Double duodenal atresia noticed as an intraabdominal cyst in the fetus

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ABSTRACT

Double duodenal atresia is a rare disease and twenty-two cases have been reported in the English literature. We report here a case of double duodenal atresia that had been suspected by fetal ultrasonography as polyhydramnios and a large cyst at the right upper abdomen. Two membranous diaphragms at the duodenum separated the duodenal lumen and formed a cystic lesion. Surgical reconstruction was performed.

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These findings suggested that the cyst was duodenum with double atresia, and a Y-shaped biliary tract connected both the first portion and the second portion of the duodenum. At 7 days of age, the patient underwent laparotomy and the cyst was found to be duodenum with double membranous atresia and the second portion formed a closed lumen by the two membranous diaphragms and was severely dilated (Fig. 3a,b). The first membranous atresia was reconstructed by a longitudinal incision and transverse closure with partial resection of the membrane. The second atresia was reconstructed by side-to-side diamond anastomosis. The postoperative course was uneventful. The biliary tree was still tortuous and slightly dilated but she had no jaundice and liver function tests were normal.

2. Discussion

Duodenal atresia is the most common type of congenital intestinal atresia and it is usually treated by the established surgical procedure without any complications. However, in some cases special caution is needed at surgery. The wind sock web-type atresia is a well-known one that needs special attention not to miss the true atretic site [1].

Double duodenal atresia is another type in which special caution needs to be taken. Twenty-two cases of double duodenal atresia have been reported in the English literature [2–12] but there was a very wide variation in atretic types including membranous diaphragm, striated atresia, and disconnected atresia.

Duodenal perforation occurred before the operation in two cases [2,3] among the cases of double duodenal atresia. Excessive bile juice retention was a major cause of perforation in those cases because the biliary tract opened to the separated duodenal space. In our case, the biliary tree opened to the cystic-form duodenum but it formed a Y-shaped tree and bile juice could slowly flow out to the first portion of the duodenum and stomach through another bile tree; this mechanism might have prevented perforation of the cyst.

In one case report, fetal ultrasonography revealed three abdominal cysts [4]. One cyst was the stomach, one was the dilated first portion of the duodenum, and one was the separated second portion. In our case, the first portion of the duodenum was mildly dilated and the second portion was prominently dilated because bile juice was secreted mainly to the separated cystic duodenum through the above-mentioned special biliary tree and the first portion of the duodenum was not exposed to high pressure. Therefore, in our case ultrasonography and MRI in the fetus could not show triple cysts. It is very difficult to diagnose double duodenal atresia in fetal life, but polyhydramnios co-existing with cysts detected at the right upper abdomen by fetal ultrasonography suggested the disease. Duplication cyst of the duodenum is one differential diagnosis in this situation, but duplication cyst coexisting with duodenal atresia is extremely rare.

Fig. 1. a) A fetal ultrasonogram at 27 weeks of gestation. A cyst of 3 cm in diameter is situated near the right kidney. The stomach and the first portion of the duodenum were detected but were not much dilated. b) MRI taken at 27 weeks of gestation. A cyst was detected near the right kidney (arrow), but another cyst was not detected. The stomach and first portion of the duodenum were also detected but they were not dilated. Polyhydramnios was also pointed out.

Fig. 2. a) An abdominal roentgenogram taken at 1 day after birth showed some gas in the stomach and duodenum but no intestinal gas. The first portion of the duodenum was mildly dilated. b) Ultrasound examination of the abdomen revealed a cyst at the right upper abdomen. The wall of the cyst was thick. c) The contrast medium study of the upper gastrointestinal tract showed a mildly dilated first portion of the duodenum and a tortuous biliary tree (arrows). The cyst was vaguely stained by the contrast medium and it was later confirmed by abdominal CT taken just after this examination.
In our case, the biliary tree was tortuous and slightly dilated. These abnormalities may have been caused by high pressure in the biliary tree due to the semi-closed lumen including cystic-form duodenum. Currently, the patient has no jaundice and liver dysfunction but close observation will be needed in the future because cases of choledochal cyst associated with duodenal atresia have been reported, although it is rare [13].

Conflict of interest statement
All authors have no conflict of interest.

References

Fig. 3. a) Schematic picture of the duodenum and biliary tree in this patient. The Y-shaped biliary tree opened to the first portion and the second portion of the duodenum and a double membrane separated the second portion of the duodenum to form a large cyst. b) Intraoperative picture. A large cyst (arrow) was the second portion of the duodenum and the lumen was closed by the two membranous diaphragms.