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Mucinous Cystic Neoplasm of Pancreas: A Rare Case Report

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Abstract
Mucinous cystic neoplasms of the pancreas are defined by the world health organization (WHO) as cystic epithelial neoplasms that occur almost exclusively in women. Mucinous cystic neoplasms account for 5-7% of primary pancreatic tumors, with a female-to-male ratio of 20:1 and with a mean age between 40 and 50 years. Approximately 70–90% of mucinous cystic neoplasms arise in the body or tail of the pancreas.
Case Description: *A-38-years female patient presented with epigastric pain for 3 months. There was no palpable mass. Computer tomography revealed a well-defined thick-walled, cystic lesion in the body and tail region of the pancreas measuring 50x50x45 mm. This mass was extending into the lesser sac and closely abutting the lesser curvature of the stomach. The radiological impression was a pancreatic pseudocyst. Ultrasound guided-FNAC was performed but it was inconclusive. The mass was excised with the tail of the pancreas and spleen. The specimen was sent for Histopathological examination for confirmation of diagnosis.*
Conclusion: *Mucinous cystic neoplasm is a rare entity and it is very important to distinguish it from the Intrapapillary ductal neoplasm and solid pseudopapillary neoplasm of the pancreas. The presence of ovarian stroma with focal mucinous lining helps to clinch the diagnosis and one should carefully look for dysplasia.*

Introduction

Mucinous cystic neoplasms of the pancreas are defined by the World Health Organisation (WHO) as cystic epithelial neoplasms which can be unilocular or multilocular that occur almost exclusively in women. Mucinous cystic neoplasms account for 5-7% of primary pancreatic tumors, with a female-to-male ratio of 20:1 and with a mean age between 40 and 50 years. Approximately 70–90% of mucinous cystic neoplasms arise in the body or tail of the pancreas [(1-2)]. MCNs of the pancreas are characterised by cystic epithelial cavities composed of columnar mucin-producing epithelium. This tumor occurs predominantly in women and there is no communication with the pancreatic duct system. These tumors are classified according to the grade of dysplasia as adenoma, borderline, and non-invasive or invasive carcinoma. MCNs are characterized by two distinct components: 1) An inner mucinous epithelial layer and, 2) an Outer dense cellular ovarian-type stromal layer[(3-8)] Histologically, the cystic spaces seen in this neoplasm are lined by a single layer of mucin-producing columnar cells. The underlying stroma is generally hypercellular and forms a characteristic, ovarian-like fibrillary tissue, with nuclear expression of estrogen receptors and/or progesterone receptors.

Case Report

A 38 years old female patient presented with epigastric pain for 3 months. There was no palpable mass. Computer tomography revealed a well-defined thick-walled, cystic lesion in the body and tail region of the pancreas measuring 50x50x45 mm. (Figure 1a)

This mass was extending into the lesser sac and closely abutting the lesser curvature of the stomach. The radiological impression was a pancreatic pseudocyst. Ultrasound guided-FNAC was performed but it was inconclusive. The mass was excised with the tail of the pancreas and spleen. (Figure 1b)

The specimen was sent for Histopathological examination for confirmation of diagnosis. Microscopic examination reveals showed a cyst with predominantly denuded epithelium, and was focally lined by cuboidal to columnar epithelium. The epithelial cells at places showed the presence of intracytoplasmic mucin with basally located nuclei. The wall had a densely cellular stroma (ovarian-like). (Figure 2a,b,c,d) No significant cytological atypia /nuclear stratification/nuclear crowding was noted.

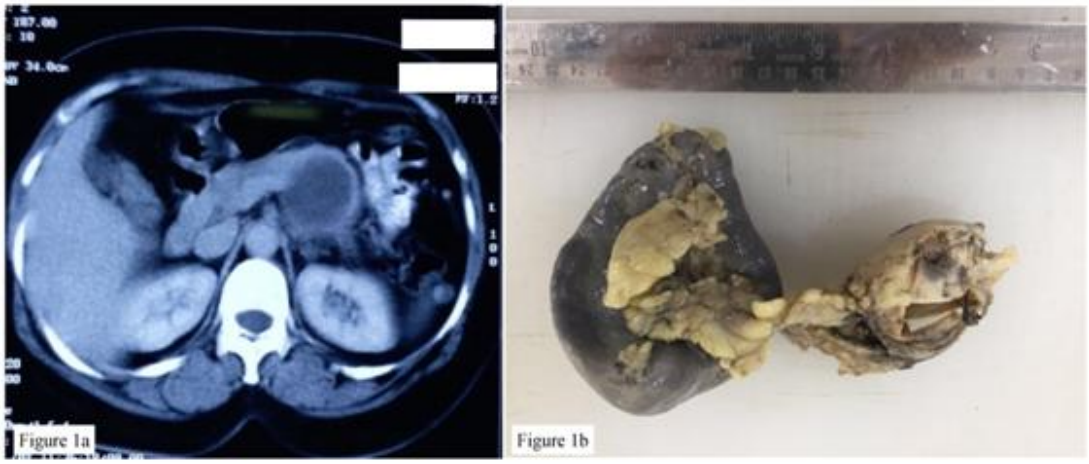

Fig 1: a) CT scan revealed a well-defined thick walled, cystic lesion in the body and tail region of the pancreas measuring 50x50x45 mm. b) Pancreatectomy with splenectomy specimen

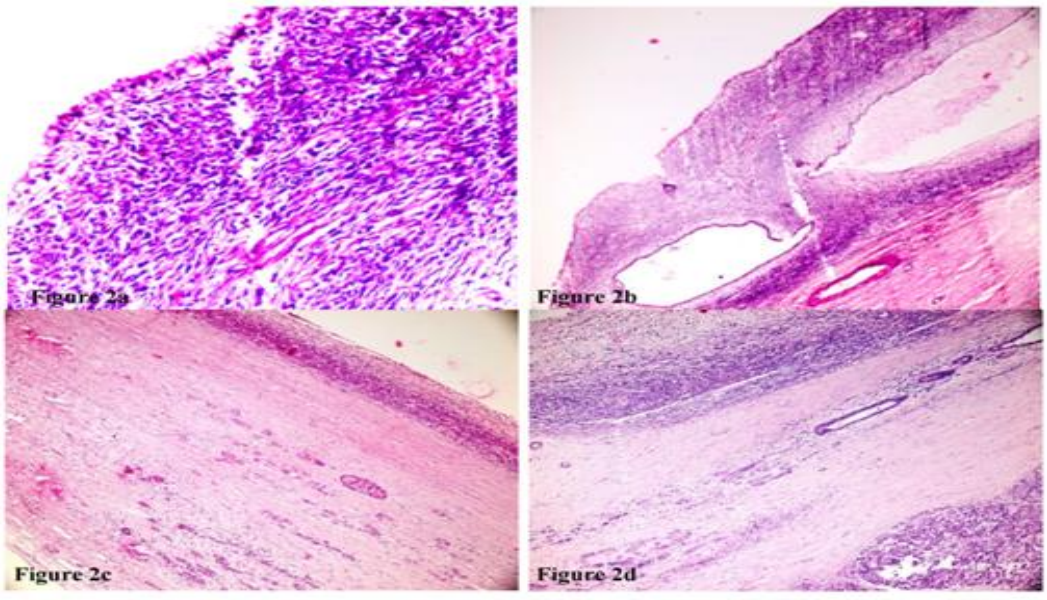

Fig 2: a&b)H&E stained smears shows a cyst lined by columnar epithelium(2AX400,2BX200). c)H&E stained smear shows a cyst lined by cuboidal epithelium(X200). d) H&E stained smears showing the wall of cyst with dense cellular ovarian like stroma along with normal pancreas(X200)

Discussion

There are different pancreatic cystic lesions that occur in the pancreas[9]

Pseudocyst of pancreas
Serous cystadenoma
Mucinous cystic neoplasms
Intraductal papillary mucinous neoplasm
Solid pseudopapillary neoplasm

Mucinous cystic neoplasm needs to be differentiated from other cystic lesions of the pancreas. The most common cystic lesion is a Pseudocyst of the pancreas and the rarest is mucinous cystic neoplasms. Less than 20% of MCN are associated with invasive carcinoma.[1-2]

In the present study, the pathological finding showed a cyst with predominantly denuded lining. Mucinous epithelium could be identified focally. The epithelium was supported by a conspicuous ovarian-like stroma. The overall features were consistent with a mucinous cystic neoplasm of the pancreas. These findings help in distinguishing from other cystic lesions of the pancreas. The main differentials, in this case, include intra-ductal papillary mucinous neoplasm and solid pseudopapillary neoplasm[9]. The present tumor had ovarian stroma with focally mucin-secreting epithelium. This stroma showed overgrowth of epithelial component, hence benign mesenchymal tumor-like inflammatory myofibroblastic tumor or mucinous-cystadenoma with sarcomatous component has to be distinguished. MCN with sarcomatous stroma is a rare variant often associated with a malignant sarcomatous stroma. This sarcomatous stroma is extremely hypercellular showing marked atypia and pleomorphism with numerous mitotic figures[10]. Unlike the sarcomatous stroma, the stroma of the present case did not show any significant cellular atypia or mitoses. Moreover, the inflammatory myofibroblastic tumor can be excluded from the viewpoint that no significant mixture of chronic inflammatory cells was seen in the present case. There are recent studies that argue that since pancreatic MCNs are often asymptomatic and discovered incidentally, have exceptionally low rates of malignant transformation when they are less than 4 cm in size[12]. One recently published study stated that MCNs could be easily distinguished from pancreatic Intraductal Papillary Mucinous Neoplasms (IPMNs) by their clinical and demographic, radiologic, and pathologic features. The study showed that most MCNs are slow growing and non-invasive, are cured by surgical resection, and have an excellent prognosis (even in cases with invasive disease) with an 80% 10-year survival rate following resection[13]. With the advancement in radiological imaging increasing numbers of cystic pancreatic neoplasms are now being diagnosed. Numerous ongoing current studies are attempting to identify tumors that may be treated non-surgically[14]. Since the accurate diagnosis of MCNs and their differentiation from other pancreatic cystic neoplasms is crucial for the accurate management of these tumors, novel biomarkers, and molecular diagnostic tools which can differentiate between the cystic pancreatic lesions need to be researched. These may prove very useful in such differentiation and facilitate early and accurate diagnosis[15]. Although so far, no definite molecular markers have been identified, a search is actively underway. It is important to emphasize that the pathologist has a critical role in both the preoperative assessment of pancreatic cystic neoplasms as well as in the accurate postoperative diagnosis and thus in determining the prognosis, further treatment, and follow-up of pancreatic cystic neoplasms including MCNs[16].

Conclusion

According to the current WHO classification of the pancreas, this case belongs to mucinous cystadenoma which is a rare entity and has to be differentiated from the other cystic neoplasm and one should always look for dysplasia in each of the cases. Since these tumors are rare, premalignant, and have strict diagnostic criteria, they must always be considered in the differential diagnosis of pancreatic mucinous cystic lesions.

Larger studies incorporating a greater number of patients and more detailed follow-ups need to be done.

Conflict of Interest

There is no conflict of interest among the authors in our case report.

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