ORIGINAL RESEARCH PAPER

Gastroenterology

KEY WORDS: Ampulloma, Ampulla of Vater, Ectopic pancreas, Pancreatico-duodenectomy.

ECTOPIC PANCREAS PRESENTING AS AMPULLOMA WITH OBSTRUCTIVE JAUNDICE: CASE REPORT AND REVIEW OF LITERATURE.

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ABSTRACT

Pancreatic tissue situated away from the normal pancreas, with one's own ductal system and vascular supply had been defined ectopic pancreas. These are found incidentally either in imaging or autopsy with 90% of them situated in stomach, duodenum and upper jejunum with less common sites like Meckel's diverticulum, Gallbladder, Umbilicus, CBD, Spleen, Fallopian tube, intracranial and Mediastinum. Their presence in the Ampulla of Vater had been previously reported in only 23 case reports and series worldwide with clinical presentation similar to periampullary tumor. Our patient, a 58-year-old female, presented with upper abdomen pain, obstructive jaundice, pruritis, pale stools and weight loss with abnormal blood values. Imaging in the form of abdominal ultrasound, CECT, MRI, MRCP and EUS (endoscopic ultrasound) showed prominent ampulloma of Vater with underlying obstruction of bile duct with dilated CBD and intrahepatic biliary radicals. Surgical resection of ampulloma in the form of Pancreatico-duodenectomy was done and HPE showed ectopic pancreas within the ampulla. Ectopic pancreas has varied symptoms according to its location with literature evidence of malignant potential. Preoperative biopsy and peroperative frozen section histopathology evaluation of indeterminate lesions of ampulla are warranted for exact diagnosis. Local Surgical excision is considered only in symptomatic individuals.

Case Report:

A 58-year-old female patient, presented with complaints of epigastric abdomen pain for three months, deep yellow discoloration of eyes and high colored urine for a month, generalized pruritis and pale stools for two weeks with weight loss of about 5kg in that same period.

On physical examination, she had epigastric abdomen tenderness, jaundice and acholic stools per rectum. On blood investigations she had normal hematology and total count, altered LFT with high bilirubinemia; total bilirubin 5.6 mg/dl (normal 0.2 to 1.2mg/dl), with direct bilirubin 4.1 mg/dl (normal 0-0.6 mg/dl), ALP 390 U/L (Normal 20-120 U/L), SGOT 122 U/L (normal 5-40 U/L), SGPT 104 U/L (normal 5-40 U/L) and normal coagulation profile.

Tumor markers Serum CEA was 3.95 ng/ml (normal 0-5 ng/ml), CA 19-9 value of 17.88 U/L (normal 0-37 U/L) and AFP was 3.4 ng/ml (normal 0-20 ng/ml). Imaging with abdominal ultrasound showed dilated CBD of 1.8cms, dilated extrahepatic with intrahepatic biliary radicals and multiple cholelithiasis. MRI with MRCP showed ill-defined mass lesion in the ampulla with upstream dilatation of CBD, CHD along with intrahepatic biliary ducts and multiple cholelithiasis Figure 1.

This ill-defined ampulloma was hyperintense on T1 and hypointense on T2 weighted images. CECT showed as hypodense ampulloma of size 5 x 6mm with arterial enhancement on intravenous contrast administration and upstream dilation of CBD, CHD with intrahepatic biliary ducts and multiple 6 to 7 radio dense calculi of size 0.8 to 1.5cms within the gallbladder Figure 1.

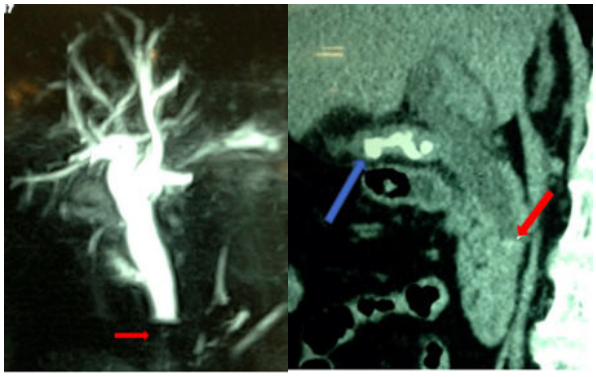

Figure 1. Mrcp And Contrast Ct Abdomen Shows Ill-defined Ampulloma (red Arrow) With Dilated Cbd & Hepatic Ducts And Multiple Gb Stones.

Upper gastro-duodeno-scopy showed a prominent ampulloma whose biopsy was normal without any conclusive pathology. EUS showed prominent hypoechoic lesion of size 1.1 x 0.8cms within the submucosa of ampulla with muscularis propria invasion and normal surface mucosa, CBD was dilated above the mass lesion with 1.5cms diameter. EUS guided FNA from the ampulloma revealed benign epithelial cells arranged in sheets and clusters with background neutrophils, lymphocytes with no overt malignant cells. The pathological diagnosis was indeterminate. Considering the clinical and symptom complex, a malignant periampullary tumor cannot be excluded, hence she was offered radical surgical resection.

Pancreatico- duodenectomy was done and the gross specimen on evaluation showed bulky ampulloma (figure 2). Postoperatively she tolerated diet from 3rd POD, drains removed on 5th POD and she got discharged on 8th POD.

The HPE showed Ectopic pancreas with components of pancreatic ducts with acini (Figure 3). On routine post-surgery follow up, patient was relieved of her symptoms, and lab values were normal within a month. At present, she is symptom free for almost two years.

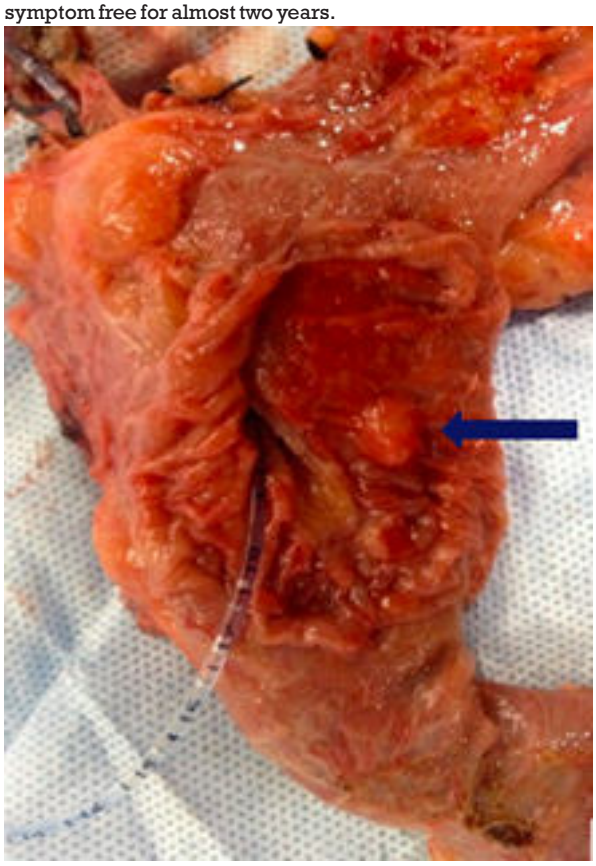

Figure 2. Whipple Pancreatico-duodenectomy specimen shows prominent Ampulloma (blue arrow) in the Ampulla of Vater region, separate from orthotopic pancreas and distal CBD.

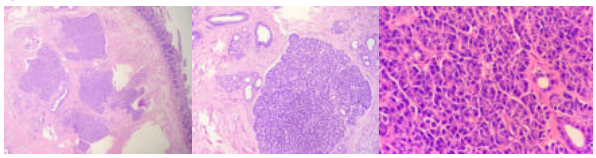

Figure 3. HPE of Ampulloma showing pancreatic ducts with acini of the ectopic pancreas (Magnification A: 10X ; B:100X ; C: 400X) interlacing with smooth muscle bundles separate from normal pancreas (Heinrich type II)

DISCUSSION:

The term heterotopic or ectopic pancreas indicates well-developed and organized normal pancreatic tissue, present outside its orthotopic position, devoid of any anatomical and vascular connections with pancreas proper. Its reported incidence in gastrointestinal tract was frequent, especially the distal stomach, anywhere in duodenum or proximal jejunum, however rarely seen at unusual places such as Meckel's diverticulum, gallbladder, umbilicus, mediastinum, fallopian tube and rarely Ampulla of Vater[1]. In 1727, Shultz reported the first identified case on ectopic pancreas. The incidence frequency of such reported cases had been 0.55% to 13.7%, 0.25%, and approximately 1.2% in autopsy, abdominal surgery, and gastrectomy operation respectively[1]. They may rarely complicate as acute as well chronic pancreatitis, chronic pseudocyst formation, intraluminal bleeding, bowel perforation, intussusception, gastric outlet obstruction, and, rarely, malignant transformation. There had been so far 23 case reports or series highlighting heterotopic pancreas arising from Ampulla of the Vater [1-23] and 2 case reports of ampullary Acinar cell carcinoma as progression from ectopic pancreas with symptomatic obstructive jaundice[24,25]. Ectopic pancreas from gastrointestinal tract are usually asymptomatic, rarely it produces nonspecific clinical symptoms, depending on its location, size, infiltration of deep layers or overlying mucosa[26]. Common symptoms were abdomen pain (45.5%) and epigastric discomfort (12%) and those arising from ampulla rarely causes bile duct obstruction and jaundice. In our case, the symptoms were intermittent abdomen pain, jaundice, weight loss with deranged LFT suggesting a possibility of peri-ampullary malignancy.

On imaging with ultrasonography, CECT scan or endoscopy, ectopic pancreas mostly present as submucosal swelling covered by normal mucosa, that can be easily mistaken as gastrointestinal stroma tumour (GIST) or leiomyoma[28]. On MRI, they appear hyperintense or isointense lesions compared to orthotopic normal pancreas on unenhanced T1-weighted images and isointense or hypointense lesions on T2-weighted images. On dynamic MRI, heterotopic pancreas appears isointense compared to orthotopic pancreas on arterial phase images[27]. In our patient CECT and MRI/MRCP did not help in establishing the diagnosis. An infrequent though characteristic finding in endoscopy of ectopic tissue within the submucosa is the presence of central umbilication[5]. At EUS, the ectopic tissue was usually seen to arise from submucosa as heterogenous, hypoechoic lesion with irregular indistinct margins mimicking leiomyoma[5,10]. More often preoperative endoscopic or radiological diagnosis of ampulloma with ectopic pancreas are difficult. Similarly, the ampulloma of our patient was smooth, bulky, hypoechoic, with size 1.1 x 0.8cm on EUS and no central umbilication noted on endoscopy. EUS guided FNA (fine needle aspiration) has been valuable in diagnosis of upper GIT lesions[30]. As these ectopic tissues are mostly located in submucosal region (76%) with sporadically appearing in the muscular (15%) and subserosa (9%), there is always high risk of false negative biopsy results[29]. In these submucosal lesions, endoscopic biopsy is seldom useful too. In our case, EUS-FNA showed benign epithelial cells.

The best treatment for a well diagnosed ectopic pancreas is local excision when sufficient, rather than radical surgery[17]. However, pancreaticoduodenectomy was done in around 64% of the previous series due to difficulties in establishing preoperative diagnosis or due to suspicion with underlying malignancy[17]. It was evident that Ampullectomy provided complete cure, as no further treatment was needed due to benign nature of pathology, diagnosed either by EUS [5] or peroperative frozen section biopsy [4] or rarely pre-operative endoscopic biopsy [22], from previous literature. In our patient, with an indeterminate periampullary tumour with underlying suspicion of malignancy, we offered her pancreaticoduodenectomy. Frequency of malignant transformation from ectopic pancreas was studied by Guillou et al. between 1975 and 1991. He found that out of 146 patients including surgical and autopsy specimens, there was 0.7% incidence of the malignant transformation, hence concluding an extremely rare phenomenon[32]. Similarly in a study done by Makhlouf et al. on 109 patients, between 1970 and 1997, who presented with pancreatic heterotopia, only two patients reported adenocarcinoma[33]. In one separate published case report, an Ampulloma mimicked cholangiocarcinoma [11]. In all malignant transformation of ectopic pancreas, patients invariably presented with specific symptoms. The histopathological classification of ectopic pancreas was first described by Heinrich in the year 1909. He classified them into three types, with type I composed of all type pan-cells, type II composed of ducts with acini and no islets of Langerhans, and type III composed of ducts only. This classification was later modified by Gaspar Fuentes et al in 1973 into four types as type I (total heterotopia) composed of all cell types, type II (canalicular heterotopia) composed of ducts only, type III (exocrine heterotopia) composed of acinar cells only and type IV

(endocrine heterotopia) composed of islets of Langerhans only[31]. In our case, HPE showed pancreatic ducts and acini suggestive of Heinrich Type II heterotopia.

CONCLUSION:

Ectopic pancreas presenting as ampulloma with progressive obstructive jaundice, pruritus or weight loss is an entity mimicking periampullary malignancy which remains difficult to diagnose, in spite of advances in imaging and endoscopic techniques. Asymptomatic patients can be observed if the benign lesion can be ascertained by imaging and EUS guided biopsy. If symptomatic, local excision by ampullectomy is recommended and a morbid procedure like Pancreatico-duodenectomy should be avoided. Though rare, Ampulloma due to ectopic pancreas should be considered a differential diagnosis of periampullary tumours, presenting in young to middle aged patients.

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