

CASE REPORT

Giant Acinar Cell Carcinoma of Pancreas Presenting In a Young Patient

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ABSTRACT

Context Acinar cell carcinoma is rare disease of exocrine pancreas with an indolent course and favorable tumor biology. It usually occurs in elderly male patient. However, its occurrence in young patient with large size is rarer. **Case report** A twenty-eight-year-old male patient presented with abdominal pain and mobile epigastric lump for 6 months and jaundice for 2 week duration. Contrast enhanced computed tomography revealed giant (15×13 cm) heterogeneously enhancing tumor arising exophytically from pancreatic head and compressing splenoportal axis and common hepatic artery, which was successfully managed with artery first pancreaticoduodenectomy. Histopathological examination with immunohistochemistry revealed acinar cell carcinoma of pancreas. **Conclusion** Differential diagnosis of acinar cell carcinoma of pancreas should be considered in young patient with large tumor size. Prognosis is better than adenocarcinoma of pancreas because of its favorable tumor biology. Surgical resection with curative intent along with multimodality regimen is preferred treatment modality.

INTRODUCTION

Acinar cell carcinoma (ACC) of pancreas is a rare entity, comprising only 1% of exocrine pancreatic tumor with an indolent course. It usually presents in elderly male of 50 to 70 year age group as large tumor (5 to 10 cm) at time of diagnosis [1]. Anecdotal reports of tumor in children and young adults have been reported. Here we report an interesting case in a young male with giant (>10 cm) ACC in head of pancreas.

CASE REPORT

A twenty-eight-year-old young gentleman presented with epigastric pain and progressively increasing lump abdomen of 6 months, along with jaundice of 2 weeks duration. The abdominal lump was hard, 18×14 cm, mobile and occupying epigastric and right hypochondrium. The laboratory investigation revealed hemoglobin of 8.5 gm/dl; blood sugar 210 mg/dl; serum albumin 3.2 gm/dl; INR 1.3; total bilirubin 7.0 gm/dl; alkaline phosphatase 233U/L and mildly elevated serum amylase 232 IU/L and serum lipase 136 IU/L. Contrast enhanced CT abdomen showed a well delimited, heterogeneously enhancing mass originating from head of pancreas measuring 15×13 cm, and involving splenoportal (SP) axis and common hepatic artery with proximal biliary and pancreatic duct dilation (**Figure 1**). Tumor markers including CEA, CA19.9 and chromagranin A were within normal limit.

The patient presented late to us with new onset jaundice as he was managed at other centre without any form of intervention in view of locally advanced and unresectable tumor. However, we attempted for surgical intervention as the patient was young, well preserved with mobile tumor and CT features were suggestive of resectable tumor.

Patient subsequently underwent upfront pancreaticoduodenectomy with SMA first approach without preoperative biliary drainage. Intraoperatively, there was a huge mass arising exophytically from head of pancreas pushing surrounding structures. SP axis (2 cm length) was laterally involved which required en bloc tangential excision and primary repair. Cut section of the specimen revealed a fleshy mass composed of soft tan tissue with areas of necrosis (**Figure 2**). Postoperatively, patient was discharged satisfactorily on postoperative day 12. Histopathological examination showed solid growth with uniform atypical cells arranged predominantly in acinar and microgranular pattern with sparse stroma. The cells had abundant granular and eosinophilic cytoplasm with prominent nucleoli. On immunohistochemistry, the tumor cells were positive for cytokeratin and negative for chromagranin, synaptophysin and CEA receptors suggesting acinar cell carcinoma of pancreas (stage pT3N0M0, AJCC 7th ed) (**Figure 3**). Postoperatively patient refused to undergo adjuvant therapy. At 18 months of follow up patient is alive with no recurrence.

DISCUSSION

Pancreatic ductal adenocarcinoma (PAD) is the most common malignant pancreatic neoplasm. ACC in pancreas is very rare, occurring uniformly in elderly population. Giant ACC of 15 cm in a young patient is even rarer. The

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Abbreviations ACC Acinar cell carcinoma; CEA carcinoembryonic antigen

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Figure 1. Contrast enhanced CT abdomen showing well circumscribed tumor of pancreatic head involving splenoportal axis.



Figure 2. Cut section of tumor from pancreatic head showing fleshy and variegated appearance.

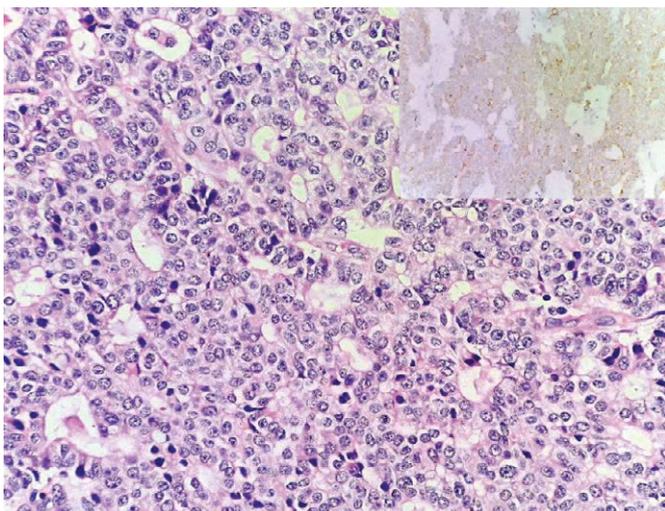


Figure 3. (H&E, x100) Photomicrograph showing tumor cells arranged in acinar pattern with cytokeratin stain positivity (*inset*) on immunohistochemistry.

prognosis seems to be better than PAD at all stages of disease due to its favorable tumor biology and early stages at presentation [1, 2].

ACC usually presents with nonspecific abdominal symptoms such as pain abdomen, abdominal lump, nausea/vomiting and weight loss. Jaundice, although seen in the present case, is less frequent with ACC. The Schmid's triad, a paraneoplastic syndrome characterized by polyarthralgia, subcutaneous fat necrosis, and eosinophilia due to lipase hypersecretion by tumor is very rare and is typical of ACC [3].

Diagnosis is generally made after surgery at receipt of histopathology report. Preoperatively it's difficult to differentiate from solid tumors like neuroendocrine tumor of pancreas and pancreatoblastoma. Moreover, FNAC is not sensitive enough to differentiate among them, as the tumor is not very cellular. Grossly the tumors are usually solid with areas of hemorrhage and necrosis. On microscopy, tumors are arranged in acinar pattern which is the hallmark, and stain positive on immunohistochemistry for cytokeratin, trypsin, chymotrypsin and lipase. Moreover, these tumors do not express chromogranin A or synaptophysin [3, 4].

Upfront surgical resection is the best possible curative treatment for suspected ACC that is considered radiologically resectable [5]. Even though the tumor appears large on CT scan, it is unlikely to involve surrounding major vessels as the tumor is not infiltrative in nature.

The role of multimodality regimen is not well defined because of its rarity. However, recent evidence indicates that partial response to prolonged disease stabilization can be achieved with 5-FU and platinum or GEMOX regimen based chemotherapy and should be the standard of adjuvant therapy [6]. Indeed with this approach, one multi-institutional series of 17 patients has showed 5-year survival of 53%, with median overall survival of 61 months, a prognosis markedly better than that observed with PAD (5-year and median survival of 20% and 18 months respectively) [7].

CONCLUSION

To conclude, ACC is a rare disease of elderly, however it can present in young patient with giant tumor. Prognosis is better than adenocarcinoma of pancreas because of its favorable tumor biology. Surgical resection with curative intent along with multimodality regimen is preferred treatment modality.

Conflict of Interest

The authors declare that they have no conflict of interest.

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