IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 17, Issue 01 Ver. VII January. (2018), PP 33-39
www.iosrjournals.org

Intraductal Papillary Mucinous Neoplasm of Pancreas An Incidental Finding- A Case Report.

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Abstract
Background :*Intraductal papillary mucinous neoplasms of pancreas are cystic lesions which are premalignant and characterised by cystic dilatation of pancreatic ducts.Most of the cases are asymptomatic and found incidentally. . Intraductal papillary mucinous neoplasms currently accounts for 20-50% of all cystic pancreatic lesions.It is important to distinguish this lesion from mucinous adenocarcinoma of pancreas as IPMNs has excellent prognosis than the latter.We present a case report of intraductal papillary mucinous neoplasm found incidentally.*
CaseReport:*70 year old male presented with difficulty in breathing for 3 months which was post traumatic. MRI abdomen revealed pancreatico-pleural fistula extending from tail of pancreas to pleural space through defect in dome of diaphragm. Subsequently distal pancreatectomy with splenectomy was done. Histopathological examination revealed an intraductal papillary mucinous neoplasm –foveolar subtype which was confirmed by immunohistochemistry.*
Conclusion:*Intraductal papillary mucinous neoplasms are cystic tumors of the pancreas that are diagnosed increasingly often. It is a diagnosable precursor of pancreatic cancer especially in symptomatic intraductal papillary mucinous neoplasm.*
Keywords : *cystic lesions of pancreas,histologic subtypes, intraductal papillary mucinous neoplasm.*

Date of Submission: 26-12-2017 Date of acceptance: 16-01-2018

I. Introduction

Intraductal papillary mucinous neoplasms of pancreas are cystic lesions which are premalignant and characterised by cystic dilatation of pancreatic ducts[1,2].Most of the cases are asymptomatic and found incidentally. True incidence is not known because many intraductal papillary mucinous neoplasms are small and asymptomatic .Typical age presentation is in the fifth to seventh decade with male predominance . [2]The male to female ratio for main duct intraductal papillary mucinous neoplasm is about 1.1 to 3:1 and for branch duct intraductal papillary mucinous neoplasm is about 0.7 to 1.8 :1 [3].Intraductal papillary mucinous neoplasms currently accounts for 20-50% of all cystic pancreatic lesions[4]It is important to distinguish this lesion from mucinous adenocarcinoma of pancreas as IPMNs has excellent prognosis than the latter[4].We present a case report of intraductal papillary mucinous neoplasm found incidentally.

II. Case Report

70 year old male presented with difficulty in breathing for 3 months which was post traumatic. MRI abdomen revealed pancreatico-pleural fistula extending from tail of pancreas to pleural space through defect in dome of diaphragm. Subsequently distal pancreatectomy with splenectomy was done.2.1 GROSS: Received distal pancreatectomy with splenectomy specimen with pancreas measuring 6x5.5x2.5cm and spleen measuring 10x6x4 cm.Cut surface of pancreas showed a fistulous tract and numerous dilated ducts . One duct showed a grey white mass with tiny papillary excrescence measuring 1.5x1x0.5 cm.Cut surface of spleen showed congestion.(Fig 1 and 2).

Fig 1: distal pancretectomy with splenectomy.

Fig 2: grey white mass with papillary excrescences.

2.1 Microscopic Features

Histopathology showed pancreatic parenchyma with a neoplasm arising in the pancreatic duct.The neoplastic cells were arranged predominantly in papillary and focally in glandular pattern with foveolar epithelial lining.The cells are columnar with moderate eosinophilic cytoplasm, some cells showed apical mucin.The papillae showed orderly nuclear stratification .Stroma shows lymphoplasmacytic and neutrophilic infiltrates.(Fig 3,4,5 and 6). Fistula showed a tract lined by non specific inflammatory cell infiltrates.

Splenic parenchyma showed congestion.

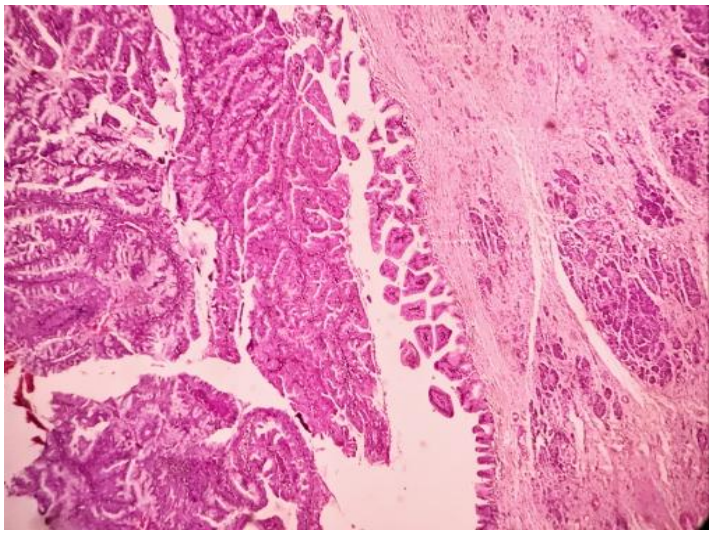

Fig:3 :pancreatic parenchyma with neoplasm,
H and E (40 X).

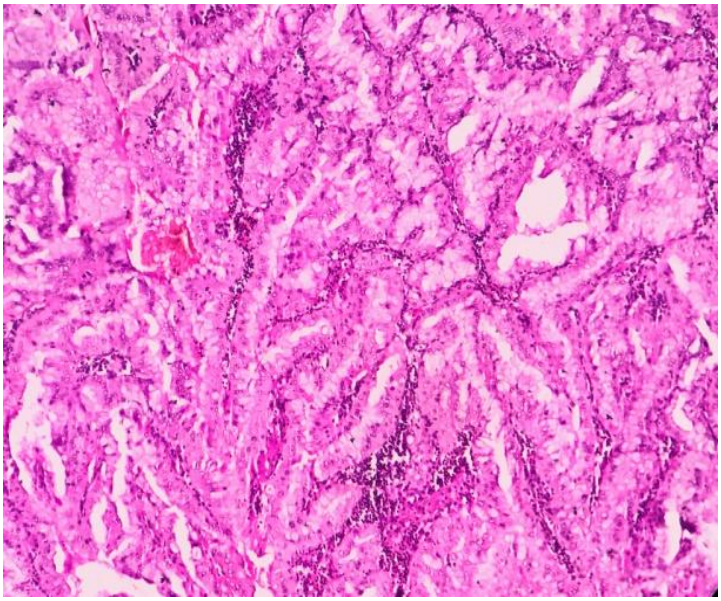

Fig :4: neoplastic cells arranged in papillary pattern.
H and E (100 X).

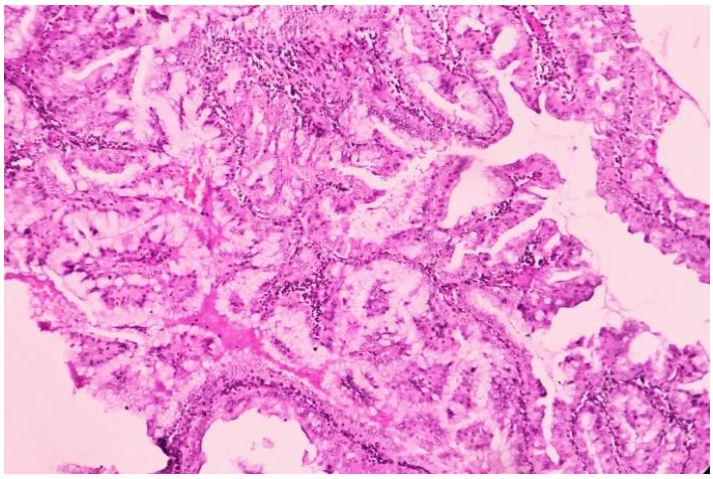

Fig :5: neoplastic cells arranged in focal glandular pattern.
H and E (100 X).

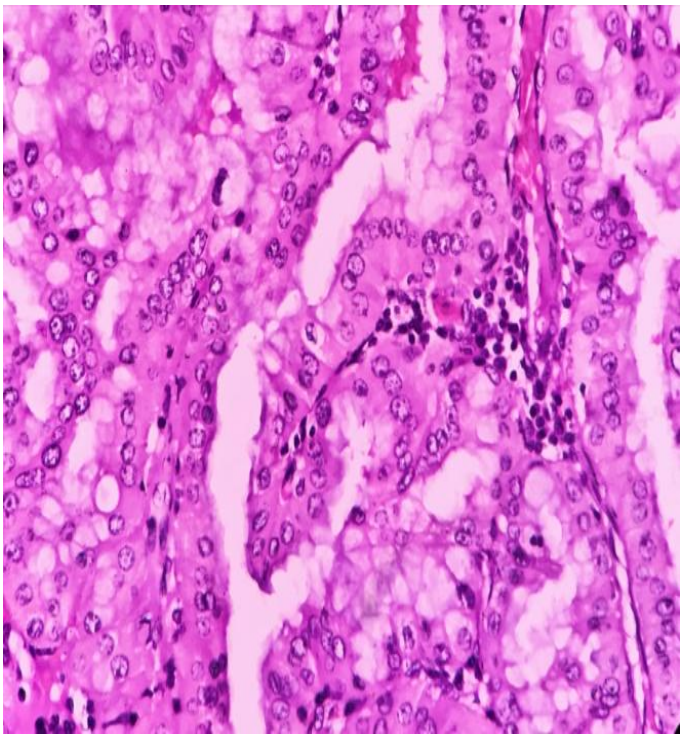

Fig :6 : columnar cells with moderate eosinophilic cytoplasm with apical mucin (H AND E)-400 X

2**.3 Immunohistochemistry** : immunohistochemistry showed CK 7 - focal cytoplasmic and membranous positivity(fig 7)

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Muc 5AC –positive(fig :8), CK 20-negative (fig :9)and CA19.9 –negative(fig :10) .

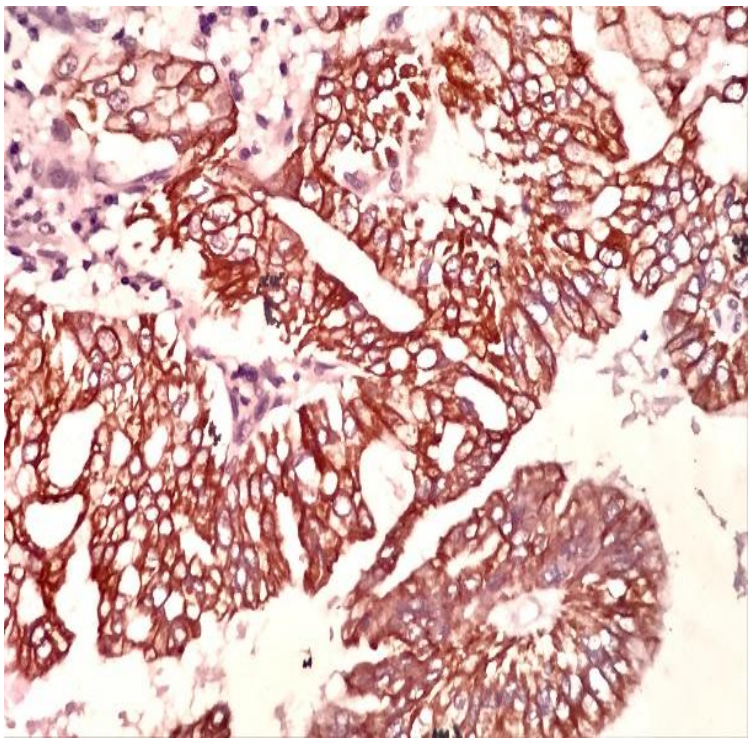

Fig :7 CK 7-cytoplasmic and membranous positive. 40x

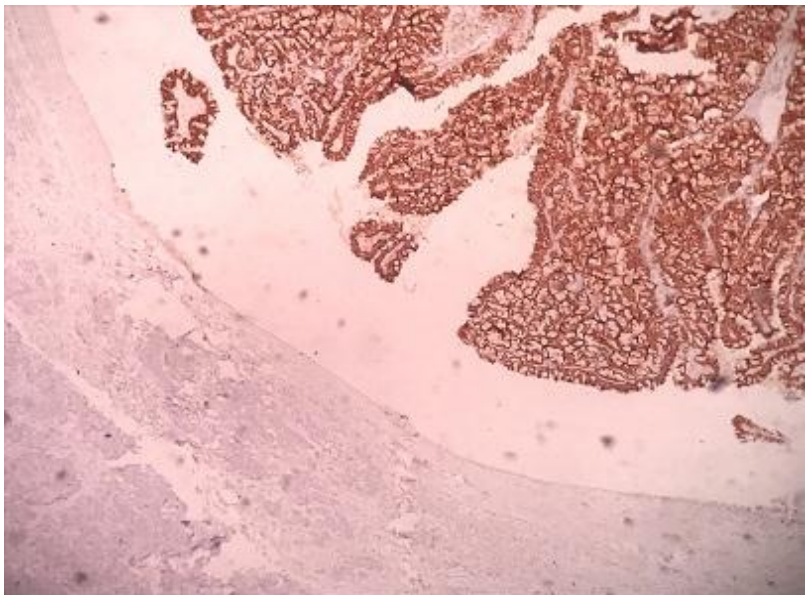

Fig:8: Muc 5 AC-positive .40X.

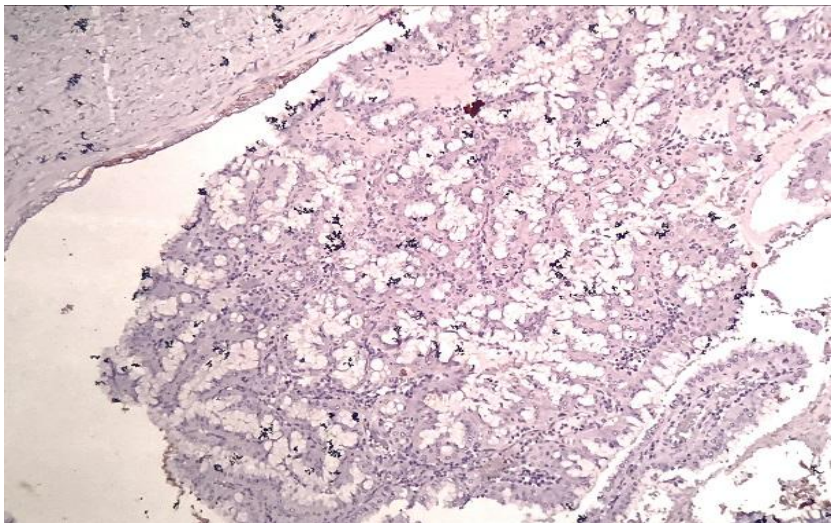

Fig 9: CK 20- negative .40X.

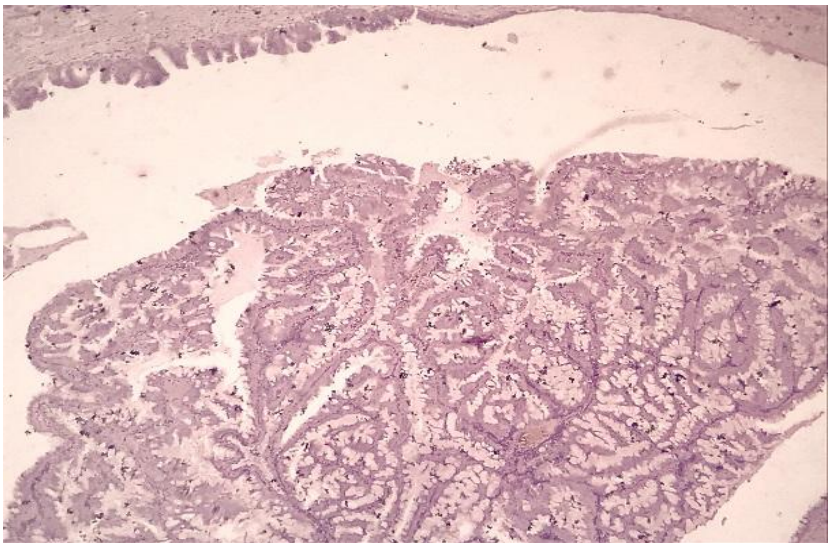

Fig 10: Ca19.9- negative .40 X.

2.4 Impression: With The Microscopic Features And Immunohistochemistry Profile ,The Mass Was Diagnosed As Intraductal Papillary Mucinous Neoplasm – Foveolar /Gastric Type With No Dysplasia.

III. Discussion

Intraductal papillary mucinous neoplasm is an intraductal tumour with mucin production and papillary epithelial proliferation leading to cystic dilatation of involved ducts. The term intraductal papillary mucinous neoplasms was coined in 1990 and given a specific entity among pancreatic neoplasms in 2000 World Health Organization classification.The incidence is more nowadays due to increasing frequency in identification of asymptomatic cystic tumours of pancreas [4].The true incidence is not known because many intraductal papillary mucinous neoplasms are small and asymptomatic. Intraductal papillary mucinous neoplasms mostly occurs in elderly males.

Clinical Features: Only one third of patients are symptomatic . [5]Symptoms include abdominal pain , weight loss , obstructive jaundice and acute pancreatitis without history of alcohol intake .3.2 IMAGING FEATURES: Computed tomography and magnetic resonance cholangiopancreatography shows the following findings such as lobulated multicystic dilatation of the branch ducts , diffuse dilatation of the main pancreatic ducts, intraductal papillary tumours ,elongated or globlike mucous plugs in the dilated ducts and bulging of the papilla into the duodenal lumen . [6]Preoperative CT IMAGING with features predictive of invasive carcinoma in intraductal papillary mucinous neoplasm are marked dilatation of main pancreatic duct, diffuse or multifocal involvement,the presence of large mural nodule or solid mass, large size of the mass and obstruction of the common bile duct. [7]

3.4 Gross: Intraductal papillary mucinous neoplasms are mostly located in the head of pancreas by about 70%, about 20% in body and tail and 5 to 10% show diffuse involvement of the gland.Most are solitary lesions with 20-40% being multifocal. [8]Dr.Matt .A.Morgan et al [9] described the TANAKA criteria based on th ductal system in which intraductal papillary mucinous neoplasms arises -Branch duct IPMN (BD-IPMN)-Main duct IPMN (MD-IPMN) with higher frequency of malignancy and dilatation of the main duct >5 mm without other cause for obstruction-5-9 mm: "worrisome feature"-showing cyst ≥ 3 cm ,thickened and enhancing cyst wall and non enhancing mural nodule with lymphadenopathy.-≥10 mm: "high risk stigmata"- showing obstructive jaundice and an enhancing solid component. -Mixed-type IPMN: appears like an advanced branch duct IPMN with main pancreatic duct dilatation (>5 mm)and has higher frequency of malignancy, similar to main duct type.

3.5 Microscopy: Histologically intraductal papillary mucinous neoplasms are classified based on the prevailing component of the epithelium into four types.

1. Foveolar / gastric subtype, which mimicks gastric foveolar epithelium, with no or minimal dysplasia.
2. Intestinal subtype,tall columnar epithelial cells with goblet cells with low grade dysplasia.
3. Pancreatobiliary subtype,which is characterised by arborizing papillae lined by cuboidal cells resembling papillary neoplasm of the biliary tract with moderate dysplasia.
4. (d)Oncocytic subtype where epithelial cells exhibit abundant cytoplasm and small basally arranged nuclei with no dysplasia . [10]

Histologically, they may demonstrate a spectrum of cellular atypia ranging from minimal mucinous dysplasia to frank invasive carcinoma. When invasive carcinomas develop in intraductal papillary mucinous neoplasms , one of two main histologic types can be encountered, namely, colloid carcinoma or the tubular adenocarcinoma. Colloid carcinoma is also designated as mucinous noncystic carcinoma.3.6 IMMUNOHISTOCHEMISTRY: Immunohistochemical markers used in intraductal papillary mucinous neoplasms are CK7. CK20,CA19.9,MUC 1,2 and 5.Four histologic subtypes are classified according to the expression of mucin.

The gastric IPMN expresses mucin type 5 (MUC5AC) ,CK 7 and is responsible for low-grade dysplasia,

The intestinal IPMN generally expresses MUC2, CK 20 and CDX 2 and is responsible for intermediate grade dysplasia. The pancreaticobiliary IPMN is the most aggressive as it expresses MUC1 and is accountable with a high-degree dysplasia.

The oncocytic type IPMN is usually positive for MUC6 and MUC 5AC. [11]

3.7 Differential Diagnosis

3.7.1 Mucinous Cystic Neoplasm Of Pancreas: Mostly occurs in females under 50 years ,does not communicate with the ducts with minimal papillae formation ,with no association with colloid carcinoma.

3.7.2 PANCREATIC INTRAEPITHELIAL NEOPLASM: Mostly undetectable clinically.Neither grossly visible nor well formed papillae are seen.No association with colloid carcinoma.MUC 2- expression- negative.

3.8 Treatment:Pancreatic resections indicated for all main duct IPMNs and other IPMNs with high risk stigmata .Endoscopic ultrasound is indicated for all cysts with worrisome features and any cyst >3cm without worrisome features. [12]3.9 PROGNOSIS:Yogi T et al described in their study with 153 patients diagnosed with IPMN- they found low/ intermediate grade dysplasia in 54.9%, high grade dysplasia in 22.2%, stromal invasion <5 mm (T1a) in 4.6%, and invasive intraductal papillary mucinous carcinoma in 18.3%.[13] Winter et al described in their study with 70 cases diagnosed with IPMN,approximately 25% of the resected cases showed intraductal papillary mucinous neoplasm associated with invasive carcinoma. Among them, 57% are tubular adenocarcinoma and 29% colloid adenocarcinoma. The overall recurrence rate observed is 24% . [14]

3.10 Recurrence And Follow Up: The Identified risk factors for tumor recurrence after surgery were tumor location, mural nodule size, presence of invasive cancer, lymph node metastasis, IPMN persistence in the pancreas remnant, and main duct dilation after surgery [14]As IPMNs are slow growing neoplasms, follow up is recommended for a longer term. In a study , the 5-yr survival of IPMT patients without invasive cancer was 94%.[15]

IV. Conclusion

Intraductal papillary mucinous neoplasms are cystic tumors of the pancreas that are diagnosed increasingly often. It is a diagnosable precursor of pancreatic cancer especially in symptomatic intraductal papillary mucinous neoplasm. Monitoring is recommended only for small, asymptomatic BD-IPMN. A better understanding of the natural history and tumor biology is needed in order to improve the current recommendations .Some of these tumors progress to invasive adenocarcinoma.

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*Dr.A.Keerthy Aberna."Intraductal Papillary Mucinous Neoplasm of Pancreas An Incidental Finding- A Case Report." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 17, no. 1, 2018, pp. 33-39.