Adenocarcinoma arising in villous adenoma of the ampulla of Vater with synchronous malignant gastrointestinal stromal tumour of the duodenum: a case report

An association between ampullary adenoma and adenocarcinoma has been reported previously. However, we believe that this is the first report of the synchronous occurrence of adenocarcinoma of the ampulla of Vater and a gastrointestinal stromal tumour (GIST).

A 41 year old woman was admitted to our hospital for the evaluation of jaundice. Her liver function tests were as follows: alanine aminotransferase, 37 U/litre (normal range, 0–31); aspartate aminotransferase, 32 U/litre (normal range, 0–32); alkaline phosphatase, 598 U/litre (normal range, 0–240); total bilirubin, 203.2 mg/litre (normal range, 1–11); direct bilirubin, 151.6 mg/litre (normal range, 0–5).

Ultrasonography, computed tomography, and magnetic resonance imaging studies showed intrahepatic and extrapapillary bile duct dilatation. Termination of the common bile duct at the distal end by a mass was noted (fig 1). The initial radiological differential diagnosis included pancreatic head tumour and peripancreatic carcinoma.

Endoscopic procedures were not performed before surgery. Pancreatodeuodenectomy, cholecystectomy, and distal gastrectomy with lymph node dissection were performed.

Grossly, there was a polypoid tumour (1.3 × 1.5 × 1 cm) at the duodenal ampulla with a solid subserosal tumour (3 × 3 × 2 cm) in the second portion of the duodenum (fig 2). On cut section, there was no transition between these two different tumours and no invasion of surrounding tissues and pancreas.

Histologically, the tumours showed a moderately differentiated adenocarcinoma associated with a villous adenoma, which was limited to the ampulla of Vater and a GIST (figs 3 and 4). The GIST was sharply demarcated from the surrounding tissue and it was mainly located at the serosa and muscular layers. Cytologically, the tumour cells had spindle shaped, blunt ended or oval nuclei, with evenly distributed chromatin and moderate pleomorphism; the cells exhibited a fascicular or storiform growth pattern, and had invaded the submucosal layer. Nine atypical mitotic figures for each 10 high power fields (HPF) were present. Immunohistochemically, the tumour cells of the GIST showed diffuse and strong positive immunoreactivity against CD117 (T95; 1/20 dilution; Novocastra, Newcastle, UK), CD34 (QBEND/10; 1/50 dilution, Dako, Glostrup, Denmark).

Grossly, there was a polypoid tumour (1.5 × 1 cm) at the duodenal ampulla (black arrows) and another solid subserosal tumour is seen in the second portion of the duodenum (white arrows).

Histologically, the tumours showed a moderately differentiated adenocarcinoma; haematoxylin and eosin stain; original magnification, ×100.

The possible cause of multiple malignancies include: reduced immunological competence, constitution, genetic factors, chemotherapy, radiation exposure, surgery, or smoking. In our patient, a family history of malignancy and other risk factors were not present. It can also be hypothesised that the duodenum was influenced by the same unknown carcinogen, resulting in a simultaneous proliferation of different cell lines (epithelial and stromal cells).

The literature includes case reports of gastric collision tumour composed of GIST intermixed with adenocarcinoma, synchronously occurring GIST and carcinoid tumour, GIST and lipoma, and GIST and mucosa associated lymphoid tissue lymphoma. To our knowledge, our case is the first
Intra-abdominal fibromatosis of the jejunum and mesentery

A 24 year old woman presented with a painless abdominal lump of six months’ duration. She had no history of colonic polyps. A mobile, non-tender, globular mass was felt in the umbilical region. A computed tomography scan showed a homogenous, non-enhancing mass, possibly arising in the small bowel mesentery.

The tumour was resected entirely with a 6 cm and was intimately related to the bowel wall. The cut surface was tan, whorled, and firm, without necrosis, cystic change, or haemorrhage. Microscopy showed loosely and haphazardly arranged spindle cells with bland, oval nuclei and minimal mitoses. The tumour infiltrated the muscularis propria and had infiltrating margins. The tumour cells were negative with antibodies to CD117, S100, CD34, and smooth muscle actin (Dako, St Louis, Missouri, USA; dilutions of 1/200, 1/50, and prediluted, respectively) and positive for desmin (Labvision, Freemont, California, USA; 1/200 dilution).

The patient has no evidence of disease 16 months after surgery.

The differential diagnosis of a bland spindle cell tumour involving the gastrointestinal tract and mesentery includes gastrointestinal stromal tumour (GIST), fibromatosis, and inflammatory myofibroblastic tumour. Inflammatory myofibroblastic tumours are more common in children and are characterised by a dense inflammatory cell component among a myofibroblastic proliferation. GIST was considered an unlikely diagnosis because of the whorled appearance on cut surface; histological evaluation confirmed this. We made a diagnosis of fibromatosis of the jejunum and mesentery.

Intra-abdominal fibromatosis, in its classic presentation, as a mesenteric mass, does not pose a diagnostic problem because of its distinctive gross and microscopic features. However, when it presents primarily as an intestinal wall tumour, the diagnosis of GIST may seriously be considered. Importantly, as many as five of 13 cases of bowel wall fibromatosis in one series had been labelled initially as low grade sarcomas, whereas 13 of 25 in another study mimicked GIST.

Distinguishing between the two entities is important because of the different treatment protocols and biological behaviour. GISTs are malignant neoplasms that may be treated by imatinib mesylate if they are unresectable or if there is a distant metastasis. They are soft, lobulated, and fleshy on cut surface. Fibroblastic cells, skinoid fibres, mitoses, and necrosis are common. Fibromatosis is a low grade neoplasm that may recur but never metastasises. Recurrence is often related to incomplete excision. Tamoxifen has been used in the management of a recurrent or unresectable tumour. GISTs usually express CD117 and often CD34 also, whereas fibromatosis is always negative for CD34 and may or may not express CD117.

However, fibromatosis is essentially a haematolymphoid and eosinosis diagnosis. The gross appearance—that is, a fibrous mass without necrosis or haemorrhage—gives a clue to the diagnosis.

References
Zoonoses: infectious diseases transmissible from animals to humans


We are constantly being bombarded with advice on how to ensure that we lead a healthy life—eat less, drink less, and take more exercise. Reading this book adds to this litany in that we are exhorted not to crack chestnuts with our teeth, to avoid eating ants and, of course, not to apply raw chopped chestnuts with our teeth, to avoid eating ants and transport media for immunofluorescence biopsies. The authors intended to provide a book based on their current practice protocols “to educate and better equip all those involved in the histopathology specimen process”, and this they have achieved.

K Kerr

Histopathology Specimens: Clinical, Pathological and Laboratory Aspects


When reviewing a book of this nature it is difficult to be critical because there are several different methods of specimen handling and processing. The method used is often dependent on personal preference and accepted protocols in individual laboratories. As the authors state in the preface, there is no one correct method; however, irrespective of the method used, maximum information must be obtained from the macroscopic and microscopic examination.

In an era where great importance is placed on the information obtained from macroscopic examination of the specimen and optimal processing of tissue for histological examination, a book to guide pathologists is welcome.

This book covers specimens from 11 anatomical regions, each including numerous specific sites and one miscellaneous section. Each section covers anatomy (including lymphovascular supply, where applicable), clinical presentation, clinical investigations, pathological conditions (both non-neoplastic and neoplastic), clinical aspects of surgical pathology specimens, and laboratory aspects of surgical pathology specimens. The sections on laboratory aspects of surgical pathology specimens provide extensive coverage of specimen types, points to consider in the description of specimens, appropriate selection of blocks, and the essentials of an adequate histopathology report. The last chapter on miscellaneous specimens and ancillary techniques discusses needle core biopsies, fine needle aspirations, cytospin and liquid based cytology, specimen photography, specimen radiography, frozen sections, immunohistochemistry, flow cytometry, in situ hybridisation, electron microscopy, molecular genetics, and proteomics.

Although there are references to fixatives in specific sections, future editions would benefit by the inclusion of sections on fixatives and optimal fixation, decalcification of specimens, lymph node identifying fluids, and transport media for immunofluorescence biopsies.