

Ectopic jejunal pancreas and congenital duodenal stenosis in a newborn patient: an unusual association

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Abstract

Congenital duodenal stenosis is one of the most common causes of neonatal obstruction, which is frequently associated with annular pancreas and Down syndrome. Ectopic pancreas is defined as an abnormally situated pancreatic mass that lacks contact with normal pancreas. Although the association between duodenal stenosis and annular pancreas is very common, the presence of an ectopic island of pancreas in such cases is very rare. We present a case of unusual association of duodenal stenosis, jejunal ectopic pancreas in a neonate with Down syndrome.

Keywords: ectopic pancreas, congenital duodenal stenosis, Down syndrome, infant.

Introduction

Ectopic pancreas is defined as an islet of pancreatic tissue that lacks any vascular or anatomic connection with the main pancreatic entity. This abnormality is relatively common, as its reported incidence is 1–13%. Nevertheless, its etiopathogenesis has not been fully clarified yet [1]. It is most frequently located in the stomach (25–38%), duodenum (17–36%) and jejunum (15–21%) submucosa, but islets of ectopic pancreas have also been reported in the liver, Meckel's diverticulum, navel, spleen or Fallopian tubes [2, 3].

Intrinsic congenital duodenal obstructions (atresia and stenosis) are often reported in association with trisomy 21 (11–39%), as well as with annular pancreas; yet, the existence of an islet of jejunal ectopic pancreas associated with congenital duodenal obstruction in Down syndrome patient has been rarely reported in literature [4, 5].

Patient, Methods and Results

Here is the case of a 24-day-old female patient hospitalized in the authors' clinic for acute dehydration syndrome due to diarrhea and vomiting. The newborn is the 6th child of a 40-year-old couple (the mother is 40-year-old and the father is 42-year-old). It was a natural birth, after 36 weeks of pregnancy, and the baby weighed 2600 g at birth. The baby's medical history includes food and gall vomiting since her birth, and normal intestinal transit time, for which reason she was originally hospitalized in another pediatric ward. It is in this pediatric ward, where the food and gall vomiting persisted, that

her permanent gastric stasis was identified, accompanied by important gastric residue, and therefore the patient was referred to "St. Mary" Emergency Children's Hospital, Iassy, Romania. On her hospitalization in our department, the patient had significant weight deficit (weight on hospitalization: 2300 g) and obvious clinical dehydration signs (depressed anterior fontanelle, persistent skin fold, dry mucous membranes, lethargy). The child also exhibited a morphotype that is specific of trisomy 21 (karyotype 47,XX,+21): low birth weight and height (-4.7SD for weight, -2.7SD for height), round face, mongoloid slanting palpebral fissures, small nose, protruding tongue, systolic murmur, transverse palmar crease (Figure 1).



Figure 1 – The clinical aspect of neonate before surgery.

As we found ourselves before an infant whose clinical symptoms included vomiting since birth, severe dehydration syndrome and typical Down syndrome appearance, our main suspicion was directed towards a duodenal obstacle. Therefore, on the 3rd day of hospitalization, after a period of hydro-electrolytic rebalancing, we performed an eso-gastro-duodenal transit using a contrast agent, which revealed a significant stomach and

first jejunal loop distension, with specific “double bubble” appearance, and delayed contrast agent transition through the second duodenal loop and through the jejunum (Figure 2).

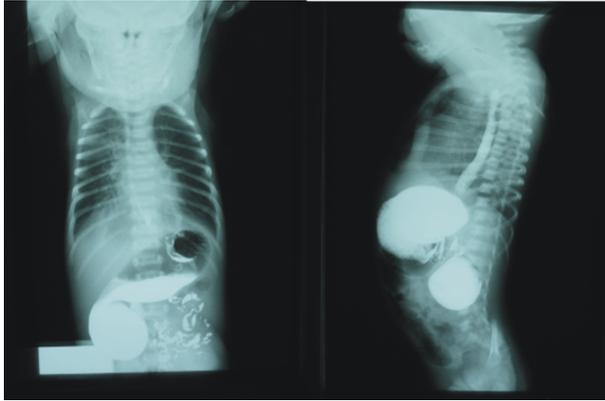


Figure 2 – Eso-gastro-duodenal transit with contrast agent: specific duodenal stenosis appearance.

As a duodenal stenosis was suspected, the patient was transferred to the Department of Pediatric Surgery, “St. Mary” Emergency Children’s Hospital, where a surgical procedure, through transverse right laparotomy approach, occurred on August 5, 2012. A duodenal stenosis area is identified through the diaphragm during the surgical procedure, located on the second section of the duodenum, beneath the ampulla of Vater.

Upon the examination of the other duodenal loops, a 1-cm sessile tumor mass is detected about 7 cm away from the angle of Treitz, on the antimesenteric border of the jejunum (Figure 3). The suspicion is of an ectopic islet of gastric or pancreatic mucosa. We proceeded to the surgical therapy of the duodenal stenosis by diamond-shaped duodeno-duodenal anastomosis, by resection of the jejunal loop carrying the tumor mass and by jejuno-jejunal termino-terminal anastomosis. Both intestinal anastomoses were performed and protected by eso-gastro-jejunal prosthetic probe. Abdominal drain tubes were also placed in the two anastomoses (right and left hypo-

chondrium). The immediate postoperative evolution was negative, as the patient exhibited the signs of a digestive fistula on the 5th day after the surgery (presence of gall secretions in the drain tubes, abdominal meteorism, abdominal wall edema), which required another surgical procedure, as we suspected a disruption of one or both digestive anastomoses (duodenal or jejunal). Another surgical procedure was performed, which revealed generalized peritonitis through two lesions of the jejunal loop downstream from the jejunal anastomosis area, punctured prosthetic tube running through the anastomosis. We checked the permeability and integrity of the two previous digestive anastomoses, which proved to be intact. We resected the borders of the two jejunal punctures and preferred a plane suture. The subsequent evolution was slowly positive, with a difficult resuming of the enteral nutrition.

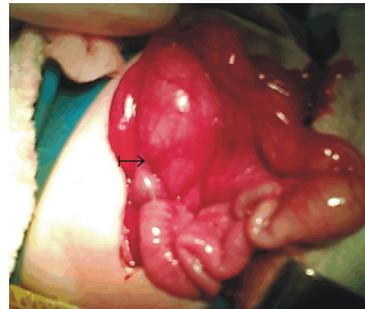


Figure 3 – Duodenal stenosis area (black arrow) with dilatation overlying the duodenum: intra-operative appearance.

A biopsy was performed on the piece of ectopic pancreas removed and on the resected jejunal loop, by paraffin processing and by means of the following dyes: Hematoxylin–Eosin (HE), PAS and immunohistochemistry (IHC) AE1/AE3 and chromogranin. The pancreatic ectopic tissue may be included in the type 1 of morphopathological types’ category; therefore, the cross section revealed intestinal wall with jejunal mucosa and exocrine and endocrine pancreatic parenchyma in the muscular and subserous layer, with infiltrates in some areas, including in the muscularis mucosae (Figure 4).

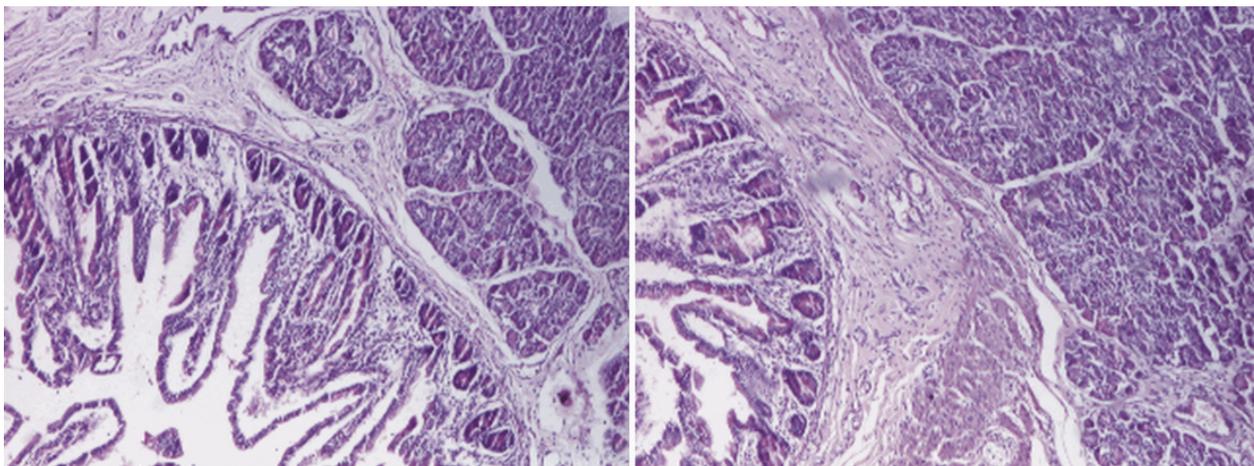


Figure 4 – Ectopic pancreas present up to the level of the muscularis mucosa and in the muscular layer of the jejunal wall. HE staining, ×40.

On some sections, a slantingly cut duct was also detected on the submucosa, located between the pancreatic parenchyma and the intestinal mucosa. Although at first it was thought to be an excretory duct connected to the

pancreatic parenchyma, this structure exhibits mucous cell villosity that would rather be suggestive of an intestinal structure, which raises the suspicion of possible associated jejunal duplication (Figure 5).

We also found areas on the pancreas where we identified secretion stasis in the small ducts. At some point, in the absence of any lesion excision, this stasis could have caused excessive accumulations, pancreatic tissue autolysis and occurrence of cysts or acute pancreatitis spurts (Figure 6).

We also performed IHC for pancytokeratin (AE1/AE3) in the epithelial cells, which was positive in the enterocytes and in the epithelial cells covering the exocrine pancreas

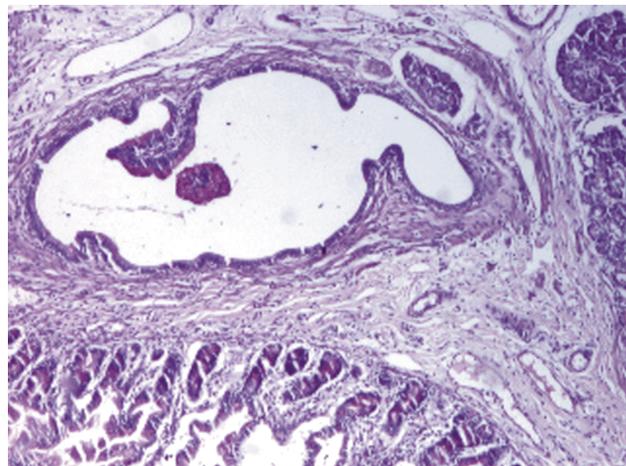


Figure 5 – Excretory duct with villosity and pancreatic acini islet. PAS staining, $\times 40$.

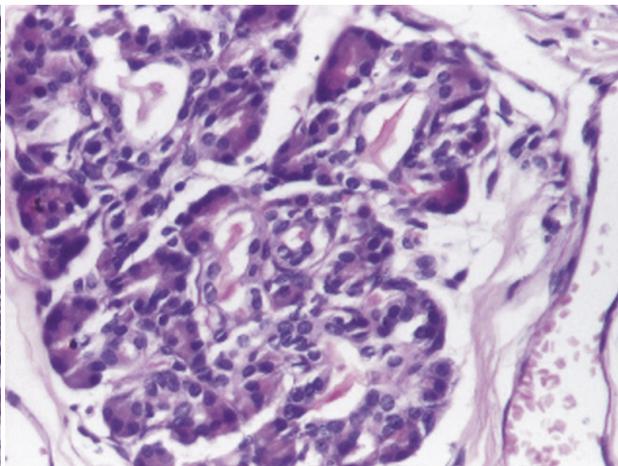


Figure 6 – Secretion stasis in the excretory ducts. PAS staining, $\times 100$.

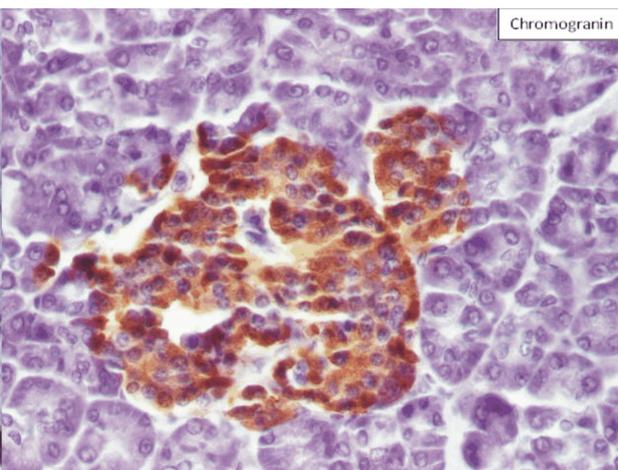
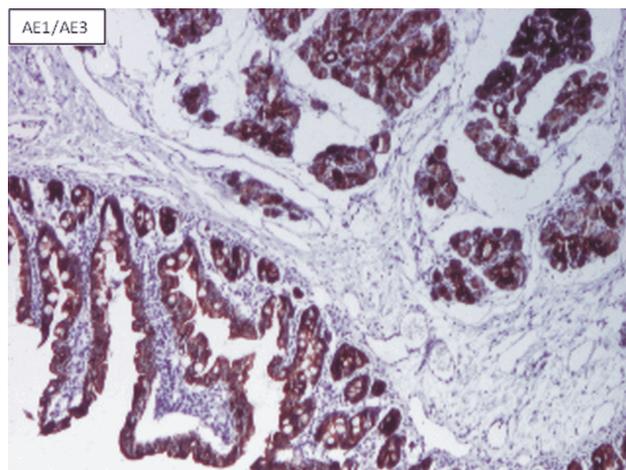


Figure 7 – Epithelial cells in the jejunum and pancreas: AE1/AE3 immunostaining, $\times 40$ (left); Chromogranin positive in the islet of Langerhans, $\times 100$ (right).

Discussion

Duodenal stenosis is one of the most frequent causes of high neonatal congenital occlusion, with a rate of occurrence of 5000–10 000/live newborns. Most of the times, duodenal obstruction is intrinsic, being revealed by the presence of an intraluminal diaphragm. About 38–55% of the intrinsic duodenal obstruction cases are accompanied by other abnormalities, of which Down syndrome is present in about 30% of the patients [4, 6]. Also, duodenal stenosis may be associated with various pancreatic and bile duct malformations, such as choledochal cyst, common bile duct duplication or stenosis and, the most frequent, annular pancreas, defined as a pancreas ring, which continues the pancreas head and by-passes the second section of the duodenum.

On the other hand, ectopic pancreas, which was first

described in 1729, is a relatively common condition consisting of the presence of pancreatic islets in other anatomic structures. This abnormality is usually identified accidentally, during various laparotomy procedures.

Ectopic pancreas is often asymptomatic, yet sometimes it may cause complications, like for instance invagination of an intestinal loop carrying ectopic tissue, spurts of acute pancreatitis or even malignization [7, 8]. Nevertheless, although it is a congenital lesion, it is very rarely reported as being symptomatic in newborns or infants. Literature actually cites only one case of symptomatic jejunal obstruction due to an islet of heterotopic pancreas located in the jejunum [9].

The embryology of ectopic pancreas has not been fully clarified yet, despite the numerous theories designed to account for its occurrence. Thus, one of these theories, supported by Skandalakis *et al.*, explains the occurrence

Subsequently, malnutrition recovery by a 3–4 g/kg protein intake and 150 kcal/kg from food and oral dietary supplements determined the resuming of the weight and height increase. Therefore, at the age of six months, the child exhibited -1.7SD for weight and -1.5SD for height, but still suffered from a certain psychomotor development retardation specific to Down syndrome.

of ectopic pancreas by an atypical metaplasia of the multipotent endodermal cells in the primitive intestine, whereas Armstrong *et al.* suggest some abnormalities in the primitive intestine rotation process and in the merging process of the two dorsal and ventral pancreatic buds [10, 11]. Abel *et al.* consider ectopic pancreas, especially the one associated with the gastric mucosa, as a variant of the involution abnormalities of the vitelline-enteric duct [12].

Pancreatic ectopic tissue may contain all or some of the structural components of a normal pancreas, including pancreatic ducts, acini or islets of Langerhans. There are four morphopathological types of ectopic pancreas: type 1 is represented by specific pancreatic tissue, type 2 contains only pancreatic ducts, type 3 includes only acinar tissue, and type 4 only cell islets [13]. The case described in this paper may be included in the type 1 category.

In our professional experience, we came across 2–3 cases of ectopic pancreas in the stomach, one in the spleen (discovered accidentally during the necropsy) and in other sections of the digestive tube. Just like in this case, there was no inflammatory reaction either in the pancreatic parenchyma or in its periphery, and the patients exhibited no clinical symptomatology related to the presence of ectopic pancreas.

This case is a rarer association between intrinsic duodenal obstruction, through the diaphragm, in the absence of an annular pancreas, and the existence of islet of type 1 ectopic pancreas located in the jejunum, possibly associated with small jejunal duplication, in a Down syndrome patient. The jejunal lesion was diagnosed by accident, during the surgical procedure, while we were checking the digestive tube to identify any associated intestinal stenoses or atresias. The preoperative exploration of the digestive tube using a contrast agent failed to reveal the existence of pancreatic heterotopia, although literature data describe a possible view of this image as a submucous jejunal loop-filling defect, which is accounted for by the absence of an adequate contrast agent transit through the jejunum, due to the duodenal obstacle [14].

The indication of jejunal lesion excision, which during the surgical procedure was thought to be an islet of gastric or pancreatic ectopic mucosa, was supported by the possible complications, such as bleeding, intestine invagination, malignization or pancreatitis spurts. The morphopathological appearance of the resection piece confirmed the lesion excision indication.

In our opinion, the postoperative complication that occurred (punctured jejunal loop downstream from the anastomosis area) was due to the quality of the digestive drain catheter and had no connection with the existence of pancreatic ectopia.

✚ Conclusions

This case represents a rare association between congenital duodenal obstruction, type 1 ectopic jejunal pancreas and Down syndrome, as our review of the literature did not find any other similar report. Although the ectopic pancreas was asymptomatic, we consider that the indication for surgical resection is mandatory in any such cases, as our morphopathological findings showed the risk for stasis, pancreatic tissue autolysis and cystic transformation.

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