

## Choledochal cyst and crossed fused ectopic kidney – a hitherto unknown association

Vikrant Sood<sup>1</sup>, Shorav Bhatnagar<sup>2</sup>

(1) Department of Pediatric Hepatology and (2) Department of Radiodiagnosis, Institute of Liver and Biliary Sciences (ILBS), New Delhi, India.

### To the editor,

Choledochal cyst is an uncommon biliary tract anomaly defined as dilatation of extrahepatic bile tree with or without intrahepatic biliary dilatation. It is one of the important differential diagnosis of jaundice and abdominal pain in the pediatric population. Though association with various developmental anomalies is already known, its combination with Crossed fused ectopic kidney has not been reported before in the literature to the best of our knowledge.

A 3 year old child was admitted with complaints of abdominal pain for the past 10 days with similar episodes in the past associated with mild icterus. On examination, mild hepatomegaly was present. Ultrasonography of abdomen showed a Choledochal cyst (Type Ic) with a crossed fused ectopic kidney. These findings were confirmed on Magnetic resonance imaging of the abdomen with cholangiopancreatography that showed a markedly dilated Common bile duct without intrahepatic biliary radical dilatation s/o Choledochal Cyst Type Ic, and Crossed Fused Ectopic Left Kidney (Figs. 1, 2). As per surgical opinion he was planned for Cyst excision with hepaticojejunostomy. But the parents refused surgery or any further investigations and the patient was lost to follow up without any surgical intervention.

Choledochal cyst has an incidence varying from 1 in 13-15,000 in the West to around 1 in 1000 in Asia, the reason of such variations being still unknown (1). It has been classified as per the commonly used but still debated Alonso-Lej classification with Todani modifications (1). Choledochal cysts are associated with many different developmental anomalies, including intestinal atresia, imperforate anus, pancreatic arteriovenous malformation, OMENS (Orbital distortion, Mandibular Hypoplasia, Ear anomaly, Nerve involvement, Soft tissue deficiency) plus syndrome, ventricular septal defect, aortic hypoplasia, pancreatic divisum, familial adenomatous polyposis etc (1).

When the kidney is located on the side opposite from which its ureter inserts into the urinary bladder, the condition is known as crossed renal ectopia. Associated anomalies include genitourinary defects (Multicystic dysplasia, patent urachus, hypospadias), Gastrointestinal defects (imperforate anus, annular pancreas), cardio-

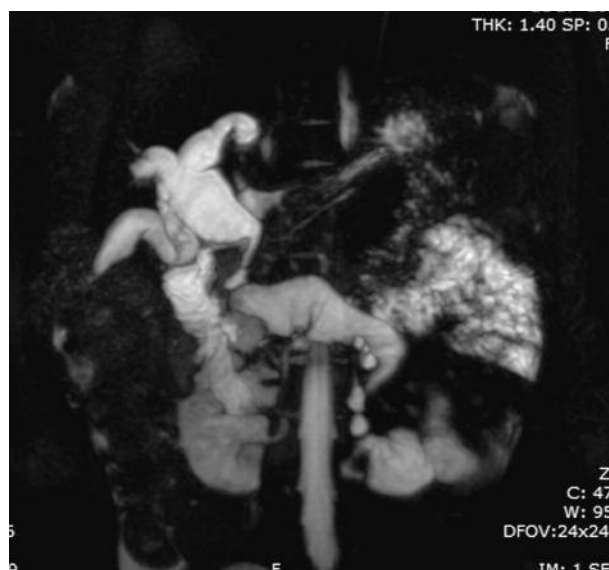


Fig. 1. — 3 D MRCP Image showing dilated proximal Common Bile Duct (CBD) extending up to primary confluence with normal distal part.

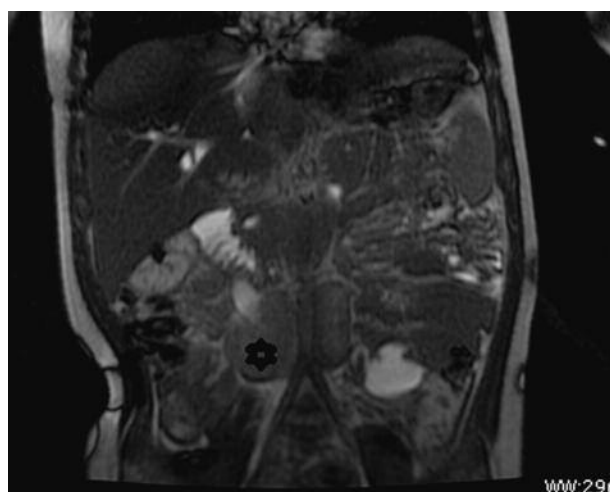


Fig. 2. — MR Coronal T2 Images Showing Ectopic fused Left Kidney (Star).

Correspondence to: Dr. Vikrant Sood, M.D., Department of Pediatric Hepatology, ILBS, New Delhi, India. E-mail: drvickyster@gmail.com

Submission date: 18/05/2012

Acceptance date: 22/06/2012

vascular defects (abdominal aneurysm, Takayasu's arteritis) etc (2,3). Usually presenting incidentally, they are diagnosed using a combination of intravenous urography, ultrasonography, and renal Computerised Tomographic angiography (4). Prognosis is usually good in absence of associated anomalies.

To summarize, the presence of a choledochal cyst should alert the treating medical professional regarding the possibility of crossed fused ectopic kidney in the same patient and vice versa. This can have significant implications regarding further management of the patient.

## References

1. SINGHAM J., YOSHIDA E.M., SCUDAMORE C.H. Choledochal cysts : part 1 of 3 : classification and pathogenesis. *Can. J. Surg.*, 2009, **52** : 434-40.
2. DEBNATH J.C., SARKER K.P., NASIR UDDIN M.D., SARKER S.K. *et al.* Crossed fused Ectopic Kidney – a Case Report. *TAJ*, 2003, **16** (2) : 79-81.
3. STUART B.B., ALAN D., PERLMUTTER, ALAN B.R. Anomalies of upper urinary tract. *In* : Campbell's Urology. 6th ed. PATRICK C., ALAN B.R., THOMAS STANEZY A., DARRACOTT VAUGHAN E. (eds). WB Saunders Philadelphia, 1992, pp. 1371-81.
4. BELEKAR D.M., DEWOOLKAR V.V., DESAI A.A., ANAM J.R. *et al.* An Unusual Case of Non-Functioning Crossed Renal Ectopia Without Fusion. *The Internet Journal of Surgery*, 2009, Volume 19, Number 2.