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CASEREPORT

A Case Report of Undifferentiated Pancreatic Carcinoma

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ABSTRACT

We report a rare case of an Undifferentiated Carcinoma of Pancreas (UCP) in a 21-year-young man, who presented with painless jaundice, dyspnea and dry cough. Weight loss for one month. No history of fever. It constitutes less than 1% of pancreatic exocrine neoplasia. Its histopathologic properties remain poorly understood. LFT shows mild increase in transaminases and alakalinephosatase. However, CA19.9 and serum electrolytes were normal. Imaging studies revealed a lobulated solid hypo enhancing mass noted in the pancreatic duodenal groove encasing the common hepatic artery, gastroduodenal artery and partly encasing main portal vein with likely involvement of part of the head and uncinate process of pancreas measuring 6.6 × 4.3 × 3.5 cm in the head of the pancreas. A EUS guided Fine Needle Biopsy (FNB) was performed and microscopy showed a cellular neoplasm composed of pleomorphic mononuclear cells (IMP3-positive; LCA, and CD68 negative) and occasional multinucleated giant cells (LCA, and CD68-positive; pancytokeratin, and EMA-negative) consistent with UCP. Evidence supports that the giant cells are non-neoplastic and of histiocytic origin.

INTRODUCTION

After colorectal carcinomas of gastrointestinal malignancies, pancreatic carcinoma is the second most common type. Undifferentiated Carcinoma of the Pancreas (UCP) is a rare malignant epithelial neoplasm, accounting for 0.5% to 7% of pancreatic tumors. Undifferentiated Carcinoma with Osteoclast-like Giant Cells (UC-OGCs) of the pancreas was first described by Rosai in 19689. UCP has a much poorer prognosis than typical ductal adenocarcinoma of the pancreas. Both genders are affected almost equally. The mean age at diagnosis is 63 years [1-3].

CASE REPORT

A 21-year-young man, who presented with painless jaundice, dyspnea and dry cough since 10 days. Weight loss since a month. No history of fever. The Liver function test shows slight increase in transaminase and alakaline phosphatase. Tumor markers, including CA-19.9 and CEA were normal. Imaging studies revealed a lobulated solid hypo enhancing mass in the pancreatico-duodenal groove encasing the common hepatic artery, gastroduodenal artery and partly encasing main portal vein with likely involvement of part of the head and uncinate process of pancreas measuring 6.6 × 4.3 × 3.5 cm. Also, there was loss of fat planes of HOP with the periportal soft tissue mass and mild hepatomegaly. Mild bilateral pleural effusion also seen.

A EUS guided biopsy was performed and Microscopically, the neoplasm was composed of non-cohesive, pleomorphic, and bizarre cells without glandular formation. Cells are round to oval with vesicular to hyper chromatic nuclei, inconspicuous nucleolus and moderate cytoplasm. Cells show marked a typia. Occasional multinucleated giant cells were scattered. Lymph vascular emboli or perineural invasion not identified.

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The surrounding fibrous stroma was scant compared with that of conventional invasive ductal adenocarcinoma of the pancreas. No evidence of spindle cells, sarcomatoid or osteoid changes.

The transition between adenocarcinoma and undifferentiated components was not observed. Immunohistochemically, most neoplastic cells were positive for IMP3 and mCEA, EMA and pancytokeratin were negative. Other markers like Synaptopyhsin, chromogranin, CD 56, CD20, LCA, CD30, SALL4, s-100, HMB-45 and CA 19.9 were also negative in the tumor cells ruling out other differentials of Neuroendocrine tumor, Lymphoma, Germ cell tumour and Melanoma respectively. Focal macrophages show immunoreactive for LCA and CD68. The final diagnosis was an undifferentiated carcinoma of the pancreas. However, osteoclastic-like giants cells or sarcomatoid or osteoid change not identified in our case [4].

Undifferentiated carcinoma of pancreas with/without osteoclast-like giant cells is a rare epithelial neoplasm composed of pleomorphic mononuclear cells showing no glandular structure or other features to indicate a definite differentiation. The histogenesis of these cellular populations remains unclear. Some authors favoring mesenchymalorigin, and others favoring epithelial origin. In a study was showed that UC-OGC's are variants of Pancreatic Ductal Carcinoma (PDC) due to the presence of shared mutations in KRAS and other critical tumor suppressor genes commonly associated with PDC (TP53, CDKN2A and SMAD4) [5].

UCP is more common in middle and elderly patients. The average age is 63-65 years. Slight female prepondance noted. The clinical symptoms are atypical, and two-thirds of cases in the literature reported abdominal or back pain, one-third presented with painless jaundice5,8 and steatorrhea, and some other Seven of patients had elevated CA19.9 levels (range 41.26–392 U/mL) and 2/8 presented with elevated CEA (range 29.6–196 ng/mL). UCP is invasive and usually has a poor prognosis. In our case, patient was a 21 years old male

UCP is more commonly to occur within the body and tail of the pancreas, account for about 70% of reported cases. In our case the mass was in the head of pancreas and radiologically, it was lobulated hypo enhancing mass with loss of fat planes whereas in adenocarcinoma of the pancreas which is uniformly hypo echoic and UCP commonly appears as a heterogeneous mass

with distinct hyper- and hypo echoic regions [6].

On other hand, undifferentiated carcinoma pancreas with osteoclastic-like giant cells (UC-OGC) comprises of benign giant cells in a background of atypical pleomorphic mononuclear malignant cells. They are commonly considered to be of benign histiocytic origin and in our case, it is supported by occasional multinucleated giant cells shows lack of pleomorphism, and immunoreactivity with CD68. It is hypothesized that OGC recruitment is a result of chemotactic factors produced by neoplastic cells. These giant cells typically arranged in nodules, sheets or few are singly scattered and associated with areas of hemorrhage. They can fill and the OGC component can replace ducts within the pancreas and comprise the majority of the tumor and even resemble osteoclastoma pattern with less amounts of the malignant cells. One third of cases reported in the study had presence of metaplastic, mature bone tissue. Many of the cases in the study showed associated pancreatitis. OGC and pancreatic intraepithelial lesions were observed in 47% of the UC-OGC cases. In our case, osteoclastic like gaint cells or metaplastic or mature bone tissue had not identified. No evidence of pancreatic intraepithelial lesions or associated pancreatitis in our case [7-9].

UCP usually shows lymph-vascular and perineural invasion. Lymph-vascular invasion was present in 63% and Perineural invasion was present in 32% of cases as compared to 86% in PDCs ($p<0.0001$). However our case showed no evidence of lymph-vascular or perineural invasion. Found that lymph node metastasis in UC-OGC was less as compared to PDC. It was present in 23% of cases of UC-OGC and 64% in PDC ($P<0.0001$) [10].

DISCUSSION

In reported cases, cytokeratin-positive ductal structures have been reported to comprise 5% to 80% of the tumor. Our case was characterized by atypical pleomorphic cells shows immunoreactivity with IMP3 and negative PANCK, EMA, mCEA, CA19.9. LCA, CD20, CD30 and SALL4 negative rules out Lymphoma and Germ cell tumour. Other differentials diagnosis includes Neuroendocrine tumor and Melanoma and ruled out with negative Synaptophysin, Chromogranin, CD56, S-100 and HMB-45 respectively. Occasional giant cells were immunoreactive for CD68 and LCA. The differential diagnosis of pancreatic undifferentiated carcinoma with osteoclast-like giant cells includes cystic lesions, such as pancreatic cystadenomas, cystadenocarcinomas, serous and mucinous cystic tumors, and pancreatic pseudocysts, and solid pancreatic tumors, such as ductal pancreatic carcinomas or neuroendocrine tumors. Because of the rarity of this tumor, it is difficult to determine a treatment modality and the treatment of choice is surgical resection, if possible.18 UC-OGC has a more favorable prognosis with a 5-year survival rate of 59.1% compared to 15.6% for PDC. Median survival in study was 7.7 years compared to 1.6 years in patients with PDCs ($p<0.0009$). Recent case reports on this disease suggest that outcomes are improving, although survival is very short for some patients.

CONCLUSION

Undifferentiated carcinoma of pancreas is a rarely encountered neoplasm. This tumor can display various clinical characteristics, and its histogenesis is controversial. It is important to differentiate UCP from other pancreatic malignancies due to the relative improvement in prognosis. Due to its rarity, therapeutic guidelines are limited. Further studies may help establish treatment modalities and possible molecular biomarkers.

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