

PRIMARY BILIARY CIRRHOSIS IN A 37 YEAR OLD NIGERIAN MALE: A CASE REPORT

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ABSTRACT

The typical patient with primary biliary cirrhosis (PBC) is a middle aged woman presenting with fatigue, pruritus, and markedly elevated serum alkaline phosphatase (ALP) levels. PBC has a higher incidence and prevalence among Caucasians and Hispanics. We reported this case to reinforce the fact that the “rarities” in our environment may not be so uncommon after all. This article reports a case of PBC in a Nigerian man who presented with pruritus, hyperpigmentation and jaundice. The patient met all 3 diagnostic criteria and responded remarkably to Ursodeoxycholic acid (UDCA) tablets.

Primary biliary cirrhosis would be an unusual diagnosis to consider in a young male of African descent. Data on PBC among Africans has remained scanty. A high index of suspicion is key in any patient presenting with jaundice, pruritus and cholestasis.

Keywords: Primary biliary cirrhosis, Male

INTRODUCTION

Primary biliary cirrhosis (cholangitis) (PBC) is an autoimmune liver disease predominantly affecting middle aged women^{1,2}. It is characterized by progressive intrahepatic duct destruction leading to cholestasis with its complications, cirrhosis and portal hypertension³. Cirrhosis usually develops only late in the course of the disease; hence the term primary biliary cholangitis may be preferred.⁴

The prevalence of PBC is highest in northern Europe and the United States of America^{1, 2} The female-to-male ratio is 9:1³. The onset age is usually 30 - 60 years².

We reported an unusual case of primary biliary cirrhosis in a 37 year old Nigerian man with no known risk factors for PBC.

CASE REPORT

A 37 year old Petrol station attendant presented with a 3-week history of recurrent fever, jaundice and generalized pruritus. He also noticed generalized hyperpigmentation and weight loss. He had no personal or family history of an autoimmune disease and past medical history was not significant. There was no prior use of orthodox or herbal drugs. Alcohol history was not significant and he had never used tobacco/ psychoactive substances.

He was hyperpigmented, icteric, had shiny nail tips, with grade 2 digital clubbing. The abdomen and other systems were normal.

An assessment of obstructive jaundice? Cause was made by the admitting surgical team. Liver function tests. (Table 1)

Table 1: Initial investigation results

At presentation	Results	2 weeks after presentation	Results
ALP	52 I.U/L (≤ 270)	ALP	74 I.U/L (≤ 270)
ALT	34 I.U/L (≤ 40)	ALT	54 I.U/L (≤ 40)
AST	38 I.U/L (≤ 40)	AST	69 I.U/L (≤ 40)
Total bilirubin	10.8 mg/dl (0.6-1.2)	Total bilirubin	28.6 mg/dl (0.6-1.2)
Conjugated bilirubin	7.8 mg/dl (0.3-0.6)	Conjugated bilirubin	22.7 mg/dl (0.3-0.6)
Clotting profile	16 seconds (10-14)	Albumin	3.6 g/dl (3.3-5.2)

Normal values are in parentheses

Urinalysis: 3+ bilirubin, HBsAg, Anti HCV and retroviral screening: negative, Abdominal ultrasound and Computed tomography scans were normal, Full blood count: reduced haematocrit (34%) and peripheral blood film: normochromic, normocytic anaemia.

Pruritus persisted after 2 weeks on cholestyramine 4g q.d.s, Ketotifen slow release tablets 2mg daily, Cefuroxime tablets 500mg b.d and methylprednisolone cream b.d. The review of the gastroenterology team was sought 6 weeks later due to persistence of symptoms and the initial assessment was: obstructive jaundice? Cause to exclude (i) Biliary cirrhosis (ii) Extrahepatic biliary obstruction. A liver function test was requested (Table 2). Pruritus subsided after commencement of tabs Prednisolone 30mg daily for 2 weeks.

Table 2: Investigation results 6 weeks after presentation

Investigations	Results
ALP	1323 I.U/L (≤ 270)
ALT	103 I.U/L (≤ 40)
AST	62 I.U/L (≤ 40)
Total bilirubin	2.5 mg/dl (0.6-1.2)
Conjugated bilirubin	1.3mg/dl (0.3-0.6)
Clotting profile	16 seconds (10-14)

Normal values are in parentheses

Antimitochondrial antibody (AMA) assay was 2.823 units. [Normal is < 1.0 unit]. Tumour markers were all within normal limits. Other autoimmune markers could not be assayed due to patient's financial constraints. A

gastroscopy was requested to exclude varices but he was unable to afford the procedure while screening for osteoporosis was unavailable as at the time of review of the patient. Liver histology was in keeping with Ludwig stage one disease (numerous bile plugs

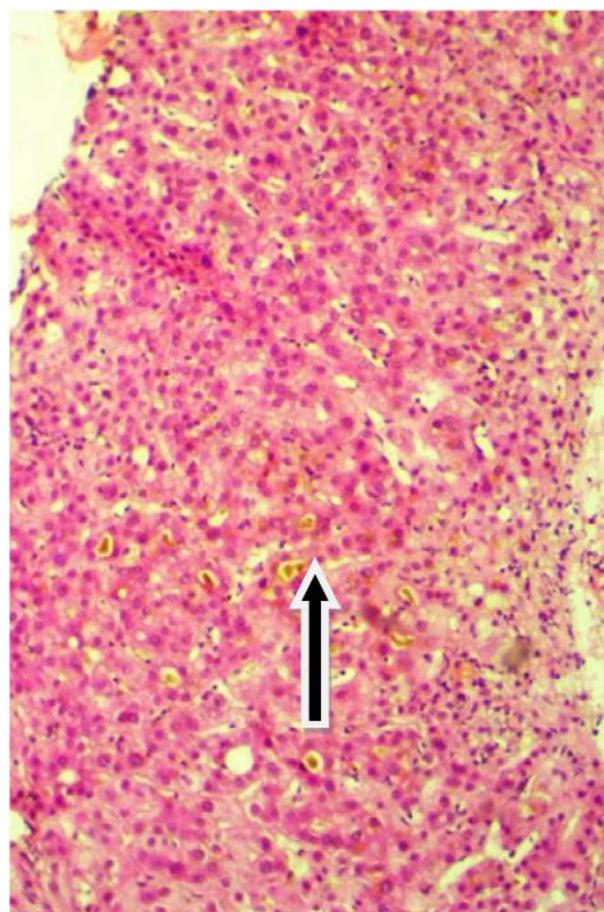


Fig. 1: Micrograph showing liver histology with cords of hepatocytes with bile plugs x40 magnification. H&E stain used

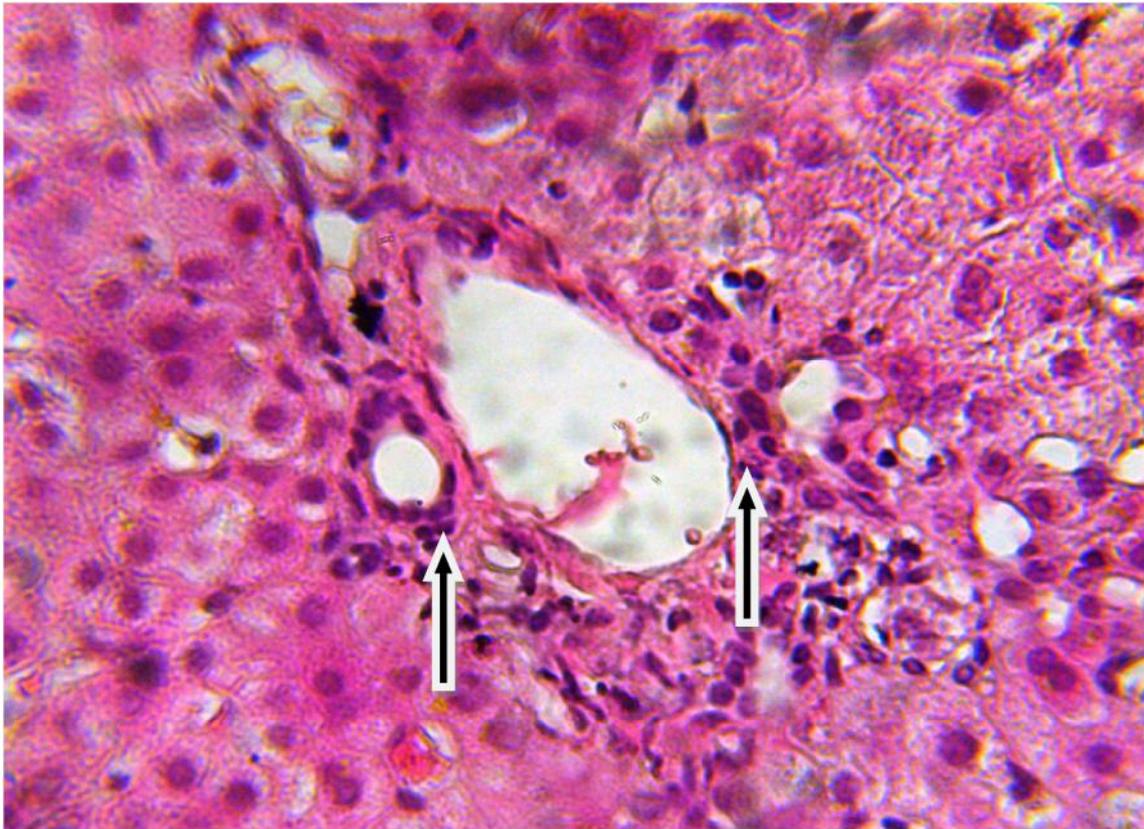


Fig. 2: Micrograph showing liver histology with portal areas with ductular proliferation & mild lymphocytic inflammation x10 magnification. H&E stain used

within dilated bile canaliculi and focal areas of feathery degeneration. Also seen were areas of portal tract oedema and residual normal portal triads.

He was subsequently discharged on tabs UDCA 1g daily but lost to follow up. He resurfaced 6 months later and had now commenced tabs UDCA (Ursofalk®) {batch no. 15H27667L; Exp 08/2019} 3 months earlier and had improved remarkably. All symptoms had completely resolved and a repeat liver function test was normal. The patient was adequately counseled on the possible course of the condition and a feasible follow up plan was subsequently instituted.

DISCUSSION

Features of PBC pointing to an autoimmune pathogenesis include the intense immune response to an intracytoplasmic antigen, presence of AMA and involvement of T lymphocytes in the destruction of bile ducts³.

We reported this case because of its unusual occurrence in a man of African descent. PBC occurring

in males is relatively rare and is reflected by the scarcity of studies examining PBC in this group⁵. About 7–11% PBC cases are males⁵. A multicenter trial of UDCA among 535 patients found 3.9% to be of African descent⁶. To the best of our knowledge, this is the first report of PBC in a Nigerian male.

Our patient's most disabling symptom was pruritus. This occurs in 19-55% of PBC patients³. Even though females experience pruritus as a single symptom than males, jaundice, jaundice with pruritus and upper GI bleeding are commoner in men³. However, the case under review did not have portal hypertension.

Generally, non-Caucasians have been reported to have worse disease progression and poorer outcomes⁶. Possible explanations include a more rapid disease progression, less access to early care, misdiagnoses due to inadequate testing, absence of liver biopsies or presence of comorbidities which may have led to delay in treatment⁶.

The diagnosis of PBC is made when 2 of 3 established criteria are met: serum ALP $\geq 1.5 \times$ ULN, serum AMA or liver biopsy findings in keeping with

PBC³. A liver biopsy is not absolutely necessary especially in patients with serum ALP >1.5-fold elevated & AST < 5-fold elevated as this has been found to be highly predictive of PBC.² Our patient met all 3 diagnostic criteria.

The patient did not benefit from anti histamines and cholestyramine. This finding was consistent with reports in literature³. UDCA has received the most attention in the treatment of PBC due to rapid and long lasting improvements in liver biochemistries and histology³. A number of other medications have also been evaluated in many trials. They include bile acids, immunosuppressants, anti-inflammatory, cupruritic and anti fibrotic agents. The farnesoid X receptor agonist Obeticholic acid, has anti fibrotic and choloretic properties⁷ and when given in combination with UDCA results in significant improvements in serum ALP by 21-25%⁸. Indications for liver transplantation in PBC are similar to those in other liver diseases namely decompensated cirrhosis with intractable ascites and spontaneous bacterial peritonitis, recurrent variceal bleeding, encephalopathy or hepatocellular carcinoma⁹. Apart from treating the underlying disease, a key component of care in PBC entails early recognition, prevention and treatment of complications as well as proper counseling on the likely course of the illness.

CONCLUSION

Primary biliary cirrhosis would be an unusual diagnosis to arrive at in an African male presenting with jaundice and pruritus. However, the case discussed, reinforces the importance of a high index of suspicion for the “rarities” in clinical practice in a resource-poor setting.

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