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Editorial

Diminutive and Ancillary-Extrahepatic Biliary Atresia

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Biliary atresia or obliterative cholangiopathy emerges as a multifactorial disease associated with significant fibrosis and hepatocellular destruction while implicating extrahepatic and intrahepatic bile ducts. The condition configures as a common contributor to pathological infantile jaundice representing within early neonatal period.

Extrahepatic biliary atresia is frequently encountered within Asians and preponderantly arises within term infants of normal birth weight.

Extrahepatic biliary atresia involves extrahepatic and intrahepatic bile ducts and is categorized as

- Type I demonstrating atresia of common bile duct, patent right and left hepatic ducts with common hepatic duct and bile impregnated gallbladder. Nearly 5% instances represent with type I lesions.
- Type II delineating atresia within common hepatic duct and gallbladder devoid of bile. Roughly 2% subjects express type II lesions.
- Type III exemplifying atresia of porta hepatis or proximal segment of extrahepatic biliary tract. Adjoining gallbladder appears atretic. An estimated >90% subjects enunciate type III lesions [1,2].

Genetically susceptible subjects with extrahepatic biliary atresia demonstrate destruction of extrahepatic biliary system along with a distinct acute and chronic inflammatory cell exudate [1,2].

Genetic mutations with consequent laterality defects within CFIC and ZIC3 gene may emerge in ~10% instances of biliary atresia. Besides, JAG1 gene may contribute towards pathogenesis of biliary atresia [2,3].

Of obscure aetiology, biliary atresia is posited to occur due to genetic anomalies within morphogenesis and embryonal development. Diverse visceral anomalies may concur in ~20% instances. Commonly, polysplenia or asplenia configuring as biliary atresia or splenic malformation syndrome may be encountered. Additionally, congenital malformations as absent inferior vena cava, cardiac anomalies, situs inversus, intestinal malrotation, duodenal atresia or anomaly of portal vein may occur [2,3].

Cystic biliary atresia occurring in up to 10% subjects may be discerned upon antenatal ultrasonography within ~50% neonates [2,3].

Besides, defective prenatal hepatic circulation, viral infections with cytomegalovirus (CMV), Rubella or Rotavirus, intrauterine or perinatal exposure to toxin and immunological dysfunction may be enunciated.

Clinically, cholestasis with conjugated hyperbilirubinemia is encountered within infants of normal gestation. Biliary atresia represents with symptoms as persistent neonatal jaundice, dark coloured urine, pale stools and failure to thrive [2,3].

The condition is associated specific clinical phenotypes as

- Foetal, embryonal or prenatal forms encountered in up to 35% instances. Cogent clinical symptoms commence within early neonatal period. The condition may concur with extrahepatic developmental aberrations [3,4].
- Perinatal or acquired extrahepatic biliary atresia configures up to 80% lesions wherein hyperbilirubinemia ensues within weeks following birth. Associated congenital anomalies appear absent [3,4].

Grossly, explanted hepatic parenchyma appears dark green and nodular.

Upon microscopy, morphological features appear to concur with biological course of disease.

Preliminary lesions delineate nonspecific morphological features as variable cholestasis, foci of extramedullary haematopoiesis and minimal bile ductular proliferation [3,4].

Adjacent portal tracts display portal oedema, bile ductular proliferation, bile plugs, ductular cholestasis, inflammation of portal tracts with acute and chronic inflammatory cell infiltrate, prominent hepatic arteriole and portal or periportal fibrosis. Lesions of extended duration appear to progress into bridging fibrosis and cirrhosis [3,4].

Hepatic lobules expound cholestasis, minimal inflammatory exudate and presence or absence of hepatocyte giant cell transformation or extramedullary haematopoiesis [5,6].

Obstructive pattern of hepatocellular injury expounds significant bile ductular reaction, portal oedema, portal inflammation with admixture of acute and chronic inflammatory cells, bile plugs confined to ductules, portal fibrosis and lobular cholestasis.

Hilar plate exemplifies fibrosis, miniature or obstructed bile ducts and variable periductal inflammation [5,6].

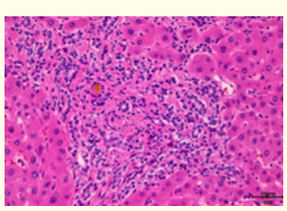

Figure 1: Extrahepatic biliary atresia depicting bile ductular proliferation, acute and chronic inflammation of portal tract, cholestasis and periportal fibrosis [11].

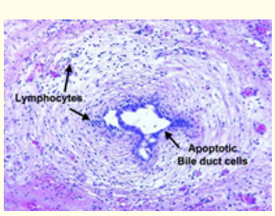

Figure 2: Extrahepatic biliary atresia displaying ductular proliferation, acute and chronic inflammation of portal tract, cholestasis and periportal fibrosis [12].

Table 1: Genes associated with biliary atresia [4].

Genes	Function
PKD1L1, CFC1, ZIC3, ZEB2, FOXA2, HNF1B, KIF3B, TTC17	Regulation of laterality, ciliogenesis, and development
ARF6, EFEMP1	Cytoskeleton and extracellular matrix modelling
ADD3	Cell contact and membrane structure
GPC1, JAG1	Hedgehog and Wnt signalling pathways
STIP1, REV1	Stress response and DNA repair

Tumour, Node, Metastasis (TNM) staging of carcinoma gallbladder as per American Joint Committee on Cancer (AJCC) 8th edition

- Stage 0: Tis, N0, M0
- Stage I: T1, N0, M0
- Stage II
  - Stage IIa: T2a, N0, M0
  - Stage IIb: T2b, N0, M0
- Stage III
  - Stage IIIa: T3, N0, M0
  - Stage IIIb: T1-T3, N1, M0
- Stage IV
  - Stage IVa: T4, N0-N1, M0
  - Stage IVb: any T, N2, M0 OR any T, any N, M1 [3,4]

Bile ductular proliferation appears immune reactive to cytokeratin CK7, CK19 or CD56/NCAM [7,8].

Extrahepatic biliary atresia requires segregation from lesions as choledochal cyst, alpha 1 antitrypsin deficiency, total parenteral nutrition associated cholestasis, neonatal hepatitis, Alagille syndrome, viral infection with cytomegalovirus (CMV) or progressive familial intrahepatic cholestasis type 3 (PFIC3) [7,8].

The condition may be appropriately discerned by ultrasonography, hepatobiliary iminodiacetic acid (HIDA) scan or percutaneous liver biopsy. Surgical tissue samples depict histological evidence of biliary tract obstruction [7,8].

Intraoperative cholangiography may be adopted as a confirmatory procedure prior to surgical intervention [7,8].

Manoeuvers as endoscopic retrograde cholangiopancreatography appear minimally beneficial for diagnostic determination [9,10].

Extrahepatic biliary atresia represents with conjugated hyperbilirubinemia and elevated serum alkaline phosphatase and Y glutamyl transferase (GGT) levels.

Upon radiography or ultrasonography, atresia or absence of gallbladder is observed. Hepatobiliary iminodiacetic acid (HIDA) scan displays significant hepatic intake with decimated to absent excretion of radioactive tracer into the intestine. However, aforesaid feature is minimally specific as absent radiotracer excretion may occur within diverse paediatric hepatic diseases with cholestasis [9,10].

Biliary atresia may be suitably alleviated with procedures as Kasai portoenterostomy. Procedures as Kasai portoenterostomy demonstrate >10 years overall survival in ~35% of subjects with native liver.

Repetitive episodes of ascending cholangitis and sepsis may occur in subjects unresponsive to Kasai portoenterostomy [9,10].

Surgical manoeuvers as liver transplantation may be beneficially adopted with curative intent.

Extrahepatic biliary atresia may progress into cirrhosis with portal hypertension wherein orthotopic liver transplantation may be necessitated. Following liver transplantation, overall survival emerges at~85% within 10 years [9,10].