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An unusual ampullary adenocarcinoma with Paneth cell and mucinous differentiation

Sir,

Carcinoma of ampulla of Vater is a relatively uncommon tumour, accounting for 6–20% of all periampullary tumours and represents 10–50% of cancers resected via pancreaticoduodenectomy.[1] Ampullary carcinoma comprises of two main histological subtypes, the pancreatobiliary type and the intestinal type which have different pathogenetic and clinical characteristics.[2] Determining the two subtypes is also important as prognosis and treatment protocols differ for these two subtypes. The intestinal subtype is correlated with a lower incidence of lymph nodal metastases, little or no invasion of the surrounding pancreatic parenchyma and a longer long-term survival after resection.[3] Paneth cells are found in the mucosa of the entire small intestine, proximal large intestine and appendix. Their neoplastic transformation, described as single case reports at sites including small intestine,[4] ampulla of Vater,[5,6] large intestine,[7] Meckel's diverticulum[8] and stomach,[9] ranges from partial Paneth cell differentiation to Paneth cell-rich neoplasia. We describe a rare case of ampullary adenocarcinoma with dual differentiation comprising of Paneth cells and mucinous patterns in equal proportions. Paneth cells demonstrated features of malignancy and were clearly neoplastic. Additionally, IgG4 positive plasma cells characterised most of the tumour stroma. The current lack of data regarding this entity makes it difficult to draw prognostic conclusions and also the significance of IgG4 positive plasma cells within the tumour stroma is unclear. This dual differentiation pattern within ampullary carcinoma is exceedingly rare and is the first case of such a tumour in the literature, to the best of our knowledge.

A 58-year-old reformed alcoholic, a known diabetic and hypertensive on regular treatment for 5 years, presented with sudden onset epigastric pain radiating to the back of one day duration. He had a similar episode two and a half years previously which was diagnosed clinically as acute pancreatitis and treated conservatively. As his condition improved with empirical treatment, no radiological investigations or endoscopy were performed. He remained asymptomatic until the

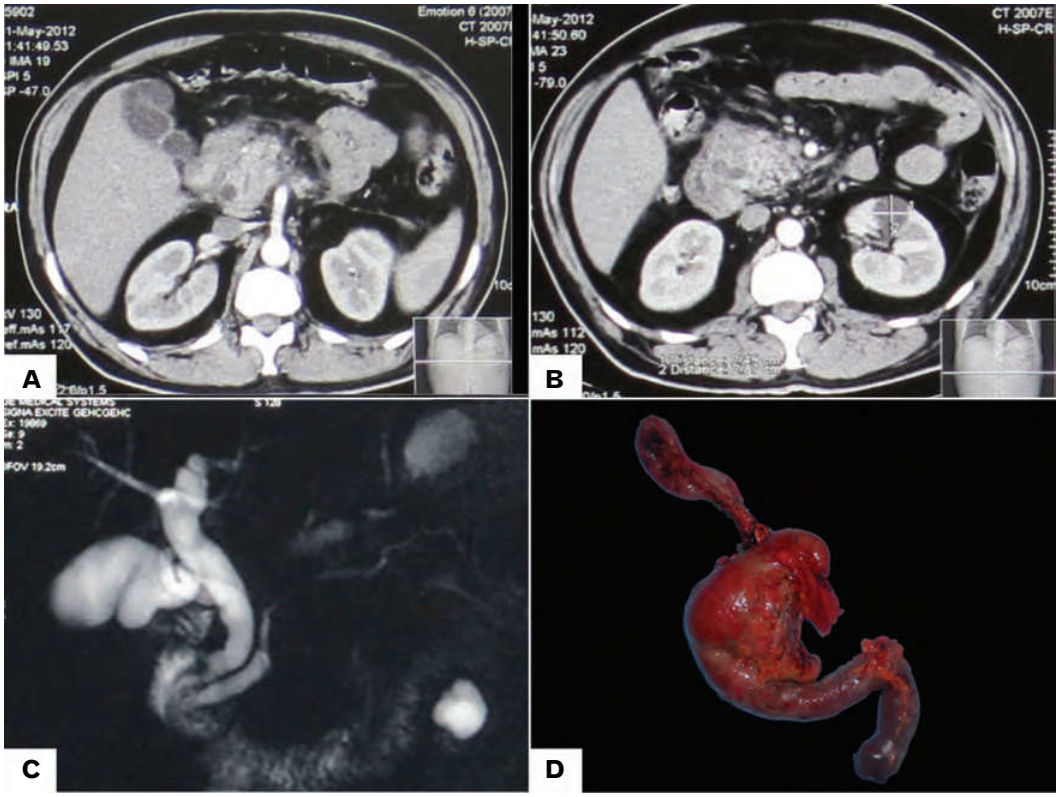

Fig. 1 Axial CECT sections through the upper abdomen demonstrate (A) dilated common bile duct and main pancreatic duct due to (B) a heterogeneously hypodense mass in the head and uncinate process of pancreas with loss of fat planes in the second part of the duodenum. (C) Coronal MRCP image demonstrating dilated common bile duct and main pancreatic duct with abrupt cut-off in the periampullary region. (D) Resected partial Whipple's specimen.