Case Report

Acinar cell cystadenoma of pancreas: A case report and review of literature

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ABSTRACT

Acinar Cell Cystadenoma (ACA) of pancreas is recently recognized pancreatic lesion by the World Health Organization (WHO) in 2010. It is a very rare benign cystic neoplasm of pancreas that only few cases (<100) have been reported in literature. Because of rare occurrence, the pathogenesis is not properly understood. Histologically it is composed of variably sized cystic spaces lined by mature acinar cells, admixed with normal pancreatic parenchyma. Here we present the histopathology of a case of acinar cell cystadenoma with review of literature.

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1. Introduction

Cystic pancreatic lesions can either be the simple cysts or cystic pancreatic neoplasms. Cystic pancreatic neoplasms can be of serous, mucinous or pseudopapillary or acinar cell type. Acinar cell cystadenoma (ACA) of pancreas is recently recognized by the World Health Organization (WHO) in 2010. Albores-Saavedra first reported the case of acinar cell cystadenoma in an autopsied specimen of pancreas in 2002. It is a rare benign cystic neoplasm of pancreas, representing around 2% of pancreatic neoplasms. As it is benign and shows acinar cystic transformation it is called as acinar cell cystadenoma. Acinar cell cystadenomas are benign cystic lesions with acinar differentiation without any atypia.

2. Case Presentation

A 21 year old male presented to Gastroscopy OPD of VS General Hospital with complaints of abdominal pain since one and half months. Abdominal examination

reveals tenderness in left hypochondriac region. Abdominal ultrasound examination revealed a cystic tumor involving head and uncinate process of pancreas. Patient was operated for Whipple procedure and specimen was sent for histopathological examination.

2.1. Histopathological examination

Specimen of Whipple procedure was received in seven parts. Largest one consisted of pancreatic head with part of stomach and duodenum, total measuring 35x5 cm in largest dimensions. The part of pancreas was measuring 4x3.5x3 cm. On cut section of pancreas area with multiple small cysts was identified measuring 3x3 cm in diameter (Figure 1) The other six portions were consisted of a gallbladder and lymphnodes.

Microscopic examination from pancreas show tumour to be composed of variable sized cysts (Figure 2) lined by the bland flattened cuboidal cells with granular eosinophilic cytoplasm (Figure 3). Cyst show focal mucinous epithelium and dense eosinophilic lamellar concentrations (Figure 4). Ductal epithelium admixed with acinar cell was seen (Figure 5). No evidence of cytological atypia or pancreatic

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Fig. 1: Grossly the cut section of Pancreas showing multiple cysts

intraepithelial neoplasm was seen. Resected surgical margins were free from tumour. Sections from lymphnodes reveals changes of nonspecific lymphadenitis. Considering the findings diagnosis of acinar cell cystadenoma of pancreas was given.

Fig. 2: Scannview showing variable sized cysts

3. Discussion

Cystic pancreatic lesions (CPLs) are now more frequently diagnosed due to high quality cross-sectional imaging studies.¹ Cystic pancreatic lesions can be non-neoplastic cysts or pancreatic cystic neoplasms (PCNs). According

Fig. 3: High power view showing acinar cells with granular eosinophilic cytoplasm

Fig. 4: Low power view showing acinar eosinophilic lamellar concentrations

to older data pancreatic pseudocysts are more common CPLs whereas recent data suggests that pancreatic cystic neoplasms are more common.²⁻⁴ The non-neoplastic cysts of pancreas are pancreatic pseudocysts (PPSs), lymphoepithelial cysts, enterogenous cysts. The four most common pancreatic cystic neoplasms are serous cystic neoplasm (SCN), mucinous cystic neoplasms (MCN), intraductal papillary mucinous neoplasm (IPMN) and solid pseudopapillary neoplasms.⁵ Acinar cell cystadenoma is very rare, newly recognized benign cystic pancreatic neoplasm.

Fig. 5: Ductal epithelium admixed with acinar cells

Conventionally it was believed that acinar neoplasms in the pancreas are always malignant.⁵ Albores-Saavedra in 2002, first reported a previously undescribed pancreatic cyst with acinar cell differentiation of the lining epithelium without cellular atypia and malignant features, as acinar cell cystadenoma in contrast with acinar cell cystadenocarcinoma.⁶ There are only few case reports of ACA in the literature.^{6,7} Age of presentation is 9-71 years (mean-42) with slight female predominance.⁷⁻¹⁰ Acinar cell cystadenomas clinically present as abdominal pain, discomfort or jaundice and sometimes found incidentally. Most cases involve the head of the pancreas.⁷ Grossly they are multiple and well circumscribed.⁷ Their cut surface shows multiple variable sized small cysts. Histologically acinar cell cystadenomas are composed of cysts lined by single or several cell layers of cuboidal cells with round, basally located nuclei and eosinophilic, periodic acid stain (PAS) positive granules in apical region of cytoplasm. Mitoses are rarely observed.^{7,9} Immunohistochemistry of ACA is similar to normal acinar cells. It is positive for pancreatic enzymes (Chymotrypsin, Trypsin, and Lipase) and epithelial markers such as AE1, AE3, and CAM 5.2. Unlike non neoplastic acinar cells, cells of acinar cell cystadenoma are CK 7 positive.^{7,11,12} MIB index is less than 1%.⁷

The etiopathogenesis of acinar cell cystadenoma is not established, but now based on results of immunohistochemistry it is assumed that acinar metaplasia could be the main pathology.⁹ Polyclonality of the X chromosome suggests that ACAs are benign non-neoplastic ballooning degeneration of acinar epithelium.¹¹ In contrast, Khor et al have suggested that ACAs are neoplastic because there is augmentation in number of specific genes.¹² No cases are reported with recurrence or malignant transformation of acinar cell cystadenoma in literature.

4. Abbreviations

ACA- Acinar Cell Cystadenoma
MCN- Mucinous Cystic Neoplasm
SCN- Serous Cystic Neoplasm
PAS- Periodic Acid Stain

5. Source of funding

None.

6. Conflict of interest

None.

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