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**Columns: Case Report**

**Pancreatic carcinosarcoma: first literature report on computed tomography imaging**

Shi HY *et al*. Case report and review of the literature

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**Abstract**

Carcinosarcoma of the pancreas is an extremely rare tumor and has a dismal prognosis. To the best of our knowledge, the histopathological features of the lesion have been illustrated in the literature, but to date, no reported cases have been documented on imaging characteristics. We report a female case of pancreatic carcinosarcoma, which is presenting as a mucinous cystadenoma on computed tomography (CT). We also summarized the CT characteristics according to the appended CT images in the reported cases. This is the first report of CT features of pancreatic carcinosarcoma in the English literature.

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**Key words: c**omputed-tomography; Carcinosarcoma; Pancreas; Neoplasm

**Core tip:** Pancreatic carcinosarcoma is an extremely rare tumor with a poor prognosis. We report a case of pancreatic carcinosarcoma in a 74-year-old woman and describe its appearance on computed tomography (CT). Detailed analysis and conclusion of the CT characteristics was performed according to the appended CT images in reported cases. This is the first report of radiological findings of carcinosarcoma originated from pancreas.

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**Introduction**

Pancreatic carcinosarcoma is an uncommon entity comprising a fairly small subset of all pancreatic neoplasm. It is histologically characterized by a mixture of carcinomatous and sarcomatous elements. The prognosis of pancreatic carcinosarcoma is very poor. From the data of reported cases, the majority of patients survived an average of only 6 mo after surgery. Because of the rarity and difficulty of diagnosis, radiological findings of the lesion have not been well illustrated in literature. Here, we report the clinical and contrast-enhanced computed tomography (CT) findings of carcinosarcoma originated from pancreas in a 74-year-old woman.

**Case report**

A 74-year-old woman was referred to our hospital for acute calculous cholecystitis. Contrast material–enhanced dual-phase multidetector row CT of the abdomen was performed and revealed a 2.2 cm x 2.0 cm well-circumscribed and cystic lesion that was located to the pancreatic tail and did not invade the adjacent viscera. After intravenous injection of contrast media, the lesion had a peripheral enhancing thick wall surrounding the non-enhancing low-attenuation area which is consistent with cystic fluid (Figure 1). Mural nodules and intratumoral septa were not seen on the CT imaging. There was no main pancreatic duct dilatation, nor were there any abnormalities in the rest of the pancreatic tissue. We diagnosed it as mucinous cystadenoma. The patient underwent a cholecystectomy and than discharged.

Follow-up CT imaging was performed 13 mo later. The lesion in the pancreatic tail grew up to 2.9 cm in diameter with a more thickened wall, and solid component appeared in the cystic lesion. The arterial phase revealed heterogeneous enhancement in the wall and solid part of the mass (Figure 2A). In the portal phase, the enhancement became more pronounced. The cystic part revealed non-enhancing low-attenuation (Figure 2B).

After nearly half a year, the patient was referred to our hospital again for intermittent abdominal pain and distention lasting for more than 2 wk. She reported no association with food. The abdomen was soft and tender at physical examination. Laboratory analysis revealed elevated CA19-9 (148‎.‎40‎ U‎/‎ml), ‎CA‎72-4 (19‎.‎19 ‎U‎/‎ml), CA24-2 (51‎.‎7‎ U‎/‎ml‎) and CEA (10‎.‎05‎ ng‎/‎ml) ‎‎levels. Liver function tests and complete blood count were all within the normal range. CT showed that the original lesion in the pancreatic tail manifested as a heterogeneous complex mass that contained cystic and mixed solid areas and measured 4.0 cm in diameter. The solid components increased and septa in the lesion aroused. The lesion showed progressive and heterogeneous enhancement (Figure 3). No evidence of metastasis was identified. The CT findings coupled with tumor markers raised the possibility of pancreatic mucinous cystadenocarcinoma. The patient underwent a distal pancreatectomy with splenectomy, followed by uneventful postoperative course.

On gross examination, the resected specimen consisted of a 9 cm × 6 cm × 3 cm segment of the pancreas with a mass in the tail and the spleen. Sectioning through the mass revealed a well-circumscribed tumor consisting of a 5 cm × 4 cm × 2 cm cystic lesions with thick wall that measured 0.1-0.3 cm. The cystic lesion contained dark-red substances.

Histologically, the tumor of pancreas showed two components separated from each other. The first component was composed of columnar mucin-producing epithelial cells with marked cellular pleomorphism and prominent mitoses, which is consistent with carcinoma. The second component revealed a sarcomatous growth pattern composed predominantly of highly cellular areas with spindle cells (Figure 4A). Immunohistochemically, the carcinoma component was strongly reactive for antibodies to cytokeratin 7, cytokeratin 19 and cytokeratin AE1/AE3 (Figure 4B). The sarcomatous component was strongly reactive for vimentin (Figure 4C). According to the morphology and the immunohistochemical staining pattern, it was diagnosed as pancreatic carcinosarcoma.

**Discussion**

The concept of carcinosarcoma is that a malignant neoplasm is composed of an intimate admixture of carcinomatous and sarcomatous elements without areas of transition between both components, and with each of these elements showing distinct immunohistochemical or ultrastructural features pertaining to their different lines of differentiation. Cases fulfilling these criteria have been reported in many organs, but rarely in the pancreas. The origin of mixed carcinosarcoma is unknown. Although controversy remains, several studies, using diverse immunohistochemical and molecular analyses, have suggested that the pancreatic carcinosarcoma could be of monoclonal origin, and that the sarcomatous component might have arisen from metaplastic transformation of the carcinomatous component[[1](#_ENREF_1),[2](#_ENREF_2)].

The carcinomatous components are varied. Pancreatic ductal adenocarcinoma is the most commonly reported, followed by mucinous cystadenocarcinoma[[3-6](#_ENREF_3)]. Okamura *et al*[[7](#_ENREF_7)] reported intraductal papillary-mucinous carcinoma (IPMC) in pancreatic carcinosarcoma, which is extremely rare and firstly reported. There are also different sarcomatous elements, including spindle cell sarcoma, leiomyosarcoma, malignant fibrous histiocytoma and osteosarcoma.

From the summary of the reported literature, we found that pancreatic carcinosarcoma is common in middle aged and elderly people, and few patients were identified incidentally. For patients with symptoms, the most common were abdominal pain, anorexia, nausea and vomiting. When tumors are located in the pancreatic head, they cause early jaundice, which is commonly seen in other malignant neoplasm. Serum CA19-9 can be elevated in some patients[[7-9](#_ENREF_7)].

Due to the rarity of this tumor, the prognosis has not been well defined. According to the reported literature, the majority of patients survived an average of only 6 mo after surgery. Zhu et al. reported that a 53-year-old woman had remained free of recurrence for 20 mo, which is the longest recurrence-free survival time recorded for this tumor[[9](#_ENREF_9)]. The pancreatic carcinosarcoma can also disseminate and recur[[10](#_ENREF_10),[11](#_ENREF_11)]. However, the strategy to improve the prognosis is still not available, because of the limited experience about it.

To our best knowledge, the imaging features of pancreatic carcinosarcoma have not been reported. By summarizing the reported cases in the literature, combined with our case, we found that the preferential location for pancreatic carcinosarcoma is the pancreatic head and tail, and the size is variable (ranging from 2.5 cm to 20 cm). It is worth noting that pancreatic carcinosarcoma can grow quickly, as observed in our case. During follow-up, the lesion in the pancreatic tail grew up from 2.2 cm to 4.0 cm in diameter and more solid component can also be seen 18 mo later. On CT images, the lesion in the pancreatic head more frequently appears as a solid mass with cystic regions and necrosis, while the one in the pancreatic tail is characterized by a cystic neoplasm with mural nodules and solid component, without accompanying ductal dilatation, though with some exception[[1](#_ENREF_1),[7](#_ENREF_7),[11](#_ENREF_11),[12](#_ENREF_12)].Tumors located in the pancreatic head can cause pancreatic main duct and intra- and extra-hepatic bile duct dilatation. After intravenous injection of a gadolinium chelate, solid component, mural nodules, and cystic wall show moderate enhancement. Unlike pancreatic ductal adenocarcinoma, carcinosarcoma of pancreas often has well-circumscribed border and seldom invade the adjacent organs, extrapancreatic nerve and vascular. However, they are easily metastasized to liver and peritoneum, which is the main cause of death[[1](#_ENREF_1),[10](#_ENREF_10)].

The predominant differential diagnosis with pancreatic carcinosarcoma is pancreatic ductal adenocarcinoma. Compared with ductal adenocarcinoma that is poorly vascularized, pancreatic carcinosarcoma has more vascularity. In addition, extrapancreatic perineural and vascular invasion, atrophy of the pancreatic parenchyma and duct dilatation are less common in carcinosarcoma. When pancreatic carcinosarcoma manifest as a cystic tumor, it is difficult to distinguish it from mucinous cystadenoma or cystadenocarcinoma. They share many features in common such as focal thickening of the wall, heterogeneous content, mural nodules, and so on. However, the preponderance of calcification in septa and cystic wall enable one to readily distinguish this entity from pancreatic carcinosarcoma, because intratumoral calcification in pancreatic carcinosarcoma has not been described so far.

Given the tumor’s rarity, it is difficult to establish more classical imaging findings. Nevertheless, a few characteristic imaging features have been described in this paper. We believe that the characterization of imaging features of pancreatic carcinosarcoma can increase the awareness of this entity among radiologists.

**COMMENTS**

***Case characteristics***

A 74-year-old woman was referred to our hospital for acute calculous cholecystitis and occasionally discovered a lesion in the pancreatic tail; then the patient was referred again to our hospital for intermittent abdominal pain and distention lasting for more than 2 wk.

***Clinical diagnosis***

Physical examination showed tenderness on the upper abdominal region.

***Differential diagnosis***

Pancreatic mucinous cystadenoma or cystadenocarcinoma.

***Laboratory diagnosis***

Laboratory analysis revealed elevated CA19-9 (148‎.‎40 ‎U‎/‎ml), CA‎72-4 (19‎.‎19‎ U‎/‎ml), CA24-2 (51‎.‎7 ‎U‎/‎ml‎) and CEA (10‎.‎05‎ ng‎/‎ml) ‎‎levels.

***Imaging diagnosis***

Computed tomography showed that a mass in the pancreatic tail manifested as a heterogeneous complex mass that contained cystic and mixed solid areas and measured 4.0 cm in diameter.

***Pathological diagnosis***

Histological examination demonstrated characteristic histological findings of an intimate admixture of carcinomatous and sarcomatous elements.

***Treatment***

The patient underwent a distal pancreatectomy with splenectomy.

***Related reports***

The histopathological features of pancreatic carcinosarcoma have been illustrated in the literature, but to date, there are no reported cases have been documented on imaging characteristics.

***Experiences and lessons***

The predominant differential diagnosis with pancreatic carcinosarcoma is pancreatic ductal adenocarcinoma, mucinous cystadenoma or cystadenocarcinoma.

***Peer review***

This study determines the histopathological features of the carcinosarcoma lesions of the pancreas as this is an extremely rare tumor with a poor prognosis. The authors report the computed tomography (CT) appearance of pancreatic carcinosarcoma, which is presenting as a mucinous cystadenoma, in one female patient. Detailed analysis and conclusion of the CT characteristics were done according to the appended CT images in other reported cases. The authors claim that this is the first report of CT features of pancreatic carcinosarcoma in English literature.

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Figure 1A.tifFigure 1B.tif

A B

**Figure 1** **computed tomography imaging of the abdomen at the first time.** The arterial phase (A) and portal venous phase (B) revealed a 2.2 cm x 2.0 cm well-circumscribed and cystic lesion that was located to the pancreatic tail, with a peripheral enhancing thick cystic wall that surrounded non-enhancing low-attenuation area which is consistent with cystic fluid. We diagnosed it as mucinous cystadenoma.

Figure 2A.tifFigure 2B.tif

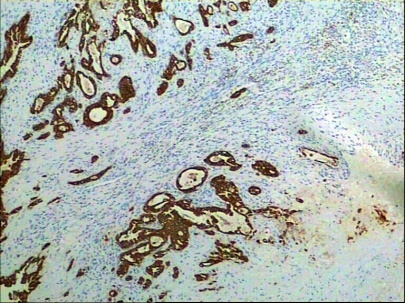
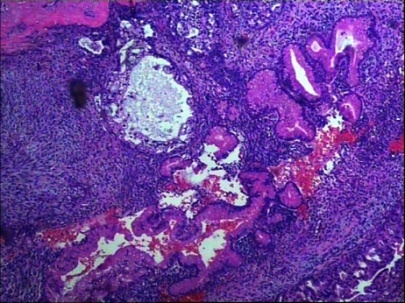
A B C

**Figure 2 Follow-up computed tomography imaging in 13 mo later**. The arterial phase (A), portal venous phase (B) and oblique sagital contrast-enhanced computed tomography image (C) showed the lesion grew up to 2.9 cm in diameter with heterogeneous enhancement in the more thickened wall and solid component.

Figure 3A.tifFigure 3B.tif

A B

**Figure 3 Follow-up computed tomography imaging after nearly half a year.** computed tomography (A: Arterial phase; B: Portal venous phase) showed that the original lesion in the pancreatic tail manifested as a heterogeneous complex mass that contained cystic and mixed solid areas that measured 4.0 cm in diameter. The solid components increased. The lesion showed progressive and heterogeneous enhancement.



A B C

**Figure 4 Histological examination of the lesion.** A: Microscopy of the tumor indicated a predominance of dual disparate sarcomatous and carcinomatous components. Spindle-shaped tumor cells and well-differentiated adenocarcinoma cells coexisted and intermingled (HE staining, × 200); B: Cytokeratin 7 immunostaining showed strong and diffuse expression in the ductal adenocarcinoma cells (× 200); C: Sarcomatous cells were immunopositive for vimentin (× 200).