CASE REPORT

One of the multifocal intraductal papillary mucinous neoplasms with the clinical characteristics of mucinous cystic neoplasm

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SUMMARY

A 74-year-old woman with multiple cystic lesions in the pancreas was first examined at a previous hospital. Many of the lesions in the head and body were diagnosed as a branch duct intraductal papillary mucinous neoplasms (IPMN), but one lesion in the tail was a simple cyst. She had no surgical treatment because there were no signs of malignancy in any of the lesions. After 3 years, solid components appeared only in the tail lesion. Because of its preoperative diagnosis as a mucinous cystic neoplasm (MCN), distal pancreatectomy was performed. Histopathological findings revealed that the cystic tumour in the tail was IPMN with minimally invasive carcinoma and the other lesion in the body was IPMN with low-grade dysplasia. They were IPMNs bridged by a non-dilated main pancreatic duct. There may be some cases in which it is difficult to diagnose between IPMN and MCN.

BACKGROUND

A variety of cystic lesions may occur in the pancreas, including intraductal papillary mucinous neoplasms (IPMN), mucinous cystic neoplasm (MCN) and serous cystic neoplasm.¹ Typical clinical and imaging characteristics of common pancreatic cysts were described in the international consensus guidelines 2012 for the management of IPMN and MCN of the pancreas.² IPMNs are often multifocal and a careful postoperative surveillance is necessary, even if the complete resection of non-invasive IPMN is achieved owing to the possibility of recurrence of IPMN or occurrence of pancreatic ductal adenocarcinoma in remnant pancreas. On the other hand, MCNs are almost always unifocal and a complete resection of a non-invasive MCN is curative.²

IPMN, referred to as 'Grape-like', is consisted of an assembly of multiple cysts. In contrast, MCN, referred to as 'Orange-like', is consisted of unilocular cyst with a common capsule. In cases of typical clinical imaging findings, it is easy to distinguish these two cystic lesions.² However, in some cases, it is difficult to distinguish between such cystic lesions in the preoperative diagnosis. Because particular cases of IPMN and almost all cases of MCN possess malignant potential, it is important for a surgical treatment to distinguish between branch duct IPMN and MCN based on preoperative clinical findings.³⁻⁴ Furthermore, some multifocal IPMN cases have different histological dysplastic findings with different grades.⁵ In this case, the imaging characteristics of one of the multiple

lesions showed the typical characteristics of MCN, but was then diagnosed as branch duct IPMN in histopathological diagnosis.

CASE PRESENTATION

In a 74-year-old woman, multiple cystic lesions of the pancreas were detected at a regular health checkup. She had no symptoms and no particular findings in her and her family histories.

INVESTIGATIONS

An MRI showed 2 cm size multiple grape-like cystic lesions in the pancreatic head and body. On the other hand, a single unilocular cystic lesion without a connection to the pancreatic duct was seen in the pancreatic tail. The two lesions in the head and body were compatible with branch duct IPMNs (figure 1A) and the lesion in the tail appeared to be simple cyst based on the MRI findings (figure 1B). No signs of suspected malignancy were seen in any lesions, and therefore the patient visited our hospital for MRI examination every 6 months.

Three years later, a follow-up MRI examination revealed solid components in the cystic lesion of the tail (figure 1B), while no changes in shape and size were seen in the lesions of the head and body (figure 1A). Abdominal contrast-enhanced CT showed a 6 cm size unilocular cystic lesion with a septal structure and enhanced solid components in the tail (figure 2A,B). Endoscopic retrograde cholangiopancreatography showed no dilation of the main pancreatic duct and no communication between the lesion in the tail and the pancreatic duct. Endoscopic ultrasonography clarified multiple, solid components (maximum diameter was 2 cm) in the cystic lesion of the tail (figure 2C). An enhanced uptake of fluorodeoxyglucose (FDG) with standardised uptake value (SUV_{max} 6.1) was detected in the solid components in FDG-positron emission tomography (FDG-PET) (figure 2D). Both carcinoembryonic antigen and carbohydrate antigen 19-9 (CA19-9) were within normal ranges.

DIFFERENTIAL DIAGNOSIS

In this case, detailed preoperative examinations showed a single, spherical, unilocular cystic lesion with a septal structure and solid components in the tail of the pancreas and no dilation of the main pancreatic duct without connection with the tail lesion. Based on these findings, we suspected mucinous cystadenocarcinoma as a preoperative diagnosis of the lesion.

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Figure 1 MRI findings of pancreatic cystic lesions at the first detection (left) and at 3 years later (right). (A) An MR cholangiopancreatography findings of the lesions in the head and body. No changes of shape or size in both lesions and no dilation of the main pancreatic duct were detected. (B) An MRI on T2-weighted imaging findings of the lesion in the tail. A solid component was seen (arrow).

TREATMENT

Subsequent distal pancreatectomy with regional lymph node dissection was performed. The pancreas was dissected above the portal vein and the pancreatic body and tail was resected with the cystic lesions en bloc. Intraoperative histological diagnosis of the stump of the pancreas was normal pancreatic ductal epithelium without dysplastic change.

OUTCOME AND FOLLOW-UP

Histopathological findings revealed that the lesion in the body was a branch duct IPMN with low-grade dysplasia (figure 3A), whereas the lesion in the tail was a branch duct IPMN with minimally invasive carcinoma (figure 3B). Furthermore, IPMN with low-grade and intermediate-grade dysplasia was detected in the non-dilated main pancreatic duct between the two lesions (figure 3C). These lesions were IPMNs bridged by non-dilated main pancreatic duct (figure 3D). An ovarian-like stroma, a representative finding of MCN, was not found in the tail lesion. The main pancreatic duct in the stump contained normal pancreatic ductal epithelium. Analysis of immunohistochemical stainings showed gastric type of mucin expression in the lesion: MUC1-negative, MUC2-negative and MUC5-positive. The

pathological diagnosis was T1, N0, M0, stage IA, based on the TNM classification, the seventh edition.

The postoperative course was uneventful. Six months after the operation, she remained in a stable condition with no symptoms of recurrence.

DISCUSSION

Among pancreatic cystic lesions, both IPMNs and MCNs show a wide spectrum of malignant transformation, ranging from low-grade dysplasia to invasive carcinoma as defined by the WHO IPMN classification.⁶ Therefore, preoperative diagnosis is important to estimate the histological grade and to clarify whether the tumour is benign or malignant for determining the surgical indication. The international consensus guidelines 2012 for the management of IPMN and MCN of the pancreas recommend surgery for all cases of main duct IPMN and some cases of branch duct IPMN, with a suspected risk of malignancy, so-called 'high-risk stigmata' (enhanced solid component and main pancreatic duct size of ≥ 10 mm).² On the other hand, surgical resection is recommended in almost all patients to avoid the risk of progression to invasive MCN because it tends to be shown in younger patients.²⁻⁷ Thus, surgical resection is

Figure 2 (A and B) Abdominal contrast-enhanced CT findings of the lesion in the tail showed septal structure and enhanced maximum 2 cm size of solid components. (C) Endoscopic ultrasonography showed multiple solid components in the cystic lesion of the tail (arrows). (D) fluorodeoxyglucose-positron emission tomography (FDG-PET) showed enhanced uptake of FDG to the standardised uptake value (SUV_{max} 6.1) in solid components (arrow).

Figure 3 Histopathological findings of the lesion in the body showed low-grade dysplasia (A) and the tail showed the minimally invasive carcinoma (B). In non-dilated main pancreatic duct connecting with these two lesions, low-grade and intermediate-grade dysplasia was found (C). Pathological mapping scheme of the body and tail of pancreas (D). Two lesions in the body and tail formed a continuous lesion with a mixture of different histological dysplastic grades.

indicated in particular cases with IPMN and almost all cases with MCN. Therefore, in the preoperative diagnosis, it is important to distinguish not only between benign and malignant IPMN, but also between branch duct IPMN and MCN. There are many clinicopathological differences between branch duct IPMN and MCN. Generally, benign or non-invasive MCN is a unifocal tumour that does not recur after complete resection.⁸ In Contrast, IPMN, even if not malignant, may occur as multifocal and recur in the remnant pancreas after complete resection. Therefore, careful postoperative observation is required.² Actually, it was reported that 14.5–41% of all branch duct IPMN are multifocal.^{3–9} While branch duct IPMN and MCN are often distinguished from each other on the basis of factors such as age, sex, location, shape and size and communication with the pancreatic duct, it is difficult to distinguish in some cases. A number of different methods for distinguishing IPMN from MCN have been attempted,^{10–11} but all have proved to be insufficient. In addition to the findings of discrimination described in the guidelines, molecular analysis of GNAS gene mutations can be useful for distinguishing among branch duct IPMN and MCN.¹²

In conclusion, we encountered a branch-type IPMN case that had the typical characteristics of MCN in the clinical image findings. The development of novel examinations for the correct preoperative diagnosis is necessary.

Learning points

- Consider the possibility of atypical clinical imaging findings in the preoperative diagnosis of pancreatic cystic lesions.
- Consider that multifocal intraductal papillary mucinous neoplasms (IPMNs) can be histologically continuous even if the main pancreatic duct is not dilated; therefore, careful attention must be paid to the histological findings in the stump of the pancreas.
- Careful observation is required for residual IPMN in the remnant pancreas.

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Competing interests None.

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