Biliary atresia splenic malformation syndrome presenting with hepatic abscesses

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Biliary atresia, a destructive inflammatory cholangiopathy, leads to liver cirrhosis and subsequent death by the age of 2 years if left untreated. Biliary atresia splenic malformation (BASM) syndrome makes up 10% of all cases of biliary atresia. Kasai hepatopancreatoenterostomy (KPE) may establish continuity of bile flow and slow down progression to cirrhosis if the procedure is performed early in infancy. We describe an 8.5-year-old boy with known BASM syndrome (polysplenia, intestinal malrotation, interrupted inferior vena cava, shortened pancreas, centralised liver and left atrial isomerism) who underwent a successful KPE at the age of 3 months. He presented with features suggestive of a late onset ascending cholangitis (AC) complicated by cholangitic liver abscesses. Resolution of the abscesses with prolonged antibiotic therapy avoided the need for percutaneous drainage. Once the abscesses resolved, the child underwent a successful cadaveric liver transplantation.


Biliary atresia (BA) is a destructive inflammatory cholangiopathy. It is a heterogeneous disease composed of 3 subgroups: group 1: isolated (perinatal) BA without associated major malformations which is the most common form of the disease (84%); group 2: BA without laterality defects but with at least one major malformation (6%); and group 3: BA occurring in association with one or more laterality malformations (10%) and includes biliary atresia splenic malformation (BASM) syndrome. Groups 2 and 3 have associated anomalies in the cardiovascular and gastrointestinal systems, while genitourinary defects predominate in group 2.[1][2] Kasai hepatopancreatoenterostomy (KPE) is a palliative surgical procedure which aims to restore bile flow. Factors determining success of KPE include age, biliary remnant anatomy, extent of liver fibrosis at surgery, number of episodes of ascending cholangitis (AC), subgroup of biliary atresia and surgical expertise.[3][4] Poor prognosis in BASM syndrome is primarily related to the severity of cardiovascular lesions.[5]

Diagnosis of AC, the most common post KPE complication, requires a high index of suspicion and is considered in the presence of >38.0°C fever with no other focus, increased clinical jaundice and bilirubin levels with acholic stools. Diagnostic confirmation may be obtained by blood cultures and liver specimen culture/histology.[2][5] AC is seen in up to 90% of patients within 1 year of KPE, with episodes beyond 2 years considered uncommon and rarely reported in long-term survivors.[1][4][6] Aetiological agents are intestinal flora pathogens. Empirical treatment with cephalexin is recommended as causative organisms are identifiable in 30% of cases.[2] Pyogenic liver abscesses of biliary origin are sequelae of acute/repeated/intractable episodes of cholangitis post KPE and are rare.[2][6]

The present report aims to educate clinicians on the heterogeneity of BA and alert them to complications that may occur after a successful KPE. Ethics permission was obtained from the University of the Witwatersrand’s Human Research Committee (ref. no. M200383). Permissions were also obtained from the head of the Department of Paediatrics and the CEO of Chris Hani Baragwanath Academic Hospital.

Case

The patient was an 8.5-year-old male with BASM syndrome who underwent a KPE and Ladd’s procedure for BA and intestinal malrotation at the age of 98 days. The KPE was successful and the child remained anicteric with pigmented stools until the age of 6 years. Progression to cirrhosis and portal hypertension was noted over the following 2 years. At the age of 8.5 years he was admitted with fever. On examination, he was drowsy, pyrexial (38.5°C), jaundiced, clubbed, and pale, and his oxygen saturation was normal. Abdominal examination revealed a right hypochondrial surgical scar, ascites, a hard nodular non-tender 6 cm hepatomegaly, a palpable (unusual in BASM) 6cm firm splenomegaly and pale stool on rectal examination. Neurological examination revealed a child with confusion and incoherent speech. Laboratory investigations were documented as follows: haemoglobin 13 g/dL; platelet count 78 ×10^10/L; white cell count 28.42 ×10^9/L (80% neutrophilia); and C-reactive protein (CRP) 131 mg/L. His liver function tests revealed the following: total bilirubin 201 µmol/L (previously 82 µmol/L); conjugated bilirubin 183 µmol/L; total protein 51 g/L; albumin 18 g/L; alanine transaminase 539 U/L; aspartate transaminase 933 U/L; alkaline phosphatase 612 U/L; γ-glutamyl transferase 69 U/L; international normalised ration (INR) 1.97; and normal a-fetoprotein levels. Ammonia was persistently high at 200 µmol/L. His blood, urine, ascitic and stool cultures remained negative. Ascending cholangitis was suspected and managed with intravenous cefotaxime. Chronic encephalopathy, worsened by cholangitis, was managed with anti-liver-failure therapy and ascites were managed with albumin infusions and diuretics. Fever persisted and tazobactam and amikacin were introduced for suspected nosocomial sepsis.

A hepatic ultrasound identified an abscess measuring 60 mm × 64 mm in the right lobe. Repeat ultrasound within a week revealed...
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multiple liver abscesses. (Fig 1A) Computed tomography (CT) of
the abdomen confirmed the presence of a larger hepatic abscess
associated with smaller cholangitic abscesses and documented features
in keeping with BASM syndrome (polysplenia, midline liver, azygous
continuation of the inferior vena cava, short pancreas and mainly
right-sided small bowel) (Fig 1B). Echocardiography revealed left atrial
isomerism and confirmed interruption of the inferior vena cava with
azygous continuation to the superior vena cava.

A pigtail insertion for drainage of the larger hepatic abscesses was
planned. In view of underlying encephalopathy, significant ascites
and clotting abnormalities, the patient was deemed too unstable for
percutaneous/surgical drainage and was managed medically. Due
to fever persistence, antibiotics were changed to meropenem and
metronidazole. Serial ultrasounds confirmed gradual resolution of the
hepatic abscesses within 3 weeks of diagnosis (Fig 1A). Although the
patient's fever resolved and his mental state improved, he remained
jaundiced. He was discharged 6 weeks after admission, on completion
of 33 days of meropenem and 14 days of metronidazole. He was
maintained on amoxicillin/clavulanic acid for AC prophylaxis. Repeat
ultrasound one month after discharge showed no evidence of hepatic
abscesses. During the following month, the child underwent a successful
cadaveric liver transplantation. Liver explant histology confirmed
cirrhosis and presence of occasional bile lakes with no abscesses.

Discussion

The aetiological heterogeneity of BA alerts medical professionals to
evaluate patients for associated abnormalities. In BASM syndrome
these abnormalities can include polysplenia (77%), asplenia (11%) situs
inversus abdominis (37%), intestinal malrotation/atriesias, left-
sided/central liver and annular pancreas or an absent pancreatic tail.
Vascular anomalies commonly involve the portal vein (61%) and inferior
vena cava (absence in 39% of cases). Cardiac anomalies can include
dextrocardia and tetralogy of Fallot.[1,2] Most children with BA ultimately
require liver transplantation, with KPE remaining the preferred initial
procedure of choice.[1,2] Proper evaluation for BASM syndrome allows
adequate pre- and postoperative surgical planning for KPE and liver
transplantation to ensure similar outcomes to isolated BA.[3] However,
the severity of the cardiac defects often determines KPE outcomes and
influences the decision regarding referral for liver transplantation.[2]

Prevention of AC with a long Roux-en-Y loop, routine use of
postoperative steroids, prolonged antibiotic prophylaxis, a high index
of suspicion for the diagnosis irrespective of the patient's age and
appropriate duration of antibiotic treatment of acute episodes may
improve native liver survival and delay the need for transplantation.
[2,5] Treatment of AC can be escalated from cefotaxime to piperacillin-
tazobactam and then meropenem if there is no clinical improvement.
Ceftriaxone is avoided, as it may lead to bile sludging. Antifungals may
be considered.[2,5] Complicated or intractable episodes of cholangitis
are considered transplant indicators, with the number of cholangitic
episodes being prognostic markers of liver transplantation, reflecting
fibrosis progression.[3,5] Prevention is especially relevant in low-income
countries where transplantation may not be feasible owing to limited
resources and lack of accessibility to liver transplant programmes.
Common practice includes cyclical use of prophylactic antibiotics

Fig. 1A. Abdominal ultrasound: Progression of the hepatic abscess (white arrow) (i) initial (60 mm x 64 mm), (ii) 3 weeks later (39 mm x 28 mm), (iii) 1.5 months
later (4.8 mm x 4.9 mm) and complete resolution (iv) 3 month later.
(amoxicillin/clavulanic acid, trimethoprim/sulfamethoxazole, ciprofloxacin) but concerns for drug resistance and contradictory preventative results require more prospective studies.\(^2,5\)

Awareness of AC complications encourages earlier radiological investigations to exclude cholangitic abscesses. Causative organisms include *Escherichia coli* (21 - 36%), *Enterobacteriaceae* and polymicrobial infections.\(^3\) Antibiotics (cefotaxime or piperacillin-tazobactam) are generally parenterally administered for 4 - 6 weeks, with antibiotic choice guided by antimicrobial sensitivity tests.\(^5\) When signs of infection persist, indications for percutaneous drainage or surgical intervention need to be assessed.\(^2\) Hepatic abscesses have been documented as post KPE complications in 3.6% \((n=1/28)\) of cases.\(^6\) On pus culture, two reported cases of cholangitic abscesses post KPE revealed a polymicrobial infection in one case and *Klebsiella* spp. in the other.\(^4\)

**Conclusion**

We were unable to compare our patient management and outcomes with published reports owing to the rarity of the condition and unconfirmed microbial cause. In the era of liver transplantation, hepatic abscesses remain rare in high-income countries. In low-income countries, individualised therapy of cholangitic abscesses can yield a good outcome, occasionally bridging time to liver transplantation.

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