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Adenocarcinoma of the duodenum is an exceedingly rare disorder. Its vague and non-specific symptoms often lead clinicians and patients to suspect other more benign differential diagnoses. Improved endoscopy and radiology have enabled more of these cases to be unearthed. Definitive surgery is the only means of potential cure, with the prognosis being significantly better for node-negative patients. We present a case of a 52-year-old man who underwent a Whipple's operation for this uncommon disease and a literature review of the subject.

Introduction

Cancer of the small bowel is rare, accounting for 1% of gastro-intestinal malignancies.¹ In a 1972 autopsy study, the incidence was 0.019 to 0.5%.² Nonetheless, the reported incidence has been increasing owing to the widespread use of endoscopy. Adenocarcinomas are the most common of small bowel malignancies, followed by carcinoid tumours, lymphomas, and leiomyosarcomas. Around half of all small bowel adenocarcinomas are located in the duodenum, making this the most favoured site.³ We report a case of a 52-year-old otherwise healthy man suffering from this commonest form of small bowel malignant tumour.

Case report

A 52-year-old man, a non-smoker but moderate alcohol drinker, presented to our unit in May 2006 with a 1-month history of epigastric fullness, anorexia, repeated vomiting, and weight loss. Physical examination demonstrated succussion splash. Baseline blood investigations showed only mildly abnormal liver function tests and a slightly elevated amylase level, compatible with chronic alcoholism. Gastric outlet obstruction was suspected. An oesophagogastroduodenoscopy was performed revealing swelling of the papilla and surrounding duodenal mucosa and narrowing of the canal. The scope could not pass through the duodenal lumen. A mucosal biopsy showed severe inflammation and mild dysplasia. A subsequent barium meal demonstrated a stricture at the junction between the second and the third part of the duodenum (D2/3) that still permitted a slow antegrade flow of contrast into the small bowel (Fig 1a). Computed tomography revealed a circumferential nodular wall thickening at the D2/3 junction, matching with a hypermetabolic focus with a maximum standardised uptake value (SUV_{max}) of 4.4 on the positron emission tomographic (PET) scan (Fig 1b). Having explained the findings, the patient agreed to undergo laparotomy. A 2-cm stenotic mass at the D2/3 junction with apparent uncinate involvement was noted (Fig 2). Whipple's operation was performed. Pathological examination of the surgical specimen found a 3-cm focally indurated region just distal to the Ampulla of Vater. On microscopy an adenocarcinoma with tall columnar cells extending through the muscularis propria was demonstrated. The common bile duct, pancreas, stomach, and peritoneum were not affected. None of the 11 sampled lymph nodes were involved. The diagnosis of adenocarcinoma, pT3N0 (American Joint Committee on Cancer [AJCC] classification), of the duodenum was established. His postoperative course was uneventful. At the 6-month follow-up the patient remained well.

Key words

Adenocarcinoma; Duodenal neoplasms; Pancreaticoduodenectomy; Prognosis

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Discussion

Adenocarcinoma of the duodenum is uncommon, accounting for less than 0.4% of all gastro-intestinal tract tumours. About 45% of these tumours arise from the third and fourth parts of the duodenum.⁴ Although most cases are sporadic, associations with familial adenomatous polyposis (FAP), Crohn's disease,⁵ Peutz-Jeghers syndrome,⁶ and neurofibromatosis¹⁷ have been reported. With early diagnosis and treatment of colonic polyposis, adenocarcinoma of the duodenum has become the leading cause of death in FAP patients. A 10-year prospective study of 114 FAP patients revealed that six died of adenocarcinoma of the duodenum.⁸ The incidence was higher in those patients with more

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十二指腸腺癌

十二指腸腺癌為一罕見疾病，其病徵模糊且非特定，常令人誤診為其他良性疾病。憑藉先進的內窺鏡和放射診斷技術，這病症較以往容易被發現。手術是唯一治療方法，尤以無淋巴擴散者之預後為佳。我們報告一名52歲患者接受惠普爾手術的病例，並就此疾病覆閱相關文獻。



FIG 1. (a) Barium meal showing a stricture at the D2/3 junction with incomplete obstruction; and (b) computed tomographic scan showing an obstructing mass in the duodenum not involving the bile duct or the pancreas

advanced duodenal polyposis at the beginning of the study.

The presentation of this disease is vague and non-specific. Patients can present with abdominal pain, bleeding, weight loss, obstruction, or jaundice. It should therefore be considered a differential diagnosis in patients presenting with epigastric discomfort. First-line investigations remain oesophago-gastroduodenoscopy and contrast studies, which can usually demonstrate the site, severity, and length of the lesion. While more proximal tumours can be picked up by oesophagogastrroduodenoscopy (with or without indigo carmine dye spraying), more distal



FIG 2. Operative specimen showing a polypoid stenotic mass with oedema of the proximal mucosa

tumours are more commonly found using radiology—contrast studies, computed tomography, and more recently PET. Nonetheless, computed tomography is required in all biopsy-confirmed cases for staging and treatment planning. A diagnostic dilemma sets in when the mucosal biopsy taken during upper endoscopy shows inflammation, as in our case. This can represent a complication of peptic ulcer disease or changes caused by reaction to a tumour in the vicinity, including carcinoma of the duodenum, pancreas, bile duct; or other lesions like gastro-intestinal stromal tumours or lymphomas. Endoscopic ultrasound may help by measuring the thickness of the lesion and allowing deeper biopsy of the lesion. Alternatively, especially when the obstruction does not allow the scope to pass, computed tomography and PET can help. The demonstration of a mass lesion necessitates an operation. An intra-operative frozen section may be required to reach a final diagnosis and guide the surgery.

Surgery remains the only potential cure for this type of tumour. Radical pancreaticoduodenectomy (Whipple's operation) is the classical curative operation and is still the treatment of choice for tumours in the first and second parts of the duodenum. Some authors advocate duodenal segmentectomy as curative treatment for lesions in the third and fourth parts.⁴ Tocchi et al⁴ described duodenal segmentectomy with intestinal derotation, allowing a macroscopically clear margin with adequate lymphadenectomy. The mortality and morbidity rates were lower than those after Whipple's operations. There is growing evidence that there is no difference in survival rates after Whipple's operations and duodenal segmentectomies. There are also sporadic case reports describing successful endoscopic resections of early tumours that have not invaded the submucosa (Tis and T1).⁹

In cases where the tumour has advanced beyond the possibility of curative resection, bypass surgery and stenting have a palliative role. Radiotherapy is not applicable as the tumour is radio-resistant and the small bowel has poor tolerance to radiation. Chemotherapy has no role in primary treatment and there is only limited information about its use as an adjuvant treatment. It is mainly reserved for recurrent disease.

The 5-year survival rate for curatively resected adenocarcinomas of the duodenum is of the order of 50 to 60%. This is better than those for other lesions in the vicinity, namely tumours of the ampulla, distal bile duct, and head of pancreas.¹⁰ Nodal involvement and the possibility of curative resection are independent prognostic factors for adenocarcinomas of the duodenum. In one

prospective study, all the patients with N0 disease given curative surgery survived at 5 years, while only 47% of those with N1 disease were still alive at 5 years.¹⁰ Node-positive patients also have a statistically higher recurrence rate than their node-negative counterparts.¹¹ None of the patients with unresectable disease survived for 5 years.

Conclusion

Adenocarcinoma of the duodenum remains a rare disease, though the prevalence appears to be rising, possibly due to improved diagnostic techniques. Surgery is the only means of cure. More evidence is needed to help define the role of segmental duodenectomy and chemotherapy in the management of this disease.

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