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CASE REPORT

Acinar cell carcinoma with fatty change arising from the pancreas

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ABSTRACT. Acinar cell carcinoma of the pancreas is a rare malignant tumour developing from acinar cells, accounting for approximately 1% of pancreatic exocrine tumours. We experienced a case of an acinar cell carcinoma with fatty change. To the best of our knowledge, this is the first case report of an acinar cell carcinoma with fatty change in the clinical literature.

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Acinar cell carcinoma (ACC) of the pancreas is a malignant tumour developing from acinar cells. It is a rare tumour accounting for approximately 1% of pancreatic exocrine tumours. ACC with fatty change is extremely rare in the literature. This is a report on a 72-year-old female who was found to have ACC with fatty change.

Case report

A 72-year-old female underwent a partial gastrectomy with Billroth I for early gastric cancer in September 2009. When a pancreatic mass was found during the 6-month follow-up CT, the patient was admitted for further investigation.

Test results showed elevated levels of alpha-fetoprotein (AFP): 1945 IU ml^{-1} (normal range: 0–7 IU ml^{-1}) and normal levels of cancer antigen 19-9: 8.5 U ml^{-1} (normal range: 0–37 U ml^{-1}) and carcinoembryonic antigen (CEA): 2.31 ng ml^{-1} (normal range: 0–5 ng ml^{-1}).

An abdominal ultrasound showed a 2.5-cm hypoechoic mass in the neck of the pancreas. The mass was ill-delimitated and heterogeneous. Contrast-enhanced CT showed a 2.5-cm mass in the neck of the pancreas with distal pancreatic duct dilatation and parenchymal atrophy. The mass was adjacent to the portal vein and showed less intense enhancement than the pancreas. No dilatation of the bile ducts, focal hepatic lesions or abdominal lymphadenopathies was found. The initial differential diagnoses were pancreatic cancer and metastasis.

Positron emission tomography (PET) showed no significant increase in fluorodeoxyglucose (FDG) uptake. MRI showed a 2.5-cm hypovascular well-defined mass at the neck of the pancreas with distal pancreatic duct dilatation and parenchymal atrophy. This mass had low signal intensity at T_2 weighted imaging with fat suppression, and had a fat component at in-phase and out-of-phase images (Figure 1). The portal vein was compressed and displaced by the mass, which remained permeable without invasion.

A Whipple procedure was performed on the patient. The specimen revealed an ill-defined lobulating mass of 1.9×1.7 cm, which was confined to the pancreas. Histologically, the mass showed an acinar pattern of cells with nucleoli (Figure 2). The tumour cells showed extensive and various degrees of clear cell change and microvesicular and macrovesicular fatty change. The tumour cells showed strong and diffuse immunoreactivity for AFP and were negative for CD56, chromogranin A and synaptophysin. Pathological diagnosis was ACC with fatty change.

Discussion

ACC is an uncommon solid epithelial exocrine tumour. There are very few reports on this type of tumour. ACC typically occurs during the fifth to seventh decades of life, with a male predilection [1]. It can occur in any part of the pancreas, but the most common site is the head of the pancreas. A diagnosis of ACC can be made on the characteristic positive periodic acid–Schiff staining and immunohistochemical results.

The most common histological patterns seen in ACC are acinar and solid [2]. In a study by Klimstra et al [3], ACCs were positive for at least one immunohistochemical marker (*i.e.* trypsin, lipase, amylase, chymotrypsin, α1-antitrypsin, keratin, epithelial membrane antigen, carcinoembryonic antigen and AFP). ACCs, unlike endocrine cell tumours, are believed to originate from transformed acinar cells [2]. ACC with fatty change showed no evidence of an endocrine differentiation and diffuse immunoreactivity for AFP.

The imaging diagnosis of ACC varies owing to the rarity of the disease: a well-marginated, large, solid mass with a varied degree of cystic components; thin enhancing

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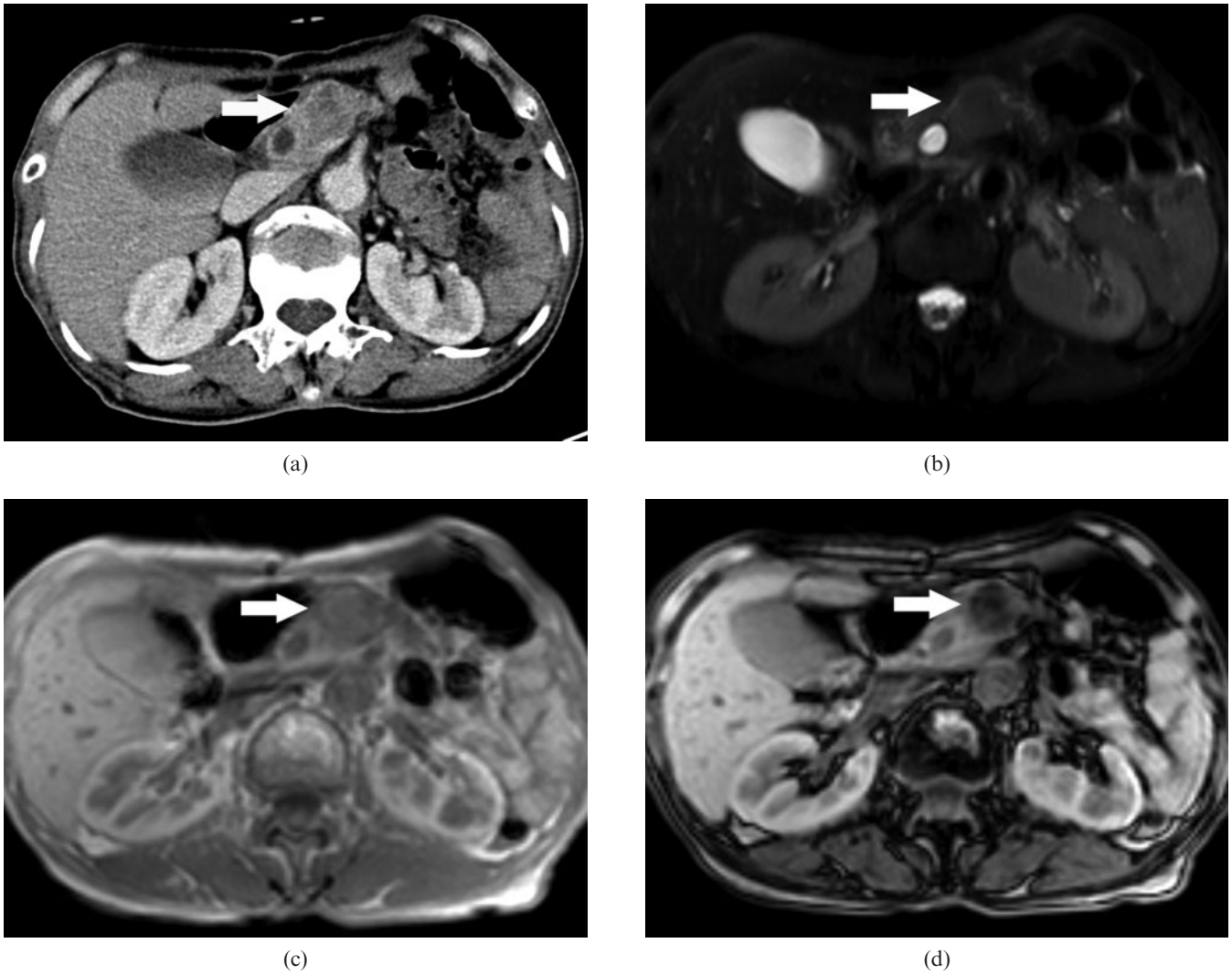

Figure 1. A 72-year-old female. (a) Contrast-enhanced CT showing a mass in the neck of the pancreas, which had less intense enhancement than the pancreas. (b) Axial T_2 weighted MRI with fat suppression showed low signal intensity in this mass. (c) In-phase and (d) out-of-phase MRI showing a fat component in this mass.

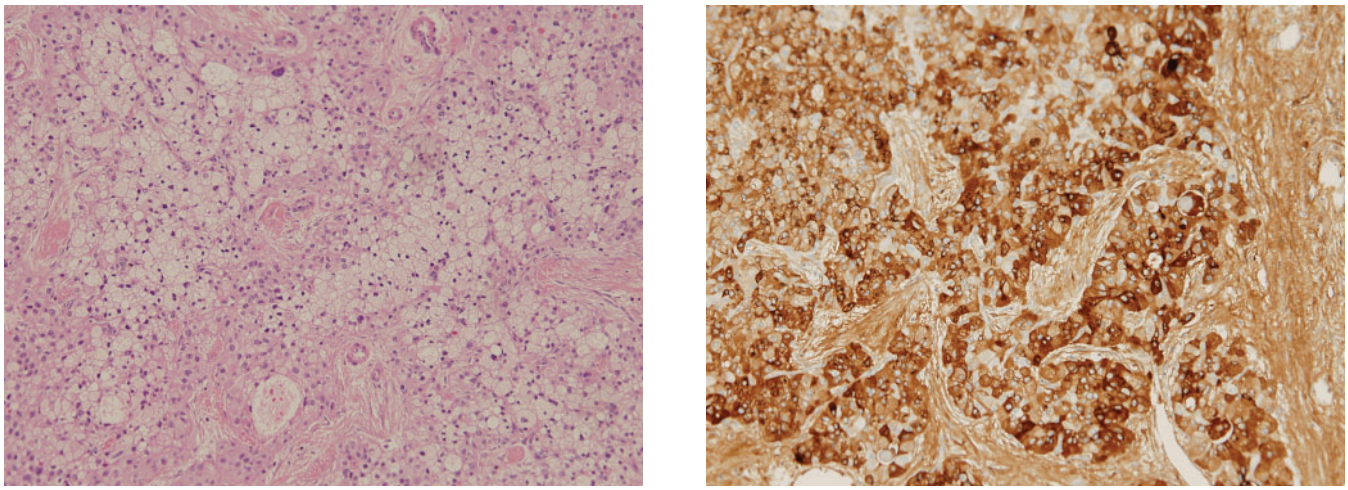

Figure 2. Microscopic findings. (a) Pancreatic mass showing an acinar pattern of cells with nucleoli. The tumour cells with extensive and various degrees of clear cell change and fatty change (haematoxylin and eosin stain) ($\times$ 100). (b) The tumour cells showed strong and diffuse immunoreactivity for alpha-fetoprotein ($\times$ 200).

capsule; occasional central calcification; intralesional haemorrhage and less intense enhancement than a normal pancreas on both CT and MRI [4, 5]. ACC is unique among pancreatic tumours as it is characterised by increased AFP levels. Our case also showed increased AFP levels and less intense enhancement than the pancreas on both CT and MRI.

Although ACC shows increased blood AFP levels and positive AFP stain, AFP can also be expressed by hepatoid carcinomas. The pathogenesis of hepatoid carcinomas of the pancreas is not fully understood. Paner et al [6] thought that the potentiality of hepatic differentiation may arise from any of the three main pancreatic cells (*i.e.* acinar, ductal and islet cells). AFP can also be expressed by pancreatic ductal carcinoma, acinar cell carcinoma, islet cell tumour and poorly differentiated pancreatic adenocarcinoma [7]. Therefore, the diagnosis of hepatoid carcinoma is mainly provided by the cancer's histological appearances on haematoxylin and eosin-stained material [8].

In our case, fat-suppressed MRI confirmed that the pancreatic mass contained a fat component. The pancreas is a very rare location for fat-containing tumours [9]. Differential diagnosis of fat-containing pancreatic tumours with direct invasion of pancreatic tissue should be mentioned (*i.e.* malignant fibrous histiocytoma, leiomyosarcoma, desmoids tumour, cystic teratoma, fibrolipoma, liposarcoma and lipoblastoma) [10–12]. However, as in our case, fat-containing tumours originating from the pancreas may be ACC with fatty change.

To our knowledge, this is the first reported case of ACC with fatty change. MRI is very important in detecting the fat component of tumours. ACC with fatty change must be considered in differential diagnosis of a fat-containing pancreatic tumour with elevated levels of AFP.

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