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**Pancreatic perivascular epithelioid cell tumor: A case report with clinicopathological feature and literature review**

Jiang H *et al.* A rare case of pancreatic PEComa

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**Abstract**

Perivascular epithelioid cell tumor (PEComa) of the pancreas is an unusual tumor deriving from mesenchyma. This paper described a case of PEComa of the pancreas, which was initially suspected as neuroendocrine carcinoma by biopsy, therefore surgical treatment was recommended due to undetermined diagnosis. The result of surgical specimen under microscope showed that the morphology of tumor cell was epithelioid or spindle-shaped, ranging in a nested pattern. Additionally, these cells had a large extent of acidophilic cytoplasm and no mitotic figures, and they expressed HMB-45, Melen-p and smooth muscle actin immunohistochemically. The pathological examination indicated PEComa originated from the pancreas, but symptoms related to tuberous sclerosis were absent. Since PEComa is extremely rare in the pancreas, it is likely to be ignored in differential diagnosis. In conclusion, our article highlighted the clinicopathological feature of PEComa and conducted a literature review focusing on PEComa so as to deepen the understanding of PEComa.

**Key words:** Pancreas; Perivascular epithelioid cell tumor; HMB-45; Immunohistochemistry; Clinicopathological feature

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**Core tip:** Perivascular epithelioid cell tumor (PEComa) of the pancreas is an unusual tumor deriving from mesenchyma. Though describing a rare case of PEComa of the pancreas, we'd like to highlight the clinicopathological feature of PEComa and conduct a literature review focusing on PEComa so as to deepen the understanding of PEComa. Besides, we also reviewed the biological behavior, prognosis and therapeutic strategy of PEComa.

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**INTRODUCTION**

Perivascular epithelioid cell tumor (PEComa) is an extremely rare tumor derived from mesenchymal tissue, with characteristics of perivascular epithelioid cell (PEC) in histology and immunohistochemistry[1]. Microscopically, the morphology of PEC is of epithelial origin, containing a bright cytoplasm or fine grained eosinophilic shapes with a positive PAS staining. Moreover, the nuclei of PEC are relatively small, and are round or oval shaped with small nucleolus; intranuclear inclusion bodies can be observed occasionally. PEC, distributing in the perivascular region in a radial pattern, is amylase intolerant. In detail, PEC shows epithelioid features in nearby vessels, while it becomes spindle-shaped distant from vessels. The proportion of epithelioid cell and spindle cell is different depending on the patient. In the immunohistochemistry staining, PEC usually expresses HMB45 and Melan-A. PEC is featured with melanosome in ultrastructure, and is rich in glycogen and cytoplasmic filaments[2].

PEComas can be classified as angiomyolipoma, clear cell “sugar” tumor of the lung, lymphangiomyomatosis (LAM) and other PEComas characterized by similar histological and immunohistochemical presentations[3]. In most cases, PEComas are benign; however, Bonetti *et al*[4] reported four abdominopelvic sarcoma of PEC in young women, raising concern about malignant PEComas that result in regional tissue infiltration, multiple metastases and even patient’s death[5-7]. As for PEComas of the pancreas, only two malignant cases were reported with liver metastasis[8,9].

Herein, we report a 50-year-old female with benign PEComas of the pancreas that cannot be definitely diagnosed preoperatively. Further, a literature review on PEComa of the pancreas with special consideration of pathological diagnosis is performed.

**CASE REPORT**

***Clinical characteristics***

A 50-year-old female patient was admitted to our hospital in November 2013 because of abdominal ultrasound (US) findings of space-occupying lesion in the head of pancreas, which cannot be diagnosed clearly. She was a lifelong nonsmoker who took no alcohol, and had no history of family inherited diseases. The patient denied any history of surgery or trauma. CT examination demonstrated no significant abnormality in the morphology and density of the pancreas, and no pancreatic duct dilatation (Figure 1A). Moreover, peripancreatic fat space is clearly demarcated and no retroperitoneal lymph nodes were enlarged. On the arterial phase, there was a relatively low density of nodule with approximately the size of 1 cm × 1.4 cm in the uncus of the pancreas. On portal venous phase and delayed phase, this nodule enhanced gradually and slightly, with a significantly lower density than the surrounding pancreatic tissue level whereas the pathologically changed border was well-defined on delayed phase. Magnetic resonance (MR) imaging (Figure 1B) showed that a round abnormal signal with the size of 1.7 cm × 1.4 cm was found in the head and uncus of the pancreas; T1WI showed low signal clearly distinctive to normal pancreatic tissue surrounding a relatively high signal. In T2WI, the mass was difficult to distinguish from the surrounding pancreatic tissue due to the equal signal. Endoscopic ultrasound (EUS) showed a hypoechoic region with size of 1.6 cm × 1.4 cm in the uncus of the pancreas. This region had clear boundaries and echo was not equal inside the low echo region. Pancreatic tail shape remained regular and pancreatic duct was not dilated. There was no dilatation of intrahepatic bile duct and enlargement of lymph nodes.

***Methods and materials***

The patient had undertaken endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) (ECHO-25G, Cook Company). Subsequently, the biopsy specimen was fixed in formalin, and then dehydrated by gradient ethanol, dewaxed by Xylol, followed by paraffin imbedding, cut section for microscopic examination, and haematoxylin-eosin staining.

Immunohistochemistry: all biopsy and surgical specimen had undergone immunohistochemical staining by EliVisionTM Plus method. Immunochemical staining was performed for all the specimen by using the following markers: CD56, Cam5.2 (Cytokeratin), Neuron Specific Endolase (NSE), Chromogranin A (CgA), Alpha-1-Antitrypsin (α-AT), CD34, CD56,S-100, TFE3, Estrogen Receptor (ER), Progesterone Receptor (PR), Calponin, Synaptophysin (Syn), Pax-8, beta-Catenin, CD117, Melan-A, HMB-45, smooth muscle actin (SMA), Epithelial Membrane Antigen (EMA), and Vimentin; heated at 37 °C for 1 h, p53, Ki-67, and kept at 4 °C overnight. The staining intensity was recorded as positive (> 10% of plasma and nucleus stained) and negative. Besides, Ki-67 was evaluated by percentage. All information of antibodies was summarized in Table 1.

***Results***

**Biopsy specimen:** Spindle-shaped nucleus can be found in some cell lumps with sporadically distributed cells in the background, suggesting that they were malignant cells derived from unidentified sources. In DNA ploidy analysis, a few DNA ploidy-abnormal cells were found.

**Cytological results:** Cytology smear showed that cells were distributed in irregular lumps, nuclear sizes varied slightly and arranged mussily. Enriched plasma and vaguely defined boundary were identified in the cell (Figure 2).

Tumor cells were epithelioid or spindle-shaped with large size and nested distribution (Figure 3). Tumor cells contained a bright cytoplasm or fine grained eosinophilic shapes with suspicion of clear cell carcinoma. Therefore, gastrointestinal stromal tumor, neuro-endocrine tumor, acinic cell carcinoma, solid pseudo-papillary tumor, and metastatic clear-cell carcinoma, should be excluded. The immunohistochemistry results were as follows: CD56 (+), CAM5.2 (-), P53 (-), NSE (-), Vimentin (-), Ki67 (2%), CgA (-), α1-ACT (-), Syn (-), Pax-8 (-), β-Catenin (positive in plasma), CD117 (-). Since PEComa of the pancreas had not been diagnosed in our hospital, this tumor was not considered as PEComa preoperatively. Based on cytological and immunohistochemical results, neuro-endocrine tumor was suspected but acinic cell carcinoma and solid pseudo-papillary tumor were not excluded, and operation was suggested.

**Surgical specimen:** A mass with the size of 2 cm × 2 cm in the head of the pancreas during operation, this mass was of medium texture with a complete capsule. Besides, the boarder was clear between the mass and adjacent tissues and metastatic lesions were not found.

Gross examination (Figure 4): the pancreas, duodenum and gall bladder were resected; the size of the pancreas head was 8 cm × 4 cm × 3 cm, a mass with the size of 2 cm × 1 cm × 1 cm that was 4cm away from the surgical margin. The transection of the mass was grayish and solid soft texture.

Microscopic examination: the boundary between tumor and adjacent tissue was clear. Tumor cells were arranged in nests and some tumor cells grew around arteries. Mounts of vessel were in mesenchyma with hyaline degeneration (Figure 5B). The morphology of tumor cell was epithelioid or spindle-shaped (Figure 5C and D), ranging in a nested pattern. Additionally, these cells had plenty of acidophil cytoplasm and no mitotic figures. There was no fat tissue found in the tumor. Lymphatic invasion was not observed in this patient.

Immunohistochemistry (Figure 6): Mel-A (+), HMB45 (+), SMA (+) CD56 (+), Ki67 (1%), CAM5.2 (-), P53 (-), NSE (-), Vimentin (-), CgA (-), α-AT (-), Syn (-), Pax-8 (-), β-Catenin (positive in plasma), CD117 (-), CD34 (positive in vessel), S-100 (-), TFE3 (-), ER (-), PR (-), Calponin (-), MSA (-).

**Diagnosis:** PEComa in the head of the pancreas.

***Follow-up***

No recurrence or distant metastases were observed in a follow-up of 14 months. However, Nagata *et al*[8] reported that benign PEComa could recur after surgery, so it is necessary to follow-up and reexamine.

**DISCUSSION**

PEComa of the pancreas is extremely rare and only twelve previous cases[2-5,8-17] were reported during the last decade. Combining these previous cases with ours, we found that these patients with a mean age of 52 years (varied from 31 to 62 years) were mostly women, including eleven females and two males. The morbidity of PEComa in female was significantly higher than that in male, suggesting that one risk factor of PEComa is related to sex hormone. Additionally, some studies revealed that progesterone receptors (PR) were expressed in PEComas immunohistochemically, especially in LAM and renal AKL[18]. However, in these thirteen cases, PR was negatively expressed in five PEComas cases and only one PEComa partially expressed ER[15]. Tuberous sclerosis (TSC) containing TSC1 or TSC2 gene deletion can be seen in some PEComas, especially in renal AKL. Located on chromatosome 9q and 16p respectively, TSC1 and TSC2 play a pivotal role in Rheb-mTOR-P7OS6K pathway[1]. Nevertheless, all the PEComas of the pancreas presented without TSC.

Abdominal pain is the main initial symptom in PEComa of the pancreas, while a few cases were asymptomatic that were found during health examination or follow-up examination for other disease. In the sites of PEComa, head of pancreas accounted for six cases, the body for five cases and uncus for two. Tumor size varied from 15mm to 100mm with an average of 23 mm, and tumor ruptured in a case.

In pathological morphology, akin to previous cases, the PEComa reported in our case was a solid homogeneous nodule with mainly clear boundary and only partially infiltrated to adjacent tissue. Histological results manifested tumor cells were mainly made up of clear cells or eosinophilic cells with nested, fascial or laminar distribution. Tumor cells were epithelioid or spindle-shaped with plenty of glycogen. Nucleolus atypia and small nucleolus can be found. Mounts of vessel were in mesenchyma, with some tumor cells growing around arteries. Adipose tissue that would make the diagnosis easier is rarely presented in PEComa. In the 12 cases of previous reports, Adipose tissue was found in two cases[8,11]. In one of the two cases occurred liver metastasis 27 mo after surgery[8]. Therefore, it seems that the biological behavior of the tumor is not related to the existence of fat tissue. Immunostaining shows that PEComa generally has both smooth muscle cells and melanoma evidence. Folpe’s[19] study indicated that HMB-45 is the most sensitive marker for melanoma cells (96%), followed by Melan-A (72%) and the microphthalmia-associated transcription factor (MiTF) (50%).In their study, all cases at least expressed one marker of melanoma cells, and 80% cases expressed smooth muscle actin, especially in epithelioid cells. In PEComa of the pancreas, only one of the 13 cases did not express α-SMA[9], and this is the only case of malignant PEComa in the pancreas with epithelioid tumor cells. Therefore, we proposed that the lack of smooth muscle expression may serve as a malignant indicator of PEComa. All three indicators previous reported in PEComa of pancreas expression are as follows:HMB-45（13/13, Melan-A（8/9）,andα-SMA（12/13）. Meanwhile, MiTF and CD63 were expressed in tumor cells. Additionally, CD117 was positive in a few cases. PEComa also expressed TFE3 and cyclin D1andexpresseddesmin to a lesser extent, and did not express S-100 and CK generally.

To date, the biological behavior and histologic origin were unknown. The pervasive concept is that PEComas are usually benign, whereas increasing reports indicated that PEComa may have malignant potential even though there is no consensus to evaluate the PEComa. In 2005, Folpe *et al*[19] suggested that tumor which meets at least two points of the following criteria should be considered as malignant PEComa based on previous reports about PEComa. The criteria are as follows: (1) tumor diameter >50mm; (2) tumors with an invasive growth pattern; (3) tumors that possess advanced nucleus and cell richness; (4) mitotic count higher than 1/50 HPF; and (5) with necrosis and vascular infiltration. However, it is difficult to confirm the accuracy of Folpe’s criteria in distinguishing malignant PEComa from the benign one due to rare cases of PEComa[5]. In these 13 cases, only one case[9] can be regarded as malignant PEComa according to Folpe’s criteria, in which liver metastasis occurred in 6 mo after surgery. But this patient had a family history of breast cancer and *BRCA2* gene mutation. The patient undertook radiotherapy and chemotherapy because of breast cancer 10 years ago. Therefore, whether malignant PEComa resulted from *BRCA2* gene mutation or chemotherapy and radiotherapy was unknown. Additionally, another invasive PEComa cannot be diagnosed as malignant one, whereas multiple liver metastases were found in 27 mo after surgery in this patient. Generally, more cases are required to be analyzed in order to evaluate the recurrent risk of PEComa and develop an effective therapy for PEComa. If histology results show some malignant features, such as mitosis index, tumor cell pleomorphism and invasive growth, close follow up is then required.

In diagnosis and differential diagnosis, PEComas, especially the type mainly presented with epithelioid cell or spindle-shaped cells, should be distinguished from pancreatic clear cell neuroendocrine tumor, solid pseudopapillary rumor, metastatic renal clear carcinoma, metastatic gastrointestinal stromal, metastatic melanoma or soft tissue clear cell sarcoma[13]. (1) clear cellular neuroendocrine carcinoma: PEComa and clear cellular neuroendocrine carcinoma are similar in morphology and featured by plenty of plasma and lipid droplets[20]. They can be distinguished by immunohistochemistry. Chromograni and Synaptophysin are positive in clear cellular neuroendocrine carcinoma while that in PEComa are negative; (2) solid pseudopapillary tumor: tumor cells are eosinophilic and neutrophilic, and papillary structure can be seen under microscope. Sometimes hemorrhagic necroses are found inside the tumor. Immunohistochemically, CK, CD56、synaptopgysin and beta-catenin are positive but HMB-45 are negative [21]; (3) clear cell carcinoma: cytokeratin is widely expressed in clear cell carcinoma while melanin is expressed in PEComa; (4) gastrointestinal stromal tumor (GIST): epithelioid cells mixed with spindle-shaped cell in both PEComa and gastrointestinal stromal tumor, but a great deal of vasoganglion and clear and eosinophilic tumor cell exist in PEComa, lacking of fibroblast-like cells. PEComa mostly do not express CD117, but some cases express CD117 in immunostaining. Besides, molecular detection shows no c-kit gene mutation. CD34 is positive in GIST, while PEComa does not express CD34[21]; (5) metastatic melanoma: owing to positive express of melanin in PEComa, differential diagnosis is necessary to distinguish PEComa from metastatic melanoma. In most cases, we can differentiate PEComa with myogenic markers from metastatic melanoma with s-100; and (6) alveolar soft part sarcoma: Because of its organ-like clear cell structure, it is also important to differentiate PEComa from alveolar soft part sarcoma. Alveolar soft part sarcoma is amalignant tumor, often contains high level nuclear atypia and clear nucleoli. Vascular invasion is common in alveolar soft part sarcoma, which is only found in malignant PEComa. Immunohistochemically, melanoma-derived markers and myogenic markers are not expressed in alveolar soft part sarcoma.

Currently, EUS-FNA serves as one of the most important preoperative diagnosis methods, and is widely used in clinical diagnosis. In these cases, 9 of 13 had performed EUS-FNA examination prior to operation. However, 5 cases cannot be diagnosed clearly. Therefore, when cell arrangement and shape akin to neuroendocrine tumor are shown under microscope, we must take into account the possibility of PEComa.

In conclusion, as an unusual tumor deriving from mesenchyma, PEComa of the pancreas is always benign. Cases such as concomitant with TSC or other syndromes have not been reported yet. Complete surgical resection is the main treatment for PEComa, whereas the necessity of resection and timing of surgical treatment are relatively limited. Further, for cases with huge inoperable tumor and multiple metastases, effective treatment is in lack since the effect of traditional radiotherapy and chemotherapy are poor. Until recently, Wagner *et al*[22] reported three cases of malignant PEComa reacted to mTOR inhibitor sirolimusin radiological examination, indicating that mTOR inhibitor may serve as a candidate for future targeted chemotherapy drug, but it also needs more cases of summary. As for benign PEComa, it is necessary to follow-up on a regular basis, but aggressive therapy is not suggested.

**COMMENTS**

***Case characteristics***

A 50-year-old female patient admitted to hospital because of abdominal ultrasound findings of space-occupying lesion in the head of pancreas, which cannot be diagnosed clearly.

***Clinical diagnosis***

Upon physical examination, the patient had no clinical abnormality and was diagnosed with the pancreas head mass according to the previous diagnosis.

***Differential diagnosis***

Pancreatic clear cell neuroendocrine tumor, solid pseudopapillary rumor, metastatic renal clear carcinoma, metastatic gastrointestinal stromal, metastatic melanoma or soft tissue clear cell sarcoma.

***Laboratory diagnosis***

Laboratory tests showed no abnormal value.

***Imaging diagnosis***

A contrast-enhanced computed tomography scan, Magnetic resonance imaging and endoscopic ultrasound all revealed a space occupying in the uncus of the pancreas.

***Pathological diagnosis***

By cytological , histological and immunohistochemical examination the pathological diagnosis was perivascular epithelioid cell tumor (PEComa) of the pancreas, 2 cm × 2 cm × 1 cm, in the head of the pancreas.

***Experiences and lessons***

PEComa originated from the pancreas is very rare and we reveals the clinicopathological features of it. Besides, caution is needed in order to differentiate this entity from other pancreatic tumors, especially pancreatic clear cell neuroendocrine tumor, solid pseudopapillary rumor, metastatic renal clear carcinoma, metastatic gastrointestinal stromal, metastatic melanoma and soft tissue clear cell sarcoma.

***Peer-review***

This article highlights the clinical characteristics of PEComa of the pancreas and offered an excellent methodology to diagnose the disease. After reviewing this manuscript, positive information of this article is worth to the readers.

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**Table 1 Characteristics of antibodies using in immunohistochemistry and staining conditions**

|  |  |  |  |
| --- | --- | --- | --- |
| **Antibody** | **Clone** | **Source** | **Dilution** |
| CD56 | 56C04 | Maxim | 1:100 |
| CAM5.2 | CAM5.2 | Maxim | 1:50 |
| NSE | E27 | Maxim | 1:100 |
| CgA | SP12 | Maxim | 1:100 |
| α-AT | Polyclonal | DAKO | 1:100 |
| CD34 | QBEnd/10 | Maxim | 1:100 |
| P53 | D0-7 | Maxim | 1:200 |
| S-100 | SP11 | Thermo | 1:100 |
| TFE3 | MRQ-37 | Maxim | 1:50 |
| ER | 1D5 | Maxim | 1:100 |
| PR | 1A6 | Maxim | 1:100 |
| Calpolin | CALP | Maxim | 1:100 |
| Syn | Polyclonal | Maxim | 1:100 |
| Pax-8 | ZR-1 | Gene Tech | 1:100 |
| beta-Catenin | CAT-5H10 | Maxim | 1:100 |
| CD117 | YR145,2E4 | Maxim | 1:100 |
| Melan-A | A103 | Maxim | 1:100 |
| HMB-45 | HMB45 | Maxim | 1:50 |
| SMA | 1A4 | Gene Tech | 1:100 |
| EMA | E29 | Maxim | 1:100 |
| Vimentin | V9 | Maxim | 1:200 |
| Ki-67 | MX006 | Maxim | 1：200 |



**Figure 1 Abdomen computed tomography-scan demonstrating.** A: CT delayed phase: a relatively low density of nodules（arrow）with approximately the size of 1 cm × 1.4 cm in the uncus of the pancreas; B: A round abnormal signal (arrow) with size of 1.7 cm × 1.4 cm was found in the head and uncus of the pancreas, T1WI showed low signal, clearly contrast to normal pancreatic tissue surrounding a relatively high signal.



**Figure 2 Cytology results.** A: Cells were irregular lumps distributed with medium sizes of nuclei, arranged disorderly, with crowded overlap; B: tumor cell with abundant cytoplasm and unclear boundary. Messily arranged spindle cell nucleus can be seen in some cell clumps. Scattered single cells were found the background.



**Figure 3 Biopsy specimen.** HE staining shows the epithelial tumor cells with bright or slightly eosinophilic granules with nested distribution.



**Figure 4 Gross examination.** Tumor with the size of 2 cm × 2 cm in pancreatic head with clear boundaries, with a cross-section of gray and solid soft texture. Metastatic lesions were not found.



**Figure 5** **Microscopic examination.** A: Microscopy showing clear boundaries between the tumor and adjacent pancreatic tissue; B: Mounts of vessel were in mesenchyma, among which were with hyaline degeneration; C: Epithelioid tumor cell; D: Spindle-shaped tumor cell with bright or slightly eosinophilic granule. A, B, C and D: HE staining.



**Figure 6 Immunohistochemistry.** A: The tumor cells expressed HMB-45; B: The tumor cells are immunophenotypically positive for the melanocytic marker Melan-A; C: The tumor cells are immunophenotypically positive for the smooth muscle marker SMA. Original magnification × 200. A, B and C: Immunostaining.