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A RARE CASE OF BILIARY ATRESIA WITH IT'S COMPLICATION OF PORTAL HYPERTENSION: A CASE REPORT

Radiodiagnosis

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ABSTRACT

Biliary atresia (BA) is a progressive, idiopathic, fibro-obliterative disease of the extrahepatic biliary tree that presents with biliary obstruction exclusively in the neonatal period. It has been listed as a rare disease by the Office of Rare Diseases (ORD) of the National Institutes of Health (NIH) - the incidence is as low as only 1 patient in 10,000 to 20,000 live births. However, Biliary Atresia is the most common surgical cause of neonatal jaundice for which Liver Transplantation is indicated. We report a rare case of Biliary Atresia in a 6 months old female child who presented with jaundice and abdominal distension.

KEYWORDS

Biliary Atresia, Ultrasonography, HIDA Scan, Portal hypertension

- **CASE REPORT**

A 6 months old female child presented with a gradual onset of abdominal distension and yellowish discoloration of body, sclera and urine over a period of 1 month. No significant family history was present.

Blood examination revealed markedly elevated Bilirubin (Total: 17.6 mg/dl, Direct: 12.2 mg/dl, Indirect: 5.4 mg/dl), elevated Alkaline Phosphatase (194 U/L) and low total proteins (5.2 gm/dl) – an indication towards hepatic involvement. Total Leucocyte Count (13200/mm^3), ESR (28 mm/hr) and Prothrombin Time (24 sec) were also elevated.

Ultrasonography of Abdomen & Pelvis revealed small atretic intrahepatic gall bladder **(Ghost GB)** [fig 1], non-visualization of Common Bile Duct in its entire course, instead a tubular echogenic structure was visualized just above portal vein **(Triangular Cord Sign)**, [fig 2].

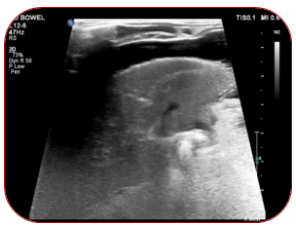


Fig 1: Small atretic intrahepatic GB (Ghost GB)

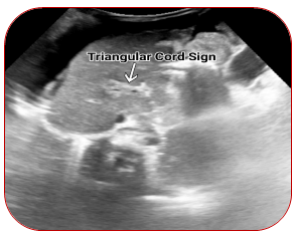


Fig 2: Triangular Cord Sign (Echogenic Periportal Fibrosis)

Other significant USG findings included cirrhotic liver (7.4 cm) [fig 3], splenomegaly (7.9 cm) [fig 4], moderate ascites, Peak Systolic Velocity of 19.6 cm/sec in Portal Vein with multiple collaterals in epigastric region, mesenteric region, at lineorenal ligament and at splenic hilum - findngs consistent with changes of Portal Hypertension.

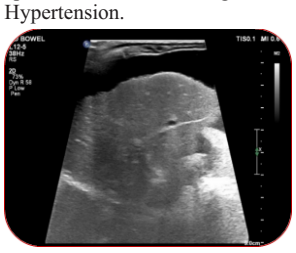


Fig 3: Cirrhotic Liver

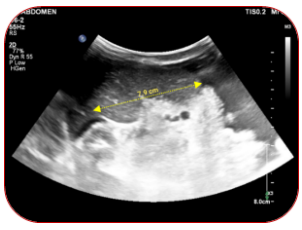


Fig 4: Splenomegaly

HepatobiliaryIminoDiacetic Acid (HIDA) scan was performed for further evaluation. Dynamic images were obtained in the anterior and right lateral projections for 45 minutes after intravenous bolus injection of 1.5 mCi 99m-Tc Mebrofenin. Appropriately timed delayed static images were obtained thereafter. Findings revealed very poor hepatic function with no appreciable excretion of tracer in either intrahepatic or extrahepatic biliary system. [fig 5a, fig 5b]

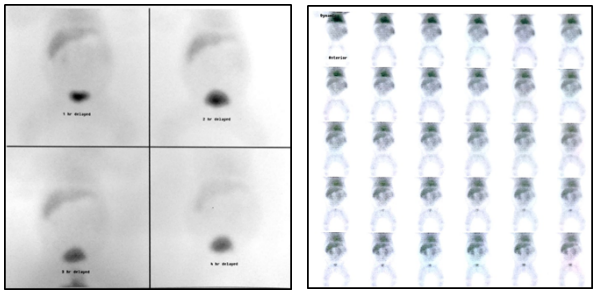


Fig 5a, Fig 5b: HIDA Scan. Renal excretion of tracer with no appreciable Hepatic excretion

Considering the age of just 6 months alongwith consistent symptoms and results of various investigations, a diagnosis of Biliary Atresia with Portal hypertension with hepatic dysfunction was established.

The patient is currently listed for Liver Transplantation.

- **DISCUSSION**

Biliary atresia (BA) is a progressive, idiopathic, fibro-obliterative disease of the extrahepatic biliary tree that presents with biliary obstruction exclusively in the neonatal period.

- **MECHANISM OF LIVER DAMAGE:** Bile is a digestive enzyme that is made in the liver. It travels through the bile ducts to the second part of duodenum, where it helps digest fats.

In biliary atresia, bile can't flow into the intestine and builds up in the liver leading to damage. The damage leads to scarring, loss of liver tissue and function, and cirrhosis.

The cause of this disease is not known. In some infants, the condition is most likely congenital, meaning present from birth due to defect in early bile duct development (particularly those with other associated anomalies).

Some may arise in the perinatal period due to an external cause such as an hepatotropic virus, reovirus 3 infection, congenital cytomegalovirus infection and autoimmunity.

- **TYPES OF BILIARY ATRESIA:**

- Depending on the associated anomalies, infants with Biliary Atresia can be grouped into three categories:
- **Biliary Atresia without any other anomalies or malformations:** 70 to 80 % of total cases. This type is also referred to as Perinatal Biliary Atresia.

These children are born without jaundice, but within the first two months of life, jaundice develops and stools become progressively acholic.

- **Biliary Atresia in association with Laterality Malformations:** 10 to 15 % of total cases. This pattern is also known as Biliary Atresia Splenic Malformation (BASM) or Embryonal Biliary Atresia.

The laterality malformations include situs inversus, asplenia or polysplenia, malrotation, interrupted inferior vena cava, and cardiac anomalies. Children with BASM have poorer outcomes compared with those with perinatal Biliary Atresia due to the associated cardiac abnormalities [10-12].

- **Biliary Atresia with other congenital malformations:** 5 to 10 % of total cases.

Associated congenital malformations include intestinal atresia, imperforate anus, kidney anomalies, and/or heart malformations.

- Depending on the extent of involvement, Biliary Atresia can be grouped into three categories [fig 6]:

- **Type I:** Obliteration of the common bile duct; proximal bile ducts are patent.
- **Type IIa:** Atresia of the hepatic duct, with cystic bile ducts found at the porta hepatis.
- **Type IIb:** Atresia of the cystic duct, common bile duct, and hepatic ducts.
- **Type III:** Involvement of the extrahepatic biliary tree and intrahepatic ducts of the porta hepatis.

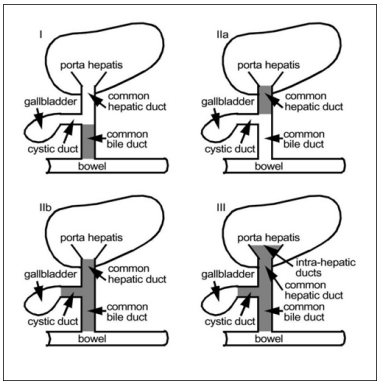


Fig 6: Classification of Biliary Atresia

- **SYMPTOMS:**

Symptoms of biliary atresia are usually evident between one and six weeks after birth. Patients develop progressive cholestasis due to inability of bile excretion from liver. When the liver is unable to excrete bilirubin through the bile ducts in the form of bile, bilirubin begins to accumulate in the blood, causing symptoms.

These symptoms include yellowish discoloration of the skin, itchiness, poor absorption of nutrients (causing delayed growth), pale stools, dark urine, and a distension of abdomen. Eventually, cirrhosis with portal hypertension will develop. If left untreated, biliary atresia can lead to liver failure.

Biliary-atresia-related cholestasis mostly does not result in kernicterus (brain damage due to high blood level of bilirubin resulting from liver dysfunction). This is because in biliary atresia, the liver, although diseased, is still able to conjugate bilirubin, and conjugated bilirubin is unable to cross the blood–brain barrier.

- **COMPLICATIONS:** If not managed in time, Biliary Atresia can lead to life threatening complications from Cirrhosis, Portal Hypertension to eventual Liver Failure.

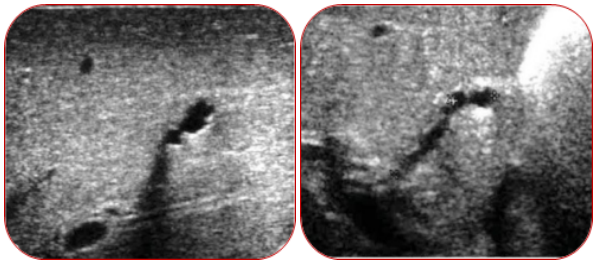
- **TREATMENT:**

Once biliary atresia is diagnosed, surgical intervention is the only option available for a definitive treatment (Kasai portoenterostomy).

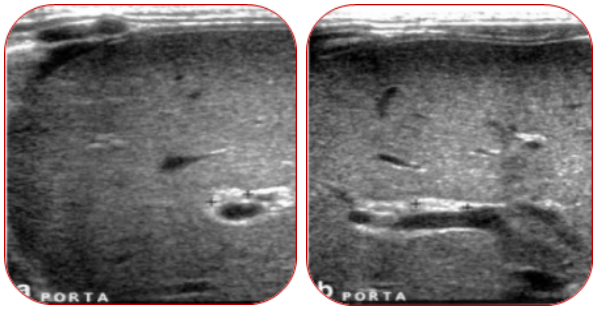
- **IMAGING MODALITIES IN BILIARY ATRESIA**
- **ULTRASONOGRAPHY:**

Ultrasonography is generally the initial investigation in patients with suspected biliary atresia. Complete visualization of the extrahepatic biliary system excludes biliary atresia, whereas nonvisualization of the common or hepatic bile ducts suggests the disease.
Ultrasonographic findings include:

- Gall Bladder Ghost Triad: This includes an atretic gallbladder with
  - Gallbladder length less than 1.9 cm
  - A thin, indistinct gallbladder wall
  - An irregular and lobular contour.



- Triangular Cord Sign: In biliary atresia, the hepatic parenchyma is often inhomogeneous, with a marked increase in periportal echoes due to fibrosis. Sonography often shows a circumscribed, focal, triangular or tubular echogenic density more than 3 mm thick located cranial to the portal vein bifurcation. This is the triangular cord sign and is due to fibrosis of the extrahepatic biliary system.



- Central Biliary Cysts and Choledochal Cysts may be associated with biliary atresia and can be depicted on sonography.

- **NUCLEAR IMAGING:**

Cholescintigraphy or hepatobiliary scintigraphy is scintigraphy of the hepatobiliary tract, including the gallbladder and bile ducts. It is known by different names depending on which radiotracer is used, such as HIDA scan, PIPIDA scan, DISIDA scan, or BrIDA scan. A radioactive tracer is injected through any accessible vein and then allowed to circulate to the liver from where it is excreted into the bile ducts and stored by the gallbladder, until released into the duodenum.

If excretion of radiotracer into the bowel is seen, biliary atresia is virtually excluded. If radiotracer excretion is absent after 24 hours, biliary atresia is suspected.

- **Magnetic Resonance Cholangiopancreatography:**

Findings in infants with biliary atresia include incomplete visualization of the extrahepatic biliary system and periportal high signal intensity on T2-weighted magnetic resonance imaging (MRI) scans (which may represent cystic dilatation of fetal bile ducts with surrounding fibrosis).

- **Endoscopic Retrograde Cholangiopancreatography:**

It is a rarely used invasive modality for evaluation of Biliary Atresia. ERCP allows direct visualization of the extrahepatic biliary tree with the injection of radiologic contrast agent into the extrahepatic biliary system through the papilla of Vater.

This technique can show obstruction in the common bile duct and enables visualization of the extrahepatic biliary system distal to the common hepatic duct and the extrahepatic biliary system.

- **Surgical And Percutaneous Cholangiography:**

Surgical cholangiography is typically performed by injecting contrast material through the gallbladder. If no communication is seen between the biliary tree and the gastrointestinal tract, biliary atresia is diagnosed.

Ultrasonography-guided percutaneous cholecystocholangiography is a relatively new technique in which radiographic contrast material is injected into the gallbladder under ultrasonographic guidance and the extrahepatic biliary system is viewed with fluoroscopy.

• CONCLUSION

The reported case provides important teaching points with regards to clinical and radiological findings of Biliary Atresia, which is a relatively rare clinical condition. Early Diagnosis and prompt management of this condition is essential since it can lead to fatal outcomes. Adequate knowledge on radiological findings of the condition in conjunction with clinical presentations not only enable a correct diagnosis and meaningful management of Biliary Atresia but also rule out other diagnostic entities, which may have a similar clinical presentation. The rarity of this condition prompted us to report this case.

• CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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