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■ BRIEF CASE REPORT ■

Heterotopic Pancreas in Omphalomesenteric Duct Remnant Results in Persistent Umbilical Discharge

Eunhyang Park · Hyojin Kim · Kyu Whan Jung[1] · Jin-Haeng Chung

Departments of Pathology and [1]Pediatric Surgery, Seoul National University Bundang Hospital, Seoul National University College of Medicine, Seongnam, Korea

Umbilical discharge in infancy is a common pediatric problem and usually attributed to infection or an umbilical granuloma. However, it is important to investigate if such discharge is due to an underlying congenital abnormality such as umbilical hernia ulceration, urachal remnant, or omphalomesenteric duct remnant, because corrective surgical intervention may then be required. Omphalomesenteric duct remnant can cause umbilical discharge generally through patency between the gut and umbilicus. However, though rare, umbilical discharge may be due to the presence of heterotopic pancreas. The prevalence of omphalomesenteric duct remnant is only 2% of the population, and most of them remain asymptomatic. The present case is an infant with persistent umbilical discharge caused by heterotopic pancreatic tissue in a remnant omphalomesenteric duct. To the best of our knowledge, this is the first such case report in Korea.

CASE REPORT

A 3-month-old female infant presented with persistent umbilical discharge since birth. The infant was born through normal vaginal delivery following an uneventful gestational period and had no congenital anomalies. She had been gaining weight well and had no family history of genitourinary or gastrointestinal problems. Ultrasonography of the abdomen revealed an isoechoic tract posterior to the umbilicus, and the diagnosis of urachal remnant was suspected. On physical examination, small droplets of clear fluid constantly discharged from a normal-looking umbilicus. Laboratory examination results were within normal limits. Under general anesthesia, an incision was made below umbilicus. Surgical exploration showed a fibrous sinus posterior to the umbilicus which was attached to the inner aspect of the umbilicus and the outer wall of the ileum by a fibrous band. Fibrous tissue was excised close to both ends, and the rest was ligated by suture tie. The excised specimen was a 7×6×5-mm-sized whitish fibrous tissue. Histologically, the excised specimen included pancreatic tissue with some small intestinal mucosa and fibrous extracellular components (Fig. 1). Both exocrine and endocrine pancreatic tissues were observed, including acini, ducts, and islets of Langerhans. Acini were separated into lobules by connective tissue. Intercalated ducts were lined by simple low cuboidal epithelium (Fig. 2). The patient was discharged without any postoperative complications and is currently alive without any sequelae.

DISCUSSION

The omphalomesenteric duct is a long narrow tube that connects the yolk sac to the midgut lumen of the developing fetus. It normally regresses during the 5th to 9th weeks of fetal development, but a part or all of it may persist postnatally and result in various abnormalities including a Meckel's diverticulum, an umbilical fistula, an omphalomesenteric duct cyst, an umbilical sinus, or an umbilical polyp. Meckel's diverticulum often contains heterotopic gastric or pancreatic mucosa which can result in some clinical manifestations such as massive rectal bleeding. However, heterotopic tissue in other types of remnant omphalomesenteric duct which present on the umbilicus has been rarely reported. To our knowledge, there have been 13 cases of

Corresponding Author
Jin-Haeng Chung, M.D.
Department of Pathology, Seoul National University Bundang Hospital,
82 Gumi-ro 173beon-gil, Bundang-gu, Seongnam 463-707, Korea
Tel: +82-31-787-7713, Fax: +82-31-787-4012, E-mail: chungjh@snu.ac.kr

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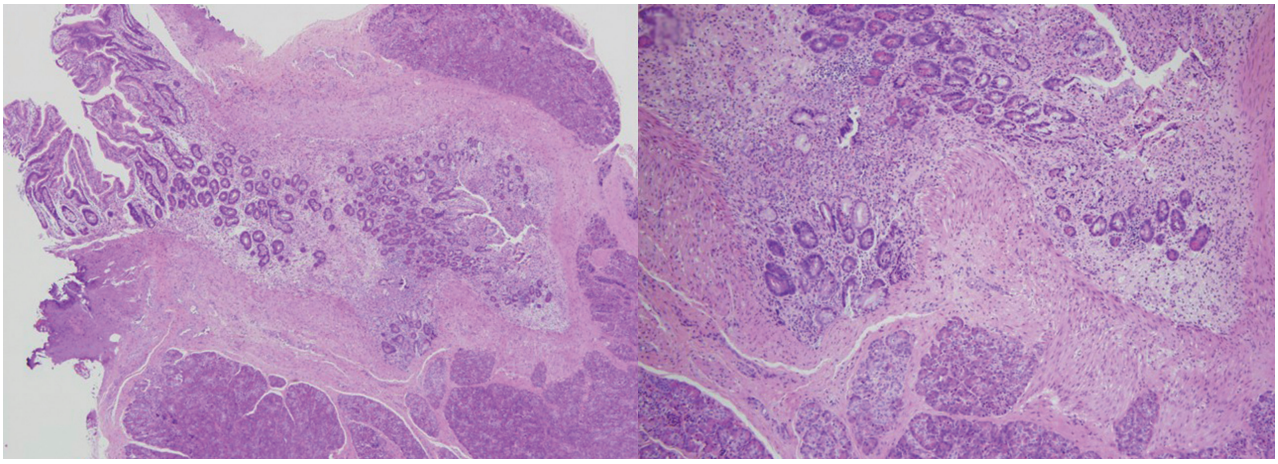

Fig. 1. Pancreatic tissue (upper and lower) with some small intestinal mucosa (middle) and fibrous extracellular components.

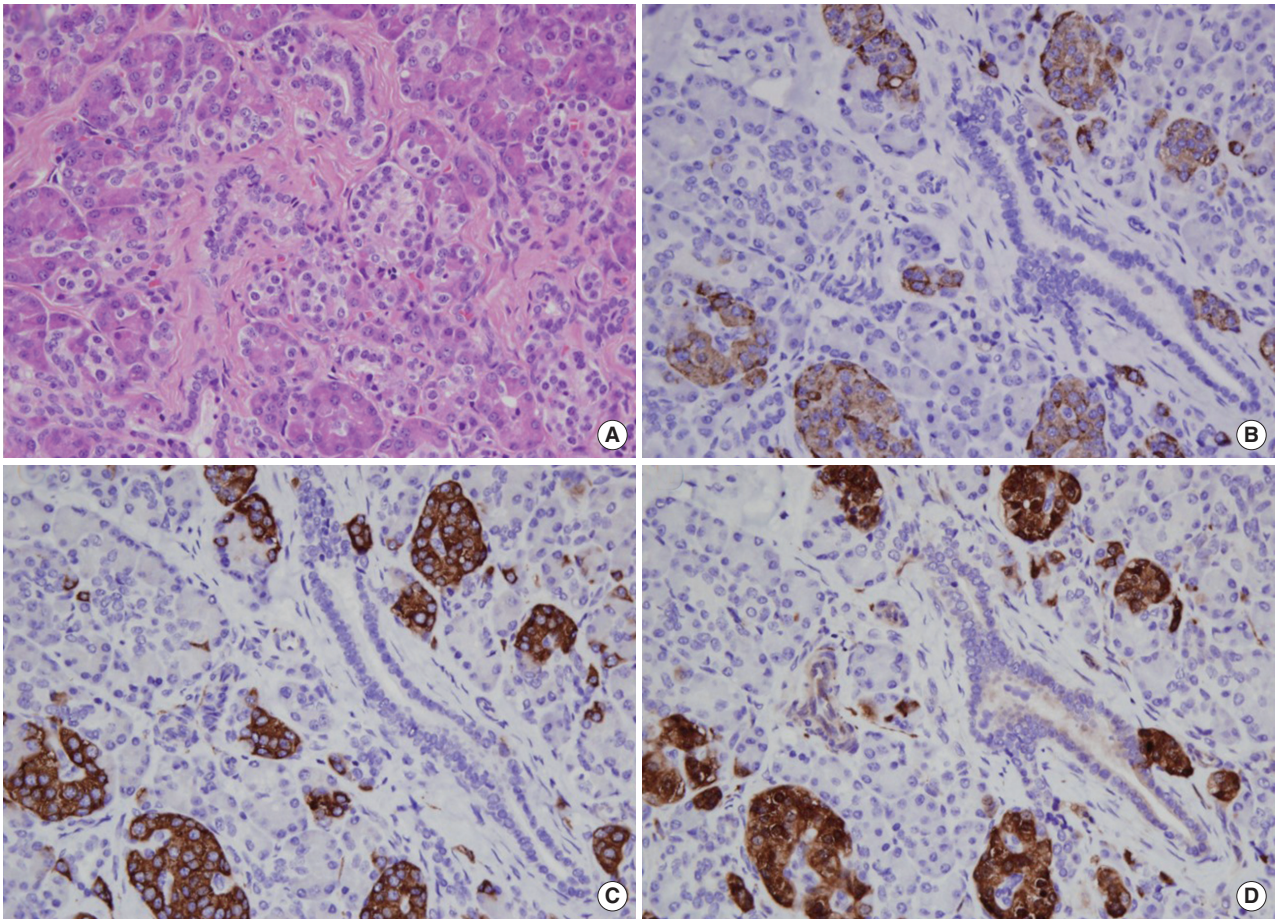

Fig. 2. Acini are separated into lobules by connective tissue, and intercalated ducts are lined with simple low cuboidal epithelium. Pancreatic tissue including acini, ducts, and islets of Langerhans (A). Expression of chromogranin (B), synaptophysin (C), and neuron-specific enolase (D) in islets of Langerhans.

heterotopic pancreatic tissue in the umbilicus (Table 1).[1-4]

Various explanations have been offered for heterotopic pancreas in the umbilicus, but there is no universally accepted theory about the cause of this aberrant tissue.[5] The three influential pathogeneses include misplacement theory,[6] in which embryonic tissue is located in an inappropriate place and develops

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Table 1. Cases of heterotopic pancreatic tissue at the umbilicus reported in the English literature

No.	Age/Sex	Mass	Discharge	Size	Site	Reference
1	12 yr/F	N/A	N/A	N/A	Umbilical subcutaneous tissue	Wright (1900), cited by Harris and Wenzl[1]
2	22 yr/M	–	+	N/A	Umbilical cyst	Trimingham (1943), cited by Harris and Wenzl[1]
3	6 mo/M	–	N/A	3-mm nodule	Umbilical nodule	Steck and Helwig (1964), cited by Avolio *et al*.[2]
4	13 mo/M	+	+	12×9×5 mm	Umbilical mass	Caberwal *et al*. (1977), cited by Avolio *et al*.[2]
5	60 yr/M	+	–	N/A	Umbilical polyp	Kondoh *et al*. (1994), cited by Avolio *et al*.[2]
6	8 mo/M	+/–	+	N/A	Umbilical mass	Avolio *et al*.[2] (1998)
7	15 mo/M	–	+	N/A	Umbilical mass	Avolio *et al*.[2] (1998)
8	6 mo/M	–	+	N/A	Urachus	Perez-Martinez *et al*. (1999), cited by Lee *et al*.[3]
9	3 mo/M	–	+	1-cm cyst	Umbilical cyst	Tan *et al*. (2000), cited by Lee *et al*.[3]
10	7 wk/M	–	+	N/A	Umbilical cyst	Tan *et al*. (2000), cited by Lee *et al*.[3]
11	8 days/M	+	+	26×20×7 mm	Umbilical mass	Lee *et al*.[3] (2005),
12	18 mo/M	+	+	N/A	Umbilical mass	Silva *et al*. (2010), cited by Sharma *et al*.[4]
13	2 yr/M	+	+	12×12×10 cm	Umbilical mass	Sharma *et al*. (2013), cited by Sharma *et al*.[4]
14	3 mo/F	–	+	7×6×5 mm	Umbilical cyst	Present case

F, female; N/A, not available; M, male; +, present; –, absent.

into mature pancreatic tissue; metaplasia theory,[7] stating that endodermal tissues migrate to the submucosa during embryogenesis and transform into pancreatic tissue; and the totipotent cell theory,[8,9] in which totipotent endodermal cells lining the gut or omphalomesenteric duct differentiate into pancreatic tissue. The misplacement theory proposes that, during rotation of the foregut, several elements of the primitive pancreas become separated and eventually form mature pancreatic tissue along the length of the gastrointestinal tract. In this theory, the heterotopic rests are prone to drop off from the dorsal primordium and develop in the distal part of the stomach and proximal part of the duodenum, the most common sites of heterotopic pancreas. While it cannot explain other rarely discovered locations of heterotopic pancreas such as Meckel's diverticulum, ampulla of Vater, gallbladder, umbilicus, fallopian tube, and mediastinum, the totipotent cell theory is quite reliable for heterotopic pancreas in omphalomesenteric duct remnant because the cells lining the omphalomesenteric duct are known to pluripotent and can express either gastric, pancreatic, hepatic, or other terminal endoderm-derived phenotypes. While normal tissue is under the restriction to differentiate into certain cell types, tissue in this case seems to escape the normal restriction to maintain its pluripotent ability.

Because preoperative diagnosis is still a challenge, primary treatment for umbilical discharge is silver nitrate application. However, if symptoms are persistent despite this intervention, other differential diagnoses like patent urachus and omphalomesenteric duct remnant should be considered for early and relevant management. If heterotopic tissue is present, as in the presented case, severe local excoriation can occur and may lead to severe complications when not treated appropriately. Limited local excision has been shown to be a safe and adequate procedure to address this affliction. Awareness of this finding in biopsy can aid with appropriate treatment decisions for the patient.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

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