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"Pancreatic Undifferentiated Carcinoma with Osteoclast-Like Giant Cells (UC-OGC)" on Cytology: A Rare Case Report

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Abstract **Case Report**

Undifferentiated carcinoma of the pancreas with osteoclast-like giant cells (UC-OGCs) is a rare entity, less than 1% of all pancreatic malignancies. Recent studies quoted that the prognosis of these cancers is more favorable and displays a less aggressive with slow metastasis and lymph node spread compare to patients with an associated conventional pancreatic ductal adenocarcinoma. We report a case of undifferentiated carcinoma with osteoclast-like giant cells of pancreas, which was diagnosed on aspiration cytology. To the best of our knowledge only twelve cases have been reported on so far and only two cases reported on cytology.

Keywords: Undifferentiated cytology malignancies.

INTRODUCTION

Pancreatic neoplasms are relatively common gastrointestinal malignancies, with the most common type being adenocarcinoma of the pancreas. However there are several types of pancreatic cancer that are much less common, including pancreatic giant cell tumors (PGCT) which are rare non-endocrine tumors of the pancreas with an incidence of less than 1% of all pancreatic tumors[1]. They were first described in 1954 by Sommers and Meissner [2]. There are three types of pancreatic giant cell tumors: osteoclastic, pleomorphic, and mixed; however since 2010, the World Health Organization has grouped them together as undifferentiated carcinoma with osteoclast- like giant cells [3, 4].The osteoclastic variant has a better prognosis than the other two subtypes, as well as pancreatic adenocarcinoma [5]. PGCT usually affects patients in the 6th to 7th decade of life, with an equal male to female ratio [6]. PGCT mostly involve the body and tail of the pancreas, unlike pancreatic adenocarcinoma which mainly involves the head [6, 7]. GCT measures around 5–6 cm at presentation in 60–80% of cases [3]. Common clinical presentations of PGCT are nonspecific abdominal pain, distension and a palpable mass, whereas jaundice is the most common presentation of pancreatic adenocarcinoma.

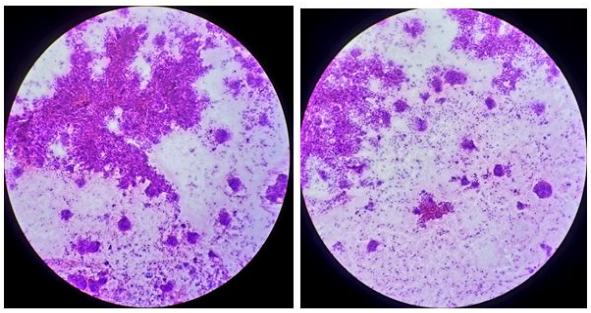
CASE PRESENTATION

A 72-year-old male presented to the surgery department with the complaints of upper abdominal pain since 6 months. The pain was insidious in onset with radiation to the back. There was no history of vomiting, jaundice or weight loss. Physical examination revealed a lump of 4 cm × 4 cm in the left hypochondrium extending to the epigastrium and umbilical region. Laboratory investigation revealed a normal level of amylase and lipase.

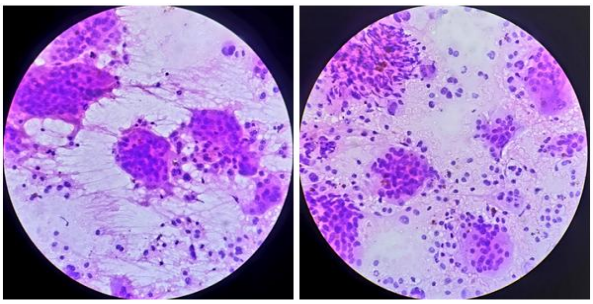
Ultrasound of the abdomen and pelvis which showed enlarged and diffusely edematous pancreas with altered texture and mixed echoic lesion measuring 3.8x4.6x4.8cm in anterior part of body and tail region of pancreas. Sonological findings were suggestive of Pseudo-pancreatic cyst. Computed tomography (CT) of the abdomen and pelvis revealed a 5.75x5cm heterogeneously enhancing lesion in neck and body of pancreas likely neoplastic pathology.

Ultrasound (US)-guided FNA was performed. Aspirate was highly cellular, composed of tumor cells arranged in collections exhibiting moderate degree of nuclear and cellular pleomorphism. The individual tumor cells have round to oval nuclei, fine granular chromatin, prominent nucleoli in some and moderate amount of cytoplasm with vacoulations at places. In addition, a large number of multinucleated osteoclast-

like GCs seen. Overall morphology suggested UC-OGC.



Fine-needle aspiration cytology shows combination of pleomorphic malignant cells with admixed multinucleated osteoclast-like giant cells with hemorrhagic background. Malignant cells demonstrate moderate nuclear pleomorphism.



Fine-needle aspiration cytology shows multinucleated osteoclast-like giant cells.

DISCUSSION

Undifferentiated carcinoma of the pancreas is a rare, aggressive tumor. A variety of other terms has been used to describe this; however, a duct epithelial origin is now established and recognized as a variant of pancreas ductal adenocarcinoma (PDAC) [8]. These tumors can be generally subdivided into two categories: Pure osteoclast-like GC tumors and those with a component of a more conventional neoplasm. Most often, the more conventional component is a PDAC, although intraductal papillary mucinous neoplasm, mucinous cystadenocarcinoma, and PanIN-3 have also been reported. Notable, studies have found that patients with pure UC-OGC survived longer than patients with an associated conventional PDAC [9, 10]. Muraki *et al*. also reported a trend toward better survival for patients with pure UCOGC [11]. Their case series support a favorable prognosis with a 5-year survival rate of 59.1%, and median survival of 7.67 years, which was incomparably better than that of PDACs (15.6% and 1.59 years). Thus, a lack of ductal differentiation seems to predict an improved prognosis. The clinical symptoms varies from abdominal pain, loss of appetite, nausea, and steatorrhea to rarely asymptomatic individuals diagnosed incidentally. The body and tail of the pancreas are the most common sites for UC-OGCs, however, they can arise from any portion of the pancreas. The size ranges from 3 to 12 (mean – 5 cm). Despite being larger tumors, they appear to be less invasive than PDACs, with less likelihood of perineural invasion and nodal metastasis. Tumor markers, CEA and CA 19-9, are indistinct as they are less commonly elevated and not significant to arriving at a diagnosis. The diagnosis is usually made on a surgical specimen. In some cases, EUS-FNA has shown to be effective, and an accurate diagnosis had been achieved by cytology test [12, 13]. Cytological findings for pancreatic aspirate composed of an obviously malignant cellular proliferation containing benign appearing osteoclast-like GCs. Smears are typically hypercellular with two cell populations; atypical mononuclear cells and osteoclast-like GCs. The mononuclear cells appear singly or in small clusters and range from medium-sized polygonal epithelioid cells with clear cytoplasm to large bizarre sarcomatoid cells with dense and/or spindled cytoplasm [14]. The mononuclear cells intermingled with the OGCs which may be few in number or can be numerous. The GCs often contain 10 or more bland appearing, centrally clustered, and slightly overlapping nuclei with even chromatin and occasionally prominent nucleoli. The cytoplasm is abundant and dense and may contain phagocytic material [11]. Histological examination demonstrates it is composed of 3 cell types-Mononuclear neoplastic cells, Mononuclear histiocytes (nonneoplastic) and Multinucleated giant cells (osteoclast-like giant cells) (nonneoplastic).

In the majority of reports, the mononuclear cell population expresses immunostaining for epithelial membrane antigen and keratin which demonstrates an epithelial origin. Conversely, epithelial markers are rarely express by osteoclastic GCs and show staining consistent with a histiomonocytic origin (CD68) [11]. Genetically, UC-OGC of the pancreas resembles PDAC, with both entities harboring frequent KRAS, TP53, CDKN2A, and SMAD4 mutations [11, 15, 16]. However, the low proliferative index of the GCs by Ki67 staining and their lack of driver mutations, it is safe to conclude that these osteoclastic cells are innocent recruits to this peculiar neoplasm [17, 18]. Metastases are less likely in UC-OGC and when they do occur, it is through lymphatics or direct peritoneal extension. Accordingly, early diagnosis and complete surgical resection represent the best chance to cure this rare tumor. However, the effectiveness of chemotherapy and radiotherapy is not established.

CONCLUSION

UC-OGCs of the pancreas are a distinct and a rare entity, less than 1% of all pancreatic malignancies. UC- OGCT has a less aggressive course with slow metastasis and lymph node spread with a better prognosis. Because of this significant difference in clinical behavior and prognosis, it is important to identify and distinguish between the subtypes of PGCT.

Cytology plays a very pivotal role in diagnosing it which significantly impacts prognosis and patient care.

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