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Retroperitoneal Infrarenal Ectopic Pancreas - An Autopsy Case

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Abstract

Ectopic pancreas is pancreatic tissue lacking anatomical and vascular continuity with the normally located pancreas. Ectopic pancreas is relatively uncommon and usually occurs in the stomach or duodenum. We report the case of a 55-year-old female with retroperitoneal infrarenal ectopic pancreas incidentally. Retroperitoneal and Infra-renal ectopic pancreas has not previously been documented.

***Keywords**: Retroperitoneal, Infrarenal, Ectopic pancreas, Autopsy.*

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Introduction

An ectopic pancreas is defined as pancreatic tissue lacking vascular or anatomic communication with the normal body of the pancreas. Ectopic pancreas is an uncommon finding, with an estimated incidence of 0.55–13.7% according to autopsy analyses. [1] It is rarely symptomatic as it is found incidentally at laparotomy or during autopsy most of the time. Most ectopic pancreatic lesions are found in the gastrointestinal tract and frequently involve the stomach (26–38% of cases), duodenum (28–36%), jejunum (16%) and, less commonly, Meckel's diverticulum or the ileum. [1–3] Ectopic pancreas in the mediastinum and lung has also been documented. [4, 5] It is prudent to differentiate it from neoplastic etiologies, as simple surgical excision can potentially be curative. We discuss the presentation, diagnosis, and treatment of an interesting case of ectopic pancreas presenting as a gastric antral tumor. However, ectopic pancreas in the retroperitoneum only 2 cases are reported till date with an extremely unusual presentation may be misdiagnosed as adrenal tumors. Herein, we report a case of retroperitoneal infrarenal ectopic pancreas which may be the 1st case to best of our knowledge.

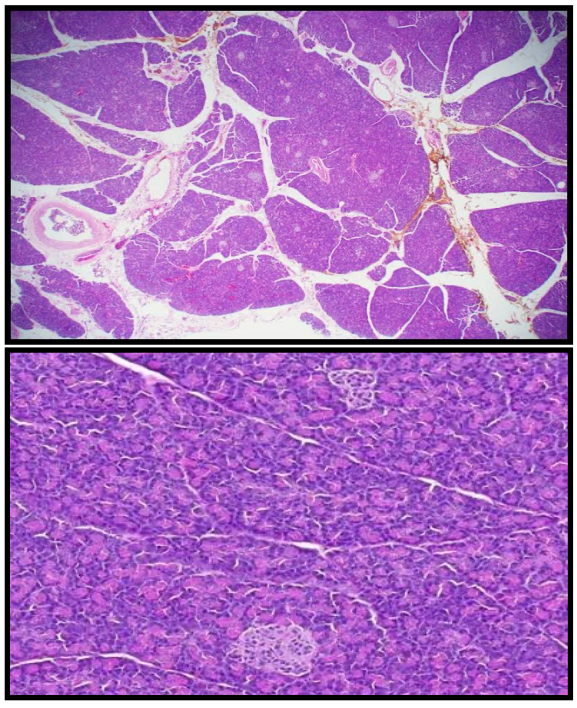
Case Report

A 55-year-old female hypertensive patient visited our hospital because of complaints of altered sensorium and distention of abdomen with pain, high grade fever, nausea and vomiting 3 months' duration. A physical examination showed no abnormal findings. His pulse rate was 74 beats per minute and his blood pressure was 150/80 mmHg. Laboratory investigations revealed Renal function tests were deranged whereas Liver function tests were normal. Clinical diagnosis was metabolic encephalopathy with diabetes mellitus in known case of Autosomal Dominant Polycystic Kidney Disease. Inspite of all medical treatment, patient succumbed to death. Clinical postmortem was performed. Bilateral Kidneys were Enlarged, polycystic, each showing multiple cysts of varying sizes filled with hemorrhagic and serous fluid. Below the lower end of kidney, a

thickened fibrofatty mesenteric mass was found measuring 7x4x2 cm and cut surface showed firm, brownish appearance, which was considered as neoplastic mass. In liver small cyst filled with seromucinous material, Stomach is filled with a huge blood clot measuring 15x12x10 cm, Duodenum shows hemorrhagic areas with necrosis at ampulla of Vater. And Cerebral vessels shows evidence of Berry aneurysm.

Figure 1: Polycystic kidneys &ectopic pancreas

Fig 2: Normal Pancreas Histology (10X) & (20X)



Discussion

Ectopic pancreas is also referred to as pancreatic heterotopia, heterotopic pancreas, and accessory or aberrant pancreas. [6] It is defined as pancreatic tissues lacking vascular or anatomic communication with the normal body of the pancreas, yet possessing histological features of pancreatic acinar formation, duct development, and islets of Langerhans with independent blood supply and ductal system. [7-9] Ectopic pancreas is a relatively rare entity with an incidence of 0.2% at laparotomy [10,11] and 0.5 to 13.7% on autopsies. [12] Multiple theories have been implicated on the embryological origin of this rare condition. One of them implicates the persistence of a duodenal evagination involved in the normal development of the pancreas. This remnant can migrate with the developing gastrointestinal tract accounting for its various locations. [13] Another theories suggests pancreatic metaplasia of the endodermal tissue in the gastric mucosa. [13] Ectopic pancreata can be found in various anatomical sites such as the stomach, duodenum, jejunum, gallbladder, esophagus, common bile duct, spleen, mesentery, mediastinum, and fallopian tubes. The stomach is the most common location (25 to 38%), with pancreatic rests most frequently found in the submucosa (75% of cases) but sporadically also in the muscularis propr. and serosa. [14] Based on Heinrich's 1909 classification and the subsequent Gasper-Fuentes modification in 1973, there are four types of pancreatic heterotopia, and they are as follows [15]

- Type I—typical pancreatic tissue with acini, ducts, and islet cells similar to the normal pancreas.
- Type II (canalicular variety)—pancreatic ducts only.
- Type III (exocrine pancreas)—acinar tissue only.
- Type IV (endocrine pancreas)—islet cells only.

The patient in our case report was a type I, with all components of the organ. Although ectopicpancreata are usually an incidental finding during autopsy or intraoperatively, they can present with nonspecific symptoms such as abdominal pain, abdominal fullness, nausea, vomiting, anorexia, weight loss, anemia, and melena. [16] Abdominal pain is the most common symptomatic presentation, and can be explained by the inflammation and irritation of surrounding tissues secondary to the secretion of pancreatic enzymes and hormones [17] Pain can also be explained as a result of hemorrhage in

the lesion due to mucosal erosion and ulcer formation, especially when it occurs in the small intestine. [18] Patients can also present with symptoms of gastric outlet obstruction as in our patient with larger lesions at the gastric antrum, especially those greater than 1.5 cm in size. [19] Published literature reports have described rare instances of malignant changes in the ectopic pancreatic tissue. [20] Radiological studies such as barium swallows can some- times assist in the diagnosis by showing nonspecific fold thickening with rounded filling defects and a typical central indentation. [21]

Endoscopic-guided fine needle aspiration is usually superficial and nondiagnostic as these tumors are covered with normal gastric mucosa. However, a positive biopsy can be diagnostic. [22] The recent advent of EUS has improved the sensitivity of diagnosis for these lesions by 80 to 100%. [23,24] Kim et al; [25] reported that EUS features of ectopic pancreata that differentiate them from mesenchymal tumors include the following: larger longest/shortest diameter ratio, antral location, mural growth pattern, third (submucosal) layer disruption, irregular margins, and intermediate echogenicity. However, none of these findings individually or collectively are diagnostic of ectopic pancreata, as they can be seen with any neoplastic lesion of the bowel wall and still require operative resection to confirm the diagnosis. At pathology, the gross appearance of an ectopic pancreas includes the presence of a characteristic central ductal orifice. [26] Histopathological examination reveals pancreatic acini, ducts, islets of Langerhans, and intervening connective tissue. The preoperative diagnosis of an ectopic pancreas is still challenging despite advanced diagnostic tools, and the frequent inability to differentiate it from neoplastic lesions warrants surgical excision. Until now only two cases of retroperitoneal pancreas reported in literature. Despite keen clinical acumen and technological advances in diagnostic tools, the final diagnosis of ectopic pancreas in our case was made after excision on histopathology in an autopsy study, hence explaining the "diagnostic dilemma."

Conclusion

In conclusion, to best of our knowledge although retroperitoneal ectopic pancreas is a rare entity, in that particularly retroperitoneal infrarenal ectopic pancreas may be first case. Clinicians should consider this as an important differential diagnosis of extramucosal gastric lesions and retroperitoneal mass as a simple excision can be potentially curative.

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