An unusual pancreatic neoplasm: the acinar cell carcinoma

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Section: Abdominal imaging
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Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 76 years, male

Clinical History:

In 2009 a man was admitted to our hospital for faint abdominal pain and weight loss. He underwent distal pancreasectomy and splenectomy for a huge pancreatic neoplasm. After two years he underwent omentectomy for peritoneal metastasis. He is currently undergoing chemotherapy for liver metastases.

Imaging Findings:

The contrast enhanced CT examination performed in 2009 showed a large exophytic mass of 20x12 cm in diameter in the left abdominal cavity originating from the body-tail of the pancreas. The mass had smooth margins and determined a dislocation of left kidney, spleen, small bowel and left colonic flexure, without any infiltration of adjacent structures. The pancreatic mass enhanced predominantly in the peripheral portion, showing some cystic and necrotic areas; splanchnic vessels were patent (Fig.1). No free fluid in the peritoneal cavity or liver metastases was observed. Histological specimen after surgical ablation revealed a pancreatic acinar cell carcinoma.

After two years follow up, a CT showed an 11 cm mass in the left abdominal cavity that showed the same CT features as the previous tumour (Fig. 2) and after omentectomy a peritoneal metastasis was found at histology. Currently the patient has multiple liver metastases (Fig. 3).

Discussion:

Pancreatic acinar cell carcinoma is uncommon and represents only 1% of pancreatic tumours. It occurs predominantly in male gender in the VI-VII decade. The distribution among head, uncinate process, body and tail of the pancreas is almost similar, although in the cephalic region it is slightly more frequent [1, 2]. Microscopically acinar cell carcinomas have two predominant cellular patterns of growth: an acinar pattern or a solid pattern. Cytoplasmatic zymogen granules determine PAS staining positivity [1, 2]. Symptoms are not specific and include faint abdominal pain, nausea, vomiting, weight loss, anaemia, and symptoms due to mass effect. More uncommon symptoms include jaundice and hypoglycaemia. In less than 10% of cases these carcinomas produce serum lipase, sometimes causing the “lipase hypersecretion syndrome”, thus resulting in arthralgia, skin nodules and subcutaneous fat necrosis [1, 3]. Due to symptom vagueness, when discovered, acinar cell carcinomas are usually large in size and this tumour has a better prognosis than pancreatic ductal adenocarcinoma: in case of complete surgical resection, the 5 years survival rate reaches 44% [4]. Some radiological features are typical of acinar cell carcinomas: they are usually large in size with well defined margins, have an exophytic growth with possible intralesional fluid areas due to necrosis and show a contrast enhancement after intravenous administration of contrast
media, although they appear hypodense in comparison with pancreatic parenchyma; there is often a lack of significant pancreatic and biliary duct dilatation, even if the mass is located in the head or uncinate process and acinar cell and this tumour, also when it has huge dimension, rarely determines vascular encasement or narrowing [3]. Differential diagnosis includes other pancreatic neoplasms and mainly the most common ductal adenocarcinoma that is usually smaller when diagnosed, it rarely has cystic areas and peripheral margins are not well defined. Differential diagnosis from other pancreatic masses, like neuroendocrine tumours, solid and pseudopapillary tumour, pancreatoblastoma (that occurs in children and infants) and sarcomas, can be difficult only considering the imaging modality [1]. Although the definitive diagnosis is based on histological specimen, the described characteristics can help radiologists to suspect an acinar cell carcinoma. Distant metastases, less frequent than in ductal adenocarcinoma, may involve the peritoneum and liver.

Take home message: a radiologist can suspect an acinar cell carcinoma in case of huge pancreatic mass with exophytic growth and without pancreatic and biliary dilatation and vascular encasement.

**Differential Diagnosis List:** Pancreatic acinar cell carcinoma, Ductal adenocarcinoma, Neuroendocrine pancreatic tumour, Solid and pseudopapillary tumours, Pancreatoblastoma

**Final Diagnosis:** Pancreatic acinar cell carcinoma

**References:**

Servet Tatli, Koenraad J. Mortele, Angela D. Levy, Jonathan N. Glickman, Pablo R. Ros, Peter A. Banks, Stuart G. Silverman (2005) CT and MRI Features of Pure Acinar Cell Carcinoma of the Pancreas in Adults. AJR 184: 511-519 (PMID: 15671372)


Description: Contrast-enhanced CT axial examination, performed in 2009, showed a huge solid mass originating from the body and tail of the pancreas with splenic vein compressed by the neoplasm but that preserved its patency (arrow). Origin: Sant'Andrea Hospital, Dept. of Radiology, Sapienza University of Rome, Rome, Italy
Description: A more caudal axial contrast-enhanced CT examination, performed in 2009, showed the normal aspect of the head of the pancreas (arrow). Origin: Sant'Andrea Hospital, Dept. of Radiology, Sapienza University of Rome, Rome, Italy
Description: Contrast-enhanced CT axial examination, performed in 2011, showed a peritoneal solid mass in the left flank (arrow) with the same CT features of the resected pancreatic tumour due to a peritoneal metastasis. **Origin:** Sant'Andrea Hospital, Dept. of Radiology, Sapienza University of Rome, Rome, Italy
**Figure 3**

**Description:** Contrast enhanced CT, performed in August 2012, detected some liver metastases (arrows). **Origin:** Sant'Andrea Hospital, Dept. of Radiology, Sapienza University of Rome, Rome, Italy