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# Intraductal papillary mucinous neoplasm of the pancreas associated with neuroendocrine tumor: A case report

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ABSTRACT

*INTRODUCTION:* Intraductal papillary mucinous neoplasm is an uncommon cystic tumor of pancreas that can be associated with ductal adenocarcinoma. Coexistence of pancreatic IPMN and neuroendocrine tumor is very rare. Here, we report the imaging features of mixed type intraductal papillary mucinous neoplasia of the pancreas with high grade dysplasia together with neuroendocrine carcinoma and perform review of the literature.

*PRESENTATION:* A 68-year old patient has been evaluated for possible IPMN that was suspected during ultrasound. MRI revealed main and side branch duct dilatations. At the head, a contrast enhancing nodular lesion was identified. Due to the presence of high risk stigmata according to guidelines, surgery was performed. Histopathological examination revealed an unusual association, including mixed type IPMN and neuroendocrine carcinoma.

*DISCUSSION:* The concomitant occurrence of pancreatic IPMN and neuroendocrine tumor has been reported in case studies and brief reviews. Yet, the imaging findings and underlying molecular mechanisms of this entity has not been fully understood. In addition to this unusual association, pancreatic intraepithelial neoplasia was also detected in the present case. Although majority of neuroendocrine tumor associated IPMNs were reported to be having low grade dysplasia, our patient had high grade dysplasia. Further studies and reviews with larger groups are needed to establish imaging features and underlying molecular mechanisms of this rare association.

*CONCLUSION:* Although the major concern during work-up of IPMN is presence of associated pancreatic ductal adenocarcinoma, the possibility of neuroendocrine tumor, in the presence of a hypervascular solid foci on imaging studies should be kept in mind.



## 1. Introduction

Intraductal papillary mucinous neoplasms (IPMN) are mucinous cystic tumors of the pancreas that originate from mucin secreting cells in the main pancreatic duct and/or its branches. IPMNs are more commonly seen in males, at the 6th–7th decade. They constitute about 20% of all cystic pancreatic neoplasia's. They are categorized as main duct, branch duct and mixed type according to macroscopic features. It is well known that patients with IPMN, especially involving the main pancreatic duct, have the potential to synchronously or met asynchronously develop pancreatic ductal adenocarcinoma (PDAC). There are certain criteria that favor the presence of malignancy defined by consensus guidelines [1]. Neuroendocrine tumor (NET) of the pancreas is a unique type of pancreatic tumor, which constitute 1–2% of all pancreatic neoplasms [2]. They most commonly occur between 3rd–6th decades, without any gender predilection. Co-occurrence of pancreatic IPMN and neuroendocrine tumor is a very rare entity and has been presented as case reports in the literature [3]. Here, we report a patient having concomitant mixed type IPMN and NET of the pancreas and make a review of the relevant literature. The work has been reported in line with the SCARE criteria [4,5].

## 2. Case presentation

A 68-year-old male with a 20 year history of type 2 diabetes mellitus was admitted to our hospital for further evaluation of a possible pancreatic IPMN that was suspected during a routine abdominal ultrasound examination. He was also diagnosed as hav-

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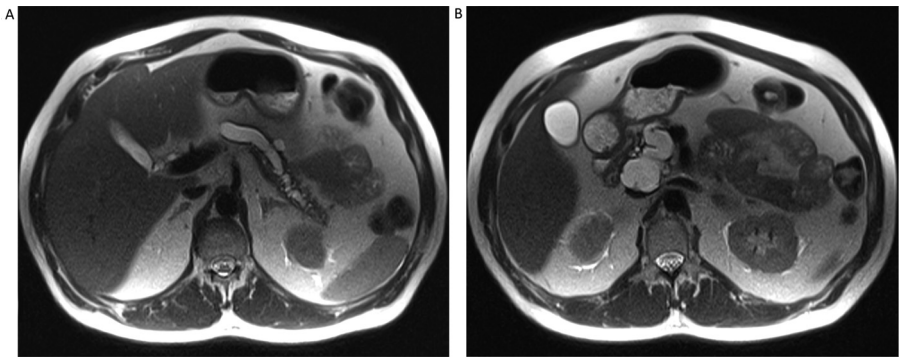


**Fig. 1. a, b.** Axial T2 weighted MR image showing pancreatic main ductal dilatation in the body and tail (a) and head (b) portions.

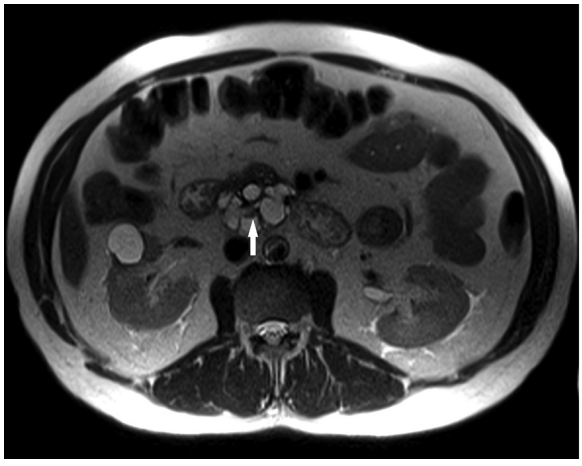

**Fig. 2.** A hypointense nodular intensity (arrow) is seen at the head of the pancreas inside the dilated duct close to the papilla level on axial T2 weighted MR image.

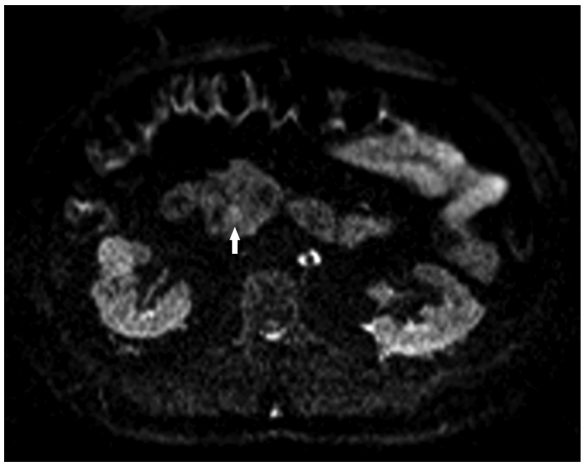

**Fig. 3.** Axial DWI image showing hyperintensity (diffusion restriction) of the nodular intensity (arrow) shown in Fig. 2.

ing diabetic nephropathy. There was a mild degree of renal failure. Magnetic resonance imaging (MRI) revealed dilatation of the main pancreatic duct throughout the gland, measuring up to 35 mm, accompanied by mild side branch dilatations (Fig. 1a, b). There was marked atrophy in the pancreatic parenchyma in all parts of the gland. The MR features were suggestive of a mixed type IPMN throughout the pancreas. A mural nodularity measuring 1.5 cm, that is similar to pancreatic parenchyma on pre-contrast images was noticed in the head region close to ampulla (Fig. 2). This nodular lesion exhibited hyperintensity on DWI (b: 800 s/mm$^2$) images with hypointensity on ADC map (restricted diffusion) (Fig. 3). After intravenous contrast administration this lesion enhanced with contrast and this finding was further verified with subtraction images that were obtained after subtraction of pre from post-contrast data (Fig. 4). As far as recent international consensus guidelines about pancreatic mucinous neoplasia's are concerned [1], the imaging features revealed the presence of both high risk stigmata's (main pancreatic duct measuring >1cm and presence of contrast enhancing solid component). Tumor markers (serum CA 19-9 and CA 125) were in normal limits.

In accordance with the imaging features, total pancreatectomy, partial duodenectomy, cholecystectomy, subtotal gastrectomy, splenectomy and peripancreatic lymphadenectomy was performed. Since mixed type IPMN with high risk stigmata is detected, total pancreatectomy was performed. Altough not seen on imaging tests, there was the probability of malignant foci on other parts of the pancreas. The surgery was done by a specialized hepatobiliary surgeon with 30 years of experience. The postoperative course of the patient was uneventful and he was discharged from the hospital at the end of the second postoperative week.

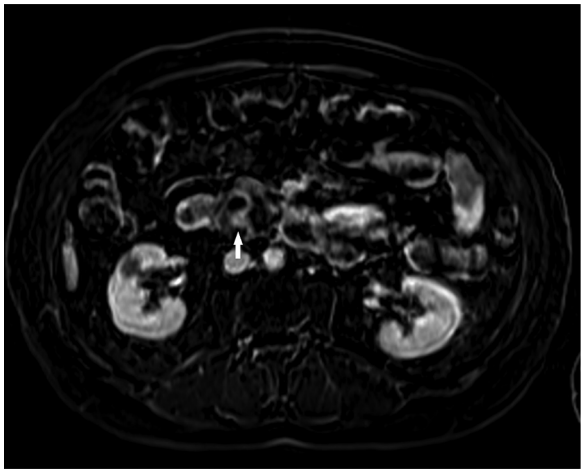

**Fig. 4.** Substracted (precontrast from post contrast) T1weighted axial image showing that the nodular lesion is enhanced markedly with contrast (arrow).

For diabetes he has been using insulin subcutaneously before the operation, after surgery insulin was continued at the same dosage, pancreatic enzyme substitute (Kreon 25.000 Unit/3 times a day) was added to therapy.

Histopathological examination revealed high grade, mixt type IPMN containing gastric, intestinal and biliary type of epithelium. At the pancreatic head adjacent to the IPMN, presence of neuroendocrine carcinoma with perineural invasion was detected, that corresponded to the contrast enhancing nodular lesion seen on