CASE REPORT

Hyaline cartilage at porta hepatis in extrahepatic biliary atresia: metaplasia or choristoma

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ABSTRACT

We report a case of a 5-month-old child who presented with the features of obstructive jaundice, anaemia and had no associated congenital anomalies. The child was suspected to have biliary atresia on hepatobiliary iminodiacetic acid (HIDA) scan and underwent Kasai portoenterostomy. Microscopic examination of the atretic gallbladder revealed islands of mature hyaline cartilage, lymphoid aggregates and fibrosis. There was an abrupt transition between the cartilage and the adjacent stroma. Only rare case reports have documented this finding with hypothesis ranging from metaplasia occurring secondary to the associated chronic inflammation or whether it is a choristoma. These cases need to be documented to elucidate the pathways of occurrence and may lead to have a better and detailed understanding of the disease process.

KEYWORDS

Gallbladder; Hyaline cartilage; Infant; Extrahepatic biliary atresia.

INTRODUCTION

Biliary atresia (BA) is an obliterative cholangiopathy of unknown aetiology which causes liver cirrhosis and is implicated as the most important cause of severe liver disease of infancy. These infants may present with syndromic and non-syndromic associations [1]. A number of variants have been defined clinically, which include the syndromic group [typically BA splenic malformation (BASM)], cystic BA and cytomegalovirus (CMV) IgM-positive associated BA and isolated BA [2]. The incidence varies greatly with more cases being reported from Taiwan [2]. BA has been the most common indication for liver transplantation during childhood for the past 20 years [3].

The occurrence of hyaline cartilage at the porta hepatis in BA has been described in rare case reports and is of undetermined significance. Several hypotheses exist ranging from cartilaginous metaplasia occurring secondarily to inflammation or whether it is a true choristoma or a developmental malformation. We present one such case of BA with islands of mature hyaline cartilage seen at the portal plate in a

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5-month-old male child. Identifying these rare findings in microscopy may help to discover newer pathways of occurrence and may help to understand disease pathogenesis better.

CASE REPORT

A 5-month-old male child presented with jaundice and pale-coloured stools since birth. His birth history was unremarkable, and he was not immunised. No history of previous hospitalisation was present. On examination, the child had severe jaundice, hepatomegaly and palpable left kidney. Respiratory and central nervous system examination was within normal limits. No congenital anomalies were noted.

Haemoglobin was 10.7 g/dl with normocytic normochromic anaemia. The total leucocyte count, differential leucocyte count and platelet counts were within normal range, and the international normalised ratio was 1.93. The direct Coombs test was negative. Urinary routine examination showed the presence of bilirubin and occasional epithelial cells. Liver function tests showed serum bilirubin of 15.71 mg/dl, direct bilirubin of 11.66 mg/dl, indirect bilirubin of 4.05 mg/dl, alanine aminotransferase of 234 U/l, aspartate aminotransferase of 376 U/l, alkaline phosphatase of 540 U/l and total proteins of 5.95 g/dl. He was nonreactive for HIV 1 and 2 and negative for hepatitis A virus, hepatitis B surface antigen and hepatitis C virus.

On ultrasonographic examination, the gallbladder was not visualised. Intrahepatic biliary radicals were not dilated. Hepatobiliary iminodiacetic acid (HIDA) scan revealed hepatomegaly with impaired hepatocyte function and no tracer exacerbation in the intestines, even in the delayed images until 24 hours. These findings were in favour of biliary atresia.

Kasai portoenterostomy was done, and the patient was managed intraoperatively with fresh frozen plasma transfusions, antibiotics, Vitamin K injections and supportive medications.

A wedge biopsy of the liver and cord-like atretic gallbladder was received. Microscopic examination of 4-µm-thick sections of haematoxylin and eosin (H and E) stained slides

of the gallbladder showed mainly denuded epithelium, few glands lined by tall columnar epithelium, congested ectatic blood vessels, moderate-to-dense chronic inflammatory infiltrate comprising of lymphocytes and histiocytes (Figure 1), fibrosis, haemorrhage and few islands of cellular mature hyaline cartilage (Figure 2). There was an abrupt transition between the cartilage and surrounding stroma (Figure 3). The cartilage did not appear to obstruct the lumen of the biliary tree. No respiratory epithelium or other elements were identified.

A wedge biopsy of the liver showed distorted lobular architecture with the formation of nodules separated by bridging fibrous septa with

Figure 1. Dense chronic inflammation surrounding island of mature hyaline cartilage with no respiratory elements (arrow), 10×, H and E.

Figure 2. Dense chronic inflammation surrounding island of mature hyaline cartilage (arrows) the porta hepatis in atretic gallbladder, 4×, H and E.

Figure 3. Hyaline cartilage with abrupt transition to stroma (arrow), 40×, H and E.

dilated and congested sinusoids, bile ductular proliferation, canalicular bile plugs and marked bile stasis. Mild-to-focal moderate inflammatory infiltrate comprising of lymphocytes, neutrophils, occasional plasma cells and eosinophils was noted around the portal tracts. Extramedullary haematopoiesis, giant cell transformation and rosette formation were not seen. The reticulin stain showed portal-to-portal tract fibrosis and regenerative nodule formation.

A diagnosis of hyaline cartilage at porta hepatis with dense inflammation and changes of liver cirrhosis in a case of extrahepatic biliary atresia (EHBA) was given.

The child is on follow-up and shows a persistence of deranged liver function tests which will necessitate liver transplantation in the future.

DISCUSSION

BA is an obliterative cholangiopathy of unknown aetiology, which causes liver cirrhosis, and is implicated as the most important cause of severe liver disease of infancy which may show varied presentations and associations [1]. A number of variants have been defined clinically, which include the syndromic group (typically BASM), cystic BA and CMV IgM-positive associated BA and isolated BA [2].

Associated syndromic and non-syndromic congenital malformations may be seen with BA. These congenital malformations are usually

seen in girls exposed to certain influences during intrauterine development [3].

Several studies have proposed that the disordered development in BA begins at the porta hepatis [4]. In normal embryology, hyaline cartilage is not seen at the porta hepatis. Hence, its occurrence at the porta hepatis is of undetermined significance and has been described in rare case reports (eight till now). We present one such case with islands of mature hyaline cartilage at portal plate in a 5-month-old male child with no other associated congenital anomalies.

The cases reported till now were more common in females (four cases) with age ranging from 2-day-old infant to 150-day-old child. Two male infants, 45-day-old and 60-day-old with similar features, have been reported. In two reports, the infants were 2-month old and 3-month old, and their gender was not documented. Of the eight reported cases, four had no associated congenital infections or anomalies.

The present case is a 5-month-old male child with no associated congenital anomalies. Microscopic examination revealed islands of mature well-organised hyaline cartilage with an abrupt transition to the surrounding stromal tissue and associated dense chronic inflammatory infiltrate. No respiratory epithelium or other element was seen.

Stahlschmidt et al. [1] suggested that the presence of cartilage at the porta hepatis occurred due to defective morphogenesis of foregut rather than secondary phenomenon. Hassab et al. [5] considered the hyaline cartilage to be arising from an aberrant morphogenesis unrelated to foregut cyst. Mirkin and Knisely [6] reported two such cases (2-day-old female and 42-day-old male infant) without associated congenital anomalies and proposed that the hyaline cartilage was more likely to represent a defective morphogenesis. Kashiwagi et al. [7] considered that both BA and cartilage formation were most probably due to CMV infection early in foetal life as occurred in their case of a 60-day-old female child.

Altamirano and Drut [8] proposed that cartilage at the porta hepatis was due to alteration resulting from metaplasia of local connective tissue rather

than true heterotopia as seen in their case of a 3-month-old infant (gender not specified). Vij et al. [9] described a similar case in a 2-month-old infant (gender not specified) and agreed that the hyaline cartilage at porta hepatis was likely to represent defective morphogenesis. Gupta et al. [10] reported hyaline cartilage and squamous epithelium within biliary ductules at the porta hepatis in a 65-day-old male child with no associated congenital anomalies or evidence of viral infection and hypothesised that these unusual findings could have occurred due to metaplastic response.

CONCLUSION

EHBA is an obliterative cholangiopathy of unknown aetiopathogenesis which leads to severe liver disease and cirrhosis in infants. Islands of hyaline cartilage at the porta hepatis in BA are rarely seen. These may occur either as a metaplastic response to dense chronic inflammation, as seen in the present case, or they may be due to true heterotopia or defective morphogenesis.

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CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest.

FUNDING

None.

ETHICS

Signed informed consent for participation and publication of medical details was obtained from the parents of this child. Confidentiality of patient's data was ensured at all stages. The

authors declare that ethics committee approval was not required for this case report.

REFERENCES

1. Stahlschmidt J, Stringer MD, Wyatt J, Davison S, Rajwal S, McClean P. Histologic oddities at the porta hepatis in biliary atresia. *J Pediatr Surg.* 2008;43:1328–1332. <https://doi.org/10.1016/j.jpedsurg.2007.09.017>
2. Davenport M. Biliary atresia: clinical aspects. *Semin Pediatr Surg.* 2012;21(3):175–184. <https://doi.org/10.1053/j.sempedsurg.2012.05.010>
3. Petersen C, Davenport M. Aetiology of biliary atresia: what is actually known? *Orphanet J Rare Dis.* 2013;8:128. <https://doi.org/10.1186/1750-1172-8-128>
4. Tan CE, Davenport M, Driver M, Howard ER. Does the morphology of the extrahepatic biliary remnants in biliary atresia influence survival? A review of 205 cases. *J Pediatr Surg.* 1994;29:1459–1464. [https://doi.org/10.1016/0022-3468\(94\)90144-9](https://doi.org/10.1016/0022-3468(94)90144-9)
5. Hassab MH, Baez-Giangreco A, Afzal M, Shadi SM, Al Olayet YF. Extrahepatic biliary atresia with hyaline cartilage and multiple congenital anomalies: a case report. *Saudi J Gastroenterol.* 1996;2:156–159.
6. Mirkin LD, Knisely AS. Hyaline cartilage at porta hepatis in extrahepatic biliary atresia. *Pediatr Pathol Lab Med.* 1997;17:587–891. <https://doi.org/10.1080/15513819709168735>
7. Kashiwagi Y, Kawashima H, Ioi H, Amaha M, Nishimata S, Takekuma K, et al. Biliary atresia with cartilage formation. *Pediatr Int.* 2007;49:683–685. <https://doi.org/10.1111/j.1442-200X.2007.02434.x>
8. Altamirano E, Drut R. Extrahepatic biliary atresia with cartilaginous metaplasia in the gallbladder remnants. *J Clin Pathol.* 2008;61:965–966. <https://doi.org/10.1136/jcp.2006.045831>
9. Vij M, Bachina P, Safwan M. Portal plate with hyaline cartilage in biliary atresia. *Indian J Pediatr.* 2015;83:185–186. <https://doi.org/10.1007/s12098-015-1805-4>
10. Gupta RK, Batra VV, Sengar M, Sakhuja P, Rooge SN. Biliary atresia with hyaline cartilage at the porta hepatis: a novel finding of undetermined significance: a case report. *Ann Pediatr Surg.* 2016;12:122–125. <https://doi.org/10.1097/01.XPS.0000481345.13724.a4>