

## CASE REPORT

# Heterotopic Pancreas in the Esophagus: What Do We Know?—A Review of the Literature



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**Heterotopic pancreatic tissue is a rare phenomenon, which may occur in locations outside of the pancreas within the gastrointestinal tract. Of these locations, involvement in the esophagus is quite rare. It is paramount to improve our understanding regarding heterotopic pancreas, as some lesions may carry malignant potential. With this article, we present a case of heterotopic pancreas in the distal esophagus and review the current literature.**

**Keywords:** Heterotopic pancreas; esophagus; congenital malformation; dysphagia; benign stricture

### Introduction

Heterotopic pancreas (HP) is a rare congenital malformation, which is usually discovered incidentally. It has been previously noted in various parts of the gastrointestinal tract; however, involvement in the esophagus is rare.

### Case Report

Patient is a 59-year-old female with a past medical history of asthma, narcolepsy, prediabetes, and pancreatitis

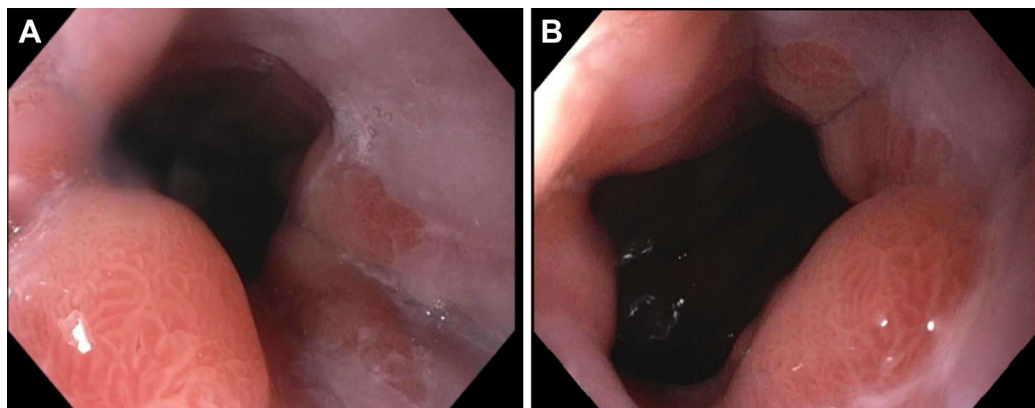
who presented with right upper quadrant pain radiating to the shoulder and nausea. Patient denied any hematemesis, melena, dysphagia or odynophagia, change in bowel movements, or weight loss. She denied any non-steroidal anti-inflammatory drugs or blood thinner use. Routine blood work was unremarkable. Esophagogastroduodenoscopy was performed, which revealed grade A esophagitis (Figure 1).

Subsequent tissue biopsy revealed evidence of active reflux esophagitis and granulation tissue consistent with focal pancreatic heterotopia (Figure 2).

Given the patient's active inflammation, she was treated with proton pump inhibitor with improvement in symptoms. Patient was told to continue routine surveillance of her esophagitis.

### Discussion

HP is a congenital malformation defined as pancreatic tissue that is not anatomically or vascularly connected to the normal pancreatic body.<sup>1</sup> HP can occur in any part of the digestive tract, but it is most commonly found in the stomach (30%), the duodenum (17%–36%),



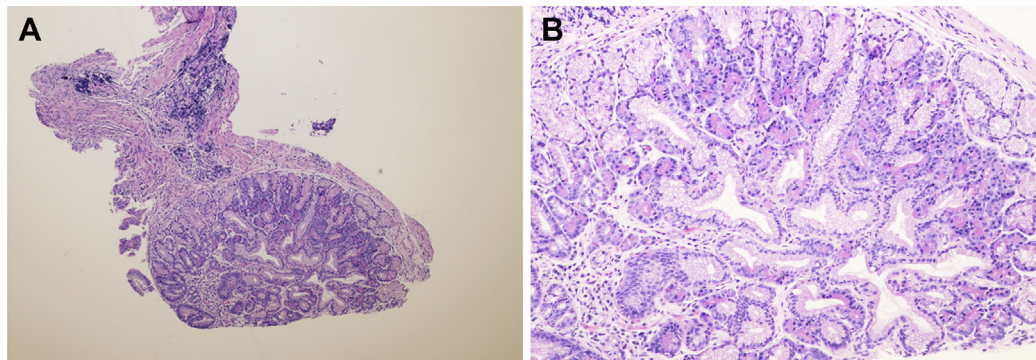
**Figure 1.** (A, B) EGD showing area of distal esophagus with visible evidence of inflammation and grade A esophagitis.

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**Figure 2.** (A, B) Histopathology demonstrating foci of acinar pancreatic tissue with surrounding inflammatory cells in the background of distal esophageal squamocolumnar epithelium.

and the jejunum (15%–21%).<sup>1</sup> It is most often diagnosed incidentally on autopsies with a prevalence between 0.5% and 13% and is more commonly found in men.<sup>2</sup>

There are several theories proposed regarding the pathogenesis of HP, including the totipotent cell theory, the pancreatic metaplasia theory, and misplacement theory. The totipotent cell theory assumes that the endodermal cells

**Table.** Review of Cases of Heterotopic Pancreas in the Esophagus Including Patient Symptomatology, Location of Lesion, Management of Lesion (if Provided) and Malignant Potential

Case	Age	Sex	Symptoms	Location	Management	Malignancy	Reference
1	24	F	Chest pain, vomiting, shortness of breath	Distal esophagus	Esophageal enucleation procedure	None	Temes et al <sup>4</sup>
2	25	M	RUQ abdominal pain	Distal esophagus	VATS	None	Lowry et al <sup>5</sup>
3	58	F	Worsening dysphagia	Distal esophagus	Thorascopic ILE	IPMN	Crighton et al <sup>6</sup>
4	47	F	Vomiting, epigastric pain	Distal esophagus	ILE, Witzel jejunostomy with pyroloplasty	None	Noffsinger et al <sup>20</sup>
5	63	M	Asymptomatic	Middle esophagus	No surgical intervention, conservative	None	Goto et al <sup>7</sup>
6	34	M	Worsening odynophagia and dysphagia	Distal esophagus	Partial fundoplication	None	Ulrych et al <sup>8</sup>
7	26	F	Nausea and epigastric pain	Distal esophagus	Laporoscopic excision followed by Partial Dor fundoplication	None	Gananadha et al <sup>9</sup>
8	25	M	Vomiting and abdominal pain	Distal esophagus	Enucleation via right thoractomy	None	Salo et al <sup>10</sup>
9	45	M	Progressive dysphagia	Distal esophagus	Thoracoabdominal esophagogastrectomy	Anaplastic cancer	Roshe et al <sup>11</sup>
10	43	M	Upper GI bleeding	Distal esophagus/lesser curvature of stomach	Enucleation of tumor from esophagus	None	Razi et al <sup>12</sup>
11	52	M	Intermittent dysphagia	Distal esophagus	No surgical intervention, conservative	None	Shalaby et al <sup>13</sup>
12	60	M	Dysphagia and epigastric pain	Distal esophagus	Tumor resection with proximal stomach resection	Adenocarcinoma	Guillou et al <sup>14</sup>
13	38	F	Heartburn and nausea	Distal esophagus	Endoscopically assisted laparoscopic resection of tumor	None	Garn et al <sup>15</sup>
14	29	M	Dysphagia and epigastric pain	Distal esophagus	No treatment mentioned	None	Salim et al <sup>16</sup>
15	41	F	Dysphagia and epigastric pain	Distal esophagus/lesser curvature of stomach	No treatment mentioned	None	Rodriguez et al <sup>17</sup>
16	70	M	Heartburn, nausea and abdominal bloating	Distal esophagus	Endoscopic resection	None	Shamoon et al <sup>18</sup>
17	73	F	Epigastric pain, vomiting	Distal esophagus	Endoscopic resection	None	Filip et al <sup>19</sup>

GI, gastrointestinal; ILE, ileoscopy; RUQ, right upper quadrant; VATS, video assisted thorascopic surgery.

of the bowel wall have the potential to differentiate into pancreatic tissue. The pancreatic metaplasia theory stipulates that during embryogenesis, pancreatic tissue migrates to intestinal submucosa. The misplacement theory assumes that pancreatic tissue is randomly lost throughout the developing primitive foregut.<sup>2</sup>

Although heterotopic pancreatic tissue may not be in close proximity to the pancreas, it is susceptible to similar pathology, such as acute or chronic inflammation.<sup>3</sup> Several reports have demonstrated pseudocyst and fibrotic features of heterotopic pancreatic tissue. Furthermore, evidence of premalignant lesions and adenocarcinoma has also been identified, although most cases were from stomach and duodenum.<sup>3</sup>

HP in the esophagus is a rare entity. It has previously been reported only 17 times in the medical literature<sup>4–19</sup> (Table).

Of the 17 cases reported, 16 of them were found in the distal esophagus/gastroesophageal junction. Although the condition is typically asymptomatic, it can cause epigastric pain, dysphagia, and upper gastrointestinal bleeding. Based on previous reports that have suggested the presence of malignancy in some of these lesions, adequate stratification of patients into those who are surgical candidates vs those who could have active surveillance is necessary. Furthermore, given that most of the previous cases underwent surgical or endoscopic intervention, there are no clear guidelines on the management of patients with asymptomatic and benign cases such as our case. Should these patients receive antireflux therapy? Should they undergo surveillance endoscopy every 1–3 years? Should the lesion be pre-emptively excised given potential for pre-malignancy? These questions will need to be answered eventually to better serve patients with pancreatic heterotopia.

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The corresponding author, on behalf of all authors, jointly and severally, certifies that their institution has approved the protocol for any investigation involving humans or animals and that all experimentation was conducted in conformity with ethical and humane principles of research.