

Double primary tumors of the pancreas

A case report

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Abstract

Rationale: Adenocarcinoma and neuroendocrine tumors are a very rare combination of double primary pancreas tumor.

Patient concerns: A Whipple operation was initially performed on a 64-year-old man to remove an adenocarcinoma. Four years after the operation, surveillance-computed tomography revealed abnormal findings of the pancreas. Recurrent adenocarcinoma and neuroendocrine tumor involving different sites of the remnant pancreas were simultaneously detected and characterized on computed tomography and magnetic resonance imaging.

Diagnoses: The patient was diagnosed with recurrent adenocarcinoma and neuroendocrine tumor in the post-operative pancreas.

Interventions: Radical pancreatosplenectomy was performed. The patient underwent subsequent chemotherapy and radiotherapy.

Outcomes: No tumor recurrence was found during the 5 years of follow-up visits.

Lessons: The possibility of multiple primary tumors of different histological origin should be considered when multiple different pancreatic lesions are detected on images. Computed tomography (CT) and magnetic resonance imaging (MRI) play key roles in the management of multiple tumors in the pancreas.

Abbreviations: ADC = apparent diffusion coefficient, CT = computed tomography, DWI = diffusion-weighted image, IPMN = intraductal papillary mucinous neoplasm, MRI = magnetic resonance imaging, NET = neuroendocrine tumor.

Keywords: adenocarcinoma, multiple primary, neoplasm, neuroendocrine tumors, pancreas

1. Introduction

A double-primary neoplasm in the pancreas are uncommon. Most studies of double-primary pancreatic tumors focused on pancreatic ductal adenocarcinoma, either derived from or occurring concomitantly, with an intraductal papillary mucinous neoplasm (IPMN) or association between a pancreatic neuroendocrine tumor (NET) and IPMN.^[1–4] A simultaneously separated adenocarcinoma and NET in the pancreas are very rare.^[5] We report a rare case of recurrent adenocarcinoma and NET in the remnant pancreas detected 4 years after a Whipple operation. This study was approved by the Dankook University Hospital

Institutional Review Board. Informed written consent was obtained from the patient for publication of this case report and accompanying images.

2. Case report

A 64-year-old man presented with abnormal findings of the pancreas detected by surveillance computed tomography (CT). The patient had a history of a curative Whipple operation due to an adenocarcinoma in the pancreatic head with an invasion of the duodenum 4 years ago. Subsequently, the patient underwent radiological surveillance, with a routine follow-up CT, and there was no evidence of tumor recurrence during the following 4 years. The CA 19-9 levels were normal, and there was no evidence of pancreatitis during the follow-up period. However, a CT scan now revealed dilatation of the main pancreatic duct, with abrupt narrowing, which was not evident on the prior CT scan (6 months ago). No focal obstructive lesion was detected at the point of abrupt narrowing of the organ (Fig. 1A). The patient underwent abdominal magnetic resonance imaging (MRI) (Ingenia 3T, Philips Healthcare, Best, The Netherlands) for further evaluation. A dynamic contrast enhancement study was performed, with gadolinium ethoxybenzyl diethylenetriamine pentaacetic acid (Gd-EOB-DTPA).

The MRI revealed a 1.2 cm ill-defined, poorly enhanced lesion in the pancreatic body, with dilation of the upstream main duct in the images that received dynamic contrast enhancement (Fig. 1B). This lesion showed increased signal intensities on diffusion-weighted image (DWI), with high *b* values ($b > 500 \text{ s/mm}^2$) (Fig. 1C), and a relatively low apparent diffusion coefficient (ADC) value, compared with the remaining pancreatic paren-

Editor: N/A.

The authors have no conflicts of interest to disclose.

The present research was conducted by the research fund of Dankook University in 2016.

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Medicine (2018) 97:50(e13616)

Received: 22 August 2018 / Accepted: 19 November 2018

<http://dx.doi.org/10.1097/MD.0000000000013616>

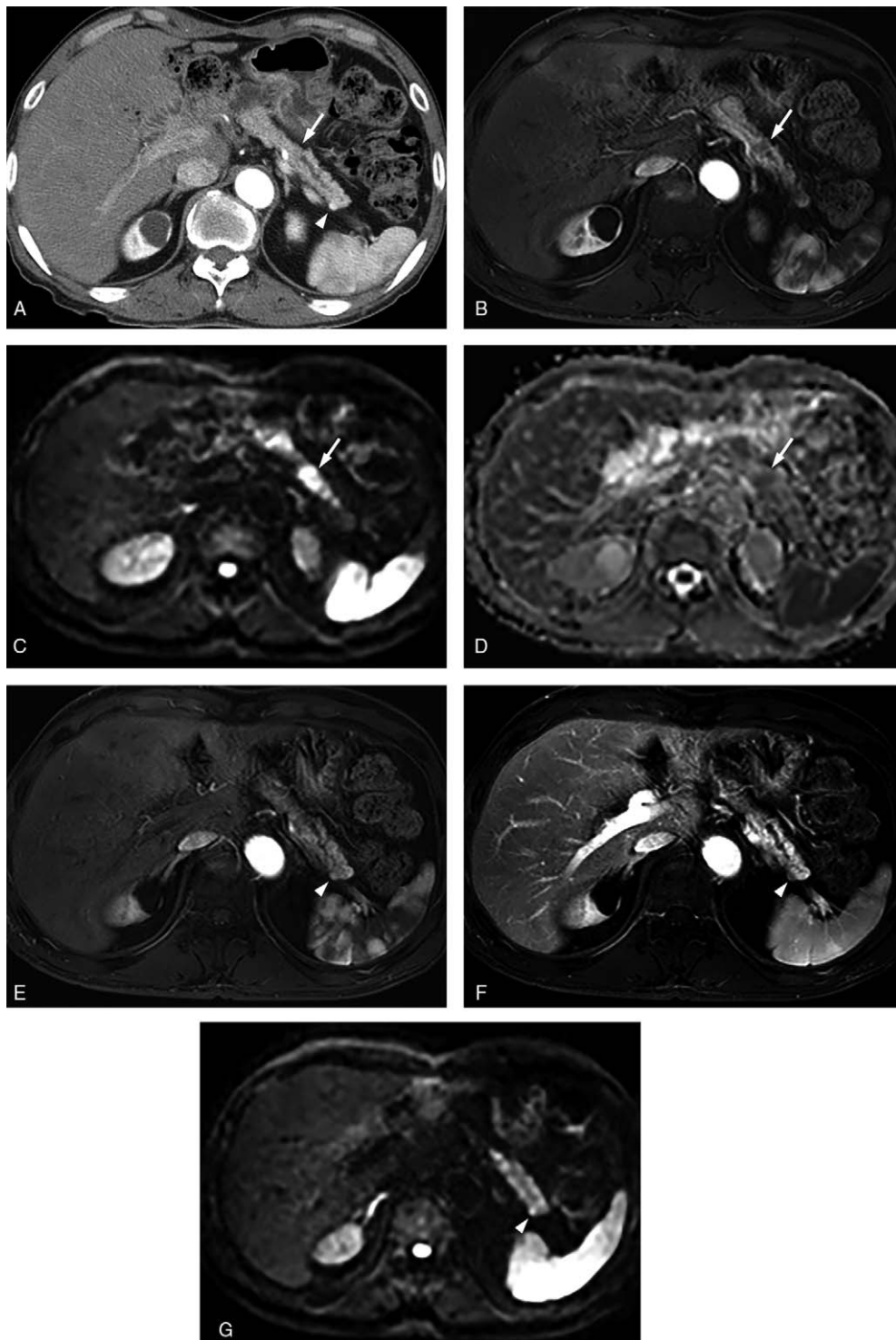


Figure 1. (A) The arterial phase computed tomography (CT) scan reveals dilatation of the main pancreatic duct with abrupt narrowing (arrow). A small enhancing nodule (arrow head) is defined in the pancreatic tail portion. (B) A 1.2 cm poorly enhanced mass (arrow) is seen in the pancreatic body in the arterial phase image under dynamic contrast enhancement study on magnetic resonance imaging. The mass (arrow) shows high signal intensity in the axial diffusion-weighted image at high b -value ($b = 800 \text{ s/mm}^2$) (C), with a low apparent diffusion coefficient value compared with the remaining pancreatic parenchyma on the apparent diffusion coefficient map (D). (E) A small well-circumscribed enhancing nodule (arrow head) is defined in the pancreatic tail portion on arterial phase image under dynamic contrast enhancement study on magnetic resonance imaging. (F) The nodule (arrow head) shows lower signal intensity compared with the surrounding pancreas in the portal venous phase. (G) The nodule (arrow head) reveals a high signal intensity in the axial diffusion-weighted image at high b -value ($b = 800 \text{ s/mm}^2$). CT = computed tomography.

chyma (Fig. 1D). Furthermore, a 5-mm enhanced nodule was newly detected in the pancreatic tail portion on surveillance CT (Fig. 1A). There was no matched lesion in the pancreatic tail portion on a preoperative MRI performed 4 years previously as part of the Whipple operation. A newly detected small pancreatic nodule showed enhancement in the arterial phase under dynamic contrast enhancement study on MRI (Fig. 1E). It was distinguished from the surrounding pancreas by a relatively lower signal intensity in the portal venous phase (Fig. 1F). This nodule was not evident in the correlated area on the axial T2-weighted image and the pre-contrast T1-weighted image. The DWI revealed a sustained hyperintensity of the nodule, with increasing value of the *b* factor (Fig. 1G). However, the ADC value was not apparently lower than that of the remaining pancreas. Radical pancreatosplenectomy was performed. Pancreatic adenocarcinoma in body portion pathologically identical to a tumor that was resected 4 years ago was confirmed. The small nodule in the pancreatic tail was confirmed as grade 1 NET. The patient underwent subsequent chemotherapy and radiotherapy. Concurrent 5-fluorouracil (5-FU) chemotherapy and external beam radiotherapy was started 4 weeks after the operation. The patient was treated with 4500 cGy radiation in 25 fractions over 5 weeks concomitant with 5-FU 300 mg/m² per day intravenously. No tumor recurrence was found during the 5 years of follow-up.

3. Discussion

The incidence of co-occurring adenocarcinoma and NET, at different locations in the pancreas, is extremely rare.^[5] This present case showed an adenocarcinoma and NET in the pancreas occurring at the same time in different locations of the organ. The incidence of multiple primary malignancies in patients with pancreatic NET is high, compared with the general population. Sporadic pancreas NET is associated with a wide range of other tumors. It may be the result of accumulated growth stimulation by the secreted hormones or a genetic alteration that leads to tumorigenesis.^[6] But the pathophysiology correlating with NET and adenocarcinoma is still unclear, and further studies are needed.

Pancreatic NET is rare, but the incidence appears to be rising with improved diagnostic techniques. Many nonfunctioning pancreas NETs are incidentally detected on the image, as in this case. Arterial enhancement is a typical feature of a pancreatic NET image.^[7] Both CT and MRI showed arterial enhancement of this small pancreatic NET. And this small NET showed high signal intensity on DWI. However, due to the small tumor size, it was difficult to determine the lower ADC value of the lesion compared with the remaining pancreas. The NETs generally show varying ADC values as a result of variable underlying histopathologies.^[8] Differential diagnosis of small pancreatic NET included intrapancreatic accessory spleen, solid pseudopapillary neoplasm, or acinar cell carcinoma, as in this case.

Pancreatic adenocarcinoma is still a disease associated with a high recurrence rate and poor survival despite recent advances in operative techniques and adjuvant treatment strategies. Isolated local recurrence of pancreatic adenocarcinoma confined to the remnant pancreas is uncommon and amenable to repeat pancreatectomy.^[9] The CT is routinely used for surveillance at

the 3-month interval during the first 2 years after operation and then 6-month intervals in our hospital. Surveillance CT was effective in detecting recurrence, and curative repeated resection was eligible in this case. Local recurrence in the remnant pancreas presented as dilatation of main pancreatic duct with abrupt narrowing on surveillance CT. But CT did not reveal obstructive tumor because the pancreatic adenocarcinoma was isoattenuated relative to the background pancreatic parenchyma. The MRI enabled the depiction of the poorly enhanced adenocarcinoma, in contrast to the well-enhanced pancreatic parenchyma, reflecting the hypovascular nature of the tumor. The ADC value of the mass was relatively low compared with normal pancreatic parenchyma. This is a well-known feature of fibrotic, highly-cellular adenocarcinoma.^[10] The CT and MRI findings were typical for pancreatic adenocarcinomas.

This case shows an extremely rare combination of a double-primary pancreas tumor, adenocarcinoma, and NET. The possibility of multiple primary tumors of different histological origin should be considered when multiple different pancreatic lesions are detected on images.

Author contributions

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