ISSN - 2394-7721

American Journal of Oral Medicine and Radiology

Journal homepage: www.mcmed.us/journal/ajomr

BILIARY ATRESIA ASSOCIATED WITH CHOLEDOCHAL CYST

Kanishka S Patil[1*], Arjun Bahuddur[2], Subramanyam[3]

[1]Resident, Department of Radiodiagnosis, SSMC, Tumkur, Karnataka, India.
[2]Associate Professor, Department of Radiodiagnosis, Gulbarga Institute of Medical Sciences, Gulbarga, India.
[3]Pediatric Surgeon, SSMC, Tumkur, Karnataka, India.

Article Info
Received 23/11/2015
Revised 16/12/2015
Accepted 02/01/2016

Key words:-
Biliary atresia,
Choledochal cyst,
Biliary atresia.

ABSTRACT
Choledochal cyst and biliary atresia are rare but important causes of neonatal jaundice. Both present with jaundice and acholic stool in neonatal period. Treatment and prognosis of both entities are very different. We are presenting a case in which choledochal cyst co-existed with biliary atresia; Patient had portoenterostomy because of associated biliary atresia. We conclude that choledochal cyst and biliary atresia are two entities with similar antenatal and postnatal presentation; they should be differentiated as management and prognosis are different

INTRODUCTION

Choledochal cyst is a rare congenital anomaly with reported incidence of 1 in 100000-150000, while the incidence of biliary atresia is 1 in 10000-15000 live births. Presence of both choledochal cyst and biliary atresia in a patient is extremely rare. Though both present with jaundice and acholic stool in neonatal period, their treatment and prognosis is different. We are reporting a case of biliary atresia with cyst at portahepatis who was diagnosed as simple case of choledochal cyst, which ultimately required portoenterostomy.

Case report

1month 10 day old male baby presented to the hospital with h/o passing white coloured stools, dark yellow coloured urine and yellowish discolouration of the skin and of eye since 1 week. No similar complaints before (since birth), no similar complaints in mother /family.FTND Cried immediately after birth. No abnormality in antenatal period. Suck –normal, cry normal, sleep wake cycle normal. On examination: HR- 120B/MIN RR- 46CPM, SPO_2 – 96-100%, Cvs- S1, S2 HEARD NO MURMURS. RS –nvbd, no additional sounds, bilateral equal air entry. P/A- normally distended and soft, no organomegaly. Sclera and skin – yellowish discolouration.

LFT:

- Sr total bilirubin – 9.8gms/dl (6.1gms/dl– 1mth)
- Sr direct bilirubin -7.0gms/dl (2.7gms/dl—1mth)
- Sr indirect bilirubin -2.8gms/dl (3.4gms/dl -1mth)
- SGOT –(AST)- 153.6IU/lt
- SGPT-(alt)- 71.5IU/lt
- Sr .ALP 130IU/lt
- Sr total protein – 6.7g/dl
- Sr albumin – 3.8gm/dl
- Sr.globulin-2.9gm/dl

Other blood investigations

- Prothrombin time -18.2secs (increased n-12-16sec)
- Prothrombin INR – 1.4sec
- TLC – 15,800cells/cc
- DLC- N-24% L- 67% M-05% E-04%
- PLT- 526000cells/cc.
- Hb -8gm% Bld gp- B+ve
- Sr.sod- 127meq/l, sr pot-5.4meq/lt srcl-100meq/lt.

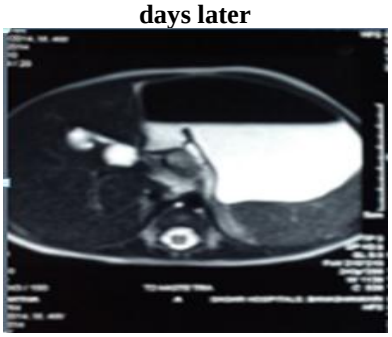
Corresponding Author

Kanishka S Patil
Email: - patilkanishka@gmail.com

Ultrasound abdomen and MRCP was done

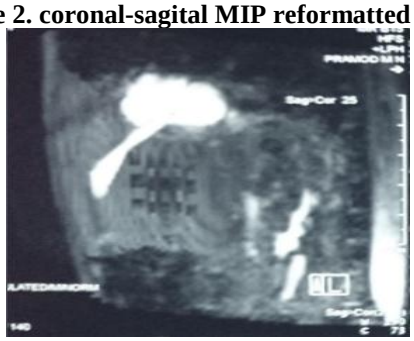
Ultrasound showed two cysts in continuity and also CBD not seen separately from cyst, cysts were intra and extrahepatic at the hila and GB not seen separately and a linear hypoechoic shadow adjacent to the Duodenum and to these cyst is seen s/o pseudogalbladder as no definite wall was seen.

Figure 1. Ultrasound abdomen and MRCP done two days later



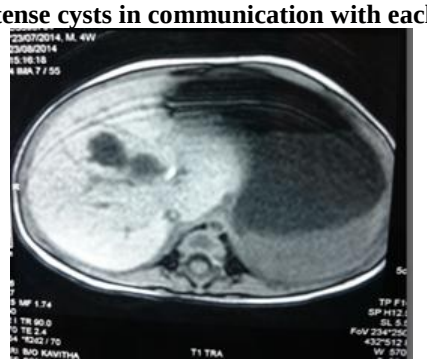
T2 weighted Haste axial image shows two hyper intense cysts with communication with each other and a linear hyper intense lesion probably a fluid collection, pseudogalbladder sign.

Figure 2. coronal-sagital MIP reformatted image



Two cysts in communication with each other and linear downward directed hyper intensity, ? doubtfull of GB., no Definite e/o GB or the CBD seen separately. no e/o IHBR .

Figure 3. T1 weighted axial image showing two hypointense cysts in communication with each other

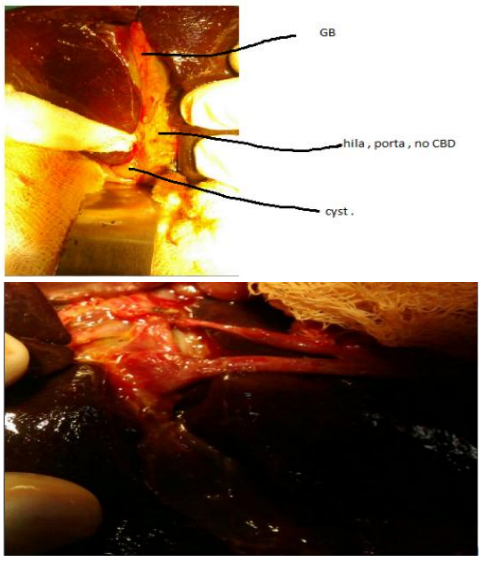


However in USG or in MRI/MRCP there was no e/o cirrhosis and we did not find a definite e/o GB or the CBD separate from the Cysts. A diagnosis of choledochal cyst with hypoplastic or aplastic GB was made with DD of a possibility of choledochalcyst with biliary atresia was made. Perop on exploration two cytic elevations were seen in the hila, the aspiration from cyst yielded a clear serous fluid with no bile within GB was collapsed and was seen as a linear tubular structure with no bile within, liver appeared dark brownish red, CBD was not present. On table diagnosis was made of choledochal cyst with biliary atresia and hypoplastic or atretic GB was made and portoenterostomy procedure was performed. Postoperatively stercobilinogen in stool became positive on 2nd day.

Histopathology

3 specimens were collected, liver biopsy, GB which measured 3cms and the portal plate. Liver biopsy showed marked cholestasis with portal inflammation. fibrosis score of 4/6 with extrahepatic hematopoesis. GB: chronic cholecystitis. Portal plate with inconspicuous ducts consistent with Biliary Atresia.

Figure 4. Per operative images.



DISCUSSION

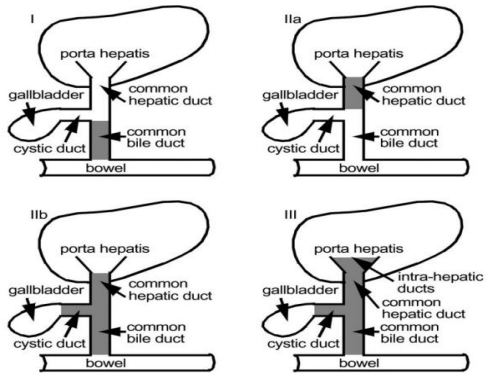
Choledochal cyst associated with biliary atresia is very rare disease; only 88 cases have been identified till now. Type I biliary atresia is the most common variety associated with choledochal cyst.

Definition: Biliary atresia is a condition in which the normal extrahepatic biliary system is disrupted. Progressive damage of extrahepatic and intrahepatic bile ducts secondary to inflammation may occur, leading to fibrosis, biliary cirrhosis, and eventual liver failure. Two types correctable nd non correctable are present and also can be classified into three types depending on the site where it's obliterated. Biliary atresia is characterized by luminal obstruction of the extrahepatic bile duct with a fibrous ductal remnant representing the obliterated duct in the porta hepat Neonates with choledochal cyst present with jaundice and acholic stools characteristic of biliary

obstruction. This clinical presentation is indistinguishable from neonatal hepatitis and biliary atresia and differentiation from these more common entities requires radiologic evaluation. The presenting symptoms and diagnostic findings of our patient were typical of those reported by other authors. Preoperative diagnosis can be made with help of ultrasonography, magnetic resonance cholangiopancreatography (MRCP) and radioisotope liver scan. Ultrasonographic criterion for the diagnosis of biliary atresia is nonvisualisation of the gall bladder or the appearance of a small gall bladder less than 1.5 cm in length. Although choledochal cyst with biliary atresia had been named as "correctable" type of biliary atresia in the past, it is known that there is no difference in treatment and prognosis between the correctable type and the "noncorrectable" type of biliary atresia. The treatment of choice is complete surgical excision of the extrahepatic bile duct and Roux-en- portoenterostomy.

In our case the intrahepatic biliary ducts were normally seen on MRI and since the presentation was after 1month and no any congenital anomalies like situs inversus, polysplenia, malrotation, intestinal atresia, and cardiac anomalies etc were detected, it can be said that it's an infantile type, correctable type and type 3 depending on site of atretion.

Figure 5. Biliary ducts



Types depending on site of attrition

1. Fetal-embryonic (or syndromic): is characterized by early cholestasis, appears in the first 2 weeks of life, accounts for 10-35% of all cases. In this form, the bile ducts are discontinuous at birth, and 10-20% of affected neonates have associated congenital defects: situs inversus, polysplenia, malrotation, intestinal atresia, and cardiac anomalies.
2. Perinatal (or acquired): accounts for the remaining 65-90% cases. This form is typically found in neonates and infants aged 2-8 weeks. Progressive inflammation and obliteration of the extrahepatic bile ducts occurs after birth. This form is not associated with congenital anomalies, and infants may have a short jaundice-free interval.

1st reported in Edinburgh Medical Journal in 1891. In 1916, the concepts of "correctable" and "noncorrectable" types of biliary atresia disease introduced. Successful surgical treatment for the correctable type was reported for the first time in 1928.

In the late 1950s, Morio Kasai reported the presence of patent microscopic biliary channels at the porta hepatis in young infants with biliary atresia-proposed kasai portoenterostomy

CONCLUSION

Cystic lesion of extra hepatic biliary apparatus should be carefully evaluated in infants because there is chances of associated biliary atresia asboth can present similarly. MRCP and radioisotope liver scan, if done simultaneously, can help to differentiate between infantile form of choledochal cyst and cystic biliary atresia but confusion can occur with the use of MRCP which tend to overestimate the patency of duct. The definitive diagnosis can only be made peroperatively. Jaundice beyond the age of 14 days need meticulous investigation and obstructive causes to be ruled out. Obstructive jaundice, timely intervention can save a great hazard of liver failure and need of liver transplantation.

ACKNOWLEDGEMENT: None

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

REFERENCES

1. Carol M. Rumack, Stephanie R. Wilson, J. William Charboneau, Deborah Levine. (2011). Diagnostic Ultrasound, 2 Vol Set 4th Edition.
2. Brian D. Coley. (2013). Caffey's Pediatric Diagnostic Imaging, 2-Volume Set, Saunders Publisher, 12 edition.
3. Seerat Aziz, Yvette Wild, Philip Rosenthal & Ruth B Goldstein. (2011). Pseudo Gallbladder sign in biliary atresia—an imaging pitfall. *Pediatr Radiol*, 41, 620–626.
4. Abu Obaidah, Nitin P. Dhende, S. B. Mane, Himanshu Acharya. (2009). Biliary atresia associated with choledochal cyst. *African Journal of Paediatric Surgery*, 6(1), 61-62.
5. Zhou L, Guan B, Li L, Xu Z, Dai C, Wang W, Xia H, Xie X. (2012). Objective differential characteristics of cystic biliary atresia and choledochal cysts in neonates and young infants: sonographic findings. *J Ultrasound Med*, 31, 833–841.

6. Nyree Griffin & Geoff Charles-Edwards & Lee Alexander. (2012). Grant Magnetic resonance cholangiopancreatography: the ABC of MRCP. *European Society of Radiology*, 3(1), 11–21.
7. AES Donia, SM Ibrahim, MSEMA Kader, AM Saleh, MS El-Hakim, MS El-Shorbagy, MM Mansour and MA Gibriel. (2010). Predictive Value of Assessment of Different Modalities in the Diagnosis of Infantile Cholestasis. *Journal of International Medical Research*, 38, 2100.