CHOLEDOCHAL CYST

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The first reported case of choledochal cyst was described by Vater 230 years ago. In 1946 Shallon, Eger and Wagner[1] reviewed the literature. In 1955 Alton and Obeid[2] added 17 more cases, making the total reported cases 201. Fleming[3] in 1947 reported a case that occured in South Africa. In the records of the Mayo Clinic, Everett Shocket *et al.*[4] found 6 cases to add to the records.

A choledochal cyst is an aneurysmal dilatation of a segment of the biliary tract. It is a disease much more prevalent in women than in men and an analysis of the above cases shows it to be 3 times commoner in females than in males. The usual age of onset is under 25 years, but many cases wait several years before surgical treatment is undertaken.

The origin of the cysts is unknown, but generally it is accepted that they are congenital, because the majority occur in children and the incidence diminishes with age. Cysts have been described in an unborn child,[5] as well as in newborn children.[6, 7, 8] Another factor supporting the congenital origin of these cysts, is that they have been described associated with other congenital abnormalities of the biliary tract and liver. Gross[9] advances the theory that there is a congenital weakness of the duct associated with a valve of the lower end causing obstruction. The congenital cyst usually involves the supraduodenal portion of the common bile-duct, and is confined to the common bile-duct. There is no associated dilatation in the hepatic duct. The cyst varies in size and may reach an enormous magnitude. Reel and Burrell[10] reported a case which contained 8 litres. The cyst wall is made up of fibrous tissue.

Symptomatology and Diagnosis

The commonest symptom-traid is a right upper abdominal tumour associated with jaundice and pain. The mass is usually large, cystic and easily palpable. Jaundice is found in 3 out of every 4 cases and is usually of the obstructive type. Pain may be slight and usually there is a fairly long history. The nature of the pain may be colicky, or it may be due to pressure on the surrounding structures, causing a 'dragging' sensation.

The mass is commonly misdiagnosed as a pancreatic cyst. Only about 50% of the cases are correctly diagnosed pre-operatively. The diagnosis may be made with this symptom-triad and X-ray studies of the case. Barium meal usually shows the duodenum pushed outwards, and forwards as can be seen demonstrated in the X-rays of the patient now described. Pancreatic cysts usually push the duodenum outwards and backwards. Cholecystogram, if concentration occurs, usually reveals the gall-bladder pushed upwards and indented, giving a comma shape. Alton and Obeid,[2] however state, that both the gall-bladder and the choledochal cyst do not concentrate even when intravenous Biligrafin is used.[2]

Treatment

Since the diagnosis is commonly not made pre-operatively, complications may occur if an attempt is made to resect the cyst without realizing what it is. The

safest procedure is to aspirate the cyst when, if bile is obtained, the condition should be recognized and a small opening is made into the sac. The contents are then aspirated and the communications to the ducts can be demonstrated. If the opening to the duodenum cannot be seen, this does not preclude the diagnosis.

Excision of the cyst is an extremely difficult and dangerous procedure and should not be embarked on, because of the high mortality. The procedure of choice is a short-circuiting operation. Alton and Obeid[2] reviewed 210 cases and found that 22 were not operated on, all of which died. The cause of death was rupture of the cyst, suppurative cholangitis and cirrhosis of the liver, The child will suffer recurrent attacks of pain and jaundice. Anastomosis with the duodenum results in a 24% mortality due to shock, ascending cholangitis, bleeding or liver failure. Roux Y anastomosis of the cyst with the jejunum would appear to be the treatment of choice, for the possibility of ascending cholangitis is avoided. The case now described is the 11th of all the collected series in which this procedure was carried out; all these cases survived.

CASE REPORT

Mrs A.M.P., a housewife aged 39, complained of jaundice and a mass in the epigastrium. She was an extremely poor witness and stated that this mass and jaundice suddenly appeared 2 weeks previously. Pain had been intermittent over the same period. There was nothing significant in her past history except that she was an alcoholic.

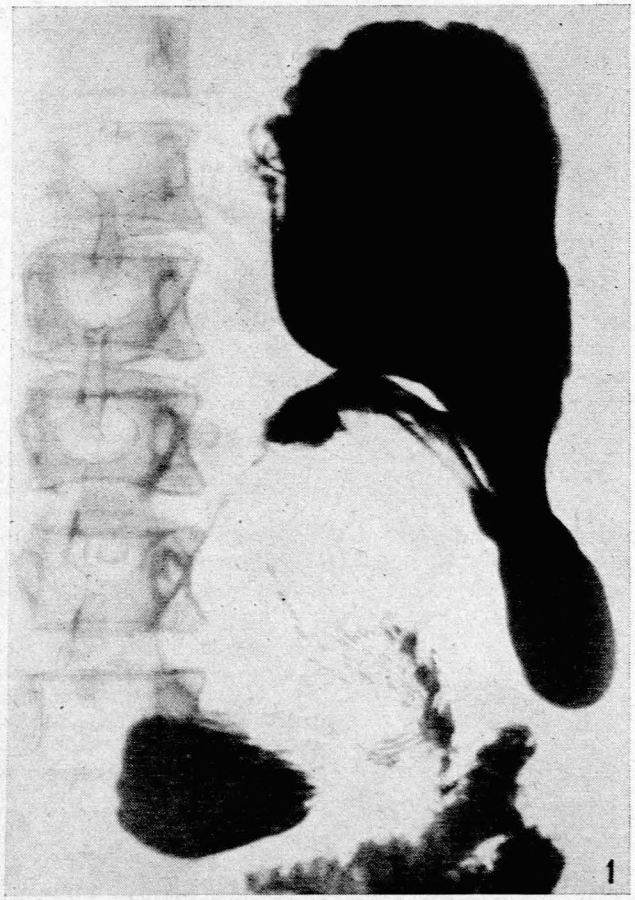

Fig. 1. Showing the 'C' curve of the duodenum greatly enlarged and the descending loop of the duodenum depressed.

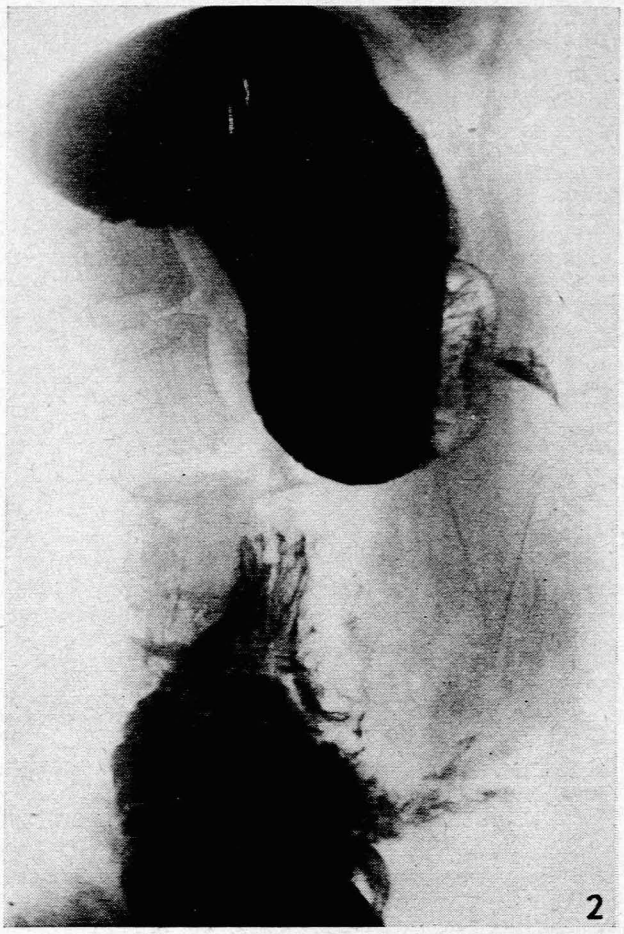

Fig. 2. The duodenum is displaced markedly forward by the choledochal cyst.

She was a thin woman, deeply jaundiced. Abdominal examination revealed a liver that was 3 finger-breaths enlarged below the costal margin and there was a large smooth cystic mass below it. The mass was 8 inches in diameter and was freely mobile, especially in the horizontal plane. Liver function and blood tests were performed with the following results (Dr. I. Bersohn, 11 August, 1955):

Hb. 14·2%. Red cells 4,840,000 per c.mm. Leucocytes 10,400 per c.mm. (neutrophils 69·5%, monocytes 4·5%, lymphocytes 23·0%, eosonophils 3·0%).

The red cells show slight anisocytosis and a tendency towards hypochromasia.

Platelets were present in normal numbers. Occasional target cells were seen.

ESR 33 mm. in 1 hour.

PCV 43·5.

PI 89% (control clotted in 11·0 sec., specimen clotted in 12·4 sec.).

Bilirubin++++., Urobilin++++., Urobilinogen present, Wallace Diamond dil. 1/30.

Thymol turbidity 1·0 unit, Thymol flocculation negative.

Colloidal-red test—negative.

Cephalin cholesterol flocculation+++.

Takata Ara reaction—negative.

Zinc sulphate turbidity—15·8 units.

Total lipid—934 mg.

Alkaline phosphatase—90 units.
Van den Bergh—prompt direct.
Bilirubin direct—3·2 mg., bilirubin total—4·6 mg.
Total protein—6·9 g. %, albumin—3·3 g. %, globulin—3·6 g. %, gamma globulin—1·54 g. %.
Serum cholinesterase—69% of the average normal activity.
Mucoprotein—163 mg. %.
Polysaccharide of mucoprotein—29 mg. %.
P/M Ratio—18.

X-rays (Dr. E. Samuel) taken on the same date revealed that the stomach was displaced towards the left side and a large mass occupied the lesser sac, displacing the stomach forwards and towards the left (Fig. 1). The 'C' curve of the duodenum was greatly enlarged and the descending loop of the duodenum was depressed and markedly displaced forwards (Fig. 2). Cholangiogram revealed a virtual obstruction of the common duct.

Operation was performed on 15 August 1954. A transverse upper abdominal incision was made and, when the peritoneum, was opened, a large mobile cyst was found pushing the duodenum forward. The duodenum was markedly stretched. The gall-bladder and common hepatic duct were markedly dilated. The gastro-colic omentum was divided and the cyst shelled easily to its attachment to the common duct. The cyst was then punctured and 20 oz. of bile evacuated. The diagnosis being made, a Roux Y anastomosis was performed 12 inches from the duodeno-jejunal flexure, and a proximal loop of 10 inches was brought up and anastomosed to the cyst.

The patient made an uneventful recovery.

The biliary fluid was sent for examination and tests for bile pigments were positive. On spectroscopic examination, methaemoglobin was detected. Biopsy of the wall of the cyst was also taken and this revealed chronically inflamed connective tissue lined on the one side by granulation tissue.

SUMMARY

1. A review of the literature on congenital cysts of the bile ducts is presented.

2. Anastomosing the cyst with the jejunum by means of the Roux Y procedure is suggested as the method of choice.

3. The dangers of excision and marsupulization of the sac are shock and ascending cholangitis.

4. An additional case of choledochal cyst is described.

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