

Case Report

Intraductal papillary mucinous neoplasm in a 26 year old lady: a diagnostic therapeutic challenge!

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ABSTRACT

Intraductal papillary mucinous neoplasm of the pancreas is a mucin producing cystic mass which originates from the pancreatic duct first defined by Ohashi et al in 1982. World Health Organization (WHO) classified mucin-producing cystic neoplasms of pancreas into 2 groups: mucinous cystic neoplasm and IPMNs. IPMN is a difficult diagnosis. Many patients may be asymptomatic for as long as 1 year and few show vague symptoms. Due to advances in radiology the incidental detection of IMPNs have increased. A 26 years old lady presented with vague pain in abdomen since 1 month with an incidental USG (A+P) finding of a cystic avascular lesion between the head and body of pancreas. CECT (A+P) and MRCP confirmed the diagnosis of branched duct IPMN with worrisome (cyst size >3 cm). Suggestive of branched duct IPMN with worrisome (cyst size >3 cm) considering the age of the patient, size of the cyst and worrisome features patient was posted for excision cyst. The tumour was well circumscribed arising from branched duct with communication with the main duct. Tumour was resected well. Histopathology revealed a benign cystic neoplasm of pancreas s/o IPMN with no e/o malignancy. In younger age group it is better to resect the tumour than timely surveillance as longer life expectancy in them provides an adequate time span for a low grade, branched duct tumour with worrisome features to undergo malignant transformation.

Keywords: Papillary, Mucinous, Radiology, Surveillance, Branched duct, Worrisome

INTRODUCTION

Intraductal Papillary Mucinous Neoplasm (IPMN) of the pancreas is a mucin producing cystic mass originating from the pancreatic ductal system first defined by Ohashi et al in 1982.^{1,3} WHO classified mucin-producing cystic neoplasms of pancreas into 2 groups: mucinous cystic neoplasm and IPMNs. WHO defined IPMNs histologically as mucin-producing, long, columnar epithelial cell lesions that cover the dilated pancreatic ducts with papillary structure.⁴ IPMNs are either invasive or non-invasive.² However, as compared to pancreatic adenocarcinomas, invasive IPMNs have better prognosis.^{4,5} The prevalence of cystic pancreatic neoplasms is 13-20% and about 25% of surgically

resected pancreatic neoplasms and 50% of incidentally detected pancreatic cysts are IPMNs. The incidence of IPMN being 2.04/ 1,00,000 population per year makes it an epidemiologically important clinical entity.^{5,6,7}

IPMNs can be classified either over ductal sites or histological characters. Depending on ductal sites: main duct IPMNs, Branched duct IPMNs and Mixed duct type and histologically according to epithelial dysplasia grade: low grade, moderate grade and high grade.¹ IPMN is a difficult diagnosis as many patients may be asymptomatic for as long as 1 year, and even though symptomatic the symptoms are bizarre. There are no gold standard methods for its diagnosis.⁸ However, due to advances in radiological techniques the incidental detection of IPMNs

have increased significantly. The question comes what is to be done over an incidentally detected IPMN, surgery or surveillance. Current guidelines emphasize on considering the prognostic features and life expectancy of the patient while we chalk out the plan of management.¹

CASE REPORT

A 26 years old lady presented with vague pain in abdomen since 1 month with an incidental USG(A+P) finding of a cystic avascular lesion between the left lobe of liver and head and body of pancreas. Patient was vitally stable. On CECT(A+P) she was found to have a tortuous dilatation of side branch of main pancreatic duct at the body of pancreas with diameter of 10.5 mm communicating with a convoluted cystic lesion of 2.6×5.4×4.2 cm in the gastric hepatic recess likely to be benign neoplastic cystic lesion of pancreas possibly IPMN (intraductal papillary mucinous neoplasm). There was no evidence of mural nodules, calcification or fat stranding.

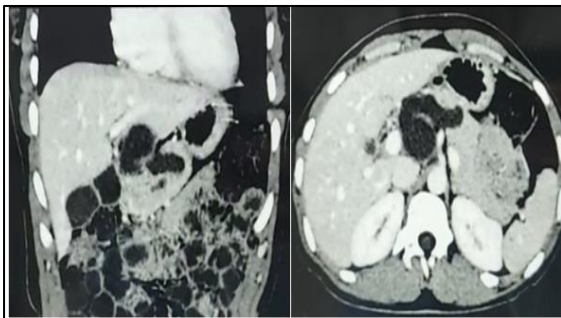


Figure 1: Coronal view and axial view of CECT(A+P) showing cystic lesion in pancreatic body.

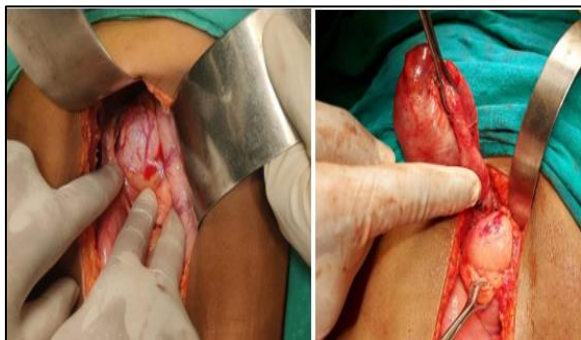


Figure 2: Intraoperative images showing cystic mass from body of pancreas.

MRCP also was s/o the similar findings. The aspirated cyst fluid appeared clear with high viscosity. Fluid amylase level was 193 U/ lit (raised) and Fluid CEA level was 126.51 ng/ml. Microscopically the fluid showed hypocellular smear with lymphocytes and macrophages and proteinaceous background. Considering the younger age of the patient and size of the cyst >3 cm being a

worrisome feature, decision was taken to post the patient for excision of the cyst.

Intra operative findings: patient was posted for exploratory laparotomy with excision of cyst under general anaesthesia. There was e/o a cystic mass of 2.6×5.4×4.2 cm arising from the body of pancreas which was originating from the branched pancreatic duct. The cyst was traced as proximally as possible and branched duct was ligated followed by retrieval of cyst. Rest of the pancreas appeared normal. Intra peritoneal drain was placed and abdomen was closed. Histopathology revealed it to be a benign cystic neoplasm of pancreas s/o IPMN with no e/o malignancy.

Postoperative period: patient was started on orals on postoperative day 3 and all sutures were removed on post op day 10 thus making her postoperative period uneventful.

DISCUSSION

IPMN is more common in the age group of 65 to 75 years with equal prevalence in male and female. The symptoms may vary from asymptomatic to pain in abdomen, nausea, vomiting, jaundice and weight loss. It is classified into main duct, branched duct and mixed type. Malignant predisposition is more with main duct IPMN (60%) then mixed followed by branched (12-30%). Investigations considered are CECT(A+P), Endoscopic USG and cyst fluid aspiration and cytology. Cyst fluid shows raised mucin, amylase and CEA level of more than 192 ng/ml.^{2,9,10-12}

Main duct IPMNs usually are subjected to resection (pancreaticoduodenectomy with triple bypass) while branched duct ones are kept under surveillance.⁹⁻¹² The features are classified into:

High risk stigmata

Obstructive jaundice with cystic lesion in head, solid component, main duct > or = 10 mm; considered for resection.

Worrisome features

Size >3 cm, main duct 5-9 mm, enhancing cyst walls and non enhancing mural nodules; further evaluation by EUS and then decision for further management.^{2,9}

If patient is planned for surveillance, a 6 monthly follow up is done for 2 years followed by annually thereafter to look for any malignant transformation.⁹⁻¹²

Current literature states that the prognostic features and life expectancy of the patient needs to be considered in management of IPMNs.⁹ Therefore in cases similar to present scenario where the age of the patient is on lower side, tumour is arising from the branched duct but patient

shows high risk and worrisome features on CECT(A+P) the mainstay of treatment should be surgical resection of the tumour. Close follow up is necessary if surveillance is planned. However as the life expectancy of the patient in this case is considerably more, one must plan for surgical intervention as the tumour though arising from branched duct and having less predisposition to malignancy as compared to main duct ones, has a longer span to undergo malignant transformation.

CONCLUSION

Age of the patient and life expectancy play a major role in the management of IPMN. In case of younger age group where the incidental detection of IPMNs are encountered it is better to consider resection of the tumour rather than timely surveillance as the longer life expectancy in such patients may tend to provide an adequate time span for a low grade, branched duct tumour with worrisome features to undergo malignant transformation. Therefore high suspicion, good radiological knowledge and timely intervention remains the key for management of IPMNs in patients with younger age group.

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REFERENCES

1. Dumlu EG, Karakoç D, Özdemir A. Intraductal Papillary Mucinous Neoplasm of the Pancreas: Current Perspectives. *Int Surg.* 2015;100(6):1060-8.
2. Tanaka M, Fernández-del Castillo C, Adsay V, Chari S, Falconi M, Jang JY et al. International consensus guidelines 2012 for the management of IPMN and MCN of the pancreas. *Pancreatology.* 2012;12(3):183-97.
3. Ohashi K, Murakami Y, Takekoshi T. Four cases of mucin producing cancer of the pancreas on specific findings of papilla of Vater. *Prog Diag Endosc.* 1982;20:348-51.
4. Longnecker DS, Adler G, Hruban RH. Intraductal papillary mucinous neoplasms of the pancreas. In: Hamilton SR, Aaltonen LA, eds. *World Health Organization Classification of Tumors.* Lyon, France: IARC Press. 2000;237-41.
5. Waters JA, Schnelldorfer T, Aguilar-Saavedra JR, Chen JH, Yiannoutsos CT, Lillemoe KD. Survival after resection for invasive intraductal papillary mucinous neoplasm and for pancreatic adenocarcinoma: a multi-institutional comparison according to American Joint Committee on cancer stage. *J Am Coll Surg.* 201;213(2):275-83.
6. Andrejevic-Blant S, Kosmahl M, Sipos B, Klöppel G. Pancreatic intraductal papillary-mucinous neoplasms: a new and evolving entity. *Virchows Arch.* 2007;451(5):863-9.
7. Lee KS, Sekhar A, Rofsky NM, Pedrosa I. Prevalence of incidental pancreatic cysts in the adult population on MR imaging. *Am J Gastroenterol.* 2010;105(9):2079-84.
8. Taouli B, Vilgrain V, O'Toole D, Vullierme MP, Terris B, Menu Y. Intraductal papillary mucinous tumors of the pancreas: features with multimodality imaging. *J Comput Assist Tomogr.* 2002;26(2):223-31.
9. Bloomgart's surgery of liver, biliary tract and pancreas. 6th edition. 959-70.
10. D'Angelica M, Brennan MF, Suriawinata AA, Klimstra D, Conlon KC. Intraductal papillary mucinous neoplasms of the pancreas: an analysis of clinicopathologic features and outcome. *Ann Surg.* 2004;239(3):400-8.
11. Conlon KC. Intraductal papillary mucinous tumors of the pancreas. *J Clin Oncol.* 2005;23(20):4518-23.
12. Turrini O, Waters JA, Schnelldorfer T, Lillemoe KD, Yiannoutsos CT, Farnell MB. Invasive intraductal papillary mucinous neoplasm: predictors of survival and role of adjuvant therapy. *HPB (Oxford).* 2010;12(7):447-55.

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