

INSPISSATED BILE SYNDROME: A CASE REPORT

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ABSTRACT

Neonatal cholestasis is a diagnostic dilemma in clinical practice.⁽¹⁾ It is a serious condition with varied etiological factors. Biliary atresia, hepatitis, and choledochal cyst are common causes in a neonate. Intrahepatic and extrahepatic causes can be easily differentiated by radiological investigations.⁽²⁾ Inspissated bile syndrome is a rare cause of neonatal jaundice.^(3,4) Predisposing factors for the development of inspissated bile in neonates include hemolysis, prematurity, parenteral nutrition, sepsis, and diuretic therapy⁽⁵⁾. Ultrasonography is valuable in diagnosing the causes of obstructive jaundice, especially in neonates where computed tomography scans cannot be done due to radiation exposure and MRI cannot be done due to the motion artifacts. Hepatobiliary scintigraphy single-photon emission computer tomography (HBS SPECT) is the most reliable diagnostic method. Generally, inspissated bile resolves spontaneously, but sometimes patients need ursodeoxycholic acid. Rarely surgical drainage is also needed.

KEYWORDS :

CASE REPORT

An 8-day-old male neonate was brought to the department of radiodiagnosis for ultrasonography. The patient had yellowish discoloration of skin and sclera. The patient also had fever and tachycardia.

It was full-term vaginal delivery with assisted techniques. The patient didn't cry immediately after birth and had a poor APGAR score at 1 min. The patient also developed cephalohaematoma.

On lab investigations total bilirubin level was 16.2mg/dl, unconjugated bilirubin was 11.5 mg/dl, and conjugated bilirubin was 4.7mg/dl. He also had raised WBC levels (20600), Alkaline phosphate -84, serum glutamate pyruvate transaminase (SGPT)-26, and serum glutamic oxaloacetic transaminase (SGOT) -61, and total protein-5.5 and albumin-3.3. Since the unconjugated bilirubin was raised liver disease was suspected and the patient was brought to ultrasonography for further evaluation.

The ultrasonography was done by using an 8-13mhz probe. On the greyscale image, echogenic material was seen distending the common bile duct (without any evidence of posterior acoustic shadowing), which was suggestive of inspissated bile (Fig. 1). The common bile duct measured 7 mm in diameter, however, Intra Hepatic Biliary Radicles were normal. The gall bladder was distended (Fig. 2), however, its wall thickness was normal. The liver measured 4.5cms, with normal echotexture and the portal vein was also normal (Fig. 3). There was no evidence of choledochal cyst or extrahepatic biliary atresia. Bilateral kidneys and ureters were normal. The visualized bowel loops were normal. Later the patient was put on ursodeoxycholic acid.



Figure 1. Grey Scale Ultrasonography Image Showing Dilated Common Bile Duct With Echogenic Sludge Within



Figure 2. Ultrasonography Image Showing Distended Gall Bladder With Normal Wall Thickness



Figure 3. Ultrasonography Image Showing Normal Echotexture Of Liver, With Normal Portal Vein And Its Bifurcation

DISCUSSION

Cholestasis refers to decreased bile formation or excretion. Neonatal cholestasis is hyperbilirubinemia that is present at birth or develops a few days after birth.⁽¹⁾

Cholestasis can cause liver dysfunction, liver transplantation may be required and eventually leading to death. Thus, early diagnosis and evaluation are important.⁽⁶⁾ Biliary atresia is the most common cause of cholestasis in 1st month of life.⁽⁶⁾ The

various causes of obstructive jaundice in neonates include choledochal cyst, Caroli's disease, and inspissated bile syndrome.⁽⁷⁾ The clinical symptoms include acholic stool, jaundice, yellow-pigmented urine and rarely bleeding due to vitamin k deficiency secondary to cholestasis. In neonates differentiating between biliary atresia and neonatal cholestasis is crucial as biliary atresia requires surgery.⁽⁸⁾

Ultrasonography is well suited for the disease of gall bladder and liver in neonates.⁽⁹⁾ Ultrasonography plays an important role in differentiating obstructive and non-obstructive causes of neonatal jaundice.⁽⁷⁾ Liver ultrasonography should be performed with 3-4 hours of fasting thus allowing the full distention of the gall bladder and minimizing the amount of the bowel gas. Higher frequency transducers of 7.5–12 MHz are also helpful in young infants because of their small size.⁽⁶⁾ In children, the normal liver size is 5-9.5 cms and the common bile duct diameter is 0.1-0.2 cms.⁽⁸⁾

Hepatobiliary scintigraphy is generally used to differentiate biliary atresia from other causes of cholestasis by demonstrating patency of the biliary tract. If 5 mg/kg/day phenobarbital is started at least 5 days prior, biliary excretion of the isotope is enhanced but has delayed diagnosis. If the uptake of the labelled substances by the hepatocytes is delayed, neonatal hepatitis should be considered.

Hepatobiliary scintigraphy single-photon emission computer tomography (HBS SPECT) is the most reliable diagnostic method compared to magnetic resonance cholangiography, HBS (hepatobiliary scintigraphy), or ultrasonography.

Magnetic Resonance Cholangio-Pancreatography is a non-invasive technique for evaluating the intrahepatic-extrahepatic bile ducts and the pancreatic duct.⁽⁶⁾ Magnetic Resonance imaging has the advantages of multiplanar and multiphase imaging, high contrast resolution, and lack of ionizing radiation, making it a preferred modality over computed tomography.⁽⁸⁾

Inspissated bile syndrome or bile plug syndrome is extrahepatic obstruction of the bile duct by sludge in full-term infants, without any congenital abnormality or hepatocellular lesion.⁽¹⁰⁾ It is a rare cause of jaundice in neonates.^{(3),(4)} In such cases sludge in the gallbladder appears as low-level echoes within the lumen. Inspissated bile is slightly more echogenic but does not give shadowing. Sometimes it is seen in the common bile duct causing partial or complete biliary duct obstruction. Inspissated bile syndrome may be associated with massive hemolysis due to Rh incompatibility, haemorrhage, or various intestinal diseases (increase enterohepatic circulation).⁽⁴⁾ Other risk factors for the formation of sludge are total parenteral nutrition, diuretic, cephalosporin, and prematurity.⁽⁸⁾

Inspissated bile may resolve spontaneously or with ursodeoxycholic acid. Rarely endoscopic retrograde cholangiopancreatography or percutaneous drainage may be needed.⁽⁸⁾

Earlier complications like cholangitis and liver abscess were treated with percutaneous transhepatic cholangiography (PTC) or cholecystography with irrigation of the biliary tree with saline.⁽²⁾

CONCLUSIONS

Inspissated bile syndrome is a rare cause of neonatal jaundice. Ultrasonography plays an important role in the diagnosis, especially in neonates. The other modalities that can be used for confirmation are hepatobiliary scintigraphy single-photon emission computer tomography (HBS SPECT), hepatobiliary scintigraphy, and magnetic resonance

cholangiopancreatography. Hepatobiliary scintigraphy single-photon emission computer tomography (HBS SPECT), is the most reliable investigation. Thus directing the clinician for proper treatment of the patient. Generally, the bile plaque passes out spontaneously or with the use of ursodeoxycholic acid. Rarely endoscopic retrograde cholangiopancreatography or percutaneous drainage has to be done.

Complications like cholangitis and liver abscess may arise which have to be treated surgically.

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