric lymph nodes</td>
<td>Resection with end-to-end anastomosis</td>
</tr>
<tr>
<td>Ileum</td>
<td>Stricture in the proximal ileum</td>
<td></td>
</tr>
<tr>
<td>Appendix</td>
<td>Inflamed appendix with yellowish plaque-like lesions in four and ulcer in one case</td>
<td>Appendicectomy</td>
</tr>
<tr>
<td>Rectum</td>
<td>Ulcer 3 cm in size about 5 cm from anal verge</td>
<td>Abdomino-peritoneal resection</td>
</tr>
</tbody>
</table>
Discussion

According to the literature,

which may occur at any age, are those common to any tumour of the gastrointestinal tract. The classical syndrome is characterised by episodic flushing of the face and upper body, associated with telangiectasia and cyanosis, chronic watery diarrhoea, wheezing and evidence of right sided valvular heart disease. The liver may be enlarged if there is metastasis, and palpation of such a liver usually produces the symptoms. None of our cases of gastrointestinal carcinoids presented with the classical syndrome. This is due to the fact that the liver was not involved and so was able to cope with the excess serotonin. Another interesting feature is the association of carcinoid with multiple endocrine neoplasia (MEN). Cytochemical studies have identified the existence of migratory neural crest cells termed APUD cells which may link carcinoid tumours with parathyroid tumours, medullary carcinoma of the thyroid and phaeochromocytoma (MEN type I) or parathyroid, pituitary and pancreatic tumours (MEN II).

There is still no direct technique available for the diagnosis of carcinoid syndrome. The best index is urinary 5-HIAA (more than 50 mg is diagnostic) but this has some drawbacks: ingestion of bananas, tomatoes, pineapples, walnuts or other serotonin containing food may lead to high levels of 5-HIAA in the serum and urine; in occult disease also, urinary 5-HIAA level is raised.

Other ancillary diagnostic procedures include the flush test, radiography, which may show filling defects, liver scan, which may show secondaries, and needle biopsy of the liver. Treatment consists of resection of all or as much of the tumour as possible even with widespread metastasis, because this gives significant palliation. Drugs in the form of 5-fluorouracil, steroids, cyclophosphamide and methysergide have been used to alleviate symptoms effectively. The overall 5 year survival rate is about 50%. Palliative and curative resections give 25% and 70% 5 year survival rates respectively.

References