CHOLEDOCHUS CYST

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Choledochus cyst is a rare congenital anomaly in which there is marked localized dilatation of the common bile duct. First described by Vater in 1723, other names given in the literature to the same lesion are choledochal cyst, cystic enlargement of the common bile duct, choledochocoele, diverticulum of the common bile duct and megacholedochus. Such terms are often qualified by the adjectives congenital or idiopathic.

As this condition is both rare and amenable to surgical treatment, we feel that any case should be reported.

CASE REPORT

T.S.K., a five-month-old female infant, was admitted on 27.9.60 to the Paediatric Unit, Singapore General Hospital, with a history of jaundice since the age of three weeks. This jaundice increased in intensity till the age of two months, when, the mother said, it regressed slightly. The baby was not unduly jaundiced at birth, was a full term baby, normally delivered, with a birth weight of 7 lb. 4 oz.

On examination, she was afebrile, but icteric, the colour characteristic of obstructive jaundice. The cardiovascular, respiratory and central nervous systems were clinically normal. However, the abdomen was distended and the liver was firm and palpable four finger-breadths below the costal margin. The spleen was two finger-breadths palpable and firm. No shifting dullness was detected in the abdomen.

The urine was highly coloured and bile was present but no urobilinogen. The haemoglobin was 11.1 Gm.%, white blood count was 11,000, with a differential count of N.50%, L.40%, E.6% and M.4%. The stools were pale. The serum bilirubin was 4.6mg.%. Haemoglobin electrophoresis revealed normal adult haemoglobin with 5.3% of foetal haemoglobin.

At this stage, it was thought that she was suffering from neonatal hepatitis rather than from biliary atresia in view of the history of intermittent obstruction, and a course of prednisolone was given when it was noticed that the stools became more coloured. She was discharged from hospital and followed up in the outpatient department.

However, she did not improve and in March, 1961, it was noticed that she was still jaundiced. The liver and spleen were still enlarged, and, just below the enlarged liver, a large circumscribed mass measuring about 3" x 2" x 1" was felt extending medially to just beyond the midline and downwards towards the umbilicus. There were a few small glands in the axilla, and, at this stage, some form of reticulosis was suspected, and the abdominal lump was thought to be an enlarged gland pressing on the bile ducts with production of obstructive jaundice. A biopsy was done on one of the axillary glands but histology revealed no signs of malignancy, reticulosis or tuberculosis.

At this stage, the possibility of a choledochus cyst was entertained as the patient was female and there was the history of intermittent episodes of increased jaundice together with the characteristics of the abdominal lump. She was then referred for surgical treatment.

On 17.4.61, under intratracheal anaesthesia, the abdomen was opened and a large choledochus cyst found, from which 400 ml. of bile-stained fluid was aspirated. As the fundus of the cyst was lying directly on the pyloric end of the stomach, and as the duodenum was rather narrow, choledocho-gastrostomy was performed. The liver at this time was dark and congested.

In the post-operative period the child became febrile and the abdomen distended gradually. As there was plainly intestinal obstruction, the abdomen was reopened on 25.4.61. The choledochus cyst was found to be empty. A band ran between the umbilical region of the peritoneal cavity to the root of the mesentery, and a loop of gut was found to have rotated on this band, causing obstruction. The loop was freed and the band excised. Ascitic fluid was prominent.

Recovery was unsatisfactory. Two days after the removal of the abdominal wound sutures, the abdomen burst. This necessitated another anaesthetic while the wound was closed.

Although the jaundice diminished, the abdomen remained distended and progress was poor. For no obvious cause, she passed many loose stools. Ascitic fluid continued to accumulate, repeated parecentesis averaged about 100 ml. per tap.

She lingered on, dying on 10.8.61.

Necropsy (P.M. 1686/61)

The body was that of a small, emaciated, mildly jaundiced, Chinese girl: the distended abdomen seemed enormous. A mass of keloid scar, 14 by

6 cm., which extended to the right of the midline beneath the costal margin, further emphasised the relative prominence of the abdomen.

The abdominal cavity contained 120 ml. of faintly turbid jaundiced fluid.

Multiple adhesions were present between the sites of the surgical wounds on the abdominal wall, the liver, the ascending and transverse colons, and several loops of small bowel. Between the coils of small bowel there were two distinct fibrous bands in the mass of adhesions, but there was no evidence of gut obstruction.

The liver, weighing 480 g , was much enlarged. The cut surface, deep green in hue, revealed numerous coarse white strands of fibrous tissue which encircled small islets of liver tissue. Along, and within, the major bile ducts there was a fair amount of pus, and several subcapsular abscesses, the largest about 0.5 cm. in diameter, were present in the left lobe.

The major intrahepatic ducts were mildly dilated and led into an extraheptic choledochus cyst some 4 by 4 by 2 cm. (Fig. 1). The gall bladder, somewhat thickened and narrow, communicated freely with the cyst by a widely patent cystic duct (Fig. 1). Posterior to the second part of the duodenum the cyst exhibited a blind recess about 2 by 2 by 1 cm. No communication with the duodenum could be established. The pancreatic duct drained normally.

The cyst wall, 5 mm. thick, was lined by inflamed tissue through which linear white streaks could be seen. The cyst contained a mass of yellowish, viscid, purulent material mixed with greenish milk curd, which drained freely on pressure into the prepyloric portion of the stomach, via the anastomosis (Fig. 1).

The lower end of the esophagus exhibited a few dilated veins. The spleen which was firm, deep red in colour, and enlarged, weighed 72 gm.

There was a large recent haemorrhage, and a smaller prior one, in the left subdural space over the parietal and temporal lobes. No bleeding point was demonstrated.

Histology

The wall of the cyst is devoid of epithelial lining, being formed by a moderately vascular granulation tissue in which moderate numbers of lymphocytes and plasma cells are seen (Fig. 2). There are numerous foci of bile-laden macrophages, and in several areas pools of golden-yellow pigment are noted.

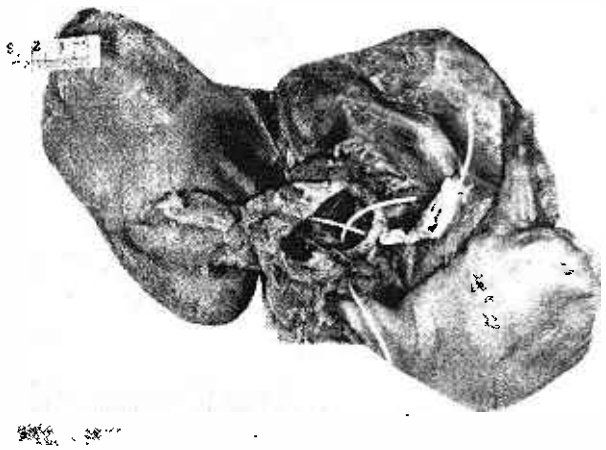

Fig. 1. On the right, the lower third of oesophagus, and the stomach, are seen. A probe passes, in a perpendicular direction from the pyloric antrum, through the anastomosis, into the opened choledochal cyst. The second probe, passing in a horizontal direction, shows the entry of the cystic duct, leading from the thickened gallbladder on the right, into the cyst. The pancreas, on the left, lies over the liver. The irregularity of the liver surface and the focal thickening of the capsule are fairly prominent.

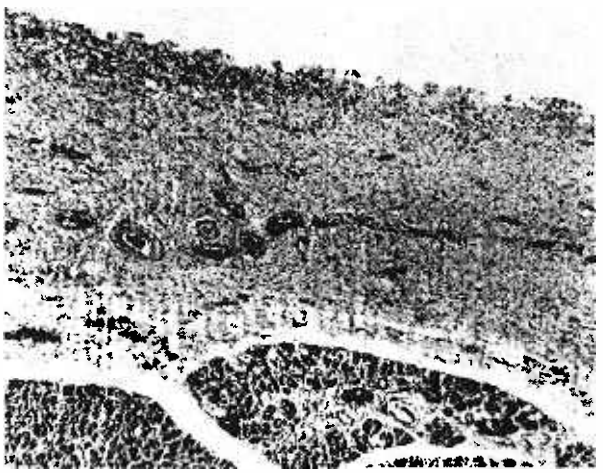

Fig. 2. The thick choledochal cyst wall is devoid of mucosa. The thickened submucosa is infiltrated with chronic inflammatory cells; the blood vessels are congested. The pancreas lies below. H. & E. x 45.

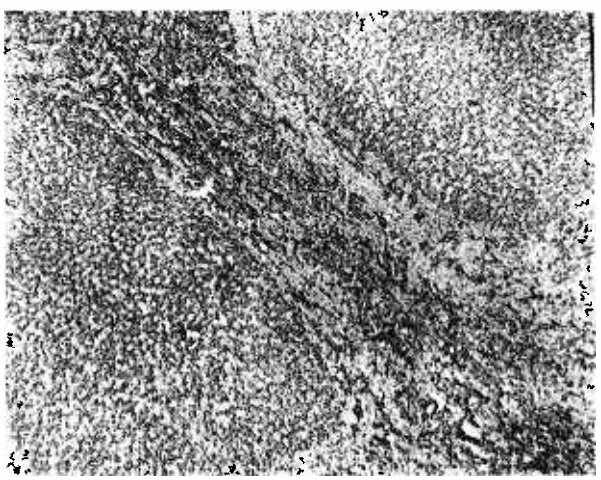

Fig. 3. Cirrhotic liver showing broad connective tissue bands separating lobules. H. & E. x 45.

The liver shows a vast increase in the periportal connective tissue which effectively isolates individual liver lobules. This connective tissue is moderately vascular and contains large numbers of bile canaliculi (Figs. 3 and 4). A fair number of these are not canalised, and are seen as solid epithelial branching cords. A few of the larger intrahepatic bile passages are filled with polymorphs and macrophages, and these show moderate periductal inflammatory change (Fig. 5). The liver cells are impregnated with bile, this being most noticable toward the periphery of the lobules.

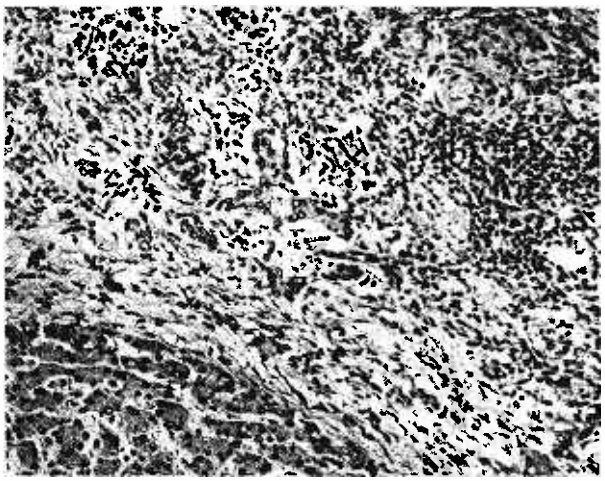

Fig. 4. Above the liver cells, lower left, in the fairly dense fibrous tissue of the biliary cirrhosis, there are numerous round cells and proliferating cords of bile duct epithelium. H. & E. x 150.

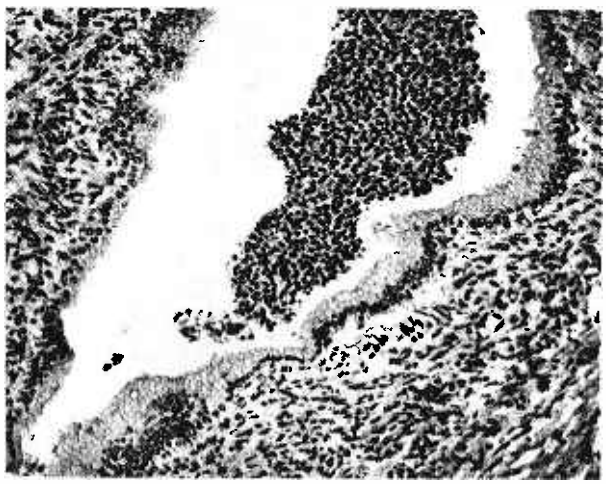

Fig. 5. Wall of large intra-hepatic bile duct shows mild inflammatory change; the lumen contains polymorphs and macrophages. H. & E. x 150.

The spleen shows a slight increase in fibrous tissue. A few bile-containing macrophages are scattered throughout the pulp.

There are no significant changes in any of the other organs examined.

Principal anatomical diagnoses:

Choledochus cyst
Ascending cholangitis
Biliary cirrhosis of liver
Multiple intestinal adhesions
Choledochocysto-gastrostomy
Subdural haemorrhage.

DISCUSSION

Incidence

Choledochus cyst is rare: Saltz and Glasser (1956) could collect only 209 cases from the world literature. There have been only two other cases in Singapore, that reported by Tan in 1952, and that by Choo in 1960.

About four females are affected for every male (Gross, 1953: Hutchins and Mansdorfer, 1944). Being generally seen in the young, some 75% of the reported cases were below 25 years of age (Attar and Obeid, 1955).

With increasing awareness of this anomaly, it seems likely that increasing numbers will be reported (Saltz and Glasser, 1956), as the clinical features are generally typical (Kelsey, 1947).

Aetiology

The aetiology of choledochus cyst is still obscure. Shallow *et al* (1943) listed 15 theories of origin, these large numbers indicating the extent of the uncertainty which still exists (Saltz and Glaser, 1956). It is true that in a certain number of cases there is demonstrable stenosis or angulation of the bile duct, but in other well studied specimens the outlet of the common bile duct shows no apparent abnormality (Gross, 1953). Congenital weakness of the duct wall would seem a reasonable cause, but Gross (1953) stressed that this alone is insufficient to produce dilatation. Some degree of obstruction is necessary; this may result from a number of lesions in the lower common bile duct.

Once the cyst attains a certain size a vicious circle begins, as drainage is further impeded by pressure from the eccentrically lying cyst (Taylor and Slaney, 1957).

Shallow *et al* (1943) and Horne (1957) concluded that the most acceptable theory is that advanced by Yotuyanagi (1936) who postulated that at the stage of physiological occlusion of the common bile duct during embryonic life, the cells of the upper segment of the duct may proliferate much more energetically than those of the lower portion, so that when recanalisation

occurs, the upper part of the duct will be abnormally wide and the lower part relatively narrow. Yotuyanagi (1936) also felt that there must be a second factor — obstruction.

Pathological Anatomy

Shallow *et al* (1943), Gross (1953) and Tsardakas and Robnett (1956), among others, have published excellent reviews and discussions on the subject, and much of the following description is based on their papers.

Characteristically, there is spherical dilatation of a part or the whole of the common bile duct, which may rarely include the junctions of the cystic and hepatic ducts with the common duct. The dilatation is a localised one and is confined entirely to the extrahepatic system. Thus, it can be easily differentiated from dilatation due to obstructive lesions of the lower common bile duct (Saltz and Glaser, 1956). Rarely the dilatation is so low that the pancreatic duct opens into it. Serfas and Lyter (1957) reported the only case in which the cyst was intraduodenal in position. The tumour mass is always retroperitoneal (Hutchins and Mansdorfer, 1944).

The cyst varies in size from that of a small cherry to a mass distending the whole abdomen. Reel and Burrell (1922) reported a case in which the cyst contained 8 litres of fluid.

The cyst wall is tough and may range from 2 to 10 mm. in thickness. Microscopically, it is made up of dense connective tissue, but occasionally, elastic and smooth muscle fibres are seen. There is generally evidence of inflammatory reaction. The lining epithelium of cuboidal or cylindrical cells may still be present, but has usually been denuded. The inner surface of the cyst may be roughened and irregular, and covered with inspissated bile pigment.

The cyst contains bile which varies in nature with the degrees of obstruction present; calculi within the cyst have been recorded (Keeley, 1948).

The cystic duct, hepatic ducts, and gall bladder may be slightly dilated. Occasionally, there are other congenital abnormalities as well, viz., cystic dilatation with congenital atresia of the bile duct (Ripstein and Miller, 1948) and double common bile duct with a cystic dilatation of the right duct (Swartly and Weeder, 1935; Weeder, 1939).

Irwin (1944) reported an unusual case in which the cyst was the seat of a carcinoma, resulting perhaps from long-standing chronic irritation.

The liver is often enlarged and cirrhotic, and there may be evidence of ascending cholangitis (Weeder, 1939; Swartly, 1943; Hutchins and Mansdorfer, 1944).

Clinical Features

Tumour, jaundice, and pain constitute the classic triad : when these appear a diagnosis of choledochus cyst should be entertained (Horne, 1957).

Attar and Obeid (1955) analysing 201 patients found that 80% exhibited a tumour, 75% were jaundiced, and 60% complained of pain. Tsardakas and Robnett (1956) stressed that the occurrence of all three components of the triad was by no means constant, being present in 63.3% of their series of 232 cases. They found that the pain is usually localized in the right upper quadrant, being cramping in nature. The jaundice, more often than not, is intermittent. The nature of the tumour has great clinical value as characteristically it is soft and rounded and movable laterally, but not vertically. If very large, it may occupy the whole abdomen.

Other signs and symptoms such as anorexia, vomiting, diarrhoea, or constipation may be present. Enlargement of the liver and spleen, and even Banti's syndrome has been observed in the advanced case. Fever is not a prominent feature, and when present, usually indicates cholangitis (Serfas and Lyter, 1957).

A striking feature, emphasized by Serfas and Lyter (1957), is the chronic and intermittent nature of the clinical course. There may be no symptoms for many years.

Sometimes there may be biliary peritonitis, usually fatal, resulting from spontaneous or traumatic rupture of the cyst (Blocker *et al*, 1937).

Investigation

A straight roentgenogram of the abdomen may show the cyst, especially if the wall is calcified (Attar and Obeid, 1955).

Cholecystography, whether oral or intravenous, is not helpful, as the gallbladder does not usually concentrate (Attar and Obeid, 1955; Serfas and Lyter, 1957). The easiest way of establishing the diagnosis in the operating room is by operative cholangiography (Tsardakas and Robnett, 1956).

A gastro-intestinal roentgenogram series is helpful, as it demonstrates the displacement of the surrounding gut by the cyst. The stomach is displaced downwards to the left, the duodenum medially and anteriorly, the coils of jejunum downwards and to the left, and the hepatic flexure and transverse colon downwards (Shallow *et al*, 1943).

Liver functions studies may be entirely normal or may indicate obstructive jaundice. However, these laboratory tests will typically fluctuate (Horne, 1957; Serfas and Lyter, 1957).

Shallow *et al* (1943) suggested that peritoneoscopy may be undertaken as a diagnostic aid in cases in which diagnosis is obscure.

Prognosis and Treatment

Conservative measures inevitably result in death from biliary cirrhosis, cholangitis, haemorrhage, or rupture of the cyst (Shallow *et al,* 1943). The treatment of choice is therefore early surgical invention (Horne, 1957).

The multiplicity of surgical procedures used in the past reflects the uncertainty felt in dealing with the condition. This uncertainty was associated, as was shown by Tsardakas and Robnett (1956), with a relatively high overall mortality rate (51%) in the 148 cases treated by operation up to 1943. The mortality rate in the 52 cases operated on between 1943 and 1956 was only 23%. This improvement, according to Tsardakas and Robnett (1956), was due to newer operative techniques and a more precise knowledge of the condition.

Present day opinion is that the treatment of choice is to anastomose the cystic dilatation and the intestinal tract, in one stage or two. The anastomosis may be a side-to-side choledochocystoduodenostomy (Gray, 1942; Shallow *et al,* 1946; Davis, 1948; Gross, 1953; Saltz and Glaser, 1956; Keith *et al,* 1957) or a choledochocysto--jejunostomy of the Roux-en-Y type (Keeley, 1948; Archambault *et al,* 1950; Attar and Obeid, 1955; Taylor and Slaney, 1957).

Comments on the present case

The apparent absence of communication between the dilated common bile duct and the duodenum places our case in the relatively small group of choledochal cysts which show definite obstruction.

As the cyst was successfully drained at the first operation, and as at necrospy there seemed to be no obstruction to the free flow of bile (Fig. 1), it would seem likely that much of the extensive biliary cirrhosis (Figs. 3 and 4) was already present at the time of first operation. The associated ascites may have predisposed to the formation of the abdominal band which required a further laparotomy to relieve the ensuing intestinal obstruction.

The burst abdomen, the ascending cholangitis (Fig. 5), and protein loss in the ascitic fluid, and possibly in the loose stools, were further obstacles on the road to recovery. The intracranial haemorrhage, probably the immediate cause of death, was perhaps a consequence of hypoprothrombinaemia.

It would seem therefore that if surgical correction is undertaken early, cirrhosis and ascending cholangitic infection are less likely to become established, with corresponding improvement in prognosis.

SUMMARY

The third Singapore case of choledochal cyst, occurring in a five-month-old Chinese girl, is presented.

The aetiology, pathological anatomy, diagnosis, prognosis and treatment of this rare condition are reviewed.

The importance of early surgical intervention is stressed.

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