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Esophageal Heterotopic Pancreas in an Asymptomatic 2-Year-Old With VACTERL Association

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Abstract: A 2-year-old male with VACTERL association and asthma presented to the emergency room due to asthma exacerbation. Chest radiography revealed lingular pneumonia and thickening of the left paraspinal line of the gastroesophageal junction. Chest computed tomography confirmed a heterogeneous fluid- and gas-filled structure at the left posterior lateral posterolateral aspect of the esophagus, which was suspected to be an esophageal diverticulum on an upper gastrointestinal series. The esophageal diverticulum was excised via left thoracoscopy, and pathological examination revealed pancreatic tissue. Heterotopic pancreas lacks anatomical, vascular, or ductal continuity with the native pancreas. It is usually asymptomatic, but when discovered, it usually occurs later in life. It has been described in the foregut, but is not as common in the esophagus, especially in the pediatric population. This case report highlights the rare occurrence, and importance of considering, esophageal heterotopic pancreas within an esophageal diverticulum in an asymptomatic patient with VACTERL association.

INTRODUCTION

Heterotopic pancreas (HP), also known as pancreatic rest, is ectopic pancreatic tissue that lacks anatomic, vascular, or ductal continuity to the native pancreas (1). The most common locations are the stomach, duodenum, and proximal jejunum (2). It is less commonly described within the ileum, esophagus, a Meckel's diverticulum, biliary tree, intestinal mesentery, and spleen (2). Although most patients with HP are asymptomatic, some may present with epigastric or abdominal pain, or on occasion, with gastrointestinal (GI) bleeding (2,3). Given its asymptomatic presentation, it is estimated to be inadvertently discovered during 1 in 500 upper abdominal surgeries and present in 0.55%–13.7% of autopsies (4). There is a male preponderance with a peak disease incidence during the fourth to sixth decades of life (3). We present the case of an asymptomatic 2-year-old male with VACTERL association and an esophageal diverticulum harboring HP. Heterotopic pancreas has been previously reported in patients with VACTERL association, specifically in those with esophageal atresia. Most cases were found in the stomach, some within the esophageal pouch (5–7). However, this is the first case report of a patient with VACTERL association presenting with an asymptomatic esophageal diverticulum harboring ectopic pancreatic tissue.

CASE REPORT

The patient is a 23-month-old male born full term, at 39 1/7 weeks gestation, via cesarean section. He was diagnosed with a high anorectal malformation, for which he underwent a sigmoid colostomy and mucus fistula on the second day of life. During initial workup, the patient underwent placement of an orogastric tube with radiographic confirmation of its location in the stomach, confirming he did not have a tracheoesophageal fistula with esophageal atresia. Given these findings, no further esophageal workup was pursued. His VACTERL workup also revealed Type I caudal regression syndrome (an anomalous L4 vertebral butterfly body, blunted conus medullaris, an absent lower sacrum and coccyx), patent ductus arteriosus, patent foramen ovale, bilateral pelvocaliectasis, and left vesicoureteral reflux. Despite these findings, his stay in the neonatology intensive care unit was uncomplicated, and he was discharged within 2 weeks.

He returned electively at 4 months of age to undergo a posterior sagittal anorectoplasty with ligation of a rectogenitourinary fistula. At 7 months of age, the patient underwent colostomy reversal. He is followed closely at the pediatric surgery clinic for bowel management due to chronic constipation.

He was diagnosed with asthma at approximately 8 months of age and prescribed albuterol as needed. The patient presented to the emergency department at 17 months of age because of persistent shortness of breath and wheezing, unresponsive to albuterol. The workup included a 2-view chest radiograph, revealing left lingular pneumonia and thickening of the left paraspinal line at the junction of the lower chest and upper abdomen. Given the latter findings, a chest computed tomography (CT) with intravenous contrast was performed, revealing a heterogeneous fluid- and gas-filled structure at the left posterolateral aspect of the esophagus, and corresponding to the convex thickened left paraspinal line evident on the chest radiograph (Fig. 1). Diagnostic considerations in this age group include a hiatal hernia, esophageal diverticulum, or duplication cyst. However, the assessment was limited due to the lack of oral contrast. He was discharged on oral antibiotics for pneumonia. He was to follow up with his primary care provider and gastroenterologist regarding the findings on the chest CT.

The patient presented for a routine pediatric surgery visit 2 months later. Chest CT findings were noted, and an upper GI series confirmed a left distal esophageal diverticulum. No prior studies existed for comparison. Given the comprehensive information obtained from these studies, and the asymptomatic nature of the diverticulum, a need for preoperative tissue biopsy was not deemed necessary. Therefore, he underwent excision of the esophageal diverticulum via minimally invasive thoracoscopic surgery allowing for a diagnostic and therapeutic approach to his care. He was discharged home on postoperative day 4 (Fig. 2).

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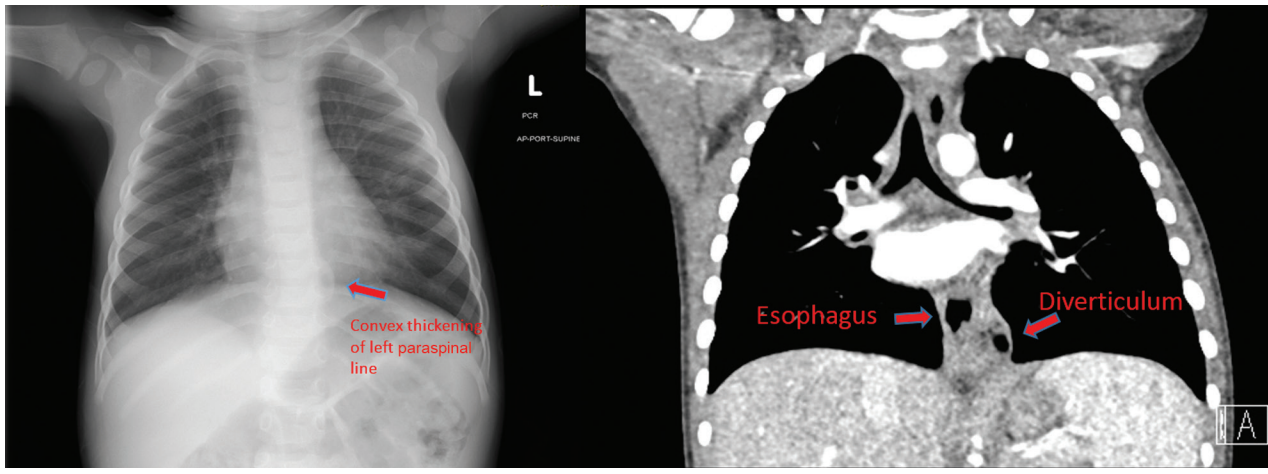

FIGURE 1. Frontal chest radiograph demonstrates focal convex thickening of the left paraspinal line at the lower chest, near the level of the diaphragm hiatus. Coronal CT image demonstrates separate foci of air within the esophagus and diverticulum.

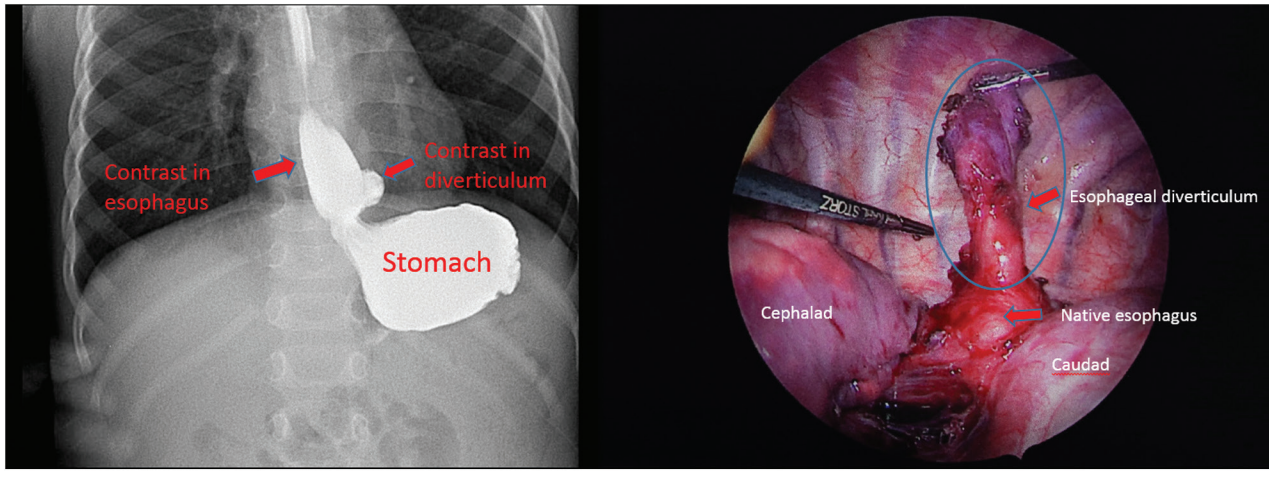

FIGURE 2. Frontal image obtained during the contrast ingestion phase of the upper GI series demonstrates contrast within the esophagus, diverticulum, and stomach. Thoracoscopic view of the esophageal diverticulum before surgical excision.

Overall, the patient continues to do well. However, he remains on treatment for his asthma and has had occasional emergency room visits postoperatively due to shortness of breath and wheezing.

Pathological assessment of the esophageal diverticulum revealed a cystic structure with gastric transitional and fundic mucosa, accompanied by submucosal lymphoid follicles, chronic inflammation, scattered cartilaginous plates, and heterotopic pancreatic tissue (Fig. 3). The presence of peripheral cartilaginous plates supports the diagnosis of a cystic diverticular foregut duplication while the presence of pancreatic acinar structures suggests the heterotopic nature of this pancreatic tissue.

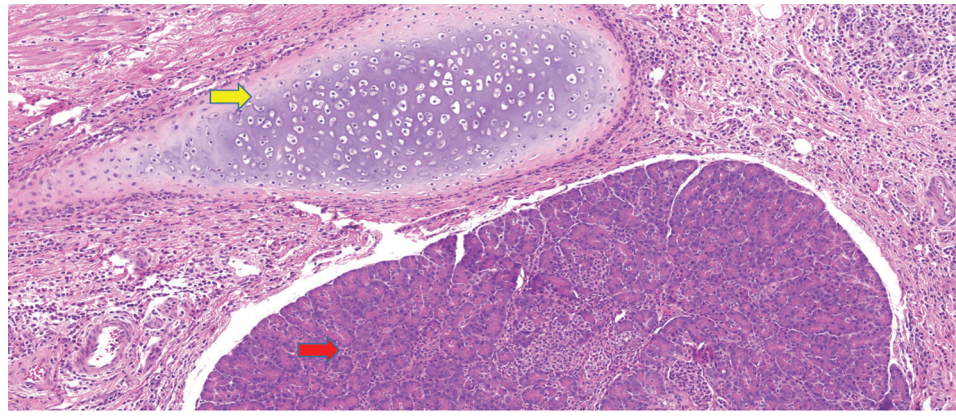

FIGURE 3. 100× total magnification. H&E stain. Scattered lobules of hyaline-type cartilage (yellow arrow) identified in regional fibrocollagenous tissues. Foci of heterotopic pancreatic tissue noted (red arrow).

DISCUSSION

Heterotopic pancreas was first described in 1727 by Jean Schultz (8,9). It is ectopic pancreatic tissue with no anatomic, vascular, or ductal connections to the main pancreatic gland (1,10). However, the embryological basis is poorly understood. The most widely held theory, the misplacement theory, postulates that small branches of the pancreatic anlagen remain on the foregut during embryonic rotation and migrate with growth (1,2,8,11). This theory accounts for heterotopic pancreatic tissue mainly located near the upper GI tract, from derivatives of the foregut (8). Given the asymptomatic nature of HP, the true incidence is difficult to determine. Heterotopic pancreatic tissue functions similarly to the native pancreas, hence, patients can develop pancreatitis, pseudocysts, cysts, bleeding from ulceration, gastric outlet obstruction, intussusception, and in some instances, malignancy (2,4,5,12). Although malignant transformation is rare, occurring in 0.7–1.8% of cases, various types of tumors have been described including intraductal papillary mucinous neoplasm (a precursor lesion), inflammatory pseudotumor, adenocarcinoma, islet tumor, cystadenocarcinoma, and anaplastic cancer (2,4,10,13,14). Unlike pediatric patients, adults harboring HP tend to have associated symptoms such as epigastric or right upper quadrant pain, dysphagia, hematemesis, nausea, vomiting, or rarely, weight loss (1–3,6). Heterotopic pancreas has been described in several locations along the GI tract. The stomach, duodenum, and proximal jejunum are the most common locations (2). It is less commonly described within the ileum, a Meckel's diverticulum, biliary tree, intestinal mesentery, and spleen (2). Esophageal HP has also been reported, although rare, with approximately 16 adult and 5 pediatric cases reported in the English literature (1,6,10,12). Of the cases reported in pediatric patients, several have been associated with variants of esophageal atresia, with HP associated with the proximal or distal esophageal pouch (6,7). These have been diagnosed during infancy, due to the nature of the esophageal congenital anomaly, with the use of upper endoscopy at the time of the operative intervention and during routine postoperative endoscopic surveillance. This case highlights esophageal HP in a toddler with VACTERL association and an asymptomatic esophageal diverticulum.

CONCLUSION

Heterotopic pancreas has been described within the stomach, duodenum, and proximal jejunum, but rarely in the esophagus. Of the cases reported in the English literature, very few have been reported in the pediatric population. We present the case of a child with VACTERL association who was ultimately discovered to have ectopic pancreatic tissue located within an asymptomatic esophageal diverticulum. Although rare, HP should be considered in patients with congenital esophageal anomalies, regardless of the symptomatology.

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