

# Acinar Cell Carcinoma of Pancreas: a Case Report and Review of Literature

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## Introduction

Acinar cells are the predominant cells of the pancreas but malignant transformation is rare with acinar cell carcinoma (ACC) accounting for 1–2% of all pancreatic neoplasms in adults and about 15% in the paediatric population [1]. Although grouped under exocrine neoplasms of the pancreas, ACCs have distinct clinical, morphological and pathological characteristics [2]. We report a case of ACC of pancreas in a 57-year-old gentleman who presented with a lesion in the tail of pancreas with liver metastasis. The management and brief review of literature is presented.

## Case Report

A 57-year-old gentleman presented with epigastric pain of 1 month duration. Physical examination did not reveal any significant findings. Serum amylase and lipase were within normal limits. CT scan of the abdomen and pelvis showed a large irregular heterogeneous mass measuring 10.2 × 8.2 cm, with early enhancement and rapid wash out arising from the tail of the pancreas, encasing the splenic vessels and extending up to the splenic hilum. There were two closely placed hypodense lesions in segment 4b of the liver, the larger one measuring 2.0 × 1.8 cm

(Fig. 1). A CT guided biopsy of the liver lesion was done which was suspicious of low-grade neuroendocrine carcinoma. With a presumptive diagnosis of a low-grade neuroendocrine tumour of the pancreas with liver metastasis, a plan for distal pancreaticosplenectomy and liver metastatectomy was made. Intra-operatively, there was a large mass of size 16 × 12 cm arising from the body and tail of pancreas, encasing the splenic vessels and splenic hilum. There was a 2 × 1 cm grey white lesion in the segment 4B of the liver. A distal pancreatectomy with splenectomy was done. Frozen section from the tumour was suggestive of a poorly differentiated carcinoma. Hence, liver metastasectomy was deferred. He had an uneventful recovery. Histopathological examination with immunohistochemical correlation revealed the tumour to be an acinar cell carcinoma of the pancreas (Figs. 2, 3). On immunohistochemistry, the tumour cells were positive for cytokeratin, beta catenin, chromogranin, synaptophysin, CD 56, CD 57 and progesterone receptors.

He subsequently underwent CT guided radiofrequency ablation (RFA) of the liver lesion. The role of adjuvant chemotherapy was discussed in the multidisciplinary tumour board. Since strong evidence of the benefit of adjuvant chemotherapy was lacking, it was decided to keep the patient on follow-up. The patient is alive and disease free 8 months after the surgery.

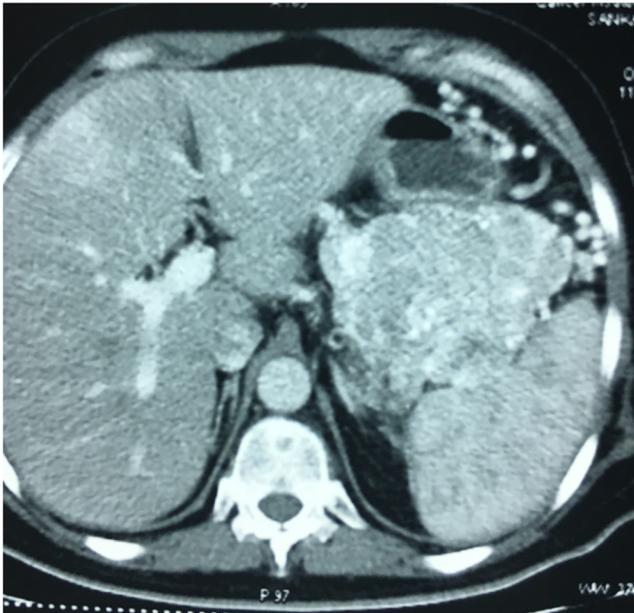
## Discussion

Acinar cell carcinoma accounts for 1% of all exocrine pancreatic tumours, although acinar cells are predominately found in the pancreas. Its benign counterpart, acinar cystadenoma is extremely uncommon [3]. In our institution, although on an average we see around 50 pancreatic/periampullary adenocarcinomas and around 5 pancreatic neuroendocrine tumours every year, this is the first patient diagnosed to have an ACC of the pancreas. ACC has a male predominance (3:1) and more

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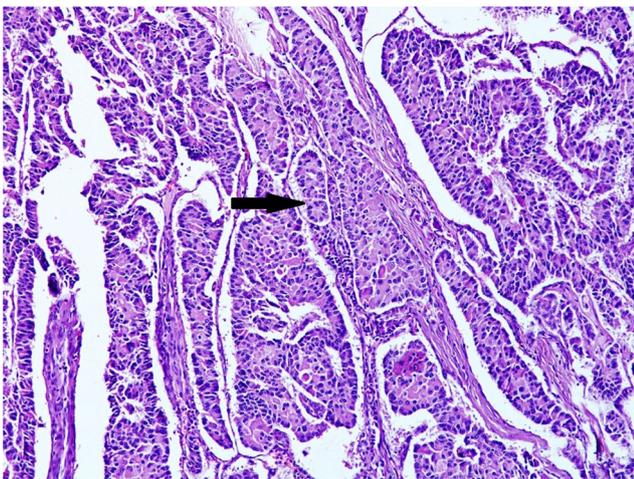
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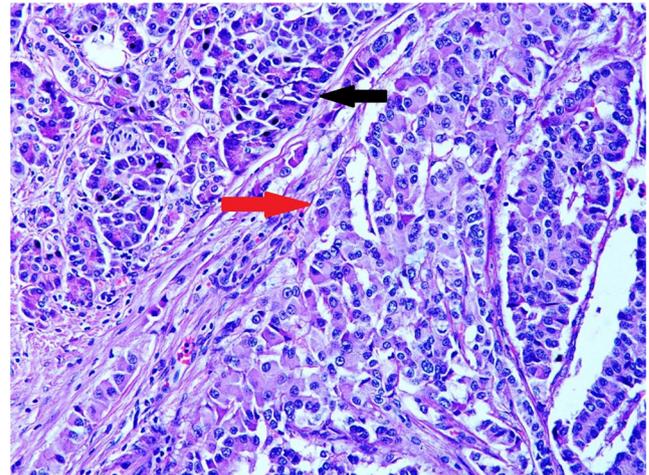
**Fig. 1** Preoperative CT scan showing necrotic tumour in the tail of pancreas and metastasis in the liver

commonly arises in the head of the pancreas [4]. They usually present as large tumours, with an average size of 10 cm at the time of diagnosis [5]. Rarely, ACC can occur outside the pancreas as a result of pancreatic acinar heterotopia/metaplasia [6]. ACC behaves like a high-grade malignancy, with up to 50% of patients presenting with metastatic disease. An additional 25% with initial organ-confined disease develop recurrence at regional or distant sites [7]. But when compared to pancreatic ductal adenocarcinoma, ACC is more often limited to the pancreas at the time of diagnosis.

Grossly, these tumours are usually solid with areas of haemorrhage and necrosis. Microscopically, acinar structures are the hallmark of this neoplasm but poorly differentiated ACC can



**Fig. 2** Photomicrograph showing tumour cells (*bold arrow*) arranged in acinar pattern, H&E 10×



**Fig. 3** Photomicrograph showing tumour cells (*red arrow*) merging with normal pancreatic acini (*black arrow*), H&E 20×

form sheets of cells without well-defined acinar structures [8]. The neoplastic cells stain positive on immunohistochemistry for cytokeratin, trypsin, lipase and chymotrypsin [9]. In some cases, immunohistochemistry for chromogranin or synaptophysin will reveal a minor neuroendocrine component, as was seen in our patient. When it constitutes more than 30% of the neoplastic cells, a diagnosis of a mixed acinar-neuroendocrine carcinoma is made. Rarely, tumours may show multiple lines of differentiation including acinar-endocrine, acinar-ductal and acinar-endocrine-ductal [9, 10].

Complete surgical resection is the best possible curative treatment for localised tumours. The role of adjuvant treatment in ACC is not well defined due to the rare nature of the disease and the small number of patients in the reported series. A systematic review reported response rates varying from 0 to 100% to a variety of single or multidrug chemotherapy [11]. In contrast to pancreatic adenocarcinoma where gemcitabine is the preferred drug, 5-fluorouracil (5-FU) appears to be the most active agent in ACC [11]. One series demonstrated partial response or prolonged disease stabilisation (>12 months) in patients treated with FOLFOX or FOLFIRINOX regimens and suggested that a 5-FU and platinum-based chemotherapy should probably be the standard of care in advanced ACC [12]. Indeed, surgery with adjuvant chemotherapy showed the highest median overall survival of 47 months in a systematic review [11]. Neoadjuvant radiotherapy is reported to produce a response rate of 25–35% in localised ACC when used alone and close to 100% response rates when used in combination with chemotherapy, suggesting that ACC could possibly be a radiosensitive tumour [11]. A few case reports describe the use of neoadjuvant chemotherapy with or without concurrent radiation to downsize borderline inoperable tumours in order to make them resectable [13–15]. However, the data regarding use of adjuvant radiation in ACC with or without chemotherapy is scarce.

The reported 5-year overall survival for non-metastatic ACC ranges from 36 to 72% [7]. The median overall survival ranges from 47 to 78 months after surgical resection and 17–25 months in metastatic disease [11, 12]. When compared to ductal adenocarcinomas, overall prognosis of ACC appears better for all the stages. For metastatic tumours, the 5-year overall survival was 17.2% for ACC versus 2.8% for ductal cell carcinoma [16, 17]. Although metastatic disease has a poor prognosis, one case report described a long-term survival of a patient with metastatic ACC after a multimodal treatment approach which included resection of the primary, RFA, external beam radiotherapy and intra-arterial brachytherapy with SIRS for liver metastasis and multiple lines of chemotherapy [18].

## Conclusion

Pancreatic acinar cell carcinomas are rare tumours that are clinico-pathologically different from pancreatic ductal adenocarcinoma. Their prognosis is better than ductal adenocarcinomas. The role of adjuvant treatment is not defined.

## Compliance with Ethical Standards

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**Conflict of Interest** The authors declare that they have no conflict of interest.

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