Case Report

A Retroperitoneal Neuroendocrine Tumor in Ectopic Pancreatic Tissue

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Abstract

Ectopic pancreas is the relatively uncommon presence of pancreatic tissue outside the normal location of the pancreas. We report a case of abdominal pain due to retroperitoneal neuroendocrine tumor arising from heterotopic pancreatic tissue between the duodenal wall and the head of the pancreas. Patient underwent surgical enucleation of the tumor.

Keywords: ectopic pancreas; neuroendocrine tumor; endoscopic ultrasonography; retroperitoneal tumor

INTRODUCTION

Ectopic pancreas (EP) is the relatively uncommon presence of pancreatic tissue outside the normal location of the pancreas. This condition is usually asymptomatic and rarely complicated by pancreatitis or malignant transformation. It is defined as pancreatic tissue abnormally situated, without connection to the normal pancreas, but provided with its own vascular and ductal systems. EP tissue has been found in both abdominal and extra abdominal locations, but is most frequently encountered in the stomach (25%-60%), duodenum (25%-35%) and rarely in mesocolon and Meckel’s diverticulum. Other terms such as heterotopic and aberrant pancreas have also been described in the literature. Retroperitoneal, EP tissue appearing as bilateral suprarenal masses has been reported once in 2009 but a neuroendocrine tumor (NET) in a retroperitoneal EP tissue has not been reported before. Gaspar Fuentes et al. modified von Heinrich’s classification of pancreatic heterotopias. Type I was total pancreatic heterotopia with all pancreatic cell types present, Type II is composed of pancreatic ducts only (the so called canalicular variety), Type III comprises acinar tissue only (exocrine pancreas) and Type IV is made up of islet cells only (endocrine pancreas). The histological classification of the heterotopic pancreas in our case is Type I because all elements were present, although the epithelial component dominated and less frequently the ductal component.

The origin of EP tissue is unknown; it is possible that during rotation of the foregut and fusion of the ventral and dorsal parts of the pancreas in early fetal life, small pieces of tissue become detached from the forming organ leading to entrapment in different locations.

CASE REPORT

A 72-year-old male was presented by dull aching epigastric and right hypochondrial pain radiating to the back. He is not known to be diabetic or hypertensive. His pulse rate and blood pressure were normal. The physical examination was unremarkable, with no palpable abdominal masses. Lab investigations were normal.

Upper endoscopy revealed a smooth bulge at the mid second part of the duodenum with normal underlying mucosa, likely due to compression from outside. Abdominal ultrasound and computed tomography (CT) showed a well-defined right lumbar mass in-between the second part of the duodenum and the head of the pancreas, but appears separated from them. Common bile duct and pancreatic duct are not dilated. No lymphadenopathy was found. Endoscopic ultrasound (EUS) showed a localized paraduodenal echogenic mass measuring 58 mm × 67 mm with a well-defined line of separation form the duodenal wall and the head of the
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pancreas (Fig. 1). It shows elasticity score 4 denoting its firm consistency (Fig. 2). EUS-guided fine needle aspiration (EUS-FNA) and cell-block preparation was carried out. Histopathological examination revealed the presence of compact groups of epithelial cells having central rounded nuclei with occasional presence of nucleoli and moderate eosinophilic granular cytoplasm separated by delicate stroma showed many small vascular spaces. Immunohistochemistry revealed positive immunostaining for synaptophysin and chromogranin (Fig. 3). The decision for surgical intervention was taken. On laparotomy, a retroperitoneal well encapsulated mass was found between the wall of the duodenum and the head of the pancreas and separated from them. Enucleation of the tumor was performed and pathological examination confirmed the FNA diagnosis.

DISCUSSION

Several theories had been implicated to describe the origin of EP tissue. The most accepted one implicates migration and/or rests of branching pancreatic tissue buds from the developing pancreas during embryogenesis. This theory is most likely when one considers gastrointestinal pancreatic heterotopia; however, pancreatic tissue occurring in more exotic, anatomically remote sites, such as the thyroid gland or fallopian tube, may have a different pathogenesis. The possibility of origin from teratomas has been advocated in these instances.

The pancreatic ectopic tissue is usually presented by gastric pain and patients may also complain of bleeding, nausea and vomiting and chest pain. The mechanism for the production of symptoms is unclear. Armstrong et al. found a correlation between the presence of symptoms, size of the lesion (greater than 1.5 cm) and extent of mucosal involvement.

EP typically appears on a radiographic barium study or endoscopic examination as a submucosal mass with central umbilication. On CT, an EP most frequently appears as a well-defined oval or round mass with smooth or serrated margins in the gastric antral wall or intestinal wall or as a mesenteric mass similar to a gastrointestinal stromal tumor or carcinoid tumor.

EUS is typically used to evaluate the submucosal lesions in the upper gastrointestinal tract. EP usually appears hypoechoic and heterogeneous with indistinct margins during EUS and the most commonly originates from the third or fourth layer or a combination of these two layers of the gastrointestinal tract. EUS also allows the use of targeted FNA biopsy, particularly helpful for the final diagnosis. Although cytological examinations are inconclusive in about

Figure 1. Endoscopic ultrasound picture of the mass

Figure 2. Elastography of the mass (elasticity score 4)

Figure 3. Tumor cells showing positive immunostaining for chromogranin (A) and synaptophysin (B) (×400)
50% of cases, in this case EUS-FNA gave the same result as the excisional biopsy as being NET.

We report, to our knowledge, the first case of a NET in a retroperitoneal EP tissue. Heterotopic pancreas should be considered in the differential diagnosis of a retroperitoneal masses as well as a submucosal mass of the gastric, duodenal, jejunal and esophageal wall and it has the same line of management as pancreatic NET, which is surgical resection that should remain the mainstay of treatment for patients with a localized disease.

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