CASE REPORT

Congenital Choledochal Cyst.

by

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Although congenital choledochal cyst, a localized dilatation of the common bile duct, is an unusual condition in many countries, it should be expected in any infant who presents the triad of jaundice, tumor and pain in the right upper abdomen. Not less than 500 cases have been reported in the medical literature. Recently it is pointed out that the congenital choledochal cyst is more common in Japan than in Europe or American countries. A personal contact with the Japanese delegation attending the meeting of the Pacific Association of Pediatric Surgeons in Van — couver, May 1971, confirmed it.

Many excellent reviewes of the history, incidence and etiology have been published but there are still problems concerning the pathogenesis, into which this paper will not go deeper. In Indonesia only two references are found, one by Zahruddin et al. (1966), of a girl, 7 years of age, with a choledochal cyst and the second by Sjaffar Juniman (1969), 2 cases, both in adult females.

The purpose of this paper is to report a case that might be worthwhile to mention for the following reasons:

a. It concerns a boy, whereas in the literature females outnumber males by 4 : 1.
b. The very young age; as a rule the congenital choledochal cyst presents very seldom the symptoms in the first few days of life.
c. The (uncommon) correct preoperative diagnosis was established.
d. The instructive course during and after surgery.

Case report

A 35-day-old Chinese boy was admitted with the following history. The mother has noticed jaundice from the first day of life, later associated with distention of the abdomen and slight colored stool. In all respects the birth was normal and family his-

tory showed no particularities. A physiological jaundice was diagnosed but when the baby got recurrent attacks of abdominal pain and the feces stayed clay colored, the mother consulted her physician. After one month of no improvement and the appearance of fever, hospitalization was advised.

Physical examination showed a boy, with a body weight of 3900 grams, who was restless, with 37.8° C temperature. There was no vomiting. A marked generalized icterus of the thin skin, the sclerae and the mucous membrane of the mouth was noticed. The abdomen was distended and tympanitic. Many large superficial veins were visible on the lower chest and upper abdomen. A large and tender cystic mass in the right upper quadrant was palpable but the lower border could not clearly be distinguished. The liver and spleen were not palpable.

Laboratory data:

Hemoglobin 60% Sahli, white blood cells 9.000/ml. Urinalysis revealed normal findings, the stool was acholic. The Kahn serologic test of the mother was negative.

The AP. X-ray of the abdomen revealed a mass occupying the right upper and lower quadrants, displacing the intestines to the left.

Discussion

The above mentioned clinical symptoms were convincing for an obstructive jaundice. The large cystic tender tumor gave suspicion to a congenital choledochus dilatation. However, jaundice due to other anomalies such as prolonged icterus neonatorum, bile duct atresia, erythroblastosis foetalis, hepatitis could not be excluded. Since exploratory laparatomy at the age under 3 months has a high mortality, every effort should first be made to arrive at a correct diagnosis before surgery.

The clinical symptoms of choledochal cyst develop mostly after the age of 6 months and the attention is called to the higher frequency in females. Therefore further tests were necessary to exclude a bile duct atresia.

Recent publications make mention of the use of sophisticated methods such as hepatobiliary scintigram and ultrasonic echogram.

On this patient gastroduodenal barium X-rays revealed the characteristic lateral location of the anteriorly displaced duodenum. The duodenum was elongated and localized directly beneath the anterior abdominal wall. Together with the triad of jaundice, tumor and pain, these pictures convinced the diagnosis of congenital choledochal cyst (see figure)

No case of a spontaneous cure of the cyst has been reported. Aspiration alone gives finally 100 percent mortality. Procedures still recomended are primarily the chelodocho-

cysto-jejunostomy, Roux-en-Y, and secondly the resection of the cyst with hepatico-jejunostomy, Roux-en-Y.

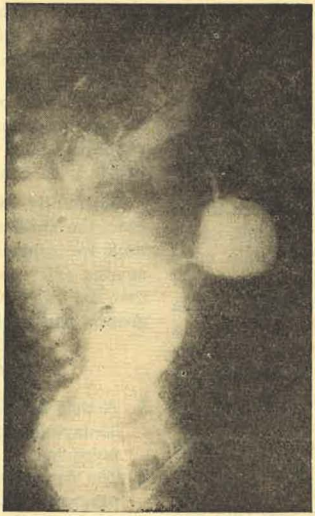
The operation on the baby was performed on the third day of admission. Through a right paramedical approach a large cystic mass occupying the right side of the abdomen with shifting the viscera to the left was found. The liver and spleen were not enlarged. To obtain more space a 500 mls liquid was aspirated which turned out to be clear bile. The cyst, lying retroperitoneally, had a thin wall. The normal gallbladder was visible between the cyst and the liver. The general condition of the baby did not permit a long time exploration, so that a one-inch-wide shunt from the cyst to the duodenum had to carried out. Since the wall of the cyst was very thin and non-contractile and the intention existed to do a post-operative cholangiogram, an external drainage was installed. The child did well and on the second day post — operation an oral feeding could be given. But soon a reflux of milk from the duodenum into the cyst and coming out from the external drainage was noticed. On the fourth day the temperature rose, probably due to cholangitis.

A relaparotomy was decided, the existing anastomosis and the external drainage were closed, a new connection between the cyst and the jejunum by a Roux-en-Y procedure was established. Not withstanding the unfavourable general condition the child did well after this second operation, although the fever persisted for another 3 weeks, the stools however, were regular and normal. Exactly one month after the last operation the child was discharged in a rather good condition.

The family lived close by; however, the child was never brought back for control. A home visit 3 months after the operation showed that the infant was quite normal in growth and development.

Summary

1. A case of choledochal cyst in a 35-day-old Chinese boy has been presented.
2. The correct diagnosis was made pre-operatively by radiologic examinations.
3. Simple anastomosis between the cyst and the duodenum has proved to be complicated by ascending infection, attributed by reflux of intestinal contents into the cyst. The correction by a choledochocysto-jejunostomy in Roux-en-Y fashion was preferable. However, the recent publication from Japan (Kasai et al.) mentions that the choice of surgical operation from choledochal cyst should be resection of the cyst followed by reconstruction by hepatico-jejunostomy in Roux-en-Y fashion.



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