Case Report

A Case of Heterotopic Pancreatic Cancer in the Jejunum

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A 78-year-old woman was admitted to our hospital because of abdominal pain, nausea, and vomiting. A plain abdominal X-ray showed both gas and a level in the small intestine. The symptoms and the level improved in response to conservative therapy, but the symptoms of intestinal obstruction recurred after she consumed a meal. Based on the symptoms and X-ray findings, the patient was diagnosed with intestinal obstruction of unknown etiology, and was operated. Intraoperatively, a tumor was palpated in the jejunum 50 cm distal to the ligament of Treitz. The tumor had invaded the entire thickness of the wall and was exposed beyond the serosa. The proximal bowel was extremely dilated, and partial jejunectomy was performed to relieve the obstruction. Gross examination of the specimen revealed a yellowish-brown protruding tumor, measuring 3×2 cm. The tumor was covered with intact mucosa and resembled a submucosal tumor. Histopathological examination showed that the tumor was developed from an aberrant pancreas composed of ducts, acini, and smooth muscle tissue. The tumor was located in the submucosa of the jejunum. The tumor consisted of moderately differentiated adenocarcinoma and was in contact with the aberrant pancreas. On the basis of these findings, the tumor was diagnosed as cancer of heterotopic pancreas in the jejunum.

It is extremely rare for aberrant pancreatic cancer to be located in the jejunum. Six cases of heterotopic pancreatic cancer in the jejunum have been reported in the literature, and our case appears to be the seventh.

Key Words: heterotopic pancreas, adenocarcinoma, jejunum

Introduction

Heterotopic pancreatic tissue has been found in several abdominal and intrathoracic locations, most frequently in the stomach or duodenum1). Heterotopic pancreas in the jejunum or ileum is rare, and malignancies in heterotopic pancreas are rare. We report a case of cancer of heterotopic pancreatic tissue in the jejunum and discuss it based on a review of the literature.

Case Report

A 78-year-old woman came to our hospital on March 8, 2005 because of abdominal pain, nausea, and vomiting. A plain X-ray of the abdomen revealed intestinal obstruction, and the patient was admitted for further examination. The physical examination on admission revealed a body height of 153 cm, weight of 37.5 kg, temperature of 35.9°C, blood pressure 106/60mmHg, and a regular pulse of 59 beats/min.
The palpebral conjunctivae and bulbar conjunctivae showed no evidence of anemia or jaundice. No physical abnormalities were observed in the chest. No tumors or the liver or spleen were palpable.

There were no abnormal laboratory findings except signs of anemia (RBC $320 \times 10^6/\mu l$, Hb 7.4 g/dl, Ht 23.0%).

A plain abdominal X-ray showed gas and a niveau in the small intestine (Fig. 1).

An abdominal CT scan showed no evidence of a mass in the abdomen.

The symptoms and niveau improved in response to conservative therapy, but the symptoms of intestinal obstruction recurred after she consumed a meal. Based on this finding, the patient was diagnosed with intestinal obstruction of unknown etiology, and surgery was performed on March 27, 2005.

Intraoperatively, a tumor was palpated in the jejunum 50 cm distal to the ligament of Treitz. The tumor had invaded the entire thickness of the wall and was exposed beyond the serosa. The proximal bowel was extremely dilated (Fig. 2), and partial jejunectomy was performed to relieve the obstruction.

Gross inspection revealed a yellowish-brown protruding tumor, measuring $3 \times 2$ cm. The tumor was covered with intact mucosa, and resembled a submucosal tumor (Fig. 3). Histopathological examination showed that the
tumor was developed from an aberrant pancreas composed of ducts, acini, and smooth muscle tissue (Fig. 4). The tumor was located in the submucosa. The tumor consisted of moderately differentiated adenocarcinoma and was developed from the aberrant pancreas (Fig. 5). On the basis of these findings, the tumor was diagnosed as cancer of heterotopic pancreatic tissue in the jejunum.

The postoperative course was uneventful, and the patient was discharged on postoperative day 17.

**Discussion**

“Aberrant pancreas” is defined as pancreatic tissue lacking anatomical and vascular continuity with the normally located pancreas, and it is usually an incidental finding at upper abdominal laparotomy or autopsy. Malignant transformation has been reported in aberrant pancreas as well as in normally located pancreas, and most cases of cancer in aberrant pancreatic tissue have been reported to be in the stomach or duodenum.

The pathogenesis of aberrant pancreas is unknown, but according to the “misplacement theory” it arises when fragments of pancreas become separated during rotation of the foregut and develop into mature elements, or it may arise as a result of pancreatic metaplasia of endodermal tissue that ends up in the submucosa during embryonic life. The misplacement theory may explain the occurrence of the heterotopia in the upper gut near the pancreas, but it does not explain its occurrence in the colon.

Aberrant pancreas usually does not produce clinical symptoms until it causes obstruction or gastrointestinal bleeding or ulceration. Our patient had abdominal pain, nausea, and vomiting due to obstruction of the jejunum. We are usually unable to diagnose cancer in aberrant pancreatic tissue preoperatively, because most jejunal tumors are malignant lymphomas, leiomyosarcomas, or carcinomas.

Pancreatic heterotopia in the gastrointestinal tract is most common in the submucosa. Pancreatic heterotopia is found at 2% to 15% of all autopsies. The most frequent sites of heterotopic pancreas are the gastric antrum (30%), duodenum (30%), jejunum (20%), and Meckel’s diverticulum (5%). Unusual locations are the colon, spleen, liver, biliary tract, mesentery, and lymph nodes.
A Case of Heterotopic Pancreatic Cancer in the Jejunum

Table 1  Review of documented cases of the heterotopic pancreatic cancer in the jejunum

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Location (distance beyond the ligament of Treitz)</th>
<th>Size</th>
<th>Type*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Persson</td>
<td>1988</td>
<td>85</td>
<td>Female</td>
<td>100 cm</td>
<td>1.5 cm</td>
<td>I</td>
</tr>
<tr>
<td>Fujiki</td>
<td>1990</td>
<td>54</td>
<td>Male</td>
<td>50 cm</td>
<td>3 × 2 cm</td>
<td>I</td>
</tr>
<tr>
<td>Satoh</td>
<td>1993</td>
<td>64</td>
<td>Male</td>
<td>110 cm</td>
<td>2 × 1.5 cm</td>
<td>I</td>
</tr>
<tr>
<td>Makhlouf</td>
<td>1999</td>
<td>71</td>
<td>Male</td>
<td>unknown</td>
<td>3.5 cm</td>
<td>I</td>
</tr>
<tr>
<td>Makhlouf</td>
<td>1999</td>
<td>61</td>
<td>Male</td>
<td>8 cm</td>
<td>1.5 cm</td>
<td>I</td>
</tr>
<tr>
<td>Arao</td>
<td>1999</td>
<td>63</td>
<td>Male</td>
<td>20 cm</td>
<td>4 × 2 cm</td>
<td>II</td>
</tr>
<tr>
<td>Sato</td>
<td>2007</td>
<td>78</td>
<td>Female</td>
<td>50 cm</td>
<td>3 × 2 cm</td>
<td>II</td>
</tr>
</tbody>
</table>

* According to the Heinrich classification.

Based on its histological features, aberrant pancreas has been classified by Heinrich into 3 types: type I, differentiated aberrant pancreatic tissue containing ducts, acini plus endocrine islets; type II, incompletely differentiated aberrant pancreatic tissue, composed predominantly of ducts plus a few acini; and type III, composed of smooth muscle tissue and ducts only, lacking acini and islets, so-called adenomyoma. Our case corresponded to type II in the Heinrich classification.

Malignancy in heterotopic pancreas is rare. Unquestionable malignant transformation was found in only 1 (0.7%) of the 146 cases of heterotopic pancreas, including in surgical and autopsy specimens, retrieved from the files of the Institute Universitaire de Pathologie of Lausanne of 109 patients diagnosed as having pancreatic heterotopia in the gastrointestinal tract between 1970 and 1997. A literature search yielded 28 reports of carcinoma arising in a heterotopic pancreas. Moreover, it is extremely rare for cancer in aberrant pancreatic tissue to be located in the jejunum. Six cases of cancer in aberrant pancreatic tissue in the jejunum have been reported in the literature so far, and our case may be the seventh. All 7 cases are reviewed in Table 1.

**Conclusion**

We have reported a case of cancer in aberrant pancreatic tissue in the jejunum and discussed its diagnosis and treatment based on a review of the literature.

**References**

2) Barbosa de Castro JJ, Dockerty MB, Waugh