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Primary Biliary Cholangitis Presenting as Decompensated Cirrhosis with Hepatic Encephalopathy in a Young Female: A Case Report

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ABSTRACT

Background: Primary Biliary Cholangitis (PBC) is a chronic, progressive autoimmune cholestatic liver disease predominantly affecting middle-aged women, characterised by immune-mediated destruction of small intrahepatic bile ducts leading to cholestasis, fibrosis, and cirrhosis. The anti-mitochondrial antibody (AMA), positive in over 95% of cases, is its serological hallmark. Rapid decompensation in younger patients with hepatic encephalopathy is uncommon and clinically challenging.

Case Summary: We report a 41-year-old female diagnosed with PBC (ANA positive, AMA positive at titre 1:100 by IFA) who presented in June 2026 with acute decompensation: drowsiness, bilateral pedal oedema, decreased urine output, vomiting, and Grade III hepatic encephalopathy. Investigations revealed severely deranged liver function (total bilirubin 25.16 mg/dL), coagulopathy (INR 2.91), hypoalbuminemia (albumin 2.1 g/dL), hyponatraemia (Na 129 mEq/L), hypokalaemia (K 1.9 mEq/L), gross splenomegaly, and moderate ascites. Fibroscan showed LSM 53 kPa confirming cirrhosis. ICU management included albumin infusion, diuretics, lactulose, rifaximin, LOLA/Heparmerz, BCAA supplementation, blood products, and antibiotics. The patient had previously been advised liver transplantation.

Conclusion: This case highlights the critical importance of early AMA screening in young women with cholestatic liver disease, timely initiation of ursodeoxycholic acid (UDCA), and early transplant listing in PBC. Rapid disease progression to decompensated cirrhosis underscores the need for vigilant follow-up.

Keywords: Primary Biliary Cholangitis; Antimitochondrial Antibody; Hepatic Encephalopathy; Decompensated Cirrhosis; Autoimmune Liver Disease; Splenomegaly; Liver Transplantation; Fibroscan

1. INTRODUCTION

Primary Biliary Cholangitis (PBC), formerly known as Primary Biliary Cirrhosis, is a chronic autoimmune cholestatic liver disease characterised by the progressive, immune-mediated destruction of small intrahepatic bile ducts. It predominantly affects middle-aged women (female-to-male ratio approximately 9:1) and has a prevalence of approximately 40 per 100,000 individuals. The disease course is variable: some patients remain asymptomatic for decades while others progress rapidly to cirrhosis and liver failure.

The anti-mitochondrial antibody (AMA), specifically targeting the E2 subunit of the pyruvate dehydrogenase complex (PDC-E2), is the serological hallmark of PBC, present in 90–95% of patients. Biochemically, PBC is characterised by a cholestatic pattern with disproportionate elevation of alkaline phosphatase (ALP) and gamma-glutamyltransferase (GGT). Anti-nuclear antibodies (ANA), particularly those with cytoplasmic or rim patterns, are seen in up to 50% of PBC patients and may indicate a more aggressive disease course.

Hepatic decompensation in PBC — manifesting as ascites, variceal bleeding, hepatic encephalopathy, and jaundice — marks a critical transition associated with poor prognosis. Fibroscan (transient elastography1) has emerged as an important non-invasive tool for liver stiffness measurement (LSM) in staging fibrosis. Liver transplantation remains the only curative option for advanced PBC, with excellent post-transplant survival. We present a case of rapidly progressive PBC decompensating to Grade III hepatic encephalopathy in a young woman, illustrating the diagnostic and therapeutic challenges involved.

2. CASE PRESENTATION

2.1 Patient Demographics and History

A 41-year-old married female presented with a known 20-month history of PBC (first diagnosed October 2024 at age 39 years), with confirmed serological markers: ANA positive (cytoplasmic pattern, titre 1:320) and AMA positive (titre 1:100 by immunofluorescence assay). Her known comorbidities included portal hypertension with splenomegaly and dyslipidaemia. She had been evaluated for liver transplantation. She had also been taking Siddha herbal medication for approximately two months prior to this admission.

Chief Complaints (3-day history):

- Drowsiness (giddiness) and generalised tiredness
- Decreased urine output with dark yellow/orange-coloured urine
- Bilateral lower limb pitting oedema
- Non-blood-stained, non-projectile vomiting — 4 episodes/day
- Altered sensorium

On general examination, the patient is drowsy and disoriented, not responding to oral commands, with bilateral icteric sclera noted. Vitals reveal a blood pressure of 110/70 mmHg, pulse rate of 100 bpm, SpO2 of 97% on room air, temperature of 98°F, and a capillary blood glucose (CBG) of 161 mg/dL. On systemic examination, the cardiovascular system shows normal S1 and S2 sounds, and the respiratory system reveals equal bilateral air entry. Per abdomen examination reveals a distended abdomen with ascites and gross splenomegaly. On CNS examination, the patient's GCS was E4V4M6 on review (10/6/26), remaining unchanged at E4V4M6 on subsequent assessment.

3. INVESTIGATIONS *Table 1. Summary of Laboratory Investigations*

Parameter	Value	Reference Range	Flag
Complete Blood Count			
Haemoglobin (g/dL)	**8.8**	12.0–15.0	**L**
Haematocrit/PCV (%)	**24.5**	36.0–46.0	**L**
RBC Count (Millions/µL)	**2.64**	3.8–4.8	**L**

Parameter	Value	Reference Range	Flag
MCV (fL)	**92.8**	83–101	
MCH (pg)	**33.4**	27–32	**H**
MCHC (%)	**35.9**	31.5–34.5	**H**
RDW-CV (%)	**16.4**	11–16	**H**
Total WBC Count (×10⁹/L)	**20.76**	4.0–10.0	**H**
Neutrophils (%)	**83.4**	20–80	**H**
Lymphocytes (%)	**8.4**	22–44	**L**
Monocytes (%)	**8.0**	2–10	
Absolute Neutrophil Count (×10⁹/L)	**17.32**	2–7	**H**
Absolute Monocyte Count (×10⁹/L)	**1.66**	0.08–0.8	**H**
Platelet Count (×10³/µL)	**84**	150–410	**L**
HbA1C	3.67%	-	L
Coagulation Profile			
PT-INR (10 June 2026)	**2.91**	0.9–1.1	**H**
PT Test (secs)	**31.3–40.8**	–	**H**
PT-INR (11 June 2026)	**2.15**	0.9–1.1	**H**
APTT Test (secs)	**46.8**	–	**H**
Other Parameters			
HbA1c (%)	**3.67**	<5.7% (normal)	**L**
Serum Calcium (mg/dL)	**8.2**	8.6–10.2	**L**
Serum Magnesium (mg/dL)	**2.8**	1.77–2.98	
Serum Phosphorus (mg/dL)	**1.5**	2.5–4.8	**L**
Serum Osmolality (mosm/kg)	**285**	280–295	
Urine Osmolality (mosm/kg)	**281.2**	50–400	
Urine Potassium (mmol/L)	**21.9**	Up to 20	**H**
Troponin I (ng/mL)	**<0.01**	<0.10	
Microbiology / Serology			
HBsAg (ELISA)	**Negative**	–	
Anti-HCV (ELISA)	**Negative**	–	
HIV 1 & 2 Antibodies (ELISA)	**Negative**	–	

SGOT: Serum Glutamic-Oxaloacetic Transaminase; SGPT: Serum Glutamic-Pyruvic Transaminase; PT-INR: Prothrombin Time-International Normalised Ratio; APTT: Activated Partial Thromboplastin Time; HbA1c: Glycated Haemoglobin; HBsAg: Hepatitis B Surface Antigen; HCV: Hepatitis C Virus; HIV: Human Immunodeficiency Viru s

3.1 Liver Function Tests — Current Admission (10 June 2026)

Parameter	Result	Reference Range	Serial Trend
Total Bilirubin	**25.16 mg/dL ↑↑**	0.3–1.1 mg/dL	2.22→4.74 (Aug25→Feb26)
Direct Bilirubin	**19.19 mg/dL ↑↑**	0.0–0.2 mg/dL	1.83→4.72
SGOT (AST)	**261.4 IU/L ↑↑**	<31 IU/L (F)	150→148
SGPT (ALT)	**151.1 IU/L ↑↑**	<34 IU/L (F)	108→85
Alkaline Phosphatase	**164.7 IU/L ↑**	42–98 IU/L	—
GGT	**101.7 IU/L ↑**	<38 IU/L (F)	—
Total Protein	**4.75 g/dL ↓**	6.0–7.8 g/dL	—
Albumin	**2.1 g/dL ↓↓**	3.35–5.2 g/dL	3.5→3.4
A/G Ratio	**0.79 ↓**	1.2:1–2:1	—

3.2 Haematology and Coagulation Profile

Parameter	Result	Reference	Clinical Significance
Haemoglobin	**7.2 g/dL ↓**	12–16 g/dL	Anaemia — transfusion required
WBC	**15.9 ×10³/µL ↑**	4–11 ×10³	Leucocytosis — infection
Platelets	**94 ×10³/µL ↓**	150–400 ×10³	Thrombocytopenia (hypersplenism)
PT	**31 seconds ↑**	11–13.5 sec	Coagulopathy
INR	**2.91 ↑↑**	0.9–1.1	Severe coagulopathy
APTT	**40.8 seconds ↑**	25–35 sec	Coagulopathy

3.3 Renal Function and Electrolytes

Parameter	Result	Reference	Status
Urea	**51 mg/dL ↑**	15–40 mg/dL	Elevated

Creatinine	0.90 mg/dL	0.6–1.1 mg/dL	Normal
Sodium (Na^+)	**129 mEq/L ↓**	136–145 mEq/L	Hyponatraemia
Potassium (K^+)	**1.9 mEq/L ↓↓**	3.5–5.1 mEq/L	Severe Hypokalaemia
Chloride (Cl^-)	101 mEq/L	96–106 mEq/L	Normal
Uric Acid	2.35 mg/dL	2.3–6.6 mg/dL	Normal

3.4 Autoimmune Serology Panel

Test	Result	Clinical Significance
AMA — Immunofluorescence Assay	**POSITIVE — Titre 1:100**	Hallmark of PBC (>95% sensitivity); M2 specificity
ANA — Immunofluorescence	**POSITIVE — Cytoplasmic pattern; Titre 1:320; Intensity 2+**	Seen in PBC; cytoplasmic/rim ANA indicates aggressive course
ASMA — Immunofluorescence	Negative (1:100)	Rules out AIH Type 1 as primary diagnosis
LKM-1 Antibody (EIA)	5.8 RU/mL (Negative <20)	Negative — excludes AIH Type 2
HBc IgM Antibody (CLIA)	0.2 Index (Negative <0.80)	No acute or ongoing HBV infection
IgG (PEG Enhanced)	12.18 g/L (6.5–16 g/L)	Normal IgG; elevated IgM is the typical PBC pattern

Figure 1. *Positive Anti Mitochondrial Antibody (AMA) by indirect immunofluorescence assay (IFA) at titre 1:100. The characteristic granular cytoplasmic fluorescence pattern of mitochondrial staining is clearly demonstrated, consistent with M2 antigen specificity in Primary Biliary Cholangitis.*

3.5 Serial Imaging and Fibro scan

Date	Modality	Key Findings	Impression
Oct 2024	CT Abdomen (Whole)	Liver 190 mm, mildly nodular, diffusely altered attenuation; Spleen 120 mm; Portal vein 10 mm; Periportal oedema; No bowel mass or ascites	Enlarged liver with parenchymal disease; Marginal splenomegaly
Aug 2025	USG Abdomen & Pelvis	Liver 14.3 cm (normal echoes); Spleen 17.8 cm grossly enlarged; Small umbilical hernia; No ascites	Gross splenomegaly; Small umbilical hernia
Apr 2026	MRCP 1.5T	Liver 180 mm, nodular, fatty attenuation, periportal oedema; Portal vein 12 mm; Spleen 160 mm; Dilated splenic vein 10 mm; Gallbladder contracted with subserosal oedema; Pancreas mildly oedematous; Peripancreatic fluid; Mild ascites	Enlarged liver + parenchymal disease; Gross splenomegaly; Mild ascites; ?Acute pancreatitis
Jun 2026	USG + Fibroscan	Liver 180 mm parenchymal disease; Spleen grossly enlarged; Portal vein 12 mm; GB contracted + subserosal oedema; Moderate ascites; Fibroscan: LSM 53 kPa, CAP 233 dB/m; UGI Endoscopy: No oesophageal varices	Decompensated cirrhosis; Portal hypertension; Moderate ascites; No varices on endoscopy

5. DISCUSSION

This case presents a compelling constellation of features characteristic of decompensated PBC. PBC is an organ-specific autoimmune disease driven by T-lymphocyte-mediated destruction of cholangiocytes lining the small intrahepatic bile ducts. The progressive biliary obstruction leads to accumulation of bile acids within hepatocytes, culminating in oxidative injury, fibrosis, and ultimately cirrhosis. The ANA and AMA positivity in this patient is consistent with the immunological profile of PBC.

Primary Biliary Cholangitis is a well-characterised autoimmune cholestatic liver disease. The diagnosis in this patient was firmly established by the combination of: positive AMA at titre 1:100 by indirect immunofluorescence (Figure 1), demonstrating the characteristic mitochondrial staining pattern; cholestatic biochemical profile with markedly elevated ALP, GGT, and direct bilirubin; progressive hepatomegaly with parenchymal disease on serial imaging; and gross splenomegaly consistent with portal hypertension. The positive ANA with cytoplasmic pattern at high titre (1:320) is a well-recognised serological feature of PBC and has been associated with more aggressive disease progression.

A critical differentiating point in this case is the negative ASMA and negative LKM-1 antibody, effectively excluding Autoimmune Hepatitis (AIH) Type 1 and Type 2 as primary diagnoses respectively,

and the negative HBc IgM excluding active hepatitis B. The normal IgG level (12.18 g/L) is also more consistent with PBC than AIH, in which IgG is typically markedly elevated. An AIH-PBC overlap syndrome, though not confirmed serologically, remains a differential consideration given the high-titre ANA. Hepatic encephalopathy (HE) in this patient was precipitated by the convergence of multiple factors: impaired hepatocellular clearance of nitrogenous waste products (particularly ammonia generated by intestinal bacteria and muscle catabolism), portosystemic shunting secondary to portal hypertension, and systemic inflammation from possible spontaneous bacterial peritonitis (SBP). The clinical picture of drowsiness, disorientation, and failure to respond to verbal commands corresponds to West Haven Grade III HE, which requires ICU-level care. The use of lactulose, rifaximin, and LOLA (Hepa-Merz) forms the cornerstone of HE management, targeting ammonia production and enhancing its clearance.

The markedly elevated total bilirubin (32.04 mg/dL), predominantly direct (24.01 mg/dL), reflects severe intrahepatic cholestasis consistent with decompensated cirrhosis superimposed on the underlying biliary pathology. Transaminase elevation (SGOT 198.1 IU/L, SGPT 145.4 IU/L), while significant, was disproportionately lower than the bilirubin, a pattern seen in advanced cirrhosis where few remaining hepatocytes are present to release enzymes.

The coagulopathy (PT-INR 2.91 on day 1, improving to 2.15 by day 2 with FFP) reflects synthetic liver failure. Coagulation factors II, V, VII, IX, and X are exclusively synthesised in hepatocytes; their depletion results in prolonged PT and elevated INR. Vitamin K was administered to address any nutritional or obstructive component, while FFP provided immediate factor replacement.

Severe anaemia (Hb 8.8 g/dL) with thrombocytopenia (platelets 84 × 10^3/μL) represents a multifactorial process: hypersplenism from splenomegaly (splenic sequestration), suppressed bone marrow erythropoiesis from chronic disease, nutritional deficiencies, and possible gastrointestinal blood loss from portal hypertensive gastropathy or varices. Leucocytosis (WBC 20.76 × 10^9/L) with neutrophilia raised the clinical suspicion of infection, and diagnostic paracentesis was planned to exclude SBP. The low HbA1c (3.67%) in the context of haemolytic anaemia is expected, as reduced RBC lifespan limits glycation of haemoglobin, making HbA1c an unreliable glycaemic marker in this population. HbA1c

Laboratory Challenge

HbA1c was found to be 3.5% despite a random blood glucose value of 161 mg/dL. In advanced cirrhosis, anaemia, hypersplenism, altered red cell survival and recent blood transfusion may result in falsely low HbA1c values. Therefore HbA1c interpretation requires correlation with direct glucose measurements and clinical status.

Hyponatraemia (Na 129 mEq/L) in cirrhosis is predominantly dilutional (hypervolaemic hyponatraemia) due to non-osmotic release of antidiuretic hormone (ADH) in response to effective circulatory volume depletion. Serum osmolality of 285 mosm/kg remained at the lower end of normal, while urine osmolality of 281.2 mosm/kg and elevated urine potassium indicated ongoing renal water retention. Aggressive diuresis with spironolactone and judicious furosemide was targeted at managing fluid overload, while fluid restriction to < 1 L/day was essential to avoid further dilution.

The clinical timeline demonstrates alarming rapidity of disease progression. Liver size increased from 190 mm (CT, October 2024) to 180 mm (MRCP, April 2026) with increasingly nodular surface, progressive periportal oedema, and portal vein dilatation from 10 mm to 12 mm over 18 months. Spleen size escalated from 120 mm (CT 2024) to 178 mm (USG, August 2025) to 160 mm (MRCP, April 2026) with further progression, indicating worsening portal hypertension. Serial bilirubin trended from 2.22 mg/dL (August 2025) to 4.74 mg/dL (February 2026) to 25.16 mg/dL (June 2026), reflecting rapid deterioration in

excretory liver function. The Fibroscan LSM of 53 kPa on current admission is markedly elevated, well above the 9.6 kPa threshold for cirrhosis in PBC, and the CAP of 233 dB/m indicates significant hepatic steatosis, possibly potentiated by dyslipidaemia and concurrent Siddha herbal medication hepatotoxicity.
Hepatic encephalopathy (Grade III) in this case is multifactorial: hypoalbuminemia (2.1 g/dL), coagulopathy (INR 2.91), hyponatraemia (Na 129 mEq/L), severe hypokalaemia (K 1.9 mEq/L), possible infection (leucocytosis, urinalysis showing bacteria and RBCs 10–12/HPF), and the underlying portosystemic shunting of decompensated cirrhosis. The management appropriately addressed each of these components with lactulose, rifaximin, LOLA/Heparmerz (ornithine aspartate), albumin replacement, electrolyte correction, and antibiotics.
Importantly, UGI endoscopy revealed no oesophageal varices despite significant portal hypertension — an uncommon finding that avoids the need for primary prophylactic variceal band ligation but mandates close endoscopic surveillance. The absence of varices with gross splenomegaly and portal vein dilatation may reflect the predominance of splenic pooling and portal-to-mesenteric rather than portal-to-oesophageal collateral development at this stage.
The use of Siddha traditional medicine for approximately two months prior to this admission deserves attention. Several herbal preparations used in traditional medicine systems have been associated with drug-induced liver injury (DILI), and their hepatotoxic potential should be considered as a possible contributing factor to acute-on-chronic liver decompensation, warranting further workup including serum ammonia and a detailed toxicology review if possible
This case also highlights a critical clinical challenge in low- and middle-income settings: the delay between diagnosis and adequate treatment. While the patient had been advised liver transplantation socioeconomic and logistical barriers likely contributed to the rapid progression to decompensation.

6. CONCLUSION

We report a case of Primary Biliary Cholangitis in a 41-year-old female who presented with acute decompensation characterised by Grade III hepatic encephalopathy, moderate ascites, and severe coagulopathy. Positive AMA by indirect immunofluorescence assay (titre 1:100) with characteristic granular cytoplasmic mitochondrial staining (Figure 1) confirmed the autoimmune aetiology. The case illustrates the rapid and potentially devastating progression of PBC in young patients, particularly in the context of undertreated disease and possible concurrent herbal hepatotoxicity. It emphasises the need for: (1) early AMA screening in young women with unexplained cholestatic liver disease; (2) prompt initiation of UDCA therapy; (3) non-invasive staging with Fibroscan; (4) early referral for liver transplantation in progressive disease; and (5) avoidance of potentially hepatotoxic traditional medicine preparations in patients with chronic liver disease.

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