An unusual case of heterotopic pancreas in the umbilical sinus of a child

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ABSTRACT

Background: Heterotopic pancreas is the presence of pancreatic tissue outside its normal location, mostly in the gastrointestinal tract. It is rare and most patients are asymptomatic adults. The presence of heterotopic pancreas in the umbilicus and the pediatric age group is even more rare.

Case Presentation: An 18-month-old boy presented with clear umbilical discharge since birth. On examination, there was a swollen and inflamed umbilicus. Exploration and excision of the umbilical lesion were done. Histopathological examination of the lesion reported as umbilical sinus with the presence of heterotopic pancreas. He was discharged well with no complications seen on the follow-up review.

Conclusions: Heterotopic pancreas is rare. Awareness of this condition is important in order for the patient to be managed accordingly.

Keywords: Heterotopic pancreas, umbilicus, pediatric, umbilical sinus, case report.

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Background

Heterotopic pancreas is the presence of pancreatic tissue outside its normal location, without anatomical or vascular connections. It is also called aberrant, accessory, or ectopic pancreas. It was first described in 1729 by Jean-Shultz. The most common sites of the heterotopic pancreas are the stomach, jejunum, duodenum, and ileum [1]. Other unusual sites reported before are gallbladder [2], mesenteric [3], and mediastinum [4]. From the literature review, less than 20 cases of heterotopic pancreas in the umbilicus have been reported. The heterotopic pancreas is rare and most patients are asymptomatic. The incidence is even more rare in the pediatric population. We report a case of a rare condition of heterotopic pancreas in a child with umbilical discharge.

Case Report

An 18-month-old boy presented to us with persistent umbilical discharge since birth. However, the parents have never sought medical treatment until now. On examination, his umbilicus was inflamed and swollen with clear discharge seen. A diagnosis of a patent urachal cyst was made. An ultrasound abdomen showed that there is no communication between the umbilicus and the dome of the urinary bladder. An exploration of the umbilicus was done; the umbilical was fully excised and reconstructed. Intra-operatively the lesion was inseparable from the umbilical skin but there was no connection between the umbilical to the bowel or bladder. The medial, lateral, and median umbilical ligaments were all obliterated. The patient was discharged well the next day. Microscopically, the tissue is composed of a sinus tract with invagination lined by stratified squamous and intestinal-type epithelium (Figures 1 and 2). The subepithelial stroma shows a heterotopic pancreatic tissue comprising of pancreatic acinar cells, ducts, and islets of Langerhans (Figure 3). Thus, the umbilical tissue was diagnosed as umbilical sinus with the heterotopic pancreas. The patient was reviewed at 2 months. The wound has healed completely and there were no complications detected. He had no other symptoms and we did not check for any other ectopic pancreatic tissue sites. A review of literature for heterotopic pancreas was done and presented below.

Discussion

The heterotopic pancreas can be histologically classified into the Heinrich system [5]. Type 1 contains acini, islets, and ducts. Type 2 contains acini and ducts only. Type 3 contains ducts alone. In the umbilicus, type 1 is most commonly encountered and this is the case for our current patient.

The heterotopic pancreas functions like a normal pancreas. Therefore, any diseases of the pancreas may also develop

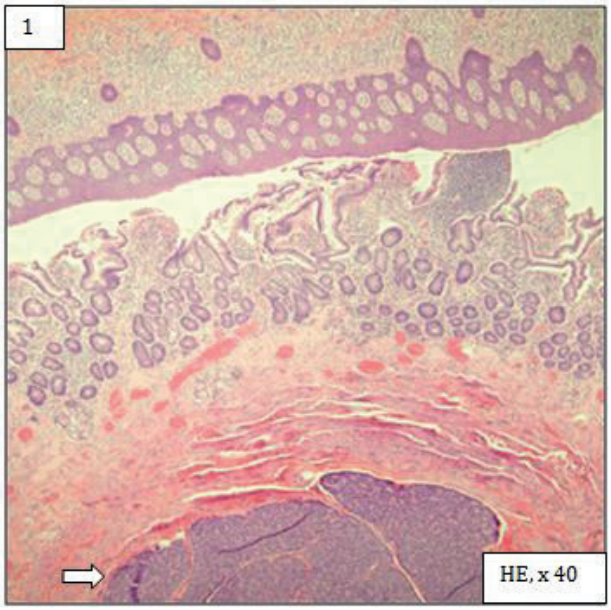

***Figure 1**. The sinus tract is partially lined by stratified squamous epithelium and the invaginated part is lined by intestinal-type epithelium. The white arrow shows a heterotopic pancreatic tissue within the stroma.*

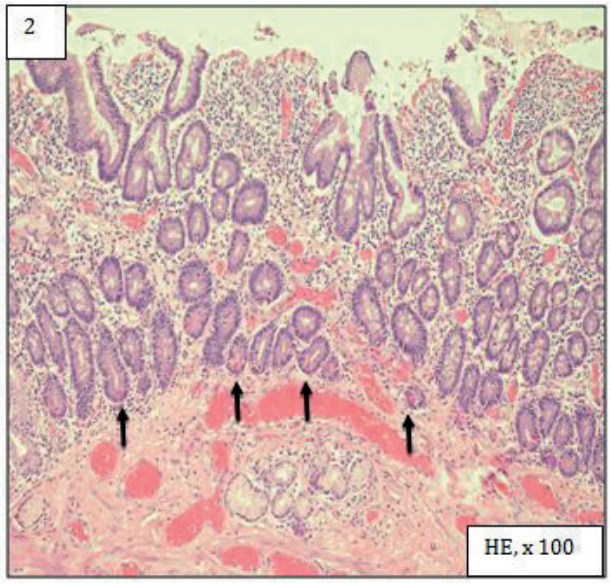

***Figure 2**. The intestinal epithelium is composed of mucin secreting columnar cells with goblet cells and occasional paneth cells (black arrow).*

in the heterotopic pancreas. Pancreatitis, pancreatic cyst, neuroendocrine tumor, and adenocarcinoma of the pancreas have all been reported to occur in the heterotopic pancreas [6]. Therefore, although this lesion is benign, a follow up is recommended if it was incompletely excised. Our patient presented with clear umbilical discharge and was initially thought to be a patent urachal cyst. This discharge could be secreted pancreatic enzymes or hormones from the heterotopic pancreatic tissue. Unfortunately, we are unable to prove this as no fluid sample was taken for analysis.

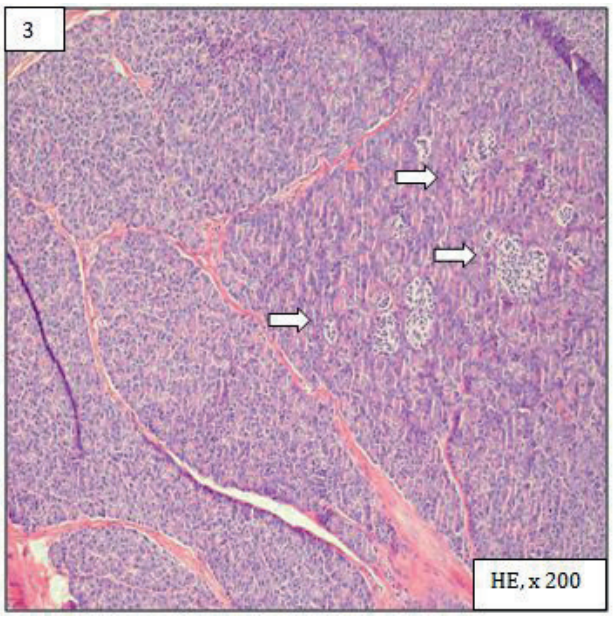

***Figure 3**. The pancreatic tissue exhibits acinar cells and islets of Langerhans (white arrows). Pancreatic ducts are not seen in this photograph.*

Patients with heterotopic pancreas are usually asymptomatic, although some patients may present with pain or gastrointestinal bleeding. Diagnosing heterotopic pancreas is difficult as it is rarely thought to be a differential diagnosis due to its rarity. Most of the time it is found as an incidental finding after surgery, as in our current case study, or after autopsy. Radiological imaging is also not specific and this condition is rarely diagnosed pre-operatively. Rezvani et al. [7] suggested that the most common computed tomographic appearance of the heterotopic pancreas is that of a small oval intramural mass with microlobulated margins and an endoluminal growth pattern. However, it is not applicable in our case as the lesion on the umbilicus and no prior CT scan was done pre-operatively. Management of the heterotopic pancreas is usually depending on the presence of symptoms and complications. Management of asymptomatic lesions found incidentally is debatable.

Conclusion

As a conclusion, the heterotopic pancreas is rare. Awareness of this condition is important in order for the patient to be managed accordingly.

What is new?

The heterotopic pancreas is rare. The heterotopic pancreas in the umbilical sinus of a child is even more rare. From the literature review, only 20 cases have been reported previously.

Funding

None.

Conflict of interests

The authors declare that there is no conflict of interest regarding the publication of this article.

Consent for publication

Consent was not obtained from the parents of this patient as this is an anonymous case report.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Boy, 18-month old
2	**Final diagnosis**	Heterotopic Pancreas in Umbilical Sinus
3	**Symptoms**	Umbilical Discharge
4	**Medications**	N/A
5	**Clinical procedure**	Exploration and excision of the umbilical lesion
6	**Specialty**	General Surgery