

*Case reports***Carcinoid tumour of the ampulla of Vater presenting as acute pancreatitis**

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**SUMMARY** The first report of a carcinoid tumour of the ampulla of Vater causing acute pancreatitis is presented. The pancreatitis resulted from ampullary obstruction and diversion of the bile through the pancreas. Endoscopic retrograde cholangiopancreatography established the correct diagnosis, making this the first occasion in which a carcinoid tumour of the ampulla of Vater has been correctly diagnosed before surgery.

Carcinoid tumours of the ampulla of Vater are rare.<sup>1,2</sup> They usually present with obstructive jaundice and are difficult to diagnose preoperatively.<sup>2</sup> We present a patient who developed pancreatitis as a result of bile duct obstruction from an ampullary carcinoid surgery.

**Case report**

A 48 year old man was admitted as an emergency with a six hour history of severe epigastric pain and frequent vomiting. Before this episode he had been well. He was a non-smoker and drank a glass of wine per day. On examination he had a tachycardia of 116 and was acutely tender in the upper abdomen. Laboratory investigation revealed a serum amylase of 3700 IU/l indicating acute pancreatitis. The pancreatitis was classified as severe<sup>3</sup> (WBC  $19900 \times 10^9/l$ , Ca 1.86 mmol/l, glucose 10.4 mmol/l,  $pO_2$  6.4 kPa). The bilirubin, AST and alkaline phosphatase were normal. Treatment included analgesia, intravenous fluids, nasogastric aspiration and oxygen. He remained hypoxic throughout the first week and developed bilateral pleural effusions. Repeated ultrasound examinations showed an enlarged heterogenous pancreas and no evidence of gall stones. His alcohol consumption was not thought sufficient to account for the pancreatitis and there-

fore he had a ERCP carried out when he had recovered from this attack, which was four weeks after the original presentation. At ERCP it was impossible to cannulate the ampulla of Vater which was enlarged, reddened and friable. It was, however, possible to cannulate the accessory papilla. When contrast was injected through the accessory duct of Santorini, it was seen to communicate with the main pancreatic duct of Wirsung and the common bile duct (Figure). Radiographs showed no evidence of stones or of bile duct dilatation and confirmed that there was total occlusion of the ampulla of Vater. Because of the abnormal appearance of the ampulla it was biopsied. A diagnosis of carcinoid tumour of the ampulla of Vater was made on histological examination of these biopsies. The tumour was argentaffin negative but was argyrophyl (Grimelius) positive.

A computed tomography scan was then carried out to assess the extent of the tumour. It suggested that there was local nodal involvement but no evidence of further spread. He was therefore submitted to laparotomy. At operation the whole pancreas was enlarged and irregular as a result of the previous pancreatitis. The liver, gall bladder, and bile ducts were normal. There was a 3 cm  $\times$  5 cm mass behind the second part of the duodenum. This was excised and frozen section examination showed a lymph node replaced by metastatic carcinoid tumour. There was no other evidence of spread. A 5 mm primary carcinoid tumour of the ampulla was identified through a longitudinal duodenotomy. Bile could be

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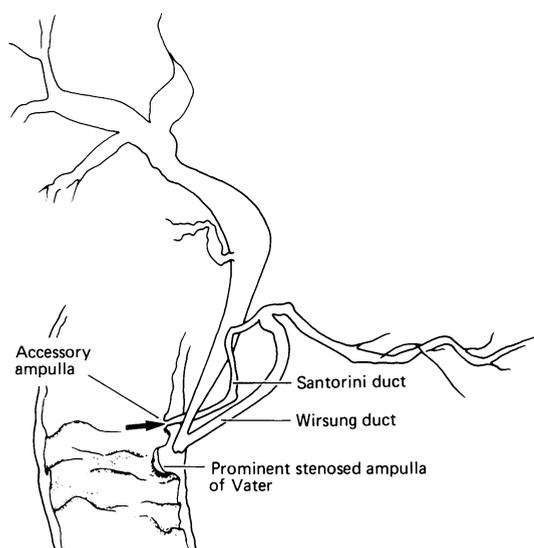


Figure Anatomical arrangement of pancreatic and bile ducts as shown by radiographs obtained by cannulation of the accessory ampulla at ERCP.

seen entering the bowel through the accessory ampulla. The tumour was locally excised and the common bile duct and pancreatic duct reimplanted in the duodenum. Subsequent histological, histochemical, and electron microscopic examination of the excision specimen confirmed the presence of a carcinoid tumour of the ampulla of Vater.

Since operation he has remained well with no further attacks of pancreatitis. A repeat ERCP two months after surgery showed the common bile duct and main pancreatic ducts were patent with no evidence of recurrent tumour.

## Discussion

Biliary tract carcinoids represent 0.2–2% of all carcinoids found in the gastrointestinal tract.<sup>2</sup> They can involve the gall bladder,<sup>4,6</sup> common bile duct<sup>1,2,8</sup> and ampulla of Vater.<sup>2,9–13</sup> Tumours involving the ampulla are reported to present with painless progressive jaundice.<sup>2,9–13</sup> The present report is the first of a patient presenting with acute pancreatitis. Obstructing lesions of the biliary tree are of course well documented as causing pancreatitis. Our patient did not become jaundiced as bile gained entry to the duodenum *via* the accessory duct after passing through the pancreas. This anatomical arrangement clearly predisposed to further attacks of acute pancreatitis.

The present case is the first in which a preoperative diagnosis of carcinoid of the ampulla has been

established. In most previous reports, the presumptive diagnosis has been carcinoma of the head of the pancreas. This is unfortunate as it tends to lead to more radical surgery with an associated increase in operative mortality.<sup>13</sup> Endoscopic retrograde cholangiopancreatography allows one to visualise and biopsy the tumour and to plan further investigation and treatment. The recommended treatment for ampullary carcinoids is wide local excision with reimplantation of the biliary and pancreatic ducts,<sup>9</sup> as done in our patient. Because of the slow progression of the disease, local resection should be carried out even when metastases are present.<sup>9,10,13</sup> Where regional metastases only are present these should also be excised.<sup>9</sup> The prognosis of these tumours is difficult to establish. They appear, however, to be slow growing and prolonged survival has been reported without treatment.<sup>7</sup>

Most biliary carcinoids are argentaffin negative and in keeping with this no carcinoid syndrome has been reported in association with these tumours.<sup>2</sup> By definition all are Grimelius positive and this establishes them as true carcinoids.<sup>2</sup>

## References

- Vitoux J, Salmon RJ, Languille D, Buffeit C, Martin E, Chaput JC. Carcinoid tumour of the common bile duct. *Am J Gastroenterol* 1981; **76**: 360–2.
- Kuiper DH, Gracie WA, Pollard HM. Twenty years of gastrointestinal carcinoids. *Cancer* 1970; **25**: 1424–30.
- Ranson JHC, Rifkind KM, Roses DF, Fink SD, Eng K, Spencer FC. Prognostic signs and the role of operative management of acute pancreatitis. *Surg Gynecol Obstet* 1974; **139**: 69–81.
- Shiffman MA, Juler G. Carcinoid of the biliary tract. *Arch Surg* 1964; **89**: 1113–5.
- Bergdahl L. Carcinoid tumours of the biliary tract. *Aust N Z J Surg* 1976; **46**: 136–8.
- Gaffney PR, Coyle LJ. Carcinoid tumour of the gallbladder associated with a meningioma. *Ir J Med Sci* 1978; **147**: 318–21.
- Little JM, Gibson AAM. Primary common bile duct carcinoid. *Br J Surg* 1968; **35**: 147–9.
- Gerlock AJ, Muhletaler CA. Primary common bile duct carcinoma. *Gastrointest Radiol* 1979; **4**: 263–4.
- McRae J, Conn HJ. Carcinoid of ampulla of Vater. *Surgery* 1959; **46**: 902–7.
- Steffani M, Urbas JE, Crockett FL. Argentaffine carcinoma (carcinoid tumour) involving the ampulla of Vater. *Ill Med J* 1970; **138**: 130–2.
- Weichert RF, Ruth LM, Krement ET, *et al.* Carcinoid islet cell tumours of the duodenum. Report of 21 cases. *Am J Surg* 1971; **121**: 195–205.
- Brown HJ. Malignant carcinoid of the ampulla of Vater. *J Okla State Med Assoc* 1960; **63**: 292–4.
- Lumsden AB, Williams ARW, Dixon JM. Carcinoid tumour of the ampulla of Vater. *Eur J Surg Oncol* 1985; **11**: 55–6.