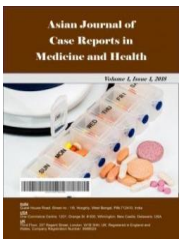

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Biliary Atresia Presenting as Post Circumcision Exsanguination

Nwafor Chukwuemeka Charles[1*] and Nwafor Nkem Nnenna[2]

[1]Department of Pathology, University of Uyo, Nigeria.
[2]Department of Radiology, Teaching Hospital, University of Uyo, Uyo, Akwa Ibom State, Nigeria.

Authors' contributions

This work was carried out in collaboration between both authors. Both authors read and approved the final manuscript.

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Case Study

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ABSTRACT

An autopsy diagnosis of biliary atresia was made in a month old male infant who died following prolonged bleeding that lasted for about 8 hours after circumcision by a non-trained local circumcision practitioner.

Keywords: Circumcision; biliary atresia.

ABBREVIATIONS

BA : Biliary atresia
KPE : Kasai prtoenterostomy
HIDA : Hepatobilliaryiminodiacetic acid

1. CASE PRESENTATION

A 4 weeks old male delivered by a traditional birth attendant, was circumcised by a non-trained local circumcision practitioner (the only person performing such in a local village). Four hours later, the child presented with complaints of continued bleeding from the site of circumcision. The nurse applied vitamin K and penicillin ointment topically and sent the baby home. Approximately 3-4 hours later the baby died.

At the autopsy venue, relevant information was collected (first contact of the parents with a

**Corresponding author: E-mail: firstcocsin@yahoo.com;*

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.

2. AUTOPSY FINDINGS

Gross anatomy findings: An apparently healthy looking male infant was seen. The head circumference was 34 cm, mid-arm circumference was 14 cm and the length was 1.9 ft. The baby was moderately jaundiced and the glands penis and site of circumcision were 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 140 g) had a smooth shiny greenish surface with a homogenous greenish cut surface (Fig. 1a-c). There was no intra-cranial bleeding or bleeding elsewhere.

Microscopic findings: The 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.

Informed written consent was obtained from the parents on with respect that the identity must remain anonymous.

3. 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 for religious, cultural and medical reasons [1-2]. 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, glandular necrosis, and glandular amputation. Post-circumcision bleeding in patients with coagulation disorders can be significant and sometimes even fatal [2,3]. Late complications of circumcision include; epidermal inclusion cysts, suture sinus tracts, chordee, inadequate skin removal resulting in the redundant foreskin, penile adhesions, phimosis, buried penis, urethrocutaneous fistulae, meatitis, and meatal stenosis [2,3]. Bleeding remains the commonest complication encountered during and after circumcision [3]. 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 bleeding disorders [4]. In situations where gentle pressure fails, the circumferential bandage may be used to aid haemostasis [2]. Pharmacological agents like 1:100, 000 adrenaline can also be used. Diathermy or suturing may be used for obvious bleeding vessels [2]. There are very few documented cases of exsanguination following circumcision. Death following circumcision is a very rare occurrence [5].

BA is the single most frequent cause of death from liver disease in early childhood [6]. BA is the complete or partial obstruction of the lumen of the extrahepatic biliary tree within the first 3 months of life, that is characterized by progressive inflammation and fibrosis of intrahepatic or extrahepatic bile ducts [7]. BA mainly involves extrahepatic ducts, though intrahepatic ducts can also be involved and are usually seen in about 1 in 10,000 births [8]. There are two major forms of biliary atresia (fetal and perinatal), based on the presumed timing of luminal obliteration [6].

The etiology of BA is not clear, though aflatoxins, toxic, infectious and autoimmune factors have been implicated [6,9]. Clinically, BA can be seen in any sex. The infants are usually delivered at full term and jaundice develops during 2nd to 3rd week, while the stool colour is usually acholic [1]. The index case was delivered at term, was jaundiced (onset not known) and passed pale coloured stool (acholic). The fetal form is a rare form and usually associated with other anomalies like interrupted inferior vena cava, polysplenia, congenital heart disease and malrotation of abdominal viscera. It is seen in about 20% of cases. The suspected cause is the aberrant intrauterine development of the extrahepatic biliary tree. The perinatal form of BA is commoner, in which a presumed normally developed biliary tree is destroyed following birth

[10]. The main cause of BA is unknown, though autoimmunity, viral infections (reovirus, rotavirus, and cytomegalovirus) and toxic factors are known to play roles [8,10]. The index case is probably a perinatal form BA, because the features listed above for fetal form were not seen.

Grossly, the liver surface is smooth and homogenously green. The case in the study was same as shown in Fig. 1a. Micro morphologic features of BA which are evident in about two-thirds of cases are marked bile ductular proliferation, portal tract oedema and fibrosis, inflammation of the hepatic or common bile ducts, parenchymal cholestasis, and progressive destruction of the intrahepatic biliary tree. If BA is unrecognized or uncorrected, cirrhosis develops within 3 to 6 months of birth [6,11]. These features were characteristically seen in the index case with some bridging fibrosis (Figs. 2a & b, 3).

Clinically these infants appear almost normal at birth with normal birth weight and postnatal weight gain. Initially, they pass normal coloured stools in the early days of to life before it progresses to acholic stools as the disease evolves [6,9]. The index case though a male, was claimed to be normal at birth by parents, though birth weight was not measured and passed normal coloured stool (yellowish brown) initially . Investigations that aid diagnosis include: elevated hyperbilirubinemia, moderately elevated aminotransferase and alkaline phosphatase levels [6,9]. Other investigations that aid diagnosis are ultrasonography, percutaneous liver biopsy and technetium 99m hepatobilliaryiminodiacetic acid (HIDA), though more definitive procedure for the diagnosis of BA currently is laparoscopic guided cholecysto cholangiography [12]. No investigation was done in the index case.

Treatment of choice is hepatoportoenterstomy (kasaiproceure) ie connecting the bowel lumen with the porta hepatitis, or else patients die from liver cirrhosis before 2 years of age [6]. The success of this procedure is dependent on the level and variability in the anatomy of BA [6]. For those that may not benefit from Kasai procedure, the other alternative is liver transplant [6,9].

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

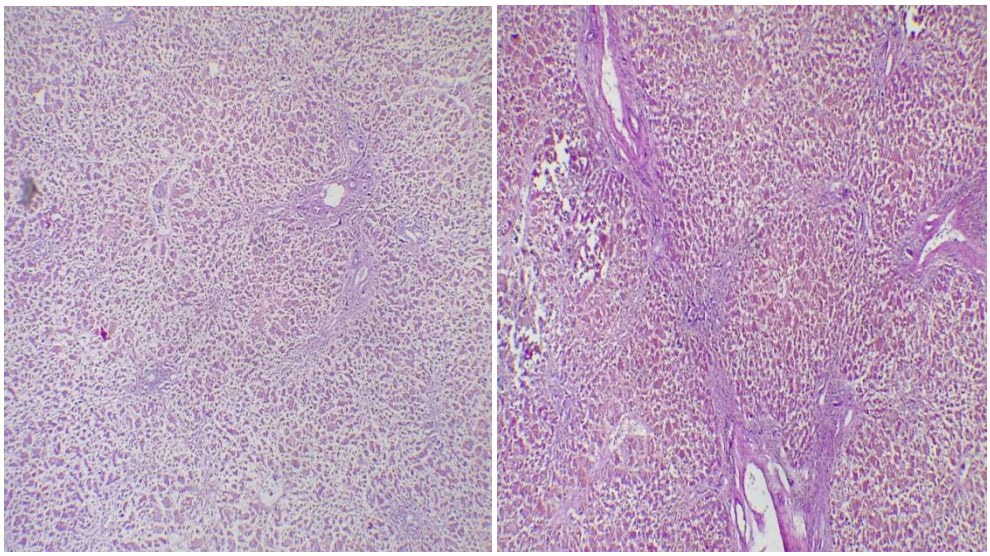

Fig. 2. A and B shows early bridging necrosis between potal tracts and parenchymal cholestasis

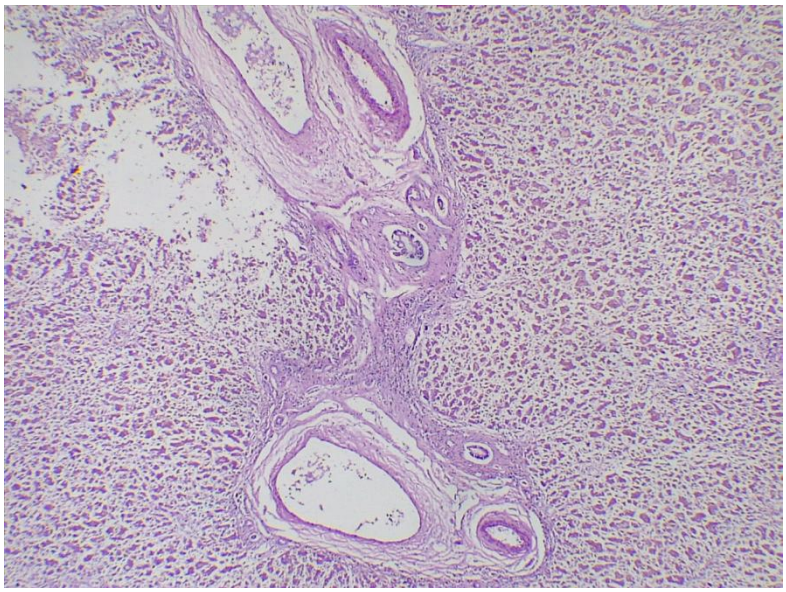

Fig. 3. Showing marked bile ductular proliferation, portal tract edema and fibrosis, and parenchymal cholestasis

It was not seen in literature were circumcision lead to death in a BA patient, because in such circumstances, the patients must have been stabilized and proper precautions taken. The index case had no antenatal care, was not delivered in a hospital and never presented to a hospital or qualified health worker.

4. CONCLUSION

In conclusion the post circumcision exsanguination seen in this case was probably due to reduced / impaired hepatic functions with compromised production of clotting factors. Though liver function test, clotting factor estimation and clotting time were not done. All health workers should be trained and legislations should pave the way. Government should promulgate a law that circumcision should be done only by medically qualified individuals with necessary clinical skills and training. If traditional / untrained circumcision practitioners must perform circumcision due to lack of personnel, they should be informed of symptoms and signs

(especially easily appreciated ones) that when seen, should discourage them from circumcising a male child.Example that no jaundiced child should be circumcised except in a medical facility. Bleeding tests are mandatory before circumcision in neonates, while prophylactic vitamin K injection might be an alternative in areas where testing is not feasible.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard or university standard, written approval of Ethics committee has been collected and preserved by the authors.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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