

CASE REPORT

Intraductal papillary mucinous neoplasm originating from a heterotopic pancreas within the jejunum: a case report

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Abstract

We report a case of intraductal papillary mucinous neoplasm arising within the heterotopic pancreatic tissue which was found incidentally in the jejunum during surgery for bowel obstruction. A 54-year-old female patient was admitted to our hospital due to sudden abdominal pain. In preoperative findings, we diagnosed bowel obstruction and performed surgery. Intra-operative findings showed adhesive intestinal obstruction, we performed synechiotomy for adhesion release. During surgery, when searching the small intestine, we coincidentally found a tumor in the jejunum and partial resected the jejunum. Pathological examination revealed a 1.2 cm × 1.0 cm × 1.0 cm white yellow nodule with cystic spaces. Histological examination demonstrated heterotopic pancreatic tissue consisting of well-formed lobules of pancreatic acini and cystically dilated ducts containing intraductal papillary neoplasm. Moreover, in immunohistochemical staining, MUC5AC was diffusely expressed, but not MUC1, MUC2 and MUC6.

INTRODUCTION

Heterotopic pancreas is rarely found in various organs, and is defined as the presence of pancreatic tissue that lacks both anatomical and vascular connection to the normal pancreas [1]. Intraductal papillary mucinous neoplasm (IPMN) is one of the tumors that occur in the pancreas; however, the incidence of this pathologic entity in heterotopic pancreatic tissue is extremely rare. It is usually asymptomatic, however, it can show size or location dependent symptoms and has the

potential of being malignant. To our knowledge, only 11 cases of IPMN have been reported in the literature, arising from the ectopic pancreatic tissue in the stomach, duodenum, Meckel's diverticulum and jejunum. In particular, only three cases of IPMN have been reported within the jejunum before the present case [2–4]. Management of IPMN occurring in the ectopic pancreas has not been reported to date, but IPMN occurring in the pancreas has been reported as a guideline [5]. Furthermore, by immunohistochemical examination, IPMN is classified into subtypes gastric,

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intestinal, pancreatobiliary and oncocytic. The vast majority of IPMNs is of the gastric type, which is MUC5AC positive but MUC1-negative, with MUC2 highlighting only the scattered goblet cells. The gastric type is typically low grade, with only a small percentage developing into carcinoma [5]. In this report, we described an extremely rare IPMN diagnosed as an histologically gastric type, which occurred in the jejunal ectopic pancreas.

CASE REPORT

A 54-year-old female patient was admitted to the hospital due to sudden abdominal pain. She had a history of hypertension, depression, gastric ulcer and uterus myoma. She underwent total hysterectomy due to uterus myoma when she was 37 years old. Routine blood and urine tests showed only elevation of serum white blood cells ($13\,700/\text{mm}^3$). Abdominal computed tomography revealed dilatation of the small intestine.

In preoperative findings, we diagnosed bowel obstruction, and performed surgery. Intra-operative findings showed adhesive intestinal obstruction, and we performed synechiotomy for adhesion release. During surgery, when searching the small intestine, we coincidentally found a tumor in the jejunum and partially resected the jejunum.

Pathological examination revealed a 1.2 cm \times 1.0 cm \times 1.0 cm white yellow nodule with cystic spaces. Histological examination demonstrated heterotopic pancreatic tissue consisting of well-formed lobules of pancreatic acini and cystically dilated ducts containing intraductal papillary neoplasm. In this area, the ducts are filled with complex tall mucinous epithelial and cuboidal cells as well as a fine and slightly complicated papillary structural area (Fig. 1A–C). We found no marked cytological or architectural atypia in these epithelia. Moreover, in immunohistochemical staining, MUC5AC was diffusely expressed, but not MUC1, MUC2 and

MUC6 (Fig. 1D). From these findings, we diagnosed this lesion as heterotopic pancreas with gastric-type IPMN and focal dysplastic change in the jejunum.

DISCUSSION

We described an extremely rare condition of IPMN arising within heterotopic pancreatic tissue, which was found incidentally in the jejunum during surgery for bowel obstruction. In 1996, the World Health Organization established criteria to classify IPMNs and to distinguish them from other mucin-producing cystic neoplasms [6]. IPMNs account for ~7% of clinically diagnosed pancreatic neoplasms. Preoperative diagnosis of heterotopic pancreas may be challenging, and this typically is an incidental finding. Heterotopic pancreatic tissue itself is uncommon, with a reported frequency between 0.55 and 13.7% [1]. Ectopic pancreatic tissues are usually asymptomatic and are found incidentally. To our knowledge, only 11 cases of an IPMN have been reported in the literature, arising from the ectopic pancreatic tissue in the stomach, duodenum, Meckel's diverticulum and jejunum. Particularly, there are only three cases report of IPMN reported within the jejunum before the present report [2–4]. Overall, 12 patients (seven males and five females) with IPMN, including our case, have been reported till date. The mean age of the patients was 63.4 years (range: 44–80 years). Four of the lesions were incidentally identified after surgery for other diseases, and symptoms experienced by other patients depended on the location and the size of the tumor.

The article 'International Consensus Guideline 2012 for the Management of IPMN and MCN of the Pancreas,' described the histological aspects of IPMN [5]. The epithelial lining of gastric-type IPMNs is composed of innocuous tall columnar cells with basally oriented nuclei and abundant pale mucinous cytoplasm.

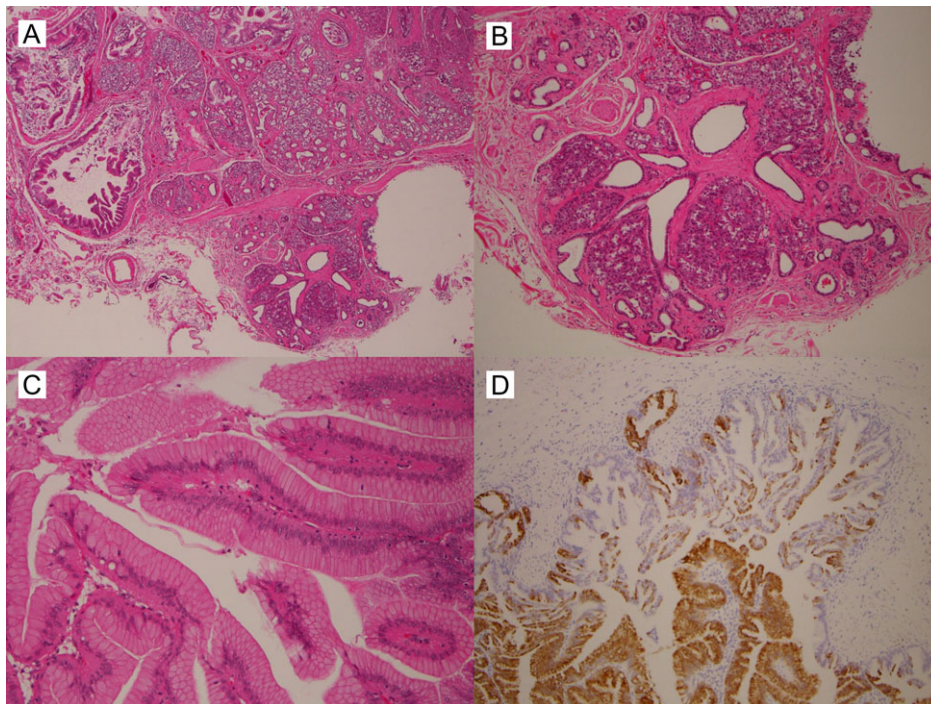


Figure 1: Microscopic findings. (A) Microscopic examination shows heterotopic pancreatic tissue consisting of well-formed lobules of pancreatic acini and cystically dilated ducts containing intraductal papillary neoplasm [hematoxylin–eosin (HE) stain, original magnification $\times 40$]. (B) Pancreas acini and dilated ducts (HE stain, original magnification $\times 100$). (C) The ducts are filled with complex tall mucinous epithelial and cuboidal cells as well as a fine and slightly complicated papillary structural area (HE stain, original magnification $\times 400$). (D) Immunohistochemical stain shows that MUC5AC was diffusely expressed (original magnification $\times 100$).

Immunohistochemical findings revealed MUC1 (-), MUC2 (-), MUC5AC (+) and MUC6 (-) proteins in the present case; therefore, we diagnosed a gastric-type IPMN.

IPMN may arise from heterotopic pancreatic tissue and can indeed carry a risk of malignant transformation. Therefore, negative margin surgical resection may be needed, where appropriate, for effective treatment control. However, the ectopic pancreatic IPMN is often found incidentally. The decision for surgical treatment can be difficult, and the physician may be hesitant to reschedule for additional treatment. Although there are no guidelines on follow-up for these patients, we completely resected the ectopic pancreas in this case. Histologically, we found no malignancy; however, frequent surveillance will be unnecessary.

In summary, we report an extremely rare case of IPMN arising within the heterotopic pancreatic tissue in the jejunum. We believe that it is important to study further IPMN cases, because ectopic pancreatic IPMN is often found incidentally and lack of guidelines makes the decision about surgical treatment difficult to determine.

CONFLICT OF INTEREST STATEMENT

None of the authors have any conflicts of interest to declare relevant to this publication.

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REFERENCES

1. Dolan RV, ReMine WH, Dockerty MB. The fate of heterotopic pancreatic tissue. A study of 212 cases. *Arch Surg* 1974;**109**:762-5.
2. Sung JY, Han JY, Choi SK, Kim L, Choi SJ, Park IS, et al. Adenocarcinoma with intraductal papillary mucinous neoplasm arising in jejunal heterotopic pancreas. *Korean J Pathol* 2012;**46**:96-100.
3. Okamoto H, Fujishima F, Ishida K, Tsuchida K, Shimizu T, Goto H, et al. Intraductal papillary mucinous neoplasm originating from a jejunal heterotopic pancreas: report of a case. *Surg Today* 2014;**44**:349-53.
4. Lee SH, Kim WY, Hwang D-Y, Han HS. Intraductal papillary mucinous neoplasm of the ileal heterotopic pancreas in a patient with hereditary non-polyposis colorectal cancer: a case report. *World J Gastroenterol* 2015;**21**:7916-20.
5. Tanaka M, Castillo C F-D, Adsay V, Chari S, Falconi M, Jang J-Y, et al. International consensus guidelines 2012 for the management of IPMN and MCN of the pancreas. *Pancreatology* 2012;**12**:183-97.
6. Sohn TA, Yeo CJ, Cameron JL, Hruban RH, Fukushima N, Campbell KA, et al. Intraductal papillary mucinous neoplasms of the pancreas: an updated experience. *Ann Surg* 2004;**239**:788-97.