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Ectopic Pancreatic Islets In Splenic Hilum : Very Rare Case Report

KEYWORDS: heterotopic, hilum, islets

Joyshree Panda	Shubhankar mishra	Debi Prasad Mishra
Junior resident, department of pathology, MKCGMedical College and Hospital, Berhampur, Odisha	Junior resident, Department of pediatrics MKCGMedical College and Hospital, Berhampur, Odisha	Professor, Department of pathologyMKCGMedical College and Hospital, Berhampur, Odisha

ABSTRACT *Ectopic pancreatic tissue (or heterotopic pancreatic tissue) refers to the situation where rests of pancreatic tissue lie outside and separate to the gland without anatomic and vascular continuity with the normal pancreas. It is common in stomach, duodenum and jejunum. Splenic hilum is an uncommon site, and more uncommon is finding of islet cells in it. A 40 year old female patient with Sickle cell anaemia trait and massive splenomegaly underwent splenectomy. Gross examination of spleen showed a nodule near the hilum, which on histosection showed pancreatic tissue having ducts, acinus and islet cells.*

Introduction

Heterotopic pancreas (HP) also referred to as ectopic pancreas, pancreatic choristoma, or pancreatic rest, is defined as the presence of pancreatic tissue in an anomalous location without any anatomic, vascular, or neural continuity with the main body of the normal pancreas. The first case of ectopic pancreas was reported by Schultz in 1729. The frequency of ectopic pancreas has been estimated as one per 500 laparotomies and 0.6% to 13.7%of autopsies [1,2]. . Ectopic pancreas may often remain asymptomatic and is diagnosed incidentally [2]. Microscopically, these pancreatic remnants may resemble normal pancreatic parenchyma with organised acini, ducts and pancreatic islets. In most instances, the islets are rich in A cells and poor in PP cells (dorsal type), but in others the reverse is true (ventral type) [3] . Every pathologic change that occurs in the orthotopic pancreas can occur in its heterotopic counterpart, including acute pancreatitis and neoplasms of either exocrine or endocrine type [4]. There are reported cases of intrasplenic mucinous cystadenomas and cystadenocarcinomas have arisen in heterotopic pancreatic tissue.[5]

Case Report

A 40 Year female having Sickle cell anaemia (heterozygous) came with history of recurrent blood transfusions and dull aching pain in left hypochondrium. The pain was intermittent and associated with vomiting occasionally. She had multiple episodes of vaso-occlusive crisis before. For which she was admitted to our hospital multiple times .On examination patient had severe pallor and mild icterus. On abdominal examination she had mild hepatomegaly and moderate splenomegaly of 5cm below the left costal margin. Complete blood count showed Hemoglobin of 4.7gm/dl TLC was 10,300 and other haematological parameters were normal. Ultrasonography of abdomen showed hepatosplenomegaly. She was given blood transfusion and advised for splenectomy. Then the patient was splenectomised and spleen was sent for histopathologic examination. Grossly the Spleen measured 20 x12 x8cm. It was well encapsulated with smooth surface. Cut section was grossly normal at different areas except near the hilum where a nodule of size 0.3cm diameter was noted in the subcapsular region.

Histology of the spleen showed congestive splenomegaly. The blood vessels showed presence of sickled red blood cells. And the section from the nodule showed the presence of pancreatic cell rest containing pancreatic duct in splenic tissue (fig 1). The cells of pancreas were arranged in small lobules with a small central lumen. A pale staining area was also identified (fig 2). Acinar cells with the islet cell of pancreas were identified (fig 3).

Discussion

Heterotopia of pancreatic tissue is a relatively frequent congenital anomaly. It may occur anywhere in the gastrointestinal tract. Most frequent locations are the stomach, duodenum or the proximal part of small intestine[2] .The lesion is also found in Meckel's diverticulum, the biliary tract, the gallbladder, the liver, the spleen, and other sites within the abdominal cavity. Heterotopic Pancreas has been noted in the stomach (24-38%), duodenum (9-36%), jejunum (0.5-27%), ileum (3-6%), and Meckel's diverticulum (2-6.5%)[6]. Ectopic (heterotopic, rest) pancreatic tissue may occur from displacement of small amounts of pancreas during embryologic development, resulting in the formation of a nodule independent from the vascular supply or anatomic connection to the pancreas. It often has a proper ductal system and circulation [7] . According to Arey and Haffer's theory the pancreas is formed in the 4th week from three primitive endodermal evaginations (buds) of the anterior intestine. The right ventral evagination fuses with the dorsal one and become the body, tail and upper part of the pancreas head. The lower part of the head and processus uncinatus stems from the left ventral evagination. Before this fusion there is a rotation of the ventral part of the pancreas, during which the buds are in close contact with the distal stomach and proximal duodenum, allowing engrafting of pancreatic germinal cells, from which histological components of the pancreas may develop One of the evaginations can also remain within the bowel wall and can be carried along with the longitudinal growth of the intestine. This allows heterotopic tissue to be formed far from the normally located pancreas. In these cases, a higher incidence of female patients between 40 and 50 years of age was observed in heterotopic pancreas that were located in gallbladder, bile ducts, splenic hilum, or liver. Preoperative diagnosis is rarely possible, either clinically or radiologically, as it is a very uncommon pathological entity, in which microscopic examination confirms the diagnosis. Most pa-

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tients are completely asymptomatic.It may be symptomatic (e.g., epigastric pain, nausea and vomiting, abdominal fullness, obstruction, ulceration, tarry stools, weight loss), but the majority of cases are incidental findings. Grossly, it may be evident as firm, pale, nodular mass. Microscopically, the usual lobular architecture is maintained with variable admixture of acini, islets and ductal structures. The presence of ectopic islets without any accompanying acini or ducts is quite uncommon . Microscopically, heterotopic pancreas has been classified into three types by von Heinrich—Type 1: Ectopic tissue with acini, ducts, and islets of Langerhans; Type 2: Ectopic tissue containing only a few acini and ducts, with absent endocrine elements—incomplete arrangement; Type 3: Ectopic tissue with only proliferating excretory ducts and absent exocrine acini and endocrine elements[2] Our case was considered to be Type 1, based on the Heinrich classification.

Conclusion

Search of literature showed only few reported case of islet cells in splenic hilum. So this case was apt for reporting, so that it can be added to literature. The pathologists must keep this in mind while reporting splenic sections for correct diagnosis and treatment.

Figures

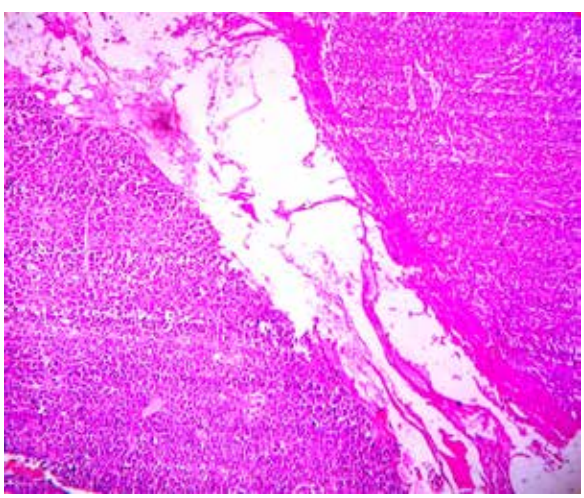

fig1; view showing splenic tissue with pancreatic tissue

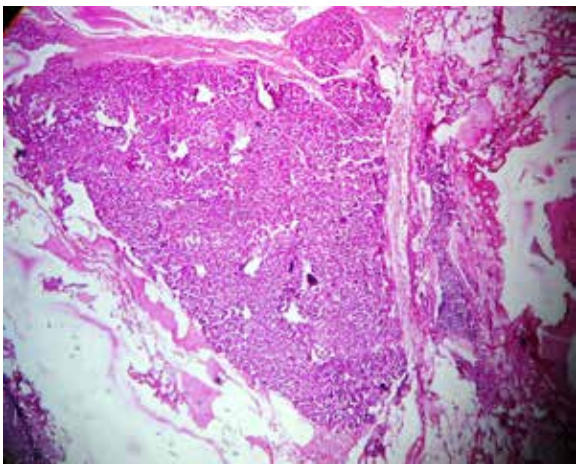

fig2; view showing pancreatic tissue beneath splenic capsule.

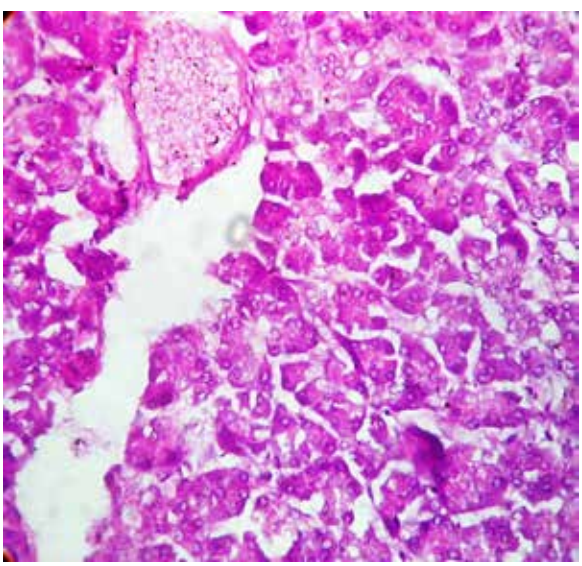

fig3; showing pancreatic ducts,acini and islet cells.

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