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Original article

BILIARY ATRESIA IS ASSOCIATED WITH POLYSPLENIA AND SITUS INVERSUS ON ULTRASOUND, A CASE REPORT STUDY.

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Abstract

Biliary atresia is a destructive, idiopathic, and inflammatory cholangiopathy that affects intra and extra-hepatic bile ducts leading to fibrosis and obliteration of the biliary tract and development of liver cirrhosis.

The polysplenia syndrome is the most common anomaly, being found among patients with atresia, and is characterized by polysplenia/asplenia associated with a midline liver, interruption of the inferior vena cava, preduodenal portal vein, situs inversus and/or intestinal malrotation.

Biliary atresia has an incidence of 1 in 10,000-15,000 live births and is more common in females than males. I report a case of a 5 months old baby boy who presented with yellow eyes since birth and abdominal distension for one week.

The diagnosis of Biliary atresia, polysplenia, and situs inversus is usually based on clinical findings. Ultrasonography and Intraoperative cholangiogram are useful tools. The most suitable recommendable treatment for this specific case is surgery.

This case study is to fill the knowledge gap among sonographers/radiographers regarding the existence of such cases to guide the management of those conditions, especially among infants.

Keywords: Biliary atresia, Polysplenia, situs inversus, Submitted: 2023-02-22 Accepted: 2023-03-28

1. Introduction

Biliary atresia is a destructive, idiopathic, and inflammatory cholangiopathy that affects intra and extra-hepatic bile ducts leading to fibrosis and obliteration of the biliary tract and development of liver cirrhosis (Mathur et al., 2014). It may be classified as embryonic or fetal and perinatal, based on the period in which it occurs. The embryonic form accounts for 20% of cases and is often associated with non-hepatic structural anomalies. The polysplenia syndrome is the most common anomaly, being found among patients with atresia, and is characterized by polysplenia/asplenia associated with a midline liver, interruption of the inferior vena cava, preduodenal portal vein, situs inversus and/or intestinal malrotation (Guttman et al., 2011; Zhan et al., 2017). Other congenital malformations can be observed, such as cardiac anomalies, annular pancreas, immotile cilia syndrome, duodenal atresia, esophageal atresia, polycystic kidney disease, cleft palate, and jejunal atresia (Rasool and Mirza, 2011). Biliary atresia has an incidence of 1 in 10,000-15,000 live births and is more common in females than males (Saggiomo et al., 2001; Nio et al., 2003)

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2. Case

Patient information; A 5 months old baby boy presented with yellow eyes since birth and abdominal distension for one week. He was born full-term, via normal vaginal delivery, and was the sixth in birth order. None of the siblings has ever presented with similar symptoms. The baby was born in Nyumazi Hospital in Adjumani District at full term and both mother and baby didn't experience any complications. The mother was discharged one day after delivery. The mother did not report any treatment given to the baby about the condition, though she reports the baby being given quinine.

2.1. Social history:

Born full term, normal vaginal delivery at Nyumazi Hospital, Adjumani District, Northern Uganda, the mother and baby were discharged one day after delivery as there was no indication of any complications. Born sixth in birth order, none of the siblings had presented with a similar condition.

2.2. Physical exam:

On general physical examination, revealed a stable baby with obvious jaundice and abdominal distension.

2.3. Diagnostic Assessment:

On the abdominal ultrasound scan, the liver, as well as the spleen, was seen on the right side. The heart apex was seen on the left side. The stomach and abdominal aorta were seen on the right side. The common bile duct and gall bladder were not visualized. The hepatic artery was hypertrophied with a diameter of 0.32cm. There were multiple small ovoid masses seen around the splenic hilum suggestive of polysplenia.

A 5 months old male referred to the ultrasound department to rule out biliary atresia had clinical features; yellow eyes since birth and abdominal distension for one week, the mother had administered quinine for unspecified duration and reason. The following findings on ultrasound were noted:

- The liver and spleen were on the right side of the abdomen,
- The heart apex was on the left side of the thoracic cavity,
- The stomach and abdominal aorta on the right side of the abdomen,
- The common bile duct and gall bladder were not visualized
- The hepatic artery was hypertrophied with a diameter of 0.32cm (normal range =<0.15cm).
- There were multiple small ovoid masses seen adjacent to the splenic hilum which were thought to be accessory spleens.
- The rest of the abdominal organs were normal

The ultrasound impression was biliary atresia (syndromic), polysplenia, and situs inversus.

2.4. Other sonographic findings

- The common bile duct and gall bladder were not visualized.
- The heart apex was on the left side of the thoracic cavity.

Biliary atresia is a congenital biliary disorder characterized by the absence or progressive obliteration of the biliary tract system. The extent of the involvement of the bile ducts varies however, the extrahepatic ducts are more affected than the intrahepatic ducts.

The etiology of biliary atresia is idiopathic, although some theories show that it can be due to viral infections, immune dysregulation in neonates affecting the hepatobiliary system, or pancreatic reflux. There is a progressive injury to the epithelial lining of the bile ducts and subsequent inflammatory response propagates it, therefore the bile duct lumen is filled with the inflammatory plug leading to fibrosis and obliteration of the bile ducts. Obliteration of the ducts can

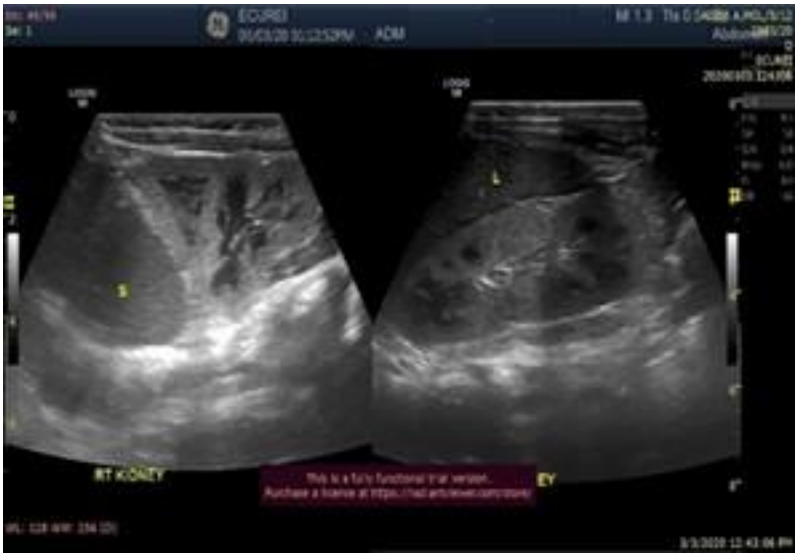

Figure 1: The liver and spleen were on the right side ofthe abdomen.

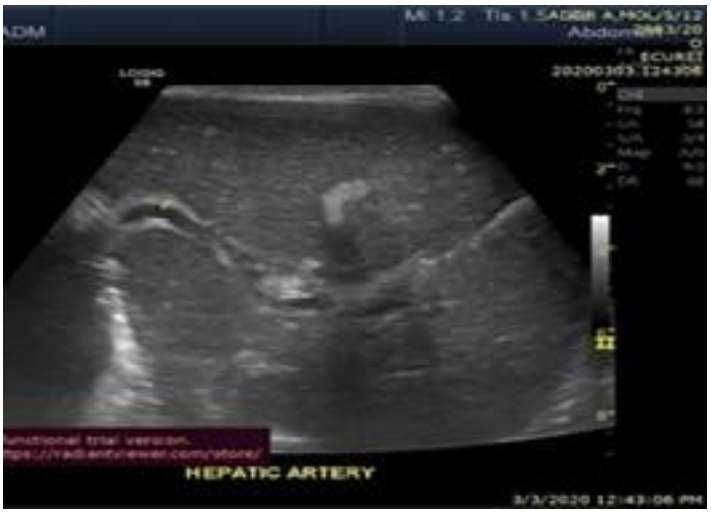

Figure 2: The hepatic artery was hypertrophied (0.3 cmnormal range =<0.15cm.

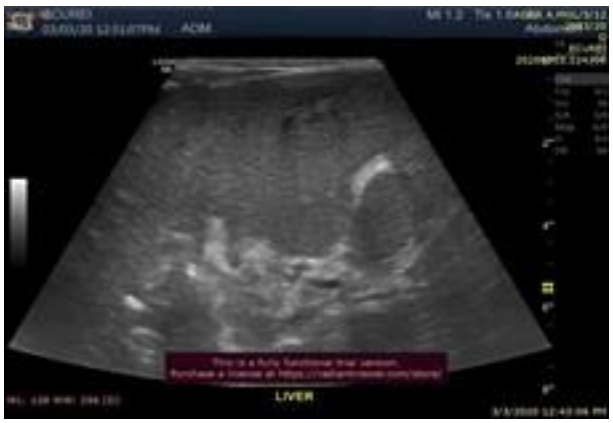

Figure 3: "Triangular Cord Sign"

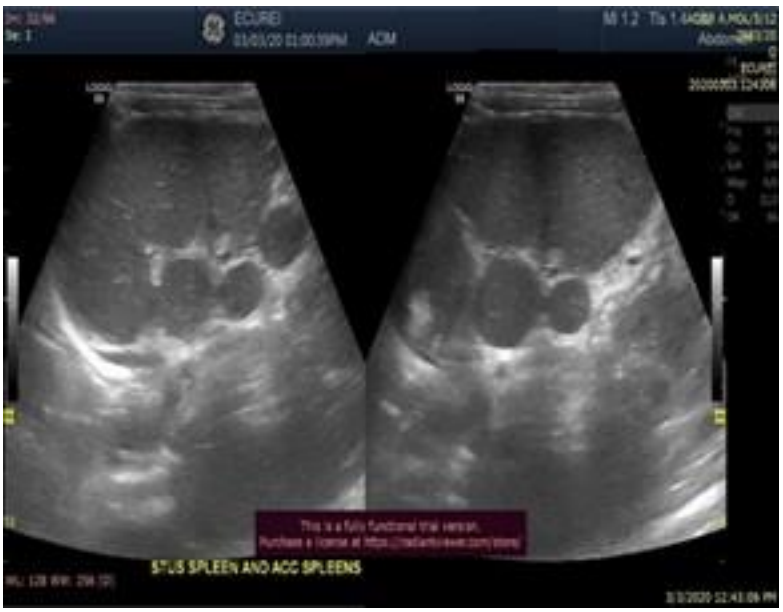

Figure 4: Multiple accessory spleens.

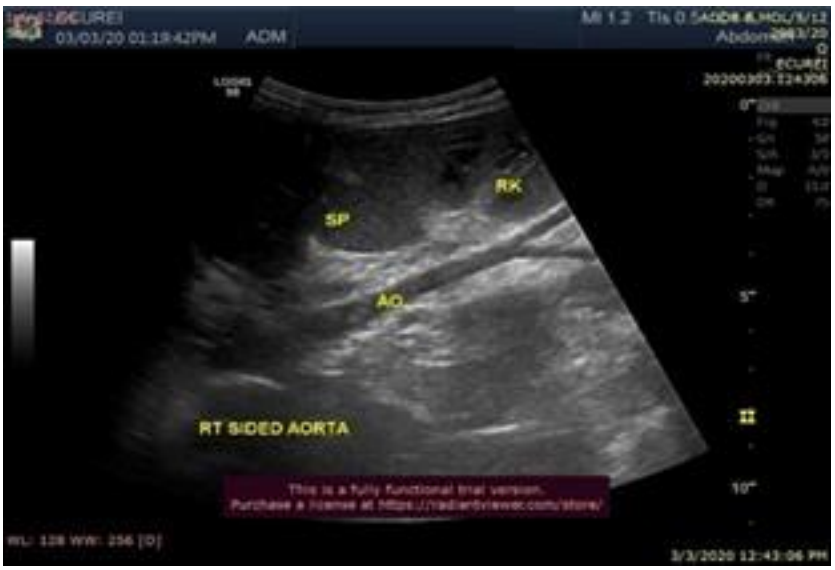

Figure 5: The stomach and abdominal aorta on the right side of theabdomen.

lead to liver injury resulting in secondary biliary cirrhosis with portal hypertension and liver failure. It can be non-syndromic biliary atresia or syndromic biliary atresia.

For this specific case, biliary atresia was associated with polysplenia and situs inversus. Polysplenia is a congenital disease manifested by multiple small accessory spleens rather than a single full-sized normal spleen. Situs inversus is when the internal organs are arranged as a mirror image of the normal anatomy. The most suitable treatment for this specific case is surgery.

2.5. Unique medical history:

Much as Biliary atresia has been reported on ultrasound in Uganda and Africa at large, its association with polysplenia and situs inversus has not yet been reported in any literature in Africa as compared to European countries which makes this case unique. This may be because such cases are rare or the sonographers/radiographers do not report them.

3. Conclusion:

This case study is to fill the knowledge gap among sonographers/radiographers regarding the existence of such cases to guide the management of those conditions especially among infants.

4. Recommendation:

The diagnosis of Biliary atresia, polysplenia, and situs inversus is usually based on clinical findings. Ultrasonography and Intraoperative cholangiogram are useful tools. The most suitable recommendable treatment for this specific case is surgery.

5. Source of funding:

The authors(s) received no financial support for this study.

6. Conflict of interest:

No conflict of interest was declared for all the authors.

7. Ethical consideration:

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