Ectopic Pancreatic Tissue Presenting as an Umbilical Mass in a Newborn: A Case Report

Wei-Te Lee, Hsing-I Tseng, Jao-Yu Lin,[1] Kun-Bow Tsai,[2] and Chu-Chong Lu
Departments of Pediatrics, [1]Pediatric Surgery, and [2]Pathology,
Kaohsiung Medical University Hospital, Kaohsiung, Taiwan.

The case of an ectopic pancreatic mass at the umbilicus in an 8-day-old male neonate is reported, the youngest patient with this condition ever reported in an English-language journal. The patient was healthy except for a protruding mass with intermittent mucous discharge at the base of the umbilical stump. Surgical intervention was performed under the impression of the umbilical mass. Pathology diagnosed an ectopic pancreas with acute hemorrhage. To the best of our knowledge, only one case of ectopic pancreas presenting as an umbilical mass with intermittent mucous discharge has previously been reported.

Key Words: ectopic pancreas, umbilical discharge, umbilical mass, newborn
(*Kaohsiung J Med Sci* 2005;21:84–7)

Ectopic pancreas has a wide range of distribution. The most common sites are related to the digestive system, particularly the stomach, duodenum, and jejunum, followed by Meckel's diverticulum and ileum [1–5]. However, ectopic pancreatic tissue has also been found in many other unusual locations including the umbilicus [1,2,5–10].

Umbilical discharge with or without an umbilical mass in previous case reports may have been due to the existence of structures derived from an omphalomesenteric (vitelline) duct remnant, patent urachus (remnants of allantoic duct), small bowel mucosal rest, ectopic pancreatic tissue, granulomas, polyps, or poor hygienic care [1,2,5–10]. Ectopic pancreatic tissue at the umbilicus with discharge has rarely been recognized preoperatively; it is usually discovered during surgery and diagnosed by histopathology.

Case Presentation

An 8-day-old male who had a protrusive umbilical mass with discharge since birth was brought to our outpatient department. Other than the umbilical mass, he was healthy with normal stool passage, appetite, and activity. On examination, a mucous-discharging polypoid lesion was found at the base of the unseparated umbilical stump, still covered with Wharton's jelly. A minor omphalocele with epithelialization was initially suspected. Routine examination of blood and umbilical discharge and abdominal ultrasound were normal. Surgical exploration revealed a 2.6 × 2.0 × 0.7 cm isolated pinkish mass at the base of the umbilical stump (Figure 1), which was excised *en bloc*. There was no continuity between the mass and the underlying viscera. The patient recovered completely and was discharged 2 days later. Histology showed a sac covered by the membrane of the umbilical cord. Both exocrine and endocrine pancreatic tissues were present in the specimen (Figure 2). Focal intense necrosis with hemorrhage was noted. The histologic diagnosis was ectopic pancreas with acute hemorrhage. At follow-up 1 year after surgery, the patient showed normal growth and development.

Discussion

The term ectopic pancreas indicates the presence of pancreatic tissue outside its normal location that lacks anatomic relation, either continuity or vascularization, with the main body of the pancreas [11]. It is generally agreed to

Received: May 3, 2004 Accepted: November 11, 2004
Address reprint requests and correspondence to: Dr. Chu-Chong Lu, Department of Pediatrics, Kaohsiung Medical University Hospital, 100 Tzyou 1st Road, Kaohsiung 807, Taiwan.
E-mail: weite@anet.net.tw

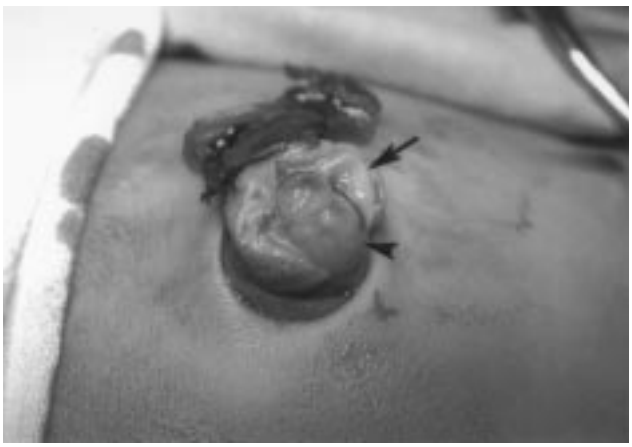

Figure 1. *Pinkish mass (arrowhead) disclosed after incision of the Wharton's jelly (arrow).*

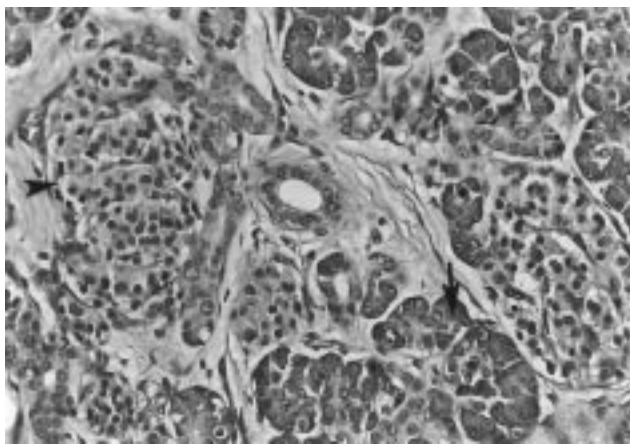

Figure 2. *Lobules of pancreatic acini (arrow) and islets of Langerhans (arrowhead). (Hematoxylin & eosin; original magnification × 40.)*

be a developmental anomaly. Various explanations have been offered for ectopic pancreas. Totipotent endodermal cells lining the gut or vitello-intestinal duct may differentiate into pancreatic tissue. Another possibility is that pancreatic cells may become transplanted or sequestered at heterotopic sites during fetal development [9]. Theoretically, they may occur anywhere from the yolk sac to the vitello-intestinal junction and result in Meckel's diverticulum, vitelline cyst and umbilical fistula, polyp, cyst, or, as in this case, cord.

The incidence of ectopic pancreas is around 1 in 500 laparotomies [12] or 0.55–13.7% of autopsy material [13,14]. About 40–68% of cases are symptomatic. When complications occur, symptoms depend on the site of the lesion [15].

To the best of our knowledge, there are only seven cases of ectopic pancreatic tissue at the umbilicus reported in the English literature (Table) [6–10]. The clinical manifestations were discharge in all cases [6–10] and mass in only one [9]. Umbilical discharge which is serous, serosanguineous, mucous, or purulent with a swelling mass expressed as a nodule, polyp, sinus, cyst, or granuloma at the umbilicus may be caused by ectopic pancreatic tissue in the remnant of the omphalomesenteric (vitelline) duct or in the patent urachus (remnant of the allantoic duct).

This case, an 8-day-old newborn, is the second reported case with both umbilical discharge and an obvious mass. It is also the youngest ectopic pancreatic tissue patient ever reported in an English-language journal.

In conclusion, we report a rare case of ectopic pancreatic tissue presenting as an umbilical mass in a newborn. Because a definitive preoperative diagnosis is rare, umbilical ectopic pancreas should be considered in the differential diagnosis of umbilical discharge and mass.

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

Year/Author	Age/Sex	Mass*	Discharge	Size	Site
1900/Wright [cited in 8]	12 yr /F	N/A	N/A	N/A	Umbilical subcutaneous tissue
1943/Trimingham [cited in 8]	22 yr /M	–	+	N/A	Umbilical cyst
1964/Steck and Helwig [6]	6 mo /M	–	N/A	3 mm nodule	Umbilical nodule
1977/Caberwal et al [9]	13 mo /M	+	+	12 × 9 × 5 mm	Umbilical mass
1999/Perez-Martinez et al [7]	6 mo /M	–	+/–	N/A	Urachus
2000/Tan et al [10]	3 mo /M	–	+	1 cm cyst	Umbilical cyst
2000/Tan et al [10]	7 wk /M	–	+	N/A	Umbilical cyst
2005/Lee et al [this case]	8 d /M	+	+	26 × 20 × 7 mm	Umbilical mass

*Ectopic pancreatic tissue (excluding granuloma and sinus) visible in the umbilical area. F = female; N/A = not available; M = male; + = present; – = absent.

References

1. Dolan RV, ReMine WH, Dockerty MB. The fate of heterotopic pancreatic tissue. A study of 212 cases. *Arch Surg* 1974;109: 762–5.
2. Lai ECS, Tompkins RK. Heterotopic pancreas: review of a 26 year experience. *Am J Surg* 1986;151:697–700.
3. Huang YC, Chen HM, Jan YY, et al. Ectopic pancreas with gastric outlet obstruction: report of two cases and literature review. *Chang Gung Med J* 2002;25:485–90.
4. Hsia CY, Wu CW, Lui WY. Heterotopic pancreas: a difficult diagnosis. *J Clin Gastroenterol* 1999;28:144–7.
5. Monig SP, Selzner M, Raab M, et al. Heterotopic pancreas: a difficult diagnosis. *Dig Dis Sci* 1996;41:1238–40.
6. Steck WD, Helwig EB. Cutaneous remnants of the omphalomesenteric duct. *Arch Dermatol* 1964;90:463–70.
7. Perez-Martinez A, Gonzalvez-Pinera J, Marco-Macian A, et al. Wet umbilicus caused by pancreatic heterotopia in urachal remains. *Pediatr Surg Int* 1999;15:143–4.
8. Harris LE, Wenzl JE. Heterotopic pancreatic tissue and intestinal mucosa in the umbilical cord. *N Engl J Med* 1963;268:721–2.
9. Caberwal D, Kogan SJ, Levitt SB. Ectopic pancreas presenting as an umbilical mass. *J Pediatr Surg* 1977;12:593–9.
10. Tan HL, Yoong A, Yu CCW. Ectopic pancreatic rests: a rare cause of persistent umbilical discharge. *Pediatr Surg Int* 2000;16:116–7.
11. Griffin AC, Brost BC, Staren ED. Symptomatic heterotopic pancreas following seat belt injury. *Injury* 2003;34:944–5.
12. Barbosa J, Dockerty MB, Waugh JM. Pancreatic heterotopia: review of the literature and report of 41 authenticated surgical cases of which 25 were clinically significant. *Surg Gynecol Obstet* 1946;82:527–42.
13. Barbosa J. *Pancreatic Heterotopia: Clinical and Pathological Studies of Surgical Cases*. Thesis, Mayo Graduate School of Medicine, University of Minnesota, Rochester, 1945.
14. Feldman M, Weinberg T. Aberrant pancreas: a cause of duodenal syndrome. *JAMA* 1952;148:893–8.
15. Anseline P, Grundfest S, Carey W, et al. Pancreatic heterotopia: a rare cause of bowel obstruction. *Surgery* 1981;90:110–3.

新生兒以臍部腫塊來表現的異位性胰臟 — 病例報告

李威德[1] 曾馨誼[1] 林釗佑[2] 蔡坤寶[3] 呂志忠[1]

高雄醫學大學附設中和紀念醫院 [1]小兒科 [2]小兒外科 [3]病理科

在一位八天大的新生兒身上，發現以臍部腫塊來表現異位性胰臟，在全世界的英文文獻中，這是年齡最小的病人。他除了在臍部殘跡內有一個腫塊合併間歇性的黏液分泌物之外，其他檢查皆正常，在臍部腫塊的診斷下做手術探查。腫塊病理報告顯示是異位性胰臟合併急性出血。經文獻查證，發現臍部的異位性胰臟在以往的報告中，只有另外一例和我們這個病人的情形相同，有明顯的臍部腫塊及間歇性的黏液分泌物。

關鍵詞：異位性胰臟，臍部分泌物，臍部腫塊，新生兒

（高雄醫誌 **2005;21:84–7**）

收文日期：**93** 年 **5** 月 **3** 日

接受刊載：**93** 年 **11** 月 **11** 日

通訊作者：呂志忠醫師

高雄醫學大學小兒科部新生兒科

高雄市三民區自由一路 **100** 號