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Research Article

Periportal Fibrosis and Contracted Gall Bladder in USG of Hepatobiliary System- As a Diagnostic Factor and Therapeutic Strategy for Biliary Atresia

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Abstract

This study aims to establish the ultrasonographic finding of periportal fibrosis and contracted gall bladder as diagnostic factor and therapeutic strategy for Biliary Atresia. Twenty-five infants were taken into study from January 2014-March 2016. All the patients had undergone ultrasonography of whole abdomen. For assisting the diagnosis 20 children had HIDA scan and percutaneous liver biopsy. The diagnosis was supported by undergoing Kasai procedure in 18 children. It was further supported by histopathological report after Kasai procedure. The age of presentation varies from 45 days to 120 days. Most of the children had clinical presentation of Biliary Atresia i.e. persistent jaundice, alcoholic stool, and abdominal mass. For further diagnosis Ultrasonography of whole abdomen was done. USG finding in 19 patients revealed periportal fibrosis ranging from 15 x 09 mm - 20 x 12 mm. In 17 patients the gall bladder was present which appeared to be irregularly contracted. In 4 patients saccular well-defined gall bladder was present. In three patients gall bladder was absent. Among 20 patients with HIDA Scan and percutaneous liver biopsy 18 patients revealed features of Biliary Atresia. Among 18 patients who had undergone Kasai procedure, 16 patient revealed absences of Biliary tract and contracted gall bladder. Two patients had common bile duct and normal looking gall bladder. The tissue from the extrabiliary tract on histopathology after Kasai procedure also revealed periportal fibrosis and chronic inflammatory cells. The findings in ultrasonography with periportal fibrosis and contracted gall bladder aid in early diagnosis and therapeutic strategy of Biliary Atresia specially in Indian subcontinent where all the people cannot effort the expensive investigations. However larger study is required to establish this relation provided USG features are carefully analysed by an experienced operator.

Keywords: *Periportal Fibrosis; Gall Bladder; USG; Biliary Atresia*

Introduction

Biliary Artesia is a congenital biliary disorder, commonly causing progressive and obstructive pathological changes in the intra-hepatic and extra-hepatic ducts secondary to inflammation and often causing progressive fibrosis and liver damage resulting in cirrhosis of liver and consequent liver failure [1].

It can be syndromic and non-syndromic. Syndromic is also known as embryonic type occur during developmental insult in differentiation of hepatic diverticulum from the foregut of the embryo. Non syndromic is also known as perinatal type occurs due to biliary obstruction of extra hepatic Biliary tree. Several Viruses such as Reo virus, Rotavirus, CMV, Epstein Barr virus are considered to be culprit [2].

It is thought to affect 1 in 10,000 - 15000 new born infants. Male are preponderance. Those infants may be normal at birth. However, symptoms develop after 2 weeks to 2 months of life. The common presentations are persistent progressive jaundice, acholic stool and hepatomegaly [3]. Favorable outcome is achieved if Biliary Artesia can be diagnosed as early as possible. Jaundice free survival may be achieved if hepatic portoenterostomy can be performed within 60 days of life. Thus, early differentiation of Biliary Artesia from neonatal hepatitis or other cause of infantile cholestatic jaundice is very important.

Till today no single pre-operative investigation has come with certainty of diagnosis. So, the evaluation of infants suspected of having Biliary Artesia has become challenging for physicians since many decades. Liver function assessment, HIDA scan, percutaneous needle liver biopsy and laparoscopic Color Doppler are thought to be useful, but their results are not individually diagnostic.

The evaluation of BA in the third world (developing countries) has become quite difficult due to limited resources i.e. socio-economic status or unavailability of imaging facilities. It is well accepted that non-invasive imaging tools like USG machine has the highest diagnostic accuracy since it avoids the unnecessary interventions [4].

The advantages of ultrasonography are its convenience, non- invasiveness and thinness of infant abdominal wall. The process can be repeated without adverse effect [1].

With advancement of high-resolution detecting head, the sonogram becomes increasingly clear. However, no general and unitive diagnostic criteria for Biliary Atresia are available at present. So, this study was therefore done to outline the ultrasonic findings in infants with cholestatic jaundice suspected to have Biliary Artesia demonstrating periportal fibrosis and contracted gall bladder. Attempts were made to correlate these findings with HIDA scan, liver biopsy, operative findings and biopsy from surgery.

Methods and Materials

Prospective, descriptive study of 25 consecutive persistently jaundiced infants was studied between Feb 2014 to March 2016 in Bangabhandhu sheikh Mujib medical university and some private clinics. All the infants were evaluated first by liver biochemistry assay especially bilirubin level-direct, SGPT, alkaline phosphates, gamma GGT and coagulation profile, followed by HIDA scan and USG Guided liver biopsy. The infants had undergone Kasai procedure. The findings were evaluated preoperatively and histopathological report.

USG

All infants underwent a detailed US examination by experienced operator. Gall bladder cannot be assessed at well fed state. So, the infants were not breast feed for 4 hours before examination. None of the infants were sedated. The sonographic exam was performed by Machine GE VOLUSON E8 Expert with abdominal probe C1-D Probe, 2 - 5 MHZ, 11 L-D Probe 4 - 10 MHZ. The gall bladder was interrogated to lay its shape and regularity of its wall. An irregular wall and a distortion of the gall bladder was considered abnormal (Figure 1). Gall bladder is poorly appreciated, lumen and walls are barely separable and the gall bladder remain same at both feeding and fasting state. The length of the gall bladder < 15 mm was considered contracted [5].

Extra hepatic bile ducts have most often become scarred cords of tissue which we have called as periportal fibrosis. These are the tissues that are presented anterior and posterior to porta hepatis.

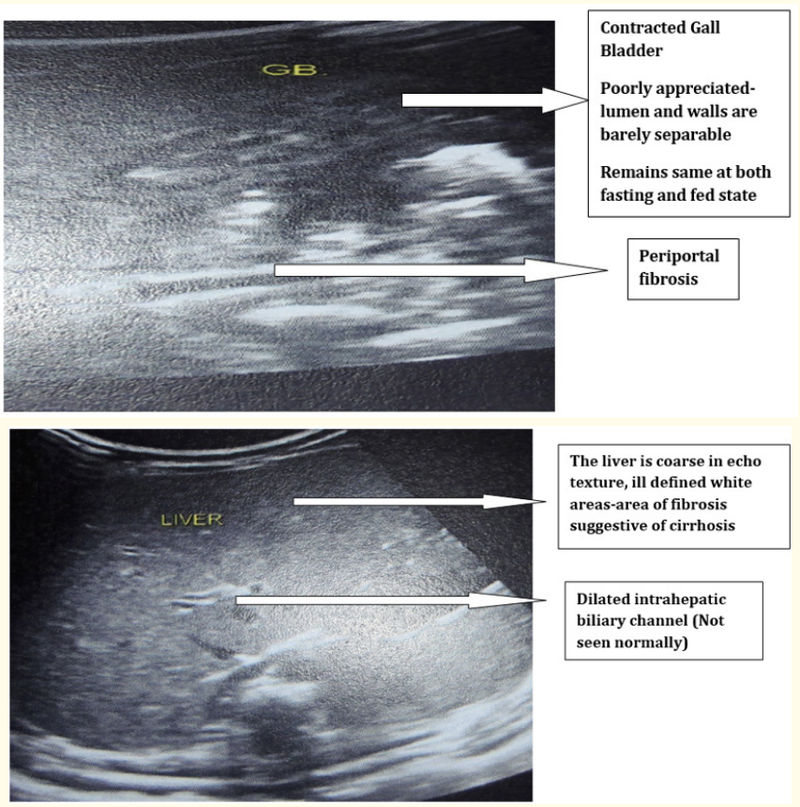

Figure 1: *Scan of Hepatobiliary System.*

Percutaneous liver Biopsy: It was done with paediatric size needle i.e. 1.6 mm internal diameter and 7 cm length. It revealed the features of cholestasis.

HIDA scan: HIDA scan was done; Common findings in favor of Biliary Artesia-Neither gall bladder nor common bile duct was visualized upto 30 minutes images. Tracer activity within the bowel loop was not upto 4 hours.

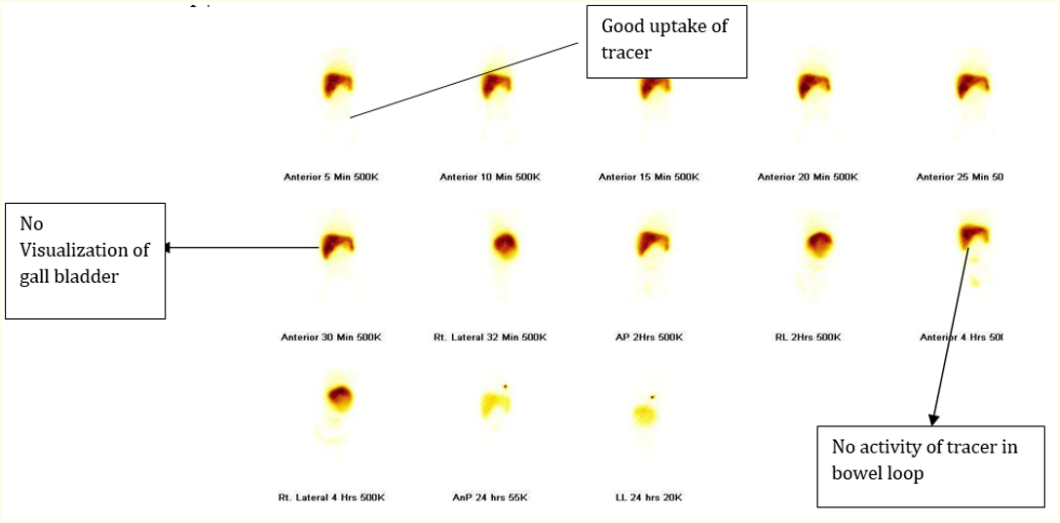

Figure 2: *HIDA Scan.*

Laparotomy: Laparotomy was done; Objective was to confirm the diagnosis by visualizing the presence of fibrotic cord in the region of porta hepatis and bile duct. The flow of bile was checked repeatedly by a normal saline soaked gauze piece for 5 minutes. The cirrhotic change of liver, remnants of extrabiliary tissue and gall bladder was observed. Kasai (Portoenterostomy) procedure was carried out in 18 patients.

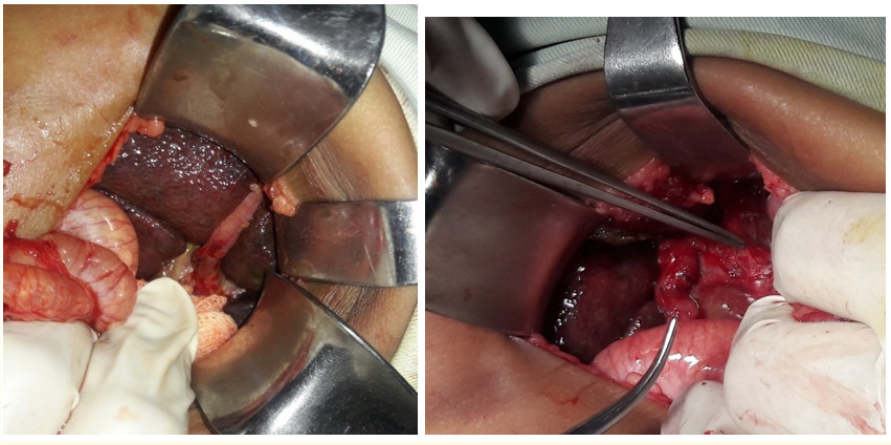

Figure 3: *Operative Picture a) Showing Cirrhotic liver and contracted gall bladder. b) Absence of extrahepatic tissue and presence of fibrotic tissue.*

Histopathology: The tissue from periportal area, gall bladder and liver were sent for histopathology. Tissue from periportal area shows fibrosis and chronic inflammatory cells. Gall Bladder revealed mild fibrosis ad moderate infiltration of chronic inflammatory cells.

Eligibility

Inclusion

- Patient with florid presentation of Biliary Artesia i.e. persistent jaundice, clay colored stool and abdominal mass.
- Patient who are willing to perform the investigations like USG of abdomen, HIDA scan, percutaneous liver biopsy
- Parent who have given consent for surgical intervention.

Exclusion

Patient not willing to participate in the study.

Operational definition

- **Periportal fibrosis:** Extra hepatic bile ducts which have become scarred cords of tissue.
- **Contracted Gall Bladder:** The maximal gall bladder length is measured and the gall bladder shape is evaluated using a high frequency transducer. When considering the gall bladder shape a discontinuous or absent mucosal lining and indistinct wall, and an irregular outer margin are regarded as abnormal i.e. lumen and walls are barely separable. The gall bladder was considered contracted when it was less than 1.5 cm long, was not detectable or detectable but had no lumen. The gall bladder remains same in both fasting and feeding state.

Results

The age range of twenty-five study subjects was within the mean of three Months (48 days - 5 months). More than half of them (68%) were less than 72 days old while remaining were more than 90 days old. Majority of them were (60%) female with all of them presenting with persistent progressive jaundice, acholic stool and abdominal distension. The onset of jaundice was less than 7 days in 52% of cases and 7 - 21 days in 25% of the cases. However, in 20% of cases mother couldn't recall the exact timing of jaundice. USG findings in 19 patients revealed periportal fibrosis ranging from 15 X 09 mm - 20 x 12 mm. In 6 patients no obvious fibrosis was seen. In 17 patients the gall bladder was present which appeared to be less than 1.5 mm contracted and no lumen were found. In 4 patients saccular well-defined gall bladder was present and in four patient gall bladder was absent.

Discussion

The cause of Biliary Artesia is not well known yet. Unless the hepatic portoenterostomy is performed within first 60 days survival without jaundice and its consequences like cirrhosis portal hypertension, is not possible. Therefore, it is very important to promptly diagnosis and do surgery to avoid progressive damage to liver. If there is progressive damage to liver early referral to liver transplant centre is advised which is not possible in developing countries like Bangladesh, Bhutan and Nepal. So surgical corrective procedure such as Kasai procedure remains the first line of treatment for most of the patient with Biliary Artesia [6].

Even with clinically available methods the differentiation of Biliary Artesia from neonatal hepatitis or other causes of infantile cholestasis remain challenging. First, there is always the diagnostic dilemma, second is the referral system. Third all the necessary investigations are not available at one centre. So, there is delay for therapeutic intervention. In our study also most of the patient presented more than 60 days. There is well known fact the outcome is not favorable after 60 days. The abdominal wall of children is thin, ultrasonic inspection is repeatable, convenient and non-invasive. Due to these advantages and improvement in ultrasonic enhancement, USG is more eminence than other diagnostic methods [7,8]. In 2003, Hee Jung., *et al.* [2] and Tan., *et al*. [9] claim that triangular cord sign and measuring the gall bladder length together are non-invasive, in-expensive and very useful marker for BA. Authors during their long clinical practice could hardly find any ultrasonologic findings as triangular cord in Bangladesh. In our study all the patient were evaluated by USG of whole abdomen after clinical and laboratory assessment suggestive of Biliary Artesia. The sonographic exam was performed by C1-D probe, 2 - 5

MHZ and 11 L-D probe 4 - 10 MHZ. Infants were not feed for 4 hours before their examination. None of the patients were sedated. Extra hepatic bile ducts which have become scarred cords of tissue were observed in 19 patients where we called periportal fibrosis. In1997 Park., *et al.* [10] gave an opinion that using ultrasonography is a new and non-invasive diagnostic tool for every year and definite diagnosis of BA.

In seventeen patient gall bladder was present which appeared to be irregularly contracted. The length was less than 1.5 cm long and lumen was seen. In 4 patients saccular well-defined gall bladder was present and. In four patients gall bladder was absent [11,12].

Contraction of gall bladder has been reported in patient BA. M Nemati., *et al.* [13] showed HIDA scan was completely sensitive in diagnosis of EHBA. Among 20 patients with HIDA scan and percutaneous liver biopsy, 18 patients revealed features of Biliary Artesia.

With all modalities mentioned above we diagnosed 18 subjects as Biliary Artesia and undergone Kasai Procedure. All patients were done under general anesthesia with endotracheal intubation with muscle relaxant with average duration of 3 hours. Sixteen patients revealed absence of biliary tract and contracted gall bladder. The two patients had common bile duct and normal looking gall bladder. However, the content of gall bladder was white bile. The tissue from the extrabiliary tract was sent for histopathology. All of them revealed periportal fibrosis and chronic inflammatory cells.

In extra-hepatic Biliary Artesia surgical corrective procedure like Kasai hepatic portoenterostomy remain the first choice for Paediatric surgeon [14]. However various treatment have been tried and could not have good results like Kasai procedure. The primary aim of the surgery is to resect the fibrous bile duct remnant followed by Roux-en-Y anastomosis of jejunum to porta hepatis [15-17].

Conclusion

We propose that periportal fibrosis and contracted gall bladder less 15 mm and doesn't change at feeding and fasting state is highly suggestive of Biliary Artesia. This is non-invasive, rapid, simple and reliable diagnostic means for evaluation of infants with persistent conjugated hyperbilirubinemia.

This study shows that simple ultrasonography findings help in diagnosis of Biliary Artesia avoiding liver biopsy and expensive investigation. This is even more helpful in Indian subcontinent where people cannot afford all the investigations. Further there is delay in making surgical intervention because all the investigations are done in different departments in different institutions. There is delay when the patient approaches for surgery.

Limitations

i) Level of awareness and education level of people regarding BA.

ii) Investigations like MRCP and intraoperative cholangiogram are not done because patient cannot afford them

iii) All Ultrasonography examinations were performed by single operator. Operator experience puts a great value in the detection and interpretation of US finding.

Recommendations

i) Mothers of babies should be alert of persistent hyperbilirubinemia and seek medical help without delay.

Biliary Artesia should be ruled out in all neonates with persistent jaundice through screening with abdominal scan.

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