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A left extra and intra-hepatic bile duct cyst with distal atresia

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Summary

A left extra and intra-hepatic bile duct cyst with distal atresia is presented. The case was treated by the excision of the extra-hepatic left bile duct cyst and primary anastomosis of the left intra-hepatic duct cyst to the common bile duct. The operative course and an eight months follow-up have been uneventful.

Key words: *Bile duct, cyst, congenital*

Introduction

Choledochal cyst or congenital common bile duct (CBD) cyst is a rare abnormality whilst common in Japan and the orient [4]. In Turkey 13 cases wih bile duct cyst, excluding the presented one, have been reported [1]. While the classification developed by Alonso-Lej, Rever, and Pessagno is widely accepted; Todani has reevaluated the entity, classifying it into six types by including the cystic dilatations in both the extra and intra-hepatic ducts [14].

A left extra and intra-hepatic duct cyst is presented in this paper, which could not be placed in one of the types in Todani's classification and may be considered as a subtype of type-4 in Todani's classification. The case was treated with the excision of the extrahepatic left bile duct cyst and primary anastomosis of the left intrahepatic duct cyst to the CBD.

Case Report

A female infant aged 2 months was admitted with a large abdominal mass, poor sucking, and vomiting since birth. She was mildly jaundiced, had transient clay-coloured stools, and looked well nourished. On admission, liver function tests were as follows: Alkaline phosphatase 200 BU/ml, SGPT 35 IU/ml and SGOT 40 IU/ml. Total serum bilirubin was 2.6 mg/dl, 2 mg of which beign conjugated. Intravenous pyelography excluded a retroperitoneal mass (Fig. 1).

An echogram confirmed a cystic mass, 20 cm in diameter which originated from the left hepatic lobe and extended to the pelvis inferiorly and to the spleen in the left upper abdominal quadrant. Laparotomy revealed a huge cystic mass attached to the hilum of the liver. Further exploration revealed a cyst of the left extra-and intra hepatic duct with distal atresia.

Treatment by excision of the left cystic hepatic duct and anastomosis of the intra hepatic cyst to the CBD was performed. A T-tube drainage was performed draining the left intra hepatic cyst which was about 2 cm in diameter. A postoperative T-tube cholangiography showed sufficient drainage of bile into the duodenum and healing of the anastomotic site (Fig. 2). The operative course and eight months of follow-up have been uneventful.

Discussion

Controversy exists concerning the correct mode of operation for congenital dilatation of the biliary system. Cystojejunostomy and cystoduodenostomy without excision of the cyst have been used most commonly [5,12]. However, anastomotic stricture, biliary stasis, cholangitis, biliary lithiasis, pancreatitis and carcinoma of the bile duct are the well known spesi-

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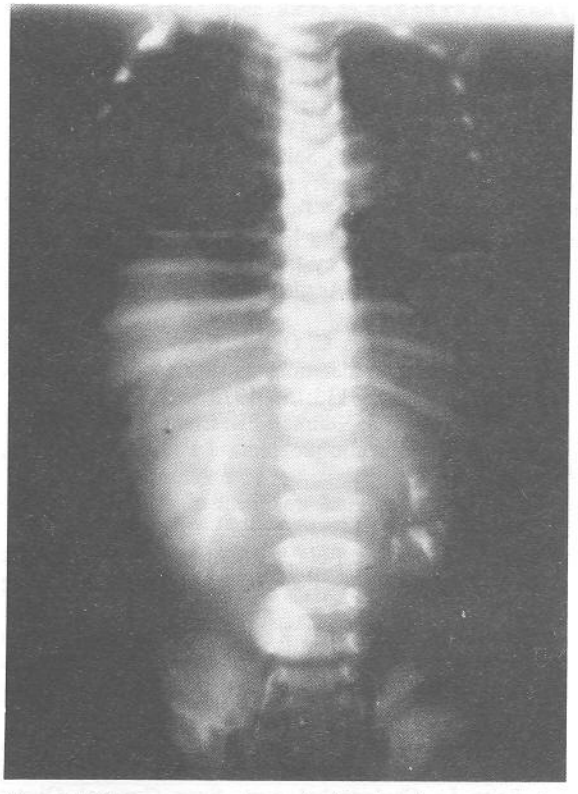

Figure 1. Intravenous pyelography showing the mass covering most of the abdomen.

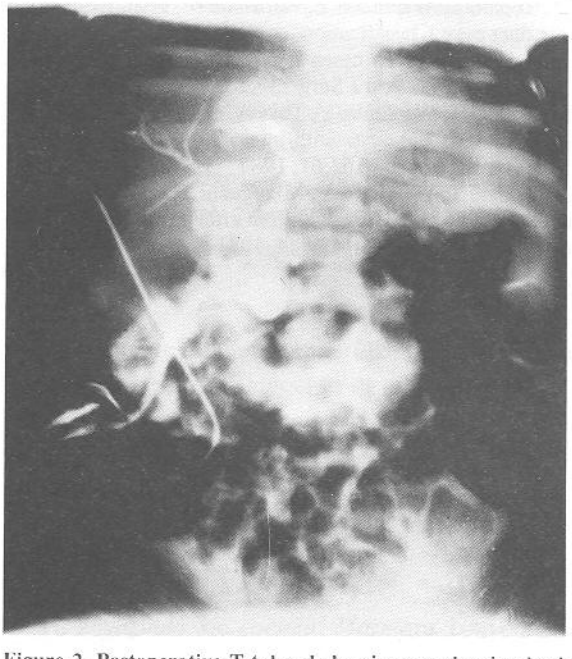

Figure 2. Postoperative T-tube cholangiogram showing healing of anastomotic site and drainage of the contrast material into the duodenum.

fic penalties for retention of the bile duct cyst (5,6,8,12). These complications are almost eliminated if the cyst is excised and bile drainage provided by a wide anastomosis of the intestine to the portahepatis (2,3,4,8,14,16).

But excision of the bile duct cyst has a higher risk factor and also a change of later anastomotic stricture (13). Late strictures can be prevented with wide anastomosis of the portahepatis to the jejunum (1,2,3,14,16). Direct anastomosis of the two ends of the bile duct after excision of the cyts is a more anatomophysiological approach which has been reported by Kasai (4), Kimura (6), Scarli (11), Klotz (7), and Longmire (9). Almost universal presence of a partial obstruction of the distal part of the CBD however, has hindered wide acceptance of direct anastomosis (10,15).

But, this procedure can be preferable, if the distal bile duct is large enough for adequate anastomosis and has sufficient muscular tissue, as was the situation in the presented patient. Left cystic hepatic duct had been excised and intra hepatic cyst reanastomosed to the CBD leaving minimal cystic tissue.

An eight-month follow-up revealed an uneventful course. Another method that could have been used in this case was a left partial hepatectomy, including the left intra-hepatic cyst, a procedure which needs more operative time than the operation applied to the presented case. Lateral hepatectomy with choledococystoduodenostomy having favorable results was performed by Todani and Caroli (cited in 11).

The classification developed by Alonso-Lej, Rever, and Pessagno is widely accepted, but this does not include the complete range of bile duct cysts (14). Cystic abnormalities of the biliary tree may be single or multiple, cystic or cylindrical, intrahepatic or extrahepatic.

Todani classified bile duct cysts into six types by including the cystic dilatations in both the extra and intra-hepatic ducts (14) (Table 1).

Todani's classification type IV-A represents and intra and extra-hepatic bile duct cyst covering choledochal dilatation. In the presented case the bile duct

Table I. Todani's classification of congenital bile duct cysts

Type I:	Common type; Ia) Choledochal cyst in a narrow sense Ib) Segmental choledochal dilatation Ic) Diffuse dilatation;
Type II:	Diverticulum type; in the whole extra-hepatic bile duct;
Type III:	Choledochocele;
Type IV-A:	Multiple cysts at the intra and extra-hepatic bile ducts;
Type IV-B:	Multiple cysts at the extra-hepatic duct only
Type V:	Intra-hepatic bile duct cyst.

cyst does not cover the CBD, so differs from type IV-A. Type IV-B includes only multiple cysts at the extra-hepatic bile duct. On the other hand, type-V covers only intra-hepatic duct cyst, these also do not confer with the presented case, since the presented case had an extra and intra-left hepatic duct cyst. It is therefore, the authors belief that, Todani's classification does not cover the presented case.

In conclusion the presented patient represents a case of bile duct cyst treated with primary excision of the extra-hepatic cyst and anastomosis of the intrahepatic cyst with CBD. This case can be considered as an isolated case of left intra and extra hepatic ductal cyst without choledochal involvement. Since it is not included in Todani's classification it is proposed that it may be well placed as a subtype of type IV-A.

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