

BILLIARY ATRESIA PRESENTING AS POST CIRCUMCISSION EXANGUINATION

ABSTRACT

A one month old male infant was circumcised by a non trained but long standing local circumcision practitioner. He bled profusely and died after 8 hours post circumcision. Autopsy findings were those of billary atresia.

Keywords :- circumcision, Billary atresia

CASE PRESENTATION

A 4 weeks old male delivered by a traditional birth attendant, was circumcised by an experienced auxiliary nurse (the only person performing such in a local village). Four hours later, the child represented with complaints of continued bleeding from the site of circumcision. She applied vitamin K injection and penicillin ointment topically and sent the baby home. Approximately 3-4 hours later the baby was brought back to her dead.

Important history gotten at the autopsy venue (first contact of the parents with a medical doctor since the child's delivery) were as follows: the child was delivered at 9 months (probably at term), the circumcision was delayed for 3 weeks because around 7 days of age, "the child was not strong enough" (in the words of the mother), was the 4th child in a family of 2 boys and 2 girls, the elder brother did not bleed much after circumcision and the stool was said to be pale coloured (non pigmented) in recent times. At autopsy, an apparently healthy looking male infant was seen. The head circumference was 34cm, mid arm circumference was 14cm and the length was 1.9ft. The baby was moderately jaundiced and

the glands penis and site of circumcision was clean (no haematoma). All the organs were present, well formed and in their normal anatomical positions. The organs were very pale and there was no excessive fluid collection in any of the serosal spaces. The lungs were hyperventilated (with prominent rib markings), while the liver (weighed 140g) had a smooth shiny greenish surface with a homogenous greenish cut surface (figure 1a-c). Microscopically, brain, heart, kidney, intestines and lungs showed features of hypoxic/ischaemic injury. Sections of the liver showed bile duct proliferation, mixed inflammatory cell infiltration of the portal tract, grade 2 bridging fibrosis and bile plugging (features of biliary atresia (BA). The autopsy diagnosis was concluded as massive haemorrhage (hypovolaemic shock) post circumcision due to BA and defective hepatocyte function.

LITERATURE REVIEW

Male Circumcision is one of the oldest and one of the most commonly performed surgical procedures in practice today. It is commonly practiced and will continue to occur for religious, cultural and medical reasons.¹⁻³ Complications of circumcision are broadly classified into early and late complications. Early complications include; bleeding (most common), pain, inadequate skin removal, and surgical site infection. Others include chordee, iatrogenic hypospadias, glanular necrosis, and glanular amputation. Post circumcision bleeding in patients with coagulation disorders can be significant and sometimes even fatal.^{2,4} Late complications of circumcision include; epidermal inclusion cysts, suture sinus tracts, chordee, inadequate skin removal resulting in redundant foreskin, penile adhesions, phimosis, buried penis, urethrocuteaneous fistulae, meatitis, and meatal stenosis.²⁻⁴ Bleeding remains the commonest complication encountered during and after circumcision.⁴ In most cases of bleeding, which is usually minor, all that is required to achieve haemostasis is gentle pressure on the area.² Excessive bleeding may be due to anomalous blood vessels or presence of

48 bleeding disorders.⁵ In situations where gentle pressure fails, circumferential bandage may be
49 used to aid haemostasis.² Pharmacological agents like 1:100, 000 adrenaline can also be used.
50 Diathermy or suturing may be used for obvious bleeding vessels.² There are very few
51 documented cases of exsanguination following circumcision. Death following circumcision is
52 a very rare occurrence, such that in a series of 500,000 consecutive circumcisions in New
53 York City, no single fatality was recorded.⁶

54 Biliary atresia (BA) is the single most frequent cause of death from liver disease in early
55 childhood.⁷ BA is the complete or partial obstruction of the lumen of the extrahepatic biliary
56 tree within the first 3 months of life, that is characterized by progressive inflammation and
57 fibrosis of intrahepatic or extrahepatic bile ducts.⁸ BA mainly involves extra hepatic ducts,
58 though intra hepatic ducts can also be involved and is usually seen in about 1 in 10,000
59 births.⁹ There are two major forms of biliary atresia (fetal and perinatal), based on the
60 presumed timing of luminal obliteration.⁷

61 The etiology of BA is not clear, though toxic, infectious and autoimmune factors have been
62 implicated.¹ Clinically, BA is mainly seen in females. The infants are usually delivered at full
63 term and jaundice develops during 2nd to 3rd week, while the stool colour is usually acholic.¹
64 The index case though a male, was delivered at term, was jaundiced (onset not known) and
65 passed pale coloured stool (acholic). The fetal form is a rare form and usually associated with
66 other anomalies like interrupted inferior vena cava, polysplenia, congenital heart disease and
67 malrotation of abdominal visera. It is seen in about 20% of cases. The suspected cause is
68 aberrant intrauterine development of the extrahepatic biliary tree. The perinatal form of BA is
69 commoner, in which a presumed normally developed biliary tree is destroyed following
70 birth.¹⁰ The main cause of BA is unknown, though autoimmunity, viral infections (reovirus,
71 rotavirus, and cytomegalovirus) and toxic factors are known to play roles.^{9,10} The index case

72 is probably a perinatal form BA, because the features listed above for fetal form were not
73 seen.

74 Grossly, the liver surface is smooth and homogenously green. The case in study was same as
75 shown in figure 1a. Micro morphologic features of BA which are evident in about two thirds
76 of cases are marked bile ductular proliferation, portal tract edema and fibrosis, inflammation
77 of the hepatic or common bile ducts, parenchymal cholestasis and progressive destruction of
78 the intrahepatic biliary tree. If BA is unrecognized or uncorrected, cirrhosis develops within 3
79 to 6 months of birth.^{7,11} These features were characteristically seen in the index case with
80 some bridging fibrosis. (Figures 2a &b, 3)

81 Clinically these infants appear almost normal at birth with normal birth weight and postnatal
82 weight gain. Initially they pass normal coloured stools in the early days of to life before it
83 progresses to acholic stools as the disease evolves. There is a slight female preponderance.^{7,9}
84 The index case though a male, was claimed to be normal at birth by parents, though birth
85 weight was not measured and passed normal coloured stool (yellowish brown) initially .
86 Investigations that aid diagnosis include: elevated hyperbilirubinemia, moderately elevated
87 aminotransferase and alkaline phosphatase levels.^{7,9} A combination of liver biopsy and
88 radiologic excretion scintigraphy are the best investigations to confirm a diagnosis of BA. BA
89 is excluded if small bowel is visualized after using isotope ^{99m}Tc-DISIDA (di-
90 isopropyliminodi-acetic acid). Ultra sound scan technique is also a good guide. In BA it
91 usually shows hypoplastic gallbladder, a cystic cavity at the porta and feature of established
92 cirrhosis early in life.¹² No investigation was done in the index case.

93 Treatment of choice is hepatoportoenterostomy (kasai proceure) ie connecting the bowel
94 lumen with the porta hepatitis, or else patients die from liver cirrhosis before 2 years of age.⁷
95 The success of this procedure is dependent on the level and variability in the anatomy of BA.⁷

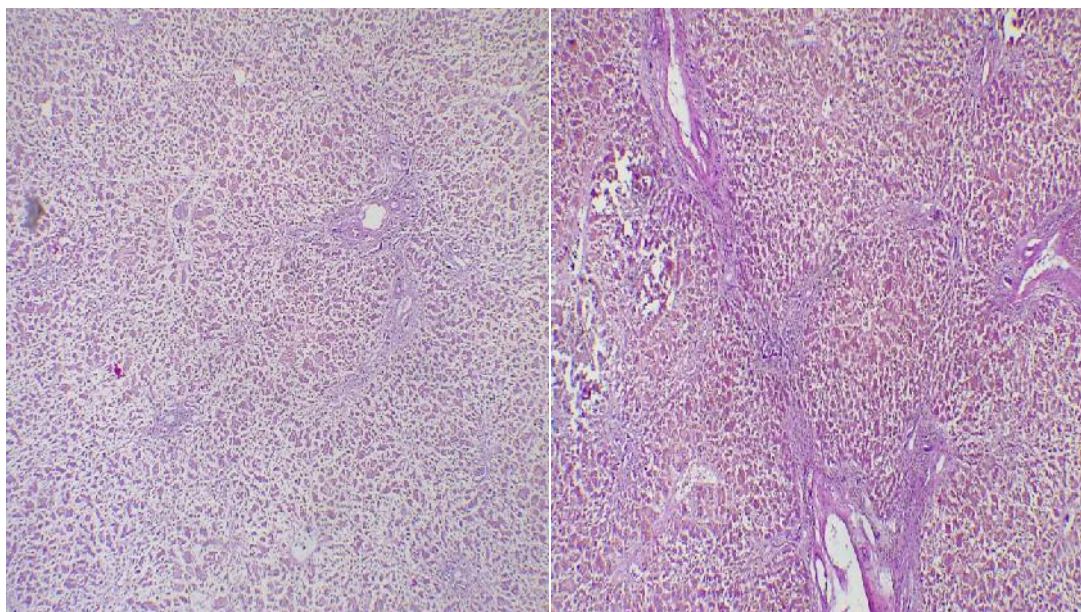
96 For those that may not benefit from Kasai procedure, the other alternative is liver
97 transplant.^{7,9}

98 It was not seen in literature were circumcision lead to death in a BA patient, because in such
99 circumstances, the patients must have been stabilized and proper precautions taken. The
100 index case had no antenatal care, was not delivered in a hospital and never presented to a
101 hospital or qualified health worker. In conclusion the post circumcision exsanguination seen
102 in this case was probably due to reduced / impaired hepatic functions with compromised
103 production of clotting factors. Traditional / untrained circumcision practitioners should be
104 informed of symptoms and signs (especially easily appreciated ones) that when seen, should
105 discourage them from circumcising a male child.

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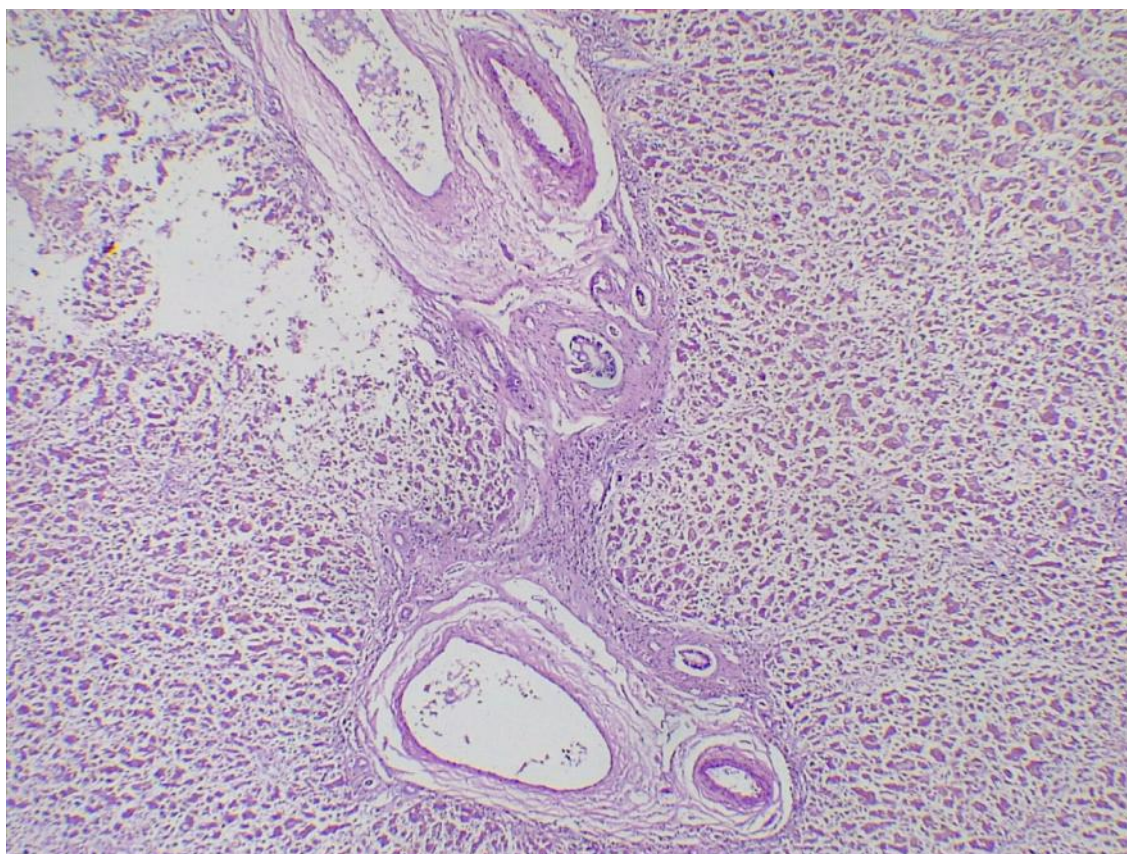
108 Figure 1: **A)** The homogenously green coloured liver with a smooth and shiny surface. **B)**
109 The inferior surface showing hypoplastic gallbladder. **C)** Hyper inflated lungs more marked
110 on the left lung.



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112 Figure 2: A and B shows early bridging necrosis between portal tracts and parenchymal
113 cholestasis

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116 Figure 3: Showing marked bile ductular proliferation, portal tract edema and fibrosis, and
117 parenchymal cholestasis

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