

CASE REPORT

Scintigraphic detection of situs inversus totalis in a patient with suspected paediatric biliary atresia: a case report

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Abstract

Congenital anomalies in infancy pose diagnostic and therapeutic difficulties. Biliary atresia is a common diagnosis in children with unrelenting neonatal jaundice with conjugated hyperbilirubinaemia. The morbidity associated with this condition is compounded by the concomitant presence of complete or partial *situs inversus* which is a condition in which major visceral organs are reversed from their original positions.

Combination of biliary atresia with situs inversus is a challenging diagnosis to make. Equivocal ultrasonographic findings not leading to a definitive diagnosis of situs inversus, not only compound the clinical features but also lead to a delay in the correct diagnosis. Scintigraphic imaging modalities especially technetium-99m hydroxy imino diacetic acid (^{99m}Tc-HIDA) scan can play a vital role in overcoming these problems.

We report a case of 2-month-old child who presented with persistent neonatal jaundice, biliary atresia, situs inversus totalis and generalized sepsis finally leading to his death.

***Key words**: Situs inversus totalis, billiary atresia, hepatobiliary scintigraphy*

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Introduction

Congenital anomalies are an area involving multidisciplinary diagnostic and therapeutic decision making. Such congenital anomalies of clinical relevance which are encountered in infancy, pose a major therapeutic challenge in view of the difficult perisurgical conditions and intraoperative difficulty due to the age of the patient as well as increased post-operative morbidity and mortality.

Unremitting neonatal jaundice with conju gated hyperbilirubinaemia should lead to a high index of suspicion for a likely diagnosis of underlying paediatric biliary atresia. Biliary atresia is a cause of disruption in the extrahepatic biliary system due to post-hepatic obliteration of bile circulation secondary to the congenital anomaly. A timely and correct diagnosis followed by recourse to surgical intervention decreases the likelihood of development of secondary biliary cirrhosis, which is an otherwise an invariable outcome.

After the identification of biliary atresia as the

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cause of conjugated hyperbilirubenemia, it is important to stratify the patients into two distinct groups depending on the presence or absence of associated situs inversus.

Imaging modalities not only play an important role in diagnosis but also provide information useful for the subsequent surgical procedure. We report a case of 2-month-old boy presenting with neonatal jaundice suspected to be due to biliary atresia who was subsequently found to have situs inversus totalis.

Case report

56-day-old male child presented with shortness of breath, jaundice, clay-coloured stool and irritability for two weeks. He was born full-term after a normal unprovoked rupture of membrane by spontaneous vaginal delivery at home in the presence of local mid-wife. He was the fourth child born out of consanguineous parentage (first cousins). He had normal APGAR score with an unremarkable antenatal history and values of birth weight and height with in 90th percentile for the age-adjusted reference chart. There was no history of birth asphyxia, cyanosis, delayed cry or feeding issues. There was no history of perinatal complications till 48 hours after birth. A suspicion of a pathologic cause of jaundice was suspected as the jaundice since birth failed to subside even after 5 days. Due to the non-availability of inpatient phototherapy, which should be instituted within 8 hours of qualifying biliary levels as per guidelines, the patient was discharged from hospital with a recommendation to acquire maximum sun exposure and advised to seek review at a tertiary care hospital. During his stay at hospital the patient was administered injection vitamin K 0.5 mg IV and multivitamin syrup and was fed with mother's expressed breast milk on crying.

The child generally remained symptom free except for the persistent jaundice, which aggravated over the next few weeks with a darker discoloration of skin and sclera. The parents sought local medical advice from the same hospital as well as private outpatient clinics nearby but there was no amelioration of symptoms.

At one and half month of age, his mother noticed gradually worsening poor feeding along with an irritable behaviour and semi formed frequent clay coloured loose stools. The child was finally taken to a tertiary care hospital at Islamabad for a paediatric consultation where he was admitted. On examination, the patient was active, mildly irritable, jaundiced with tender hepato splenomegaly and signs of inadequate hydration including sunken eyes, cold extremities and coated tongue. Serum total bilirubin level was variable with values ranging from 3.1 to 11.94 mg/dl over a period of one week along with elevated serum alanine transaminase (238 U/l, normal <40 U/l), aspartate transaminase (238 U/l, normal <49 U/l) and alkaline phosphatase levels (1059 u/l, normal 65-306 U/l). Complete blood picture revealed normal-for-age haemoglobin (11.8 g/dl) along with elevated total leukocyte count. Coagulation profile (prothrombin time 13/13 sec, activated thromboplastin time 37/33 sec) and renal function tests (urea 27 mg/dl, normal 12-50 mg/dl; creatinine 0.44, normal <1.2 mg/dl) were within normal limits. The patient was kept under observation for 10 days and during this period was subjected to diagnostic tests to determine the probable cause of persistent hyperbilirubinaemic neonatal jaundice. The child gradually developed sepsis despite intravenous antibiotic cover and high dependency care (total leukocyte count 28.1 x 109/l). A clinical suspicion of biliary atresia was raised but a subsequent ultrasonographic review was inconclusive.

The patient was finally referred to Nuclear Medical Centre, Armed Forces Institute of Pathology Rawalpindi, for review regarding possible corroboration of clinical diagnosis by scintigraphic imaging. The patient was evaluated by a hepatobiliary scan to assess the functional status of gall bladder. The patient was injected with 75 MBq of ^{99m}Tc-BrIDA (bromo-2, 4,6-trimethylacetanilido iminodiacetic acid) intravenously in a dorsal

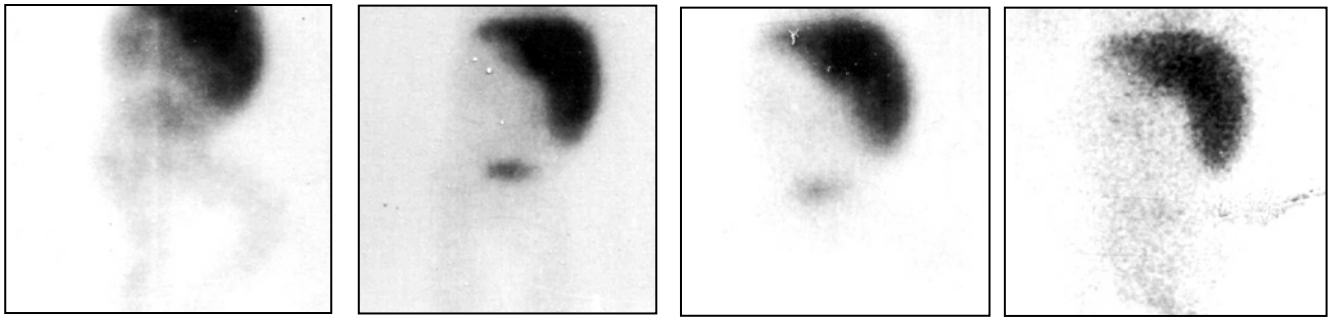

Figure 7 *From left to right: static images at 1-min, 1-hour, 3-hour and 24-hour postinjection*

metacarpal vein. An initial dynamic study was carried out for 5 min followed by serial static images at 5, 10, 15, 30, 45, 60, 120 and 180 minutes, with a final static view at 24-hour postinjection. Analysis of scintigraphic data demonstrated enlarged liver with fairly homogenous tracer uptake, lying in the left hypochondrium. The gallblader wasn't visualised and there was no intestinal activity seen on the images acquired until 24-hour postinjection with only generalized hepatic parenchymal retention of activity seen. The findings were consistent with persistent hepatogram and confirmed the diagnosis of biliary atresia with situs inversus (Figure 1).

The patient was referred for echocardiography, which demonstrated dextrocardia with double inlet left ventricle, unbalanced ventricles with hypoplastic right atrioventricular valve, ventricular septal defect, malposition of great vessels, pulmonary stenosis and patent ductus arteriosus. Ultrasonography revealed situs inversus with non-visualization of gall bladder and prominent biliary channels. Hence, after careful evaluation of history, examination, laboratory studies and imaging modalities, a final diagnosis of situs inversus totalis with biliary atresia was confirmed.

Discussion

The first case of dextrocardia was reported in a male child in 1906, followed by other such cases reported from various parts of the world [1]. Situs inversus (also sometimes called as situs transversus or oppositus), is a congenital condition in which the major visceral organs are reversed from their normal anatomical positions. The condition is called situs inversus totalis if the position of heart is also changed in addition to the positions of other organs [2]. Epidemiological data on situs inversus varies among different populations but the prevalence is less than 1 in 10,000 people [3]. Situs inversus is seen as an associated feature in 20% of cases of biliary atresia [4]. There are three main types of extrahepatic biliary atresia: type I which involves common bile duct only, type II involving the common hepatic duct and type III with atresia of either the right or left hepatic duct. Other associated anomalies in addition to situs inversus include, cardiac lesions, polysplenia, absent vena cava and a preduodenal portal vein [5,6].

Situs inversus has a strong correlation with biliary atresia as 28% of the infants with situs inversus develop biliary atresia as compared to 0.01% of general population [7]. A diagnosis of complete extrahepatic biliary atresia should caution the physician and investigator alike about the possibility of underlying situs inversus.

In our case a timely diagnosis of biliary atresia was not aggressively pursued to rule out underlying situs inversus. The diagnostic difficulty was compounded by equivocal ultrsonographic findings leading to a delay in diagnosis. The diagnosis of situs inversus was finally established by ^{99m}Tc-BrIDA scan, which not only established the actual extent of the sclerosing fibroobliteration of bile duct but also confirmed the presence of underlying situs inversus.

Limited availability and attending costs have led to an under utilization of scintigraphic imaging modalities as an adjunct in timely and correct decision-making. A timely and clinically warranted recourse to nuclear medicine imaging modalities can not only reduce the morbidity and mortality of disease but also cut costs in terms of limiting unnecessary treatment expenses incurred partly because of incomplete or late diagnosis.

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