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Annular Pancreas Associated with Downs Syndrome

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Abstract

This study reports a case of Term (GA 38 weeks) male baby with annular pancreas in association with Down syndrome which was diagnosed antenatally as duodenal obstruction and during operation annular pancreas was found in association with duodenal atresia. Due to prior knowledge of intestinal obstruction delivery was planned in a facility well equipped with NICU and Paediatric surgery team. He was offered the appropriate treatment without delay and outcome of the surgery was excellent. The treatment done was bypass duodenoduodenostomy and post-operative management in NICU.

Keywords: Downs syndrome; Pancreas

Introduction

The annular pancreas occurs in 1 in approximately 20,000 births. In children, it is more commonly associated with other congenital anomalies including Down syndrome, cardiac anomalies, duodenal atresia or stenosis and intestinal malrotation. The Annular pancreas results from abnormal rotation of the ventral pancreatic bud in its course around the posterior aspect of the duodenal anlage. The duodenum is encircled and might be obstructed by normal pancreatic tissue. Clinical presentation varies according to degree of obstruction whether as partial or complete duodenal obstruction. A duodenal diaphragm or duodenal stenosis often accompanies annular pancreas.

Case Report

A male infant was delivered by vaginal delivery at 38 weeks gestation to a Primigravida mother of 31 years age. The patient had a birth weight of 3180 grams and Apgar score of 9 and 9 at 1 and 5 minutes respectively. Routine antenatal ultrasound revealed maternal polyhydramnios. There was no family history of Diabetes or multiple gestation. Follow up scan done at 36 weeks of gestation showed abdominal findings of markedly distended fluid filled fetal stomach (measuring approx.7.98 X 3.12 X 3.45 cm, volume of 44.9 ml) with normal pylorus and dilated proximal duodenum along with Polyhydramnios. The risk of additional congenital anomalies especially Down's syndrome explained to parents. The procedure of amniocentesis was offered to confirm diagnosis of Downs's syndrome.

Neonatal and surgical team were updated. Baby was born in stable condition at term and no resuscitation was required. The infant showed characteristic facial appearance of Downs's syndrome with clinical features of bowel obstruction. On physical examination baby had brachycephaly, upslanting palpebral fissure, Hypotonia and single palmar crease.

After delivery the patient was admitted to NICU, kept nil by mouth and NGT was placed on continuous drain. X-ray abdomen revealed typical double bubble sign of duodenal obstruction. Ultrasound abdomen was performed to look for any other anomaly, but it was inconclusive. The patient was taken to operation theatre on the next day for laparotomy.

During the procedure, annular pancreas was found along with duodenal atresia. Bypass Duodenoduodenostomy was performed, and the patient was brought back to the NICU for further management. The case was managed with parenteral nutrition and kept nil by mouth post-operatively. Gradually feeding was introduced and tolerated well. Baby was discharged home at the age of 12 days in good condition taking full feeds orally for further follow up in clinic. Chromosomal analysis supported the

diagnosis of Trisomy 21. Echocardiogram performed during hospital stay was within normal limits and no associated cardiac anomaly [1-12] (Figure 1).

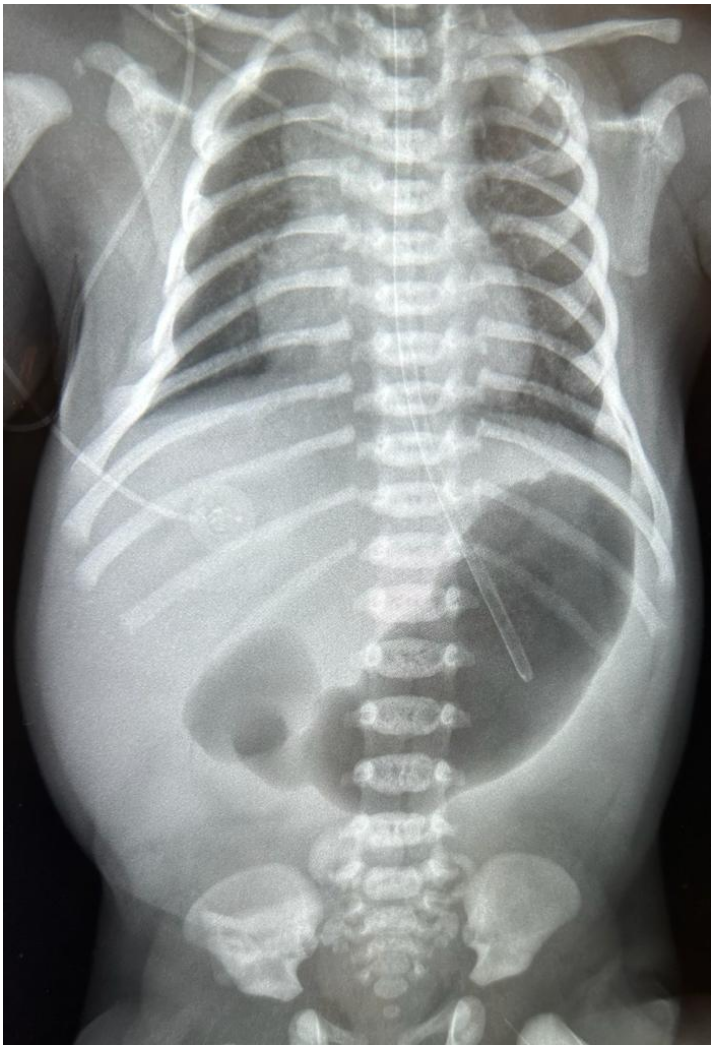

Figure 1: *Abdominal X-ray for Annular Pancreas.*

Discussion

Down's syndrome, or Trisomy 21, is the most common genetic condition of moderate intellectual disability. The condition can be suspected antenatally by characteristic features on ultrasound scan during second trimester of pregnancy and diagnosis confirmed antenatally by amniocentesis and chromosomal analysis.

It is usually suspected at birth due to characteristic facial appearance. The diagnosis is confirmed by chromosomal analysis. An affected child has 47 chromosomes and characterized by presence of 3 copies of chromosome 21. The extra chromosome 21 may result from Nondisjunction (94 %), Translocation (5%) or Mosaicism (1%). Down's syndrome is associated with multiple congenital anomalies. Among the common anomalies detected are congenital heart defects (50 %), Congenital and acquired gastrointestinal anomalies and hypothyroidism. Antenatal ultrasound scan has a key role in detecting congenital anomalies which are treatable. Once anomalies are detected, early surgical intervention offers a favorable outcome. In our case duodenal atresia was suspected antenatally and annular pancreas was detected during surgery. Baby had bypass duodenoduodenostomy and treatment was successful.

Annular pancreas is a rare congenital abnormality characterized by a thin band of pancreatic tissue surrounding the second part of duodenum leading to partial or complete duodenal obstruction.

During second month of gestation the ventral portion of the pancreas in the process of migration may become misaligned and encircle the second portion of the duodenum and fuse with the dorsal aspect of the pancreas. Such a ring of pancreas is accompanied by some degree of constriction in the duodenum almost invariably.

The age at presentation is related to degree of duodenal obstruction and coexistent malformations especially of rest of GI system.

Approximately one third of cases of Annular Pancreas present during neonatal period and half during the first year of life. Approximately two thirds of patients remain asymptomatic till later age. Annular pancreas may occur isolated or in association with other congenital malformations. The most common associated malformations are usually related to gastrointestinal system including duodenal abnormalities (web, stenosis and atresia), tracheoesophageal fistula, Hirschsprung´s disease and imperforate anus. Clinical manifestations of annular pancreas in the neonatal period present with features of intestinal obstruction including bilious vomiting, feeding intolerance, and abdominal distention. Passing meconium does not exclude the diagnosis of AP. In our case only abdominal distension and bilious aspirate was noted as feeding was not started due to prior knowledge of the condition in baby. After the neonatal period, symptoms differ which may include recurrent vomiting and chronic gastric distention. Recurrent attack of non-bilious vomiting and intolerance of solid foods are common symptoms. A plain abdominal X-ray or ultrasound in symptomatic neonates will show the classic double bubble sign but this is not a specific finding for Annular Pancreas. Although the diagnosis is usually made with an upper GI series or an abdominal CT scan in older children, surgery remains the gold standard method of diagnosis .In our case baby underwent laparotomy for duodenal obstruction and annular pancreas was found during surgery.

Pancreatic ring resection is not performed as there are associated duodenal malformations frequently associated with the condition and annulus often contains a duct which is adherent to the duodenal wall. Bypass duodenoduodenostomy or duodenojejunostomy is the surgery of choice.

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