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Unroofing and argon mucosal remnant ablation of neonatal duodenal duplication cyst

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Abstract

Background: Duodenal duplication cyst is an uncommon foregut malformation usually diagnosed at birth or during infancy. Differently from elsewhere, sited small bowel duplications cannot be removed with simple bowel resection, because of the proximity of the biliary and pancreatic ducts also possibly with abnormal course.

Case presentation: We report a duodenal duplication cyst in a newborn female requiring early surgery because of nutritional difficulties. The cyst was located adjoined to the second portion of the duodenum sharing part of its muscle wall with the bowel. It was treated by removal of all the esophytic cyst while the remaining mucosa on the common wall with the duodenum was ablated with argon plasma coagulation, preserving the bowel integrity. Early postoperative period was uneventful, and the child could be fed per os on the second day. Yearly follow-up was maintained until 16 years for the risk of recurrence and cancer change due to the incomplete excision. Clinic and echographic controls had always been stayed free from any sequelae.

Conclusions: Foregut duplications should be removed totally to prevent complications and the long-term risk of cancer, but a duodenal resection can be a harmful surgery in neonatal age. Duplication cysts that are impossible to remove totally can be treated by unroofing and argon plasma coagulation of mucosal surface remnants, avoiding the risks of major procedures also in newborns.

Keywords: Foregut cyst, Duodenal duplication, Mucosal ablation, Neonatal surgery

Background

Duodenal duplication cysts (DDC) are an uncommon form of foregut malformation. They represent no more than 2–5% of all alimentary tract duplications [1, 2] with an estimated prevalence of less than 1 per 100,000 live births [3, 4].

Most of the reported cases are diagnosed during the first two decades, but recently, this malformation has been recognized also prenatally [5, 6].

DDC are variously dimensioned and placed from pylorus to distal duodenum; their cavity can develop inside or outside the bowel lumen, and usually, their deep wall is shared with the normal bowel. They are normally coated by duodenal mucosa, but in 15% of cases, gastric or pancreatic metaplastic mucosa can cause peptic ulcer, bleeding, or perforations [4, 6, 7].

Depending on the size and site of the congenital DDC, symptoms can be delayed and are mostly related to duodenal obstruction, pancreatitis, or gastrointestinal bleeding [3, 4].

DDC can also show late neoplastic changes [8, 9]; therefore, every patient, even though asymptomatic, should be treated.

The therapy of choice for localized small bowel duplication is the complete surgical excision, together with the bowel tract to which they are attached. DDC, owing to the proximity to the biliary and pancreatic ducts, cannot be removed as every small bowel duplication

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elsewhere located; therefore, procedures alternative to bowel resection must be considered [10].

When technically feasible, DDC are accepted to be treated endoscopically by opening their wall from within the duodenum [11, 12], but there is still concern about the evolution, notably carcinoma change, of the untouched residual mucosa.

We report the case of an outgrown DDC treated with unroofing and residual mucosa ablation with argon plasma coagulation (APC) in a newborn, whose outcome was evaluated with a long-term follow-up of 16 years. Informed consent was obtained by the patient together with institutional approval.

Case presentation

The patient was a full-term girl weighting 3360 g. Prenatal ultrasound showed, since the 23rd week, a cyst in the right hypocondrium, and postnatal echography and CT scan confirmed the cyst attached to the duodenum. The perinatal period was untroubled, but after few days, the child showed persisting nutritional troubles with recurrent bilious vomitings and unsatisfying weight growing curve leading to surgery on the 13th day.

Video laparoscopic surgery was considered contraindicated for age and general conditions. Through a subcostal right incision, 4cm, DDC was found on the free peritoneal surface obstructing the 2nd portion of the duodenum, not involving the pancreas (Fig. 1a).

At the opening, it was found to be filled with mucus, not harboring stones.

The outer wall was removed by unroofing the cyst and leaving the deepest wall which was judged to be shared with the duodenum by its thickness (Fig. 1b). To fulfill the removal, the mucosa stripping was attempted, but due to a difficult and bleeding procedure, we turned to mucosa ablation with APC, preserving the bowel integrity (Fig. 1c).

Once duodenal mucosa ablation was performed by using argon, an adequate amount of air was introduced into the duodenum through a nasogastric tube to prevent any iatrogenic perforation on the duodenal wall.

The burnt surface was not covered by the omentum because the latter was not really represented, as it commonly occurs during the antenatal period. The abdomen was closed on a burnt surface, without signs of collection nor bleeding (Fig. 1d).

The *postoperative course* was uneventful. The child was fully fed per os on the 4th day, and she was dismissed on the 7th day after surgery.

Specimens from the DDC wall confirmed the macroscopic appearance of a bowel wall complete of all the layers lined with normal duodenal mucosa (Fig. 2a); histology of a fragment from the wall treated with APC showed the complete shrinking of the mucosal layer (Fig. 2b).

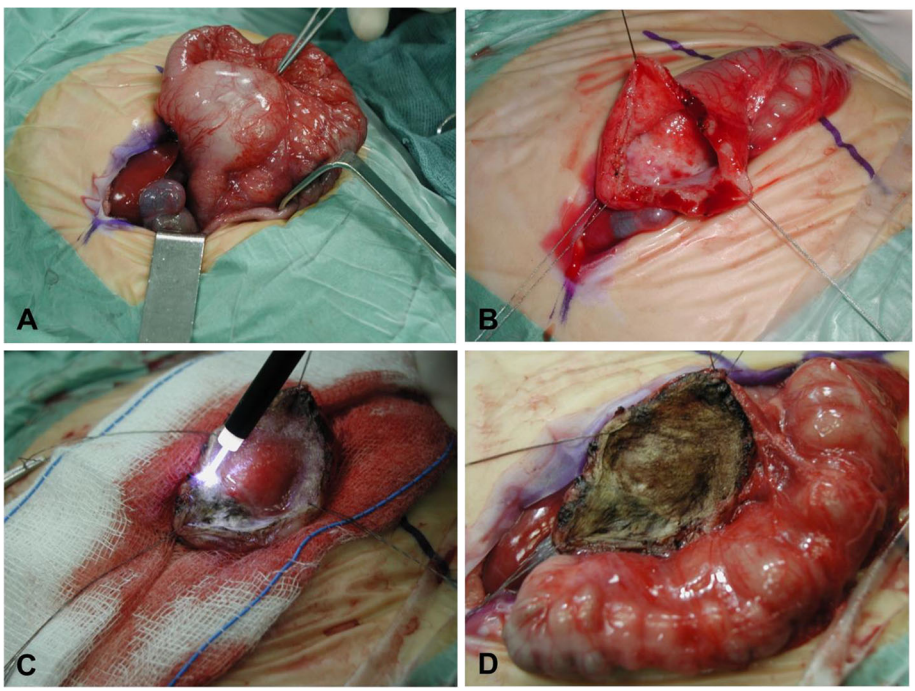

Fig. 1 Surgery pictures. **a** Duodenal duplication cyst. **b** Cyst unroofing. **c** APC coagulation of common wall mucosa remnants. **d** Completion of treatment

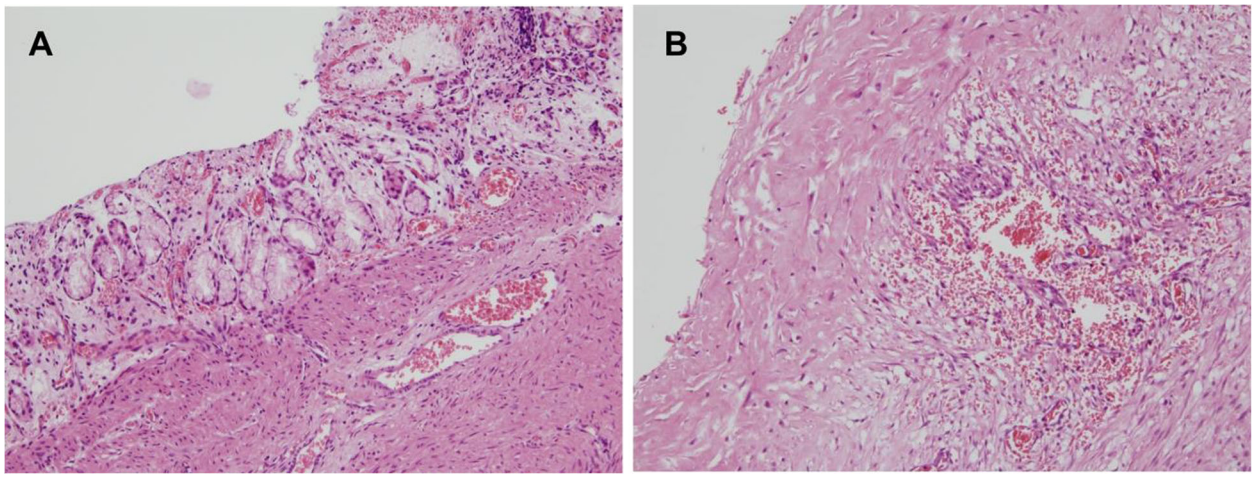

Fig. 2 Histologic study. **a** Histology of the removed wall layers of the cyst with normal mucosa lining. **b** Histology of a fragment after APC treatment showing complete ablation of the mucosa

The mid-term risk of recurrence and the long-term risk of cancer change due to the macroscopically incomplete removal of the cyst moved us to maintain yearly clinic and echographic controls on a long-term follow-up.

During a 16-year period, the patient always stayed asymptomatic and did not show any specific clinical signs related to gastrointestinal bleeding, adhesive intestinal obstruction, or pancreatitis.

Abdominal US scans performed annually ruled out recurrences or other lesions at the site of surgery. Over the whole period of time, the patient did no go through any additional gastrointestinal examination such as endoscopy, CT, or MRI due to the lack of clinical indications.

Conclusions

The management of intestinal duplications is surgical, and the optimal treatment is the complete primary excision [1, 3]. When sharing their wall with a long gastrointestinal tract or when a radical therapy could endanger other vital structures, they can be treated by a more limited surgery, matching the need for mainly benign lesions.

DDC derived from the foregut are variously sized and sited cysts; consequently, their surgical treatment although mandatory should be modulated in the aggression required.

Subtotal removal is the preferred method for inward grown cysts feasible of endoscopic treatment, privileging the lesser aggression to the safety of DDC mucosal elimination [12].

In outward grown cases, in order to prevent any further postoperative complication and mostly the long-term risk of cancer, the usual practice is the safe removal of as much as possible of the duplication wall together with the whole mucosal layer.

For these reasons, marsupialization and mucosa stripping is appropriate, but sometimes, this procedure turns out to be a troubled trick.

In our case, a decision was taken to avoid endoscopically opening the cyst into the duodenum as, in a newborn patient, it would have been difficult to safely move inside the duodenum abnormally compressed by the cyst and to locate on the duodenal wall shared with the cyst the correct area for the opening, due to the high risk of abdominal perforations.

Based on these motivations, we choose to preserve bowel integrity by ablating all the mucosa-lined remnants with the APC technique.

This method produces a no-touch, less-than-3mm-depth coagulation by a gas-carried high-frequency electric current and offers the ability to treat a large surface under good visual control of the effect, as it has been clearly demonstrated by Barrett's esophagus treatment [13–15]

The histologic study of the specimen at the border of treated DDC confirmed that APC, while preserving the underlined muscle wall, produced a definite devitalization of the mucosal surface. This result was time-honored by postoperative ultrasounds, as no collection nor cyst recurrences ever developed on a long-term follow-up of 16 years.

Cyst unroofing with APC of mucosal remnants can be a safe and efficient alternative also in newborns to achieve a complete removal of a dysonthogenetic cyst when a more radical surgery would require an unmatched major procedure. The same method could accomplish also less invasive endoscopic treatments.

Abbreviations

APC: Argon plasma coagulation; DDC: Duodenal duplication cyst

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Consent for publication
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