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Case Report

Long survival (21 years) after portoenterostomy for biliary atresia: A case report and review of complications

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Abstract

Long term survival for decades after portoenterostomy (Kasai procedure) for biliary atresia is rare and the association of portoenterostomy with liver cirrhosis is well known. Not much attention was given in the evaluation of the imaging features of cirrhosis caused by portoenterostomy as received by other known usual causes of cirrhosis. We presented a case of a Nigerian with confirmed biliary atresia at birth, that suvived portoenterostomy performed at two months of age for 21years. The cirrhotic presentation at imaging was that of prominent volume redistribution with less parenchymal changes in the liver. The long term survival and the type of cirrhotic presentation on imaging in this case is worthy of note for the record.

key words : Portoenterostomy, complication, cirrhosis, Imaging.

Introduction

Biliary atresia (BA) is the commonest cause of cholestatic disorder in the newborns[1] and it was thought to arise as a result of obstruction of bile flow due to obliteration of the extrahepatic bile ducts.[2] Treatment of BA is by Portoenterostomy (*Kasai* procedure)[3] where liver transplantation is unavailable with the attendant risk of long term complication of liver cirrhosis and portal hypertension.[4]

Alcoholism in the western world and viral hepatitis in Africa and Asia are the commonest causes of liver cirrhosis among other things.[4] Imaging presentation of cirrhosis due the above named causes are well documented in the literature. Whereas, liver cirrhosis secondary to hepatic portoenterostomy has not received such attention, perhaps due to the fact that long time survival after this procedure is rare.[5]

The purpose of this report is to present the imaging features of liver cirrhosis in a Nigerian patient with confirmed BA, 21years after portoenterostomy.

Case Report

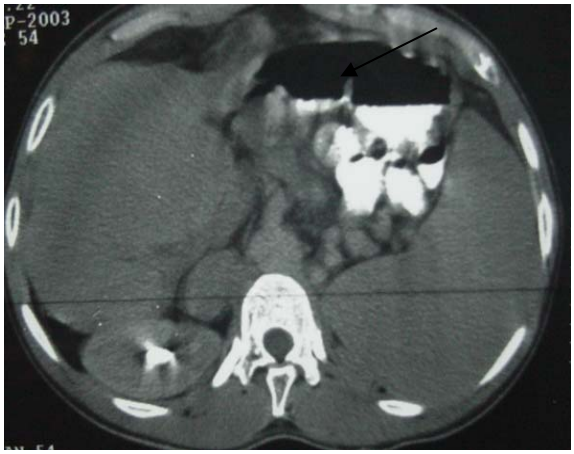
M.Y a male Nigerian was born 21years ago at a United States (U.S) based hospital. He had a low imperforate anus with a fine perineal fistula at birth, for which he underwent a perineal cutback procedure on the 2nd day of life. He did well post operatively on regular dilatation and the perineal anoplasty healed without stenosis.

Two months after birth he was readmitted at the same hospital with persistent jaundice which was noticed at the immediate neonatal period. Initial work up revealed direct hyperbiluribinaemia and elevated liver function tests. Abdominal ultrasound showed the bile ducts were not dilated. PIPIDA (Paraisopropyl iminodiacetic acid) scan revealed no GI uptake after 18hours, suggestive of obstruction.

Hepatic portoenterostomy (kasai procedure with a loop enterostomy) and liver biopsy were performed. The procedure resulted in good bile drainage with over 40cm^3 of bile drainage per day. This was despite the fact that on sections, the biliary

ducts had a diameter of only 60-80microns. He did remarkably well post operatively on antibiotics. At 3months post-op the loop enterostomy with its stoma had to be formally closed because the patient and his parents planned to leave U.S for Nigeria.While in Nigeria, he continued on weekly anal dilatation and antibiotics for the subsequent 6months and 2years respectively. Since then he lived very well without complain, until recently, at age 21years when he developed jaundice and right upper quadrant pain.When examined, he was found to be a young man, small for age, jaundice, not pale with female pubic hair distribution. No other peripheral stigmata of chronic liver disease. There was minimal pedal oedema. He had distended anterior abdominal wall veins with liver span of 8cm and splenomegaly of 6cm. No demonstrable ascites. Other systemic examinations were not remarkable and he was not in hepatic failure.He was negative for both hepatitis B and C viruses and had elevated total biluribin with predominant conjugated fraction. Also the serum biluribin was low (20g/dl) with reversal of albumin : globulin ratio and elevated transaminases (ALT>AST). The prothrombin time was prolonged (6sec above control). Full blood count and ESR were not remarkable. On abdominal CT and Ultrasound scanning, the liver was found to have a smooth parenchyma with normal density and echogenicity. However, there was volume redistribution in favour of the left lobe (hypertrophied) at the expense of the right lobe which was atrophied (Figures 1); associated with marked splenic enlargement (Figure 1) and multiple dilated vessels at the splenic hilum. The measured ultrasound right lobe : left lobe ratio and caudate lobe : right lobe ratio was 1.01 and 1.39 respectively. There was also intimate contact of the bowel loop with the hypertrophied left lobe and the hepatic hilum, most probably representing the previous portoenterostomy site (figures 1). Normal caliber portal vein was visualized and no ascites was seen in both the imaging modalities. The gall bladder and the biliary tree were not seen also.

Fig 1 **Axial upper abdominal CT scan, showing atrophied right hepatic lobe, enlarged left lobe and splenomegaly.**



Discussion

Jaundice in the neonatal period has numerous causes. The most common cause of conjugated hyperbiluribinaemia in neonates are neonatal hepatitis and biliary atresia (BA).[1] Around the time when the case presented above was born, Mabogunje[6] reported 36 histologically confirmed cases of BA in infants and children aged 2-20months from one of the tertiary health centers in Nigeria within a 13year study period, where almost half of them(15) died, 19 lost to follow up and 2 survived with persistent jaundice. In this same report, bilio-enteric anostomosis could only be done on four of them, which unfortunately did not establish bile flow. Our case was thus fortunate to have been delivered and attended to, where facilities were available more than 2 decades ago. Before the advent of liver transplant and its subsequent popularity in the treatment of BA, hepatic portoenterostomy was the treatment of choice for BA. Long term survival after this surgery is rare.[5] Infact, the famous kasai procedure of hepatic portoenterostomy for BA is most successful when performed early in life, 91% of such patients operated on before age 60days will have sustained bile drainage, while 56% of them drain bile if surgery is performed between 61-70days of age.[3,5,7,8] Our case was again lucky to have been operated within the window with the highest chance of survival (first-2months) and has now lived for 21years.

Ultrasound and Computed Tomography are the commonly used cross sectional imaging modalities in the evaluation of hepatic morphology for liver cirrhosis and its complications. Morphologically liver cirrhosis could be micronodular or macronodular and prolonged biliary obstruction was among other factors resulting to micronodular cirrhosis.[4] These nodules are of uniform size and are usually less than 3mm in diameter.[4,9] The usual compliments of signs for cirrhosis at imaging include, coarse and inhomogenous parenchyma, increased echogenicity/attenuation, surface nodularity, evidence of volume redistribution and signs of portal hypertension. The liver in our case has a homogenous and smooth parenchyma with normal echogenicity/attenuation

comparable to that of the spleen (Figures 2 & 4). However, what is prominent in our case is the volume redistribution, where the right lobe is hardly visualized due to atrophy and prominent left and

caudate lobes (Figures 1 & 2). There are also early features of portal hypertension in the form of splenomegaly and dilated portosystemic vessels at the spleenic hilum (Figures 3 & 4).

The above noted volume redistribution in favour of the left lobe may not be unconnected with the normal morphology of the intrahepatic portal system, where the right portal vein is wider and more vertically oriented when compared with its counterpart on the left, just like the division of the main bronchi in the chest. The result of this arrangement is that any insult that affects the liver, the right lobe is more likely to suffer than the left. Whereas, the non-demonstration of prominent parenchymal changes in this report, may represent the nature of presentation of cirrhosis in post-portoentrostomy cases after a prolonged period. Moreso, there is no persistent biliary obstruction which is known to be responsible for the previously mentioned micronodular parenchymal changes. It must be added here that, it may be possible to see full blown imaging changes of cirrhosis, had it been this case was imaged in his next decade or beyond. This remain to be further established.In conclusion, long term survival up to the 3rd decade after kasai procedure performed in the first-2months of life is possible, with the attendant risk of liver cirrhosis and portal hypertension. At cross sectional imaging, post-portoenterostomy cirrhosis presents more with volume redistribution than parenchymal changes. These peculiarities are worthy of note for the records.

References

1. Motalo C, Ireland JD, Hill ID, Bonie MD. Cholestatic disorders of infancy-aetiology and outcome. J Trop Pediatr 1990;36(5):218-222.
2. Kobayashi H, Stringer MD. Biliary atresia. Semin Neonatol 2003;8(5):383 – 391.
3. Kasai M, Suzuki H, Ohashi E, Ohi R, Chiba T, Okamoto T. Technique and results of operative management of biliary atresia. World J Surg 1978;2:571-580.
4. Conn HO, Atterbury CE. Cirrhosis : definition and history. In : Schiff L, Schiff ER, eds. Diseases of the liver. JB Lippincott, Philadelphia 1993;875-876.
5. Nio M, Ohi R, Miyano T, Saeki M, shiraki K, Tanaka K. Five and 10-year survival rates after surgery for biliary atresia : a report from the Japanese Biliary Atresia. J Pediatr Surg 2003; 38(7) : 997 – 1000.
6. Mabogunje OA. Biliary atresia in Zaria, Nigeria : a review. Ann Trop Paediatr 1987; 7(3) : 200-204.
7. Sangkhathat S, Patrapinyokul S, Tadtayathikom K, Osatakul S. Peri-operative factors predicting the outcome of hepatic portoenterostomy in infants with biliary atresia. J Med Assoc Thai 2003; 86(3): 224 – 231.
8. Lykavieris P, Chardot C, Sokhn M, Gauthier F, Valayer J, Bernard O. Outcome in adulthood of biliary atresia: a study of 63 patients who survived for over 20 years with their native liver. Hepatology 2005; 41(2) : 231 – 233.
9. Lefkowitch JH. Pathologic diagnosis of liver disease : major pathologic forms of liver Disease. In : Zakim D, Bayer TD (eds). Hepatology : a textbook of liver disease. WB saunders, Philadelphia 1990; 729.