Imaging Findings of Alpha-Fetoprotein-Producing Acinar Cell Carcinoma of the Pancreas

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Alpha-fetoprotein (AFP)-producing acinar cell carcinoma of the pancreas is a rare neoplasm. We present the imaging findings of two cases of pancreatic AFP-producing acinar cell carcinoma. The CT findings of this tumor revealed a hypodense mass with occasional calcification, a central necrotic area, and a well-defined enhanced wall. On MRI, the tumor showed a slightly low signal on T1-weighted images, a high signal on T2-weighted images, and poor enhancement after gadolinium injection. Percutaneous transhepatic cholangiography and drainage facilitated the assessment of the severity of the common bile duct stricture and helped relieve jaundice in one patient. Angiography provided little help in the diagnosis of this tumor except for showing the encasement of the splenic vessels. Serum AFP level is a useful marker for the diagnosis and for evaluating the response of therapy in such cases.

Key words: Alpha-fetoproteins; Pancreas, neoplasms; Carcinoma, acinar cell; CT; MRI

Alpha-fetoprotein (AFP) is considered to be specific to patients with hepatocellular carcinoma, so it is regarded as a very useful tumor marker. Serum AFP elevation has also been found in patients with non-tumorous liver diseases (e.g., acute or chronic hepatitis, liver cirrhosis), other gastrointestinal neoplasms (e.g., hepatoid adenocarcinoma of the stomach), yolk sac tumor, teratoma, or lung tumors [1-2]. Pancreatic neoplasms with a high serum level of AFP are rare, and most of these are pancreaticoblastomas that occur in children [3]. AFP-producing acinar cell carcinoma of the pancreas is very rare, and only four cases with their radiological findings have been reported in the English literature [4-6]. We therefore report two cases of AFP-producing acinar cell carcinoma of the pancreas with their imaging findings.

Case 1

Elevation of serum AFP was noted in a 70-year-old male during a routine health check-up. After admission, laboratory data of tumor markers showed a high serum level of AFP (2636 ng/ml, normal < 8.5 ng/ml). On computed tomography (CT), a lobulated heterogeneous soft tissue nodule with central necrosis and a well-enhanced peripheral wall was noted in the tail of the pancreas (Fig. 1). Angiography demonstrated tumor encasement of the distal splenic vein without definite hypervascular tumor staining. The patient underwent a distal pancreatectomy and splenectomy. Histopathological and electronic microscopic examinations revealed this tumor to be an AFP-producing acinar cell carcinoma of the pancreas. The postoperative course was smooth.
The serum AFP level rapidly decreased to 170 ng/ml after 2 weeks and returned to normal limits after 3 months.

Case 2

A 69-year-old female was admitted to our hospital on account of epigastralgia and jaundice. After admission, laboratory data of tumor markers showed a high serum level of AFP (8164 ng/ml) and CA-199 (61.62 unit/ml, normal < 34.60 unit/ml). On a CT scan, a large lobulated soft tissue mass with spotty calcification and central necrosis was noted in the pancreatic head, and it had invaded the hepatoduodenal ligament, caudate lobe of the liver, the second and third portions of the duodenum, and the hepatic flexure of the colon (Fig. 2). Dilatation of the bilateral intrahepatic bile ducts (IHDs) and the common hepatic duct (CHD) was also noted. On magnetic resonance imaging (MRI), the mass lesion showed mild low-signal intensity on T1-weighted images and slightly high-signal intensity on T2-weighted images. After contrast medium administration, the lesion showed heterogeneous enhancement with a poorly enhanced central necrotic area (Fig. 3). Multiple lymph nodes were also found in the para-aortic retroperitoneum. Percutaneous transhepatic cholangiography and drainage (PTCD) showed severe luminal narrowing of the common bile duct (CBD) with dilatation of the bilateral IHDs and CHD (Fig. 4).
A biopsy was performed, and the histopathological and electronic microscopic examinations showed this tumor to be an AFP-producing acinar cell carcinoma of the pancreas. She received combined radiotherapy and chemotherapy. After finishing the seven courses of chemotherapy, the tumor had decreased in size (Fig. 5), the jaundice had disappeared, and the patient’s serum AFP level had dropped to 30.49 ng/ml. Further chemotherapy was not performed due to severe gastrointestinal bleeding in this patient.

**DISCUSSION**

The three main components of the pancreas are ducts (4%), acinar cells (82%), and islet cells (14%). Although acinar cells occupy the majority of the normal pancreas, acinar cell carcinoma accounts for approximately 1% of all pancreatic neoplasms [7]. It occurs more commonly in men, with peak incidence in the seventh decade. The prognosis of acinar cell carcinoma is better than that of ductal carcinoma [8]. AFP-producing acinar cell carcinomas of the pancreas are very rare, comprising only 4.5% to 6% of pancreatic acinar cell carcinomas. A diagnosis of AFP-producing acinar cell carcinoma of the pancreas can be established on the basis of immunohistochemical and electromicroscopical results [9].

AFP is an oncofetal protein that is produced in the liver and yolk sac and in the fetal gastrointestinal tract. It is thus likely that tumors originating from these tissues may show AFP production. AFP is widely used as a tumor marker for malignant neoplasms, especially for hepatocellular carcinomas. AFP-producing tumors of the stomach can histologically mimic hepatocellular carcinomas and have therefore been named hepatoid adenocarcinomas [10]. Among pancreatic malignancies, serum AFP has been demonstrated to increase in pancreatoblastomas [3] and in acinar cell carcinomas [4, 6, 8]. The mechanism by which pancreatic tumor cells produce AFP is still unknown. The close embryologic relationship between the pancreas and liver, both of which are derived from the foregut endoderm, may explain the elevation of AFP in pancreatic tumors.

Reviewed in the English literature, fewer than 30 cases of AFP-producing acinar cell carcinomas of the pancreas have been reported [4, 6, 8, 9, 11]. Among these, only four cases of AFP-producing acinar cell carcinoma of the pancreas with their imaging findings were found. All these AFP-producing acinar cell carcinomas of the pancreas on CT scan showed a hypodense mass relative to the pancreatic parenchyma, a well-enhanced capsule, and a low-attenuation central necrotic area inside the tumor [4, 5, 8]. Dynamic CT (unenhanced, arterial, and venous phase images) can increase the conspicuity of enhancement between the pancreatic parenchyma and tumor, which can help narrow the differential diagnosis. The tumors in both of our cases also had central necrosis and an enhanced peripheral wall on CT scan; in addition, one tumor had calcification. However, in our second case, the tumor was very large and had invaded the adjacent organs, so the enhanced capsule wall was disrupted.

On MRI, the mass showed hypointense signal intensity compared to normal pancreatic tissue on T1-weighted images and slightly increased signal intensity on T2-weighted images [11]. Because the lesions were not very hypervascular, the use of gadolinium enhancement would be helpful in differentiating the tumor from normally enhanced pancreatic tissue.

PTCD provided more information. It showed the severity of the CBD stricture and relieved the jaundice in our second case. On account of hypovascularity of this tumor, angiography provided little help in the diagnosis except for clearly identifying the splenic venous encasement.

The differential diagnosis of acinar cell carcinomas includes pancreatic ductal carcinomas.
and islet cell tumors. Pancreatic acinar cell carcinomas tend to be large and lobulated with a fairly enhanced capsule. Hypovascularity relative to normal pancreatic tissue with some central necrosis has also been recognized inside the tumor [12]. Pancreatic ductal carcinomas are often irregular, heterogeneously enhanced, locally invasive, and nonencapsulated. Functioning neuroendocrine neoplasms tend to be hyperdense in the arterial phase, although many are small and difficult to detect [12, 13]. Nonfunctioning islet cell tumors are usually large and show nonuniform enhancement, although smaller tumors can have homogeneous enhancement and imaging characteristics similar to those of functioning tumors [13]. Calcification in islet cell tumors is not rare, but central necrosis is uncommon, presumably because of the hypervascularity of these tumors.

Surgery is the best therapeutic modality for pancreatic AFP-producing acinar cell carcinoma [14]. Chemotherapy and radiotherapy are helpful for those who can not receive an operation. The serum AFP level rapidly decreased to a normal limit after the operation, and it might increase again if tumor recurrence or metastasis occurred.

In conclusion, the imaging findings of pancreatic AFP-producing acinar cell carcinomas tend to show a well-defined hypodense mass with occasional calcification, a central necrotic area, and a well-defined enhanced wall. With these characteristics, acinar cell carcinoma can be distinguished from the locally invasive ductal adenocarcinoma of the pancreas. Serum AFP levels also played an important role in the diagnosis and in the evaluation of therapeutic effects in these cases.

REFERENCES

製造阿爾發胎兒蛋白的胰臟腺胞細胞癌的影像學發現

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胰臟的製造阿爾發胎兒蛋白腺胞細胞癌是一種很罕見的腫瘤。我們報告兩例製造阿爾發胎兒蛋白腺胞細胞癌的影像學發現。電腦斷層檢查腫瘤是呈現低密度，中間有壞死區，偶爾有鈣化，及界限清楚且顯影不錯的壁。磁振造影檢查，這類腫瘤在T1影像是呈現低訊號強度，T2影像像呈現稍微高訊號強度，打過顯影劑以後腫瘤只呈現輕微的顯影。經皮穿肝膽道攝影及引流術則可以評估總膽管狹窄的程度並且可以解決病人的黃疸問題。血管攝影的幫忙較少，偶爾可以評估腎臟血漿被腫瘤壓縮的程度。除此之外，血清中阿爾發胎兒蛋白的濃度可以用來輔助診斷，並且評估治療效果及是否有復發。

關鍵詞：阿爾發胎兒蛋白，腫瘤，腺胞細胞癌，電腦斷層，磁振造影