Case Report

# Pancreatic paraganglioma: a case report of CT manifestations and literature review

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**Abstract:** Paragangliomas are rare tumors arising from paraganglia (or chromaffin bodies) within the ganglia of the sympathetic trunk and the ganglia of the celiac, renal, suprarenal, aortic and hypogastric plexuses. These tumors can be widely distributed from skull base to the bottom of pelvis. Paraganglioma originating from pancreas is extremely rare. Here we report a 40-year-old female patient with paraganglioma located in the pancreatic head as a solid, well-demarcated mass with substantial enhancement on contrast-enhanced CT examination. Relevant literatures are also reviewed.

**Key Words:** Paraganglioma; pancreas; computer tomography

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## Introduction

A paraganglioma is a rare neoplasm originating from paraganglia (or chromaffin bodies) within the ganglia of the sympathetic trunk and the ganglia of the celiac, renal, suprarenal, aortic and hypogastric plexuses. Although it is rare, it can be found in abdomen, pelvis, and the head and neck region. In abdominal region, most paraganglias are located at adrenal glands or along aorta (the latter is known as Zuckerkandl body). To our knowledge, only 12 cases of paraganglioma arising from pancreas have been reported in English literatures, most of which are in pathological literatures.

## Case report

A pancreatic mass was revealed in a 40-year-old woman by abdominal ultrasonography at a routine health check. She did not complain any relevant symptoms. Her family history, past history, physical examination, and laboratory test were unremarkable, except slightly elevated serum CA15-3 (30.04 U/mL; normal, <25 U/mL). Abdominal CT showed a well-demarcated solid mass measured 4.5 cm ×4.2 cm in the uncinate process of the pancreas (*Figure 1A*). Contrast-enhanced CT showed that the tumor demonstrated dramatical enhancement at pancreatic and portal vein phases (*Figure 1B,C*) . Several low-density foci were revealed also at plain and enhanced scans. Mild dilation of pancreatic duct was noted at the body and tail of pancreas. No biliary dilation or liver lesions were detected. The resected mass was grey-yellow in color, with multifocal hemorrhagic areas and cystic degeneration. Microscopically, the tumor was composed of nests of cells arranged in a classic Zellballen pattern separated by a delicate vascular network (*Figure 1D*). The chief cells showed positive staining to neuron-specific enolase, chromogranin A, synaptophysin, and cytokeratin (*Figure 1E*), but showed negative response to insulin, somatostatin, and glucagon. Less than 2% of the tumor cells were positive to Ki67, indicating a low proliferation ratio. The chief cells were surrounded by S-100 protein-positive sustentacular cells (*Figure 1F*). Regional lymph nodes and the resected margins were free of tumor cells. A diagnosis of paraganglioma of the pancreas was established.

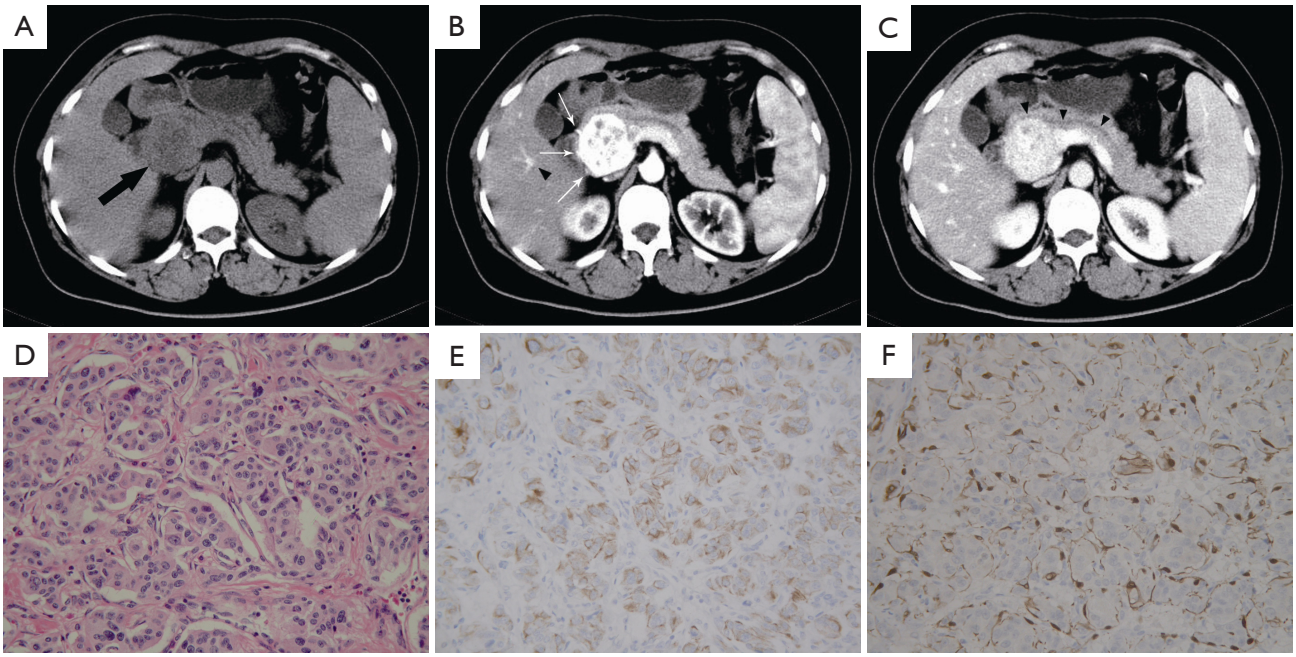


**Figure 1** A. Non-enhanced abdominal CT shows a well- circumscribed mass (thick black arrow) with a low density (37HU) in the uncinate process of the pancreas; B. The mass enhances remarkably (181HU) with isodensity to the aorta in the arterial phase. Non-enhancing patchy areas are found inside the tumor. Intratumoral vessels are obscure. Peritumoral arteries are rich and slightly thickened (thin white arrows). No obvious drainage veins of the tumor are observed. There is early contrast filling of the portal vein (black arrowheads); C. The mass is still well enhanced (135HU) in the venous phase, while the non-enhancement areas shrank compared with those in the arterial phase. The head section of the main pancreatic duct is displaced anteriorly and the main pancreatic duct shows diffuse and mild dilation (black arrowheads). No dilation of biliary ducts is observed; D. Microscopically, the tumor is composed of nests of cells arranged in a classic Zellballen pattern separated by a delicate vascular network. Nuclei of the chief cells are round to oval in shape, large, and hyperchromatic without obvious mitosis. Cytoplasm is eosinophilic in most chief cells and clear in others (Magnification ×200); E. The chief cells show positive staining to cytokeratin (Magnification ×200); F. The chief cells are surrounded by S-100 protein-positive sustentacular cells (Magnification ×200)

## Discussion

Paragangliomas originate from the neural crest cells that are widely located in adrenal medulla, carotid and aortic body, jugular foramen, middle ear, aortopulmonary region, the organ of Zuckerkandl, and other sites of the body. The adrenal gland is the most common primary site of paragangliamas, followed by priaorta region and the head and neck. Paragangliomas were also found in many other organs. Only 12 cases of pancreatic paragangliomas have been reported in English literatures, (1-10) and only two of them were focused on radiological mani-festations. In other words, the imaging features of pancreatic paragangliomas have not been well described. In our current case, the pancreatic paraganglioma appeared as a solid mass with necrotic foci or cystic changes on CT scans. Dramatical enhancement was found on contrast-enhanced CT. Although it shares similar imaging characteristics with other neuroendocrine neoplasms of pancreas, its CT findings are different from pancreatic carcinoma, which often appears as a poor enhanced mass with dilations of bile ducts and pancreatic duct as well as retroperitoneal involvement. Although the pancreatic paraganglioma is located at the head of pancreas, the biliary ducts usually show no dilation and patients show no jaundice, which are helpful in differentiating pancreatic paraganglioma from pancreatic carcinoma.

Paragangliomas at various sites share relatively identical imaging features, including clear margin, hypervascularity, and cystic areas within the tumor; the latter demonstrates cystic degeneration or hemorrhagic necrosis. In our current case, these imaging characteristics were also found. Although most paragangliomas are solitary, 8 among 13

 

cases of pancreatic paragangliomas were cystic (1-10). Cystic pancreatic paragangliomas ranged from 4.0 cm to 13.0 cm in diameter, with a median of 8.2 cm. Solid pancreatic paragangliomas ranged from 1.5 cm to 7.0 cm in diameter, with a median of 3.4 cm. Differential diagnosis between pancreatic cystic paraganglioma and pseudocyst can be difficult. According to literatures, four patients were misdiagnosed preoperatively and drainage was carried out in two of them (2,3). It is also difficult to differentiate between solid paraganglioma and other neuroendocrine tumors of the pancreas, especially those nonfunctional pancreatic paragangliomas. Kim SY *et al.* argued that early contrast filling of the prominent draining veins of solid paraganglioma and the portal vein might provide clues for differential diagnosis (4). However, prominent draining veins of solid paraganglioma were not observed in our patient. Actually, we observed plenty of slightly thicken peritumoral arteries and early contrast filling of the portal vein, which is believed to be a common phenomenon of hypervascular tumors of the pancreas. Occasionally, extra-adrenal paraganglioma arising from the retroperitoneum and abutting the pancreas may mimick a pancreatic neoplasm (11).

In conclusion, we present a rare paraganglioma arising in pancreas, whose CT findings were featured by a well-demarcated solid mass with necrotic foci or cystic changes and by the dramatic enhancement after the intravenous administration of contrast media. These findings are similar with those of paragangliomas in other sites and other neuroendocrine neoplasm of pancreas, which may be helpful to urge the radiologists to consider the possibility of paraganglioma during differential diagnosis.

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