

tic regimens remain empirical, although, sulfonamides, 5-FC and hydroxystibamide isethionate have been recommended(2).

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Cholelithiasis Associated with a Variant of Annular Pancreas

M.H. Mehta
R.V. Patel
V.P. Hathila
D.B. Dekiwadia

The association of annular pancreas with biliary tract anomalies is extremely rare. Only two cases of agenesis of gall bladder(1) and one case of biliary obstruction caused by annular pancreas(2) have been reported. We report a variant of annular pancreas associated with agenesis of common bile duct, ectasia of extrahepatic biliary tract and cholecystolithiasis.

From the K.T. Children Government Hospital, Rajkot and Department of Surgery, M.P. Shah Medical College and Irwin Group of Hospitals, Jamnagar.

Reprint requests: Dr. (Miss) Manorama H. Mehta, Medical Superintendent, K.T. Children Government Hospital, Rajkot 360 001.

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Case Report

An 11-year-old boy was admitted with recurrent right abdominal pain of 4½ years duration. He had attacks of severe colicky pain associated with vomiting at the ages of 7 years and 9 years for which he was hospitalized. At both times he was diagnosed to have intestinal colic and poor quality emergency X-rays were reported to be within normal limits. Perabdomen examination showed fullness and mild tenderness in the gall bladder region. Investigations including liver function tests were within normal limits. Plain X-rays of abdomen showed multiple gall stones and ultrasound confirmed stones lying within the lumen of markedly dilated gall bladder. Oral cholecystography demonstrated non-visualization of gall bladder.

At exploration via right upper transverse incision, gall bladder was remarkably dilated containing multiple stones. There was a very long, dilated and tortuous cystic duct running parallel to common hepatic duct. At the lower end, annular pancreas was incompletely encircling the cystic duct and second part of duodenum, the anterior bridge being mainly fibrous tissue which was released gently. Further gentle dissection and complete Kocherisation of duodenum revealed agensis of common bile duct and cystic duct, common hepatic duct and pancreatic ducts were all found joining at one point just medial to the duodenal wall. There was mild dilation of common hepatic duct. Cholecystectomy and incidental appendectomy followed uneventful recovery. Gall bladder contained clear bile which did not grow any organisms on culture. Histologically its mucosa was found to be normal without any inflammatory changes. At follow up, the patient is asymptomatic and well.

Discussion

The development of the duodenum, pancreas and bile ducts has been described as an 'embryological traffic jam' and it has been suggested that the obstruction of the duodenum in annular pancreas is not the result of extrinsic compression but is due to a developmental failure(3). Consequently, it is not surprising that intrinsic duodenal obstruction and biliary tract anomalies often occur concomitantly.

Annular pancreas on the other hand has seldom been reported in association with biliary anomalies and a recent review of experience with annular pancreas reported no associated biliary tract anomalies(4). We could not trace a single case report in which annular pancreas was associated with agencies of common bile duct, mild ectasia of common hepatic duct and gross dilatation of cystic duct and gall bladder containing multiple gall stones.

Our case demonstrates that congenital absence of common bile duct may lead to mild dilatation of common hepatic duct and in itself does not require any surgical treatment. However, long, dilated and tortuous cystic duct in association with annular pancreas at its lower end causing incomplete obstruction led to poor drainage and cholestasis. The situation is comparable with choledochal cystic lesion of paraxial extrahepatic biliary tract and therefore, surgical treatment in the form of cholecystectomy has been recommended.

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Alström Syndrome

R.A. Garg
J. Singh
B.B. Mathur

The Alstrom Syndrome is a rare autosomal recessive disorder with involvement of the retina, ear, kidney and endocrine glands. During childhood, Alstrom syndrome usually presents with blindness (due to retinal degeneration and pigmentary changes) and nerve deafness. In adults, carbohydrate intolerance and slowly progressive renal disease develops; while obesity may disappear(1,2). Males have hypogonadism with small testes and low

plasma testosterone, but normal secondary sexual characteristics(3). Females lack evidence of hypogonadism but their menstrual periods are irregular. Other manifestations include hyperuricemia, hypertriglyceridemia(3), acanthosis nigricans(4) and baldness. This syndrome has never been diagnosed in childhood and only about ten cases in adults have been reported till date.

Case Reports

Two siblings, born to Indian Muslim first cousin consanguineous parents, are being reported. Three other siblings and other members of the family were normal.

Case 1: A 13-year-old girl was referred for evaluation of blindness and diabetes mellitus. She was delivered normally at term with an uneventful perinatal period. Parents suspected poor vision at 5-6 months of age. Till about 5 years of age she was able to recognize toys but subsequently visual acuity worsened and she became completely blind by 6 years. Parents noticed that she was obese at 6 years when compared to other children of her age. She also had hearing problems. One year back she attained menarche, but her periods were irregular and varied from scanty to hypermenorrhea.

On physical examination, the patient was a short statured, mildly obese, intelligent girl. Her height and weight were 140 cm (3rd centile) and 41 kg (25th centile), respectively. Scalp and axillary hair were sparse. There was no evidence of baldness. Pubic hair and breast development were normal for her age (SMR Stage 3). She was completely blind and deaf. Extremities were normal, with no digital anomalies.

Ophthalmological examination revealed bilateral dilated and fixed pupils with horizontal nystagmus. Optic fundii showed

From the Department of Pediatrics and General Medicine, Holy Family Hospital, New Delhi 110 025.

Reprint requests: Dr. Jagdish Singh, Department of Pediatrics, Holy Family Hospital, New Delhi 110 025.

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