Case Report

A case of HIV associated cholangiopathy (CAS) - A case report

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Abstract

Diseases of the liver and biliary tree have been described with significant frequency among patients with human immunodeficiency virus (HIV), and its advanced state, acquired immunodeficiency syndrome (AIDS). Through a variety of mechanisms, HIV/AIDS has been shown to affect hepatic parenchyma and biliary tree, leading to liver inflammation and biliary strictures. One of the potential hepatobiliary complications of this viral infection is AIDS cholangiopathy, a syndrome of biliary obstruction and liver damage due to infection-related strictures of the biliary tract. AIDS cholangiopathy is highly associated with opportunistic infections and advanced immunosuppression in AIDS patients, and due to the increased availability of highly active antiretroviral therapy, is now primarily seen in instances of poor access to anti-retroviral therapy and medication non-compliance. Current published literature describes well the clinical, biochemical, and endoscopic management of AIDS-related cholangiopathy. We present a case of 23 year male, presented with the complains of jaundice and severe abdominal pain associated with vomiting which on further investigations and imaging work up turned out to be AIDS cholangiopathy.

Key Words: Human immune deficiency virus, hepatobiliary, cholangiopathy.

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INTRODUCTION

Clinical presentation

23-year male presented with complains of severe abdominal pain in right hypochondrium since 1 week with intermittent breathlessness, loose stools, vomiting, loss of appetite, yellowish discoloration of skin, sclera and urine since 1 week with clnical diagnosis of obstructive jaundice. He gave past history of jaundice – no other relevant complaints in past. There was tenderness in the right hypochondrium region with hepatomegaly. General Examination- Icterus was evident by yellowish discoloration of skin, nails and sclera. He was undernourished and cachexic as well. Patient was found to be HIV positive. Biochemical tests revealed hyperbilirubinemia as total bilirubin was 3.5 mg/dL; of which direct bilirubin was 1.8mg/dL and indirect bilirubin was 1.7 mg/dL. Liver function tests were deranged as the SGOT levels were found to be 71 IU/L and SGPT levels were 55 IU/L with significantly raised serum alkaline phosphatase levels which was found to be 338 IU/L. These findings are consistent with obstructive biliopathy.

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Ultrasonography reviles

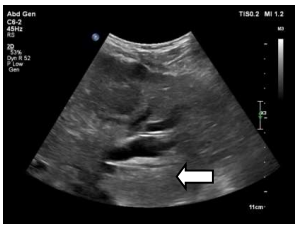
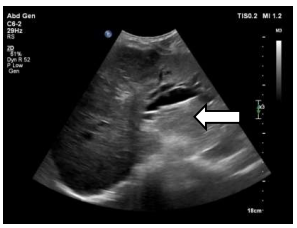
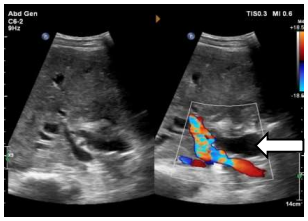

Figure 1 **Figure 2** **Figure 3**

Figure 1 and 2: longitudinal ultrasonography sections of the left lobe of liver shows markedly dilated left hepatic duct marked by arrows. Also note the heterogeneous echogenicity of the hepatic parenchyma; **Figure 3:** shows oblique ultrasonography section at the porta hepatis shows the dilated common bile duct (shown by arrow) just above the normal portal vein

Contrast enhanced CT revealed

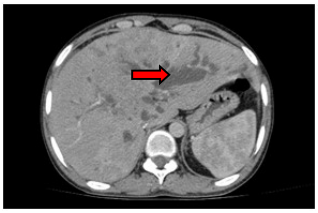
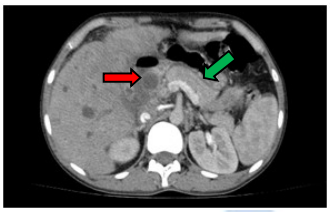
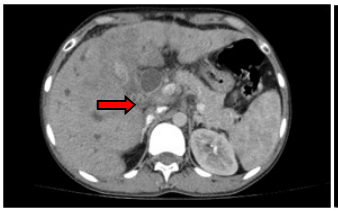
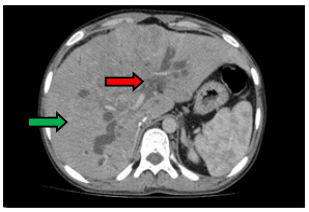

Figure 4 **Figure 5** **Figure 6** **Figure 7**

Figure 4: Axial contrast enhanced CT section shows the markedly dilated left hepatic duct and intrahepatic biliary radicals involving left lobe of liver. **Figure 5:**Contrast enhanced axial CT section shows dilated intrahepatic billiary radicals involving right lobe. Red arrow shows the gall bladder while green arrow shows normal pancreatic duct. **Figure 6:** Contrast enhanced axial CT section shows mild heterogeneity in the contrast enhancement pattern of liver parenchyma . Red arrow shows dilated CBD (8mm). **Figure 7:** Axial CECT section shows grossly dilated IHBR involving both lobes of liver shown by red arrow. Green arrow show heterogeneous density of the liver parenchyma

MRI and MRCP features

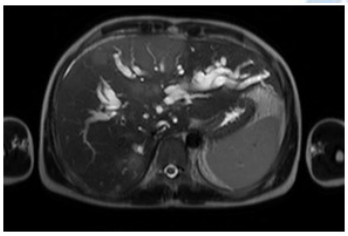
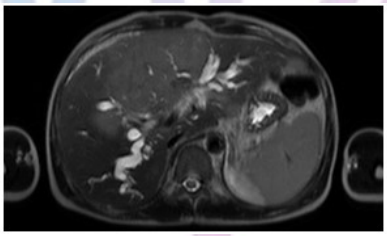
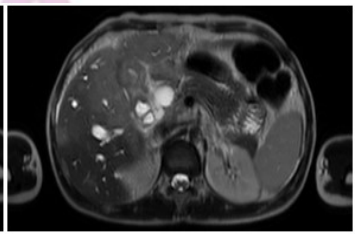

Figure 8 **Figure 9** **Figure 10**

Figure 8: Axial T2 weighted MRI image of the abdomen shows the enlarged liver with grossly dilated right and the left bile ducts and intrahepatic biliary radicals involving both the lobes of liver. **Figure 9:** Axial T2 weighted MR image of abdomen shows the heterogeneous signal intensities within the right lobe of liver which appears slightly hyperintense compared to the normal parenchyma **Figure 10:** T2 weighted axial MRI image of abdomen shows the dilated common bile duct and the minimally distended gall bladder with thick and oedematous walls.

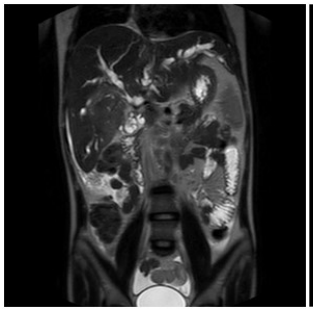
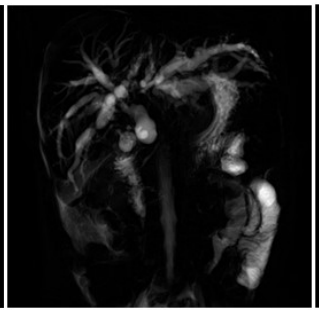
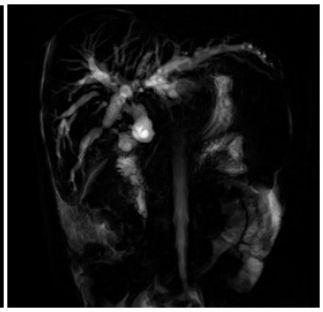

Figure 11 **Figure 12** **Figure 13**

Figure 11: Coronal T2 weighted MRI of abdomen shows grossly dilated intrahepatic biliary radicals with irregular lumen with multiple intermittent narrowed segments suggestive of strictures; **Figure 12 and 13:** MRI 3d coronal cholangiogram shows the grossly dilated bile ducts and intrahepatic biliary radicals with multiple focal strictures.

Considering the findings of heptomegaly with gross dilatation of bile ducts and intrahepatic biliary radicals due to multiple focal biliary strictures in this seropositive cachexic young male patient presenting with obstructive jaundice the diagnosis of AIDS associated cholangiopathy (CAS) was made. The lack of obstructive calculi in the biliary system and the absence of mass lesion/ parasitic infestation helps to rule out other etiologies of obstructive biliopathy.

Differential diagnosis:

1. primary sclerosing cholangitis (PSC)
2. Pyogenic cholangitis
3. Acalculous cholecystitis
4. Cholangiocarcinoma

The main differential diagnosis of AIDS associated cholangiopathy (CAS) is represented by sclerosing cholangitis[7], the morphological intrahepatic findings may be indistinguishable from primary sclerosing cholangitis,. Both diseases are characterized by focal stenosis of the bile duct interspersed with moderate segmental dilatation, giving a beaded appearance to the intrahepatic bile ducts[8]. To differentiate, it should be noted that pseudodiverticuli in the bile duct walls and high-grade extrahepatic bile duct stenosis, are typical findings of sclerosing cholangitis[7] while moderate ductal dilatation associated with irregular margins and nodules leans more toward AIDS associated cholangiopathy (CAS)[8]. Pyogenic cholangitis, another differential diagnosis, usually has a different septic context[9] and is associated with hepatic parenchymal abnormalities (eg. peribiliary micro abscesses or multifocal perfusion disorders) which the AIDS-associated cholangiopathy(CAS) lacks. Distal common bile duct fibrous stenosis secondary to passing of gallstones or chronic pancreatitis represent another possible cause of some of the imaging findings described in CAS[7]. To differentiate, it is important to review the medical history and previous studies of the patient. Finally, other eventual diagnoses to consider include alithiasic cholecystitis and cholangiocarcinoma. However, the clinical context of these is quite different. The first, although it also has a thickened gallbladder wall, occurs in severe septic patients with multiple organ failure or signs of peritonitis. In the second case, we see progressive biliary obstruction in time and with contrasted CT the appearance of infiltrated ductal masses invading the hepatic parenchyma, with delayed capture of contrast medium.

DISCUSSION

AIDS cholangiopathy is a biliary syndrome in AIDS patients which was first described by Cello in 1989[1]. It is diagnosed on clinical features, raised alkaline phosphatase, and on ultrasound and ERCP/MRCP examination. Estimated incidence of AIDS cholangiopathy in AIDS patients is 45% although many asymptomatic patients may exist. The disease is frequently present in male homosexual individuals suggesting male homosexuality as a major risk factor[2].Opportunistic infections of the biliary tree are believed to be the most common cause of AIDS cholangiopathy. The most commonly identified organisms are cryptosporidium and cytomegalovirus. Other opportunistic organisms are microsporidia, cyclospora, Mycobacterium avium complex, Isospora belli, Salmonella enteritidis, Salmonella typhimurium, Enterobacter cloacae and Candida albicans[3].

Infectious causes of AIDS-associated cholangiopathy

Bacteria, Mycobacteria	Viruses	Protozoa	Fungi
Mycobacterium Mycobacterium kansasii Mycobacterium tuberculosis Rochalimaeahenselae Rochalimaeaquintana Salmonella enteritidis Salmonella typhimurium Enterobacter cloacae Campylobacter fetus	Cytomegalovirus Herpes simplex Adenovirus HIV	Pneumocystis carinii Microsporidia Enterocytozoonbieneusi Encephalitozoon cuniculi Encephalitozoon intestinalis Cryptosporidium parvum Leishmania donovani Toxoplasma gondii Dicrocoeliumdendriticum Cyclospora cayetanensisIsospora	Histoplasma capsulatum Cryptococcus neoformans Coccidioides immitis Candida albicans

Patients of AIDS cholangiopathy usually present as right upper quadrant pain, fever and chills (clinical cholangitis), with dramatic elevation of serum alkaline phosphatase, usually with mild elevation or normal levels of serum bilirubin. Jaundice is uncommon, presenting in less than 5% of patients. Diarrhoea is a common associated feature in these patients. HIV-associated cholangiopathy is categorized into the following four types according to the ERCP findings: Type I, papillary stenosis (20% of cases); Type II, intrahepatic sclerosing cholangitis-like pattern alone (15-20%), Type III: combined papillary stenosis and intrahepatic sclerosing

cholangitis (50%); and Type IV, long extrahepatic bile duct stricture with or without intrahepatic involvement (15%)[4].Although some reports of HIV-associated cholangiopathy have included asymptomatic patients, its most common manifestation is right upper quadrant abdominal pain of biliary origin or pancreatitis[5].Diagnosis of AIDS cholangiopathy is made by clinical presentation, laboratory tests, and imaging studies. Serum alkaline phosphatase is the most commonly elevated biochemical test. Whereas hyperbilirubinaemia is uncommon. Abdominal ultrasound is usually the best initial imaging study in patients suspected of AIDS cholangiopathy. The previously established diagnostic tools for HIV associated cholangiopathy include US, CT, and ERCP. Although ERCP is considered to be the gold standard for the diagnosis and type classification of this disease[1], it is an invasive technique. MRCP is now increasingly being used as a non-invasive alternative imaging method, with high diagnostic accuracy for biliary involvement [6].

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