Intraductal Papillary Mucinous Neoplasm of the Pancreas

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PRESENTATION Of CASE

A 36-year-old non-smoker, chronic alcoholic female presented with recurrent episodes of epigastric pain and vomiting in the last 10 days. H/o similar episodes of pain 3 times, with last episode was noted 2 months back. She also had history of vomiting (3 episodes). Jaundice/melena/steatorrheas was not observed.

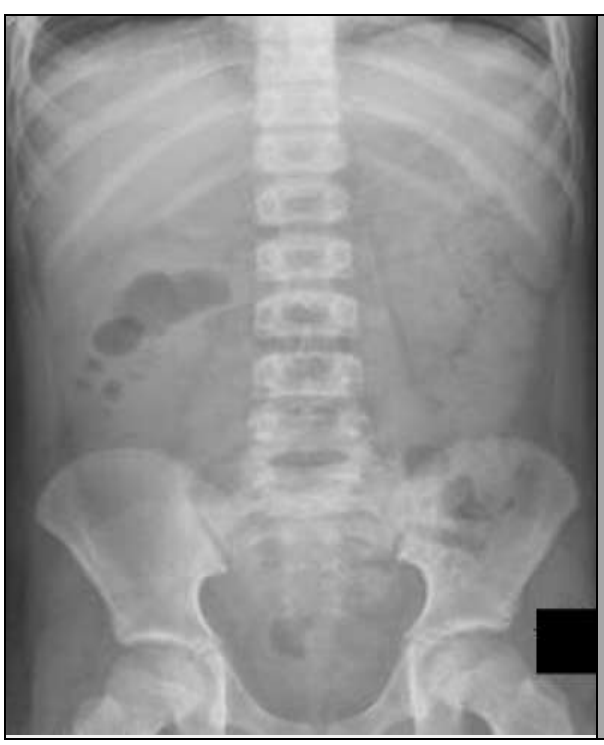

Figure 1. Erect Abdominal Radiograph Shows Normal Features

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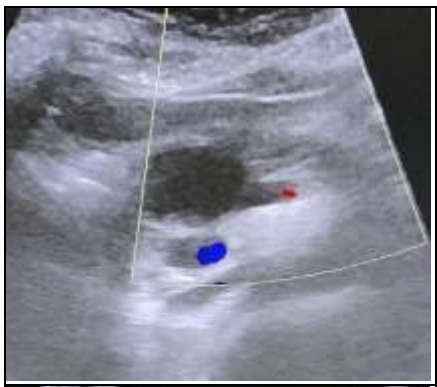


Figure 2a. Few Well Defined Anechoic Cystic Lesions are Noted in Head and Proximal Body of Pancreas

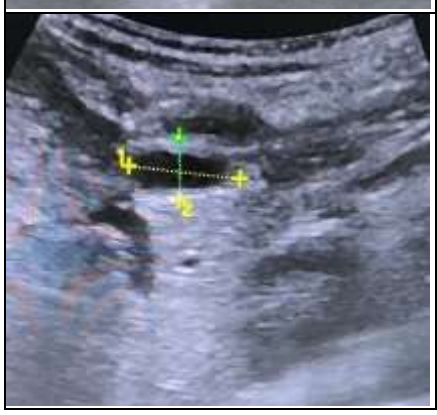


Figure 2b. One of the Lesion is Seen Communicating with Dilated Main Pancreatic Duct

Subsequently, contrast-enhanced computed tomography (CECT) of the patient was done and it showed well defined multiple cystic lesions with peripheral wall enhancement involving head and body of pancreas, largest measuring 24 X 22 X 22 mm in the head of the pancreas, the lesion was seen abutting antro-pyloric region anteriorly. Main pancreatic is mildly dilated measuring 4 mm in diameter. A well-defined heterogeneously hypodense (necrotic) para duodenal lesion, was noted inferior to uncinate process, m/s 22 X 20 X 20 mm, which is indicative of lymph nodal deposit.

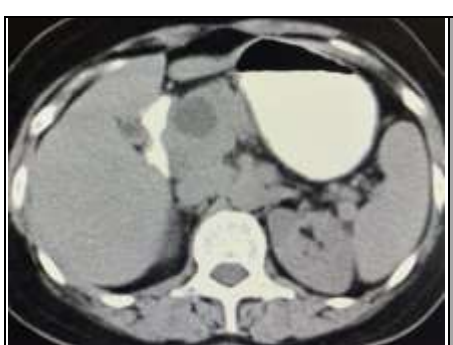


Figure 3a. Axial Sections of CT Plain Showing Well-Defined Cystic Lesions in Pancreatic Head

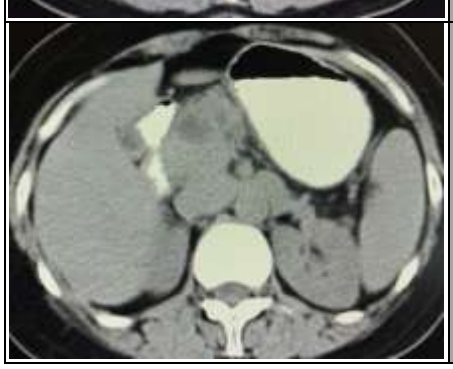
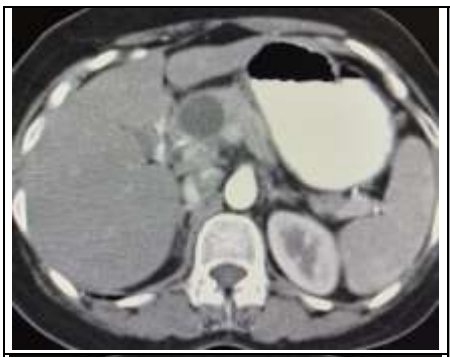


Figure 3b. Proximal Body



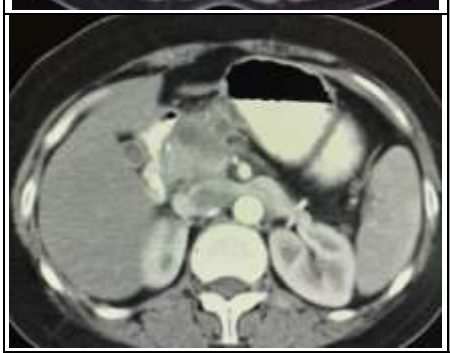
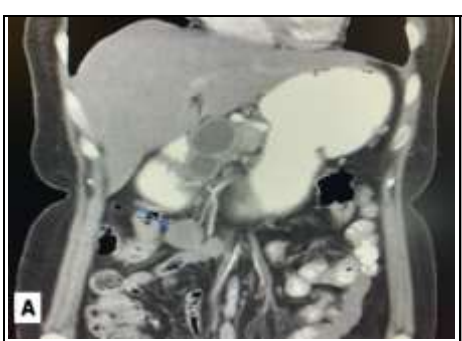


Figure 4a & b. Axial Sections CECT in Arterial Phase Showing Multiple Cystic Lesions with Subtle Wall Enhancement in Pancreatic Head and Proximal Body

Figure 4c. Coronal CECT Sections Shows the Cyst in Head of Pancreas



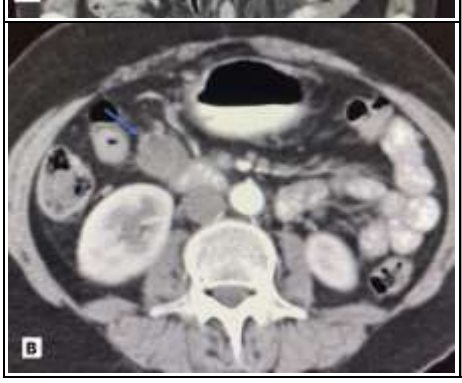


Figure 5a & b. Axial and Coronal Sections CECT in Arterial Phase Showing Para Duodenal Lymph Nodal Deposit (Blue Arrow)

On magnetic resonance cholangiopancreatography (MRCP): few cystic lesions were seen scattered in the pancreatic parenchyma, with one of the cysts showing communication with main pancreatic duct (MPD) ab.

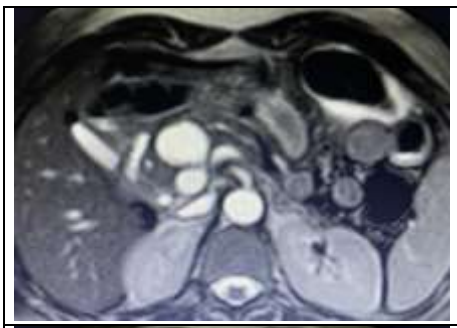


Figure 6a. Showing the Communication of the Cyst with MPD,

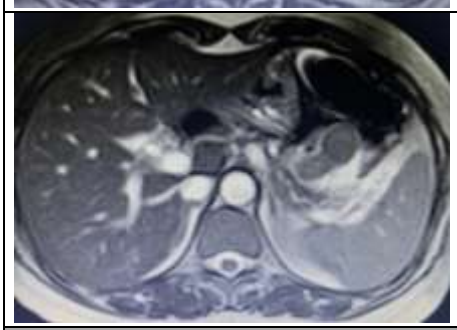


Figure 6b. Showing Mild Dilated MPD

Figure 6. T2 Axial Sections

PROVISIONAL DIAGNOSIS

Main duct intraductal papillary mucinous neoplasm of pancreas

DIFFERENTIAL DIAGNOSIS

- Pancreatitis with pseudo cyst.
- Mucinous cyst adenoma of pancreas.

PATHOLOGICAL DISCUSSION

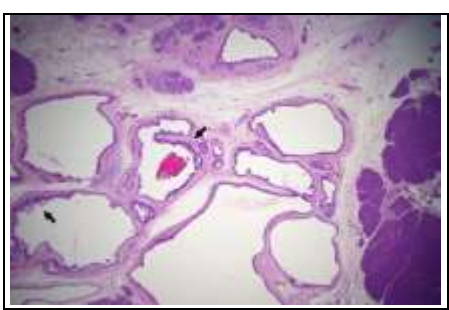


Figure 7a. Multiple Clusters of Cyst with Flattened Epithelia and Focal Micro Papillae (Arrow)

Figure 7b. Multiple Villous Structure Noted to Protrude into Cyst Lumen

- Histopathological microscopy section showed oncocytic cells with scattered goblet cells arranged in a complex papillary configuration with anatomizing fronds and solid areas showing cribriform appearance.
- Inter and intra cellular luminal spaces had mucinous material within.
- The nuclei within the cells are basally placed, small, round with eccentrically placed nucleoli and cells showed more of granular and eosinophilic cytoplasm.
- Dilated ducts lined by low grade mucinous columnar cells were found in adjacent ducts consistent with low grade pancreatic intraepithelial neoplasia.

DISCUSSION

Pancreatic cancer is the fourth most common cause of cancer-related mortality worldwide, with an incidence rate equalling that of its mortality rate.[1] There are numerous primary pancreatic neoplasms, in part due to the mixed endocrine and exocrine components. Previously thought to be rare, intraductal papillary mucinous neoplasms (IPMNs) now constitute 20 - 30 % of pancreatic cystic neoplasms.[2] IPMN is similar to chronic pancreatitis in presentation with symptoms such as relapsing abdominal pain, steatorrhea, pancreatitis and radiological findings of cystic lesions communicating with dilated pancreatic duct that can be misdiagnosed as chronic pancreatitis with pseudocysts. Pancreatic pseudocyst may simulate clinically and radiologically with other cystic neoplasms of the pancreas.[3]

IPMNs have been classified as branch duct (BD) type, main duct (MD) type, and combined or mixed type based on imaging findings. Computed tomography (CT) and magnetic resonance imaging (MRI), along with magnetic resonance cholangiopancreatography, are the most useful radiologic methods for detecting IPMNs.[4] Moreover, these imaging modalities are useful in distinguishing BD-IPMNs from other cystic lesions by showing multiplicity and a connection to the main pancreatic duct (MPD). Study results showed that MR imaging with MRCP showed acceptable diagnostic performance in the prediction of the malignant potential of IPMNs when using the image criteria of the international consensus guidelines 2012.[5]

There has been significant increase in the incidence of IPMN in the last decade and this increase is most likely due to a combination of factors.[6,3] The grouping of previously autonomous tumours, such as mucin producing tumours of the pancreas, papillary carcinomas and villous adenoma, as well as improved diagnostic imaging undoubtedly has a central role here.[6,3] Although thought to be benign pancreatic IPMN is potential preneoplastic cystic lesion of the pancreas with a number of radiological findings that favour malignancy called as "worrisome feature" and "high-risk stigmata" groups.[7] Four subtypes of IPMN are currently recognized: gastric, intestinal, oncocytic and pancreatico-biliary. This carcinoma resembles conventional ductal adenocarcinoma in both morphology, diagnosis and prognosis. The classical type of malignancy accompanying IPMN is pancreatic ductal adenocarcinoma (PDAC). The frequency of PDAC occurrence has been reported with a spectrum of 1.9 – 9.2 % in different literature data.[8] Although unrecognized at the time of

cholangiopancreatography, the finding of a 'bulging papilla with mucoid secretion' is nowadays referred to as the 'fish-eye sign', which, although observed in only about 25 % of cases, is considered by some authors to be pathognomonic for IPMN.[9] IPMNs appear to have distinct epidemiology and risk factors. Unlike typical chronic pancreatitis, patients are usually older women, smoke fewer cigarettes and consume less alcohol.[10,11]

Surgery remains the treatment of choice with excellent survival at 5 years in localized disease (70 – 100 %). Survival in patients with advanced disease is more modest and roughly estimated at 40 % in IPMN with an invasive component.[12,13]

Radiological Features-BD-IPMNs have been found to manifest as either grape-like clusters of cysts or cysts without dilatation of the MPD and usually located at the pancreatic head, especially the uncinate process.[14] Of patients with BD-IPMN, 39 % - 64 % have been found to show multiplicity.[15,16]

Identification of the communication between the cystic lesion and the MPD is one of the most reliable findings for diagnosis of BD-IPMN.[4] MRI with MRCP has shown better diagnostic performance than CT in differentiating IPMNs from other pancreatic cystic lesions by showing the ductal communication of cystic lesions with MPD.[17] CT falls short of MRCP in detecting a ductal connection, estimating main duct involvement, and identification of small branch duct cysts.[19]

MD-IPMNs appear as dilated and tortuous MPDs, with the dilatation extending into the secondary branches that sometimes appear cystic.[3] MD-IPMNs show a much higher malignant potential than BD-IPMNs (40 % - 95 % vs 12 % - 62 %).[17] According to recent International Association of Pancreatology guidelines 15, the threshold of MPD for characterization of MD-IPMN has been lowered to > 5 mm in the absence of other causes of obstruction. Dilatation of MPD around 5 - 9 mm is a "worrisome feature", while diameters ≥ 10 mm are considered "high-risk stigmata". However, the dilatation of MPD in MD-IPMN can be due to mucin, protein plugs, or focal pancreatitis without neoplastic involvement.[18]

Intralesional solid components can be observed, with calcification occurring in about 11 % of these patients.[19] Dystrophic calcifications can be seen in the mucous material in dilated MPDs.[19] Endoscopic retrograde cholangiopancreatography can show filling defects, representing tumours or inspissated mucus, within the dilated duct.[20]

Combined or mixed IPMN involves both the MPD and its BDs, with CT and MRI showing features of MD-IPMNs and BD-IPMNs.

FINAL DIAGNOSIS

Main duct intraductal papillary mucinous neoplasm of pancreas

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