

# Primary Biliary Cholangitis Presenting with Low Back Pain and Complicated by Immune-Mediated Necrotizing Myositis: A Case Report

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## Abstract

Primary biliary cholangitis (PBC) and immune-mediated necrotizing myositis (IMNM) are both rare autoimmune disorders, and their coexistence is uncommon. Low back pain (LBP) as the initial presentation of this overlap is extremely rare and may delay diagnosis. A case is presented of a 46-year-old woman with recurrent LBP and mild proximal muscle weakness, along with laboratory findings of elevated liver and muscle enzymes, positive anti-mitochondrial antibody subtype M2 (AMA-M2), anti-GP210, and anti-SSA antibodies, and negative myositis-specific antibodies. Imaging revealed paraspinal myositis, and histopathology confirmed PBC (stage 1) and IMNM. The patient responded well to a combination of ursodeoxycholic acid, corticosteroids, hydroxychloroquine, and mycophenolate mofetil, achieving normalization of laboratory parameters and complete symptom resolution after 15 months. This case underscores the need for clinicians to consider systemic autoimmune diseases in patients with atypical LBP and highlights the potential for AMA-M2–positive myositis to involve the paraspinal muscles, providing insight into a rare manifestation of PBC–IMNM overlap.

**Keywords:** Antibodies, antimitochondrial, biliary, liver cirrhosis, low back pain, myositis

## Introduction

Primary biliary cholangitis (PBC) is a chronic autoimmune liver disease characterized by progressive intrahepatic bile duct destruction and the presence of antimitochondrial antibodies, most commonly the M2 subtype. The immune-mediated necrotizing myositis (IMNM) is a distinct subtype of idiopathic inflammatory myopathy (IIM), marked by muscle fiber necrosis and severe weakness with minimal inflammatory cell infiltration. The coexistence of PBC and IMNM is rare, and LBP as the first manifestation is even more uncommon. This case involves PBC complicated by IMNM with LBP as the initial symptom, aiming to raise awareness of this unusual presentation and its diagnostic challenges.

## Case Presentation

A 46-year-old woman with no significant medical history presented in January 2024 with an 8-month history of recurrent lower back pain and bilateral upper limb weakness. Written informed consent was obtained from the patient for the publication of this case report, including clinical details and images, with measures to ensure anonymity. The study was approved by the institutional ethics committee. The onset was insidious with no identifiable trigger. Symptoms included back pain during bending or stretching, difficulty raising the upper limbs, and morning stiffness, with mild activity limitation. She denied night pain, uveitis, joint pain, rash, dysphagia, hoarseness, chest tightness, shortness of breath, oral ulcers, or gastrointestinal symptoms. She reported mild dry mouth and eyes. Diet, sleep, and bowel habits were normal, with no weight changes. There was no family history of genetic or similar diseases. The patient consulted multiple departments and was diagnosed with lumbar strain, but symptomatic treatment was ineffective.

Physical examination revealed: temperature 36.5°C, respiratory rate 18/min, blood pressure 120/82 mmHg, heart rate 76/min, height 154 cm, weight 50 kg. She was alert, well-nourished, with no scleral icterus, conjunctival hyperemia, or rash. Oral mucosa was dry, with no dental caries. Gait was normal, heart rhythm regular without murmurs, and lungs and abdomen were unremarkable. Cranial nerves were intact, spinal mobility was normal, with mild paraspinal muscle tenderness. Muscle strength was grade 5 in all limbs, except for neck flexors (grade 4). Bilateral "Figure 4" sign, sacroiliac joint tenderness, and straight leg raise tests were negative.

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**Laboratory findings:** Liver function tests showed elevated alanine aminotransferase (143 U/L, reference 7-40 U/L), aspartate aminotransferase (149 U/L, 13-35 U/L), alkaline phosphatase (289 U/L, 35-100 U/L), and gamma-glutamyl transferase (153 U/L, 7-45 U/L). Muscle enzymes were elevated: CK 549 U/L, CK-MB 13.25, lactate dehydrogenase (LDH) 256 U/L, with normal troponin I (0.020, 0-0.045). Immunological tests revealed ANA (antinuclear antibody) 1 : 1000 (cytoplasmic granular pattern), positive AMA (antimitochondrial antibody), anti-M2-3E, anti-SP-100, anti-GP-210 (Table 1), and SSA antibodies. Myositis-specific antibodies (anti-JO-1, PL-7, NXP-2, SSA/Ro52kD, SAE1/2, TIF1- $\gamma$ , MDA5, PL-12, EJ, HMGCR, Mi-2, SRP) were negative. The RF and HLA-B27 were negative. Tumor marker screening was negative (Table 2). Electrolytes, creatinine, uric acid, lipids, homocysteine, glucose, and hemoglobin A1C were normal. Chest computed tomography (CT) showed no interstitial changes or other abnormalities (Figure 1), bone density (DXA), and cardiac ultrasound were unremarkable.

**Electromyography:** Spontaneous potentials were observed at rest, with narrow and small motor unit potentials during mild contraction, short spike waves, and early recruitment or simple mixed phases during strong contraction, consistent with myogenic damage.

**Imaging:** Magnetic resonance imaging (MRI) of the bilateral thighs and lumbosacral region revealed multifocal myositis (Figure 2).

#### Pathology

**Lip gland biopsy** (Figure 3) revealed lymphocytic and plasma cell infiltration with lymphoid foci (>1 focus/4 mm<sup>2</sup>) on hematoxylin-eosin

**Table 1. Key Laboratory Findings**

Parameters	Result	Reference Range
ALT (alanine aminotransferase)	143 U/L	7-40 U/L
AST (aspartate aminotransferase)	149 U/L	13-35 U/L
ALP (alkaline phosphatase)	289 U/L	35-100 U/L
GGT ( $\gamma$ -glutamyl transferase)	153 U/L	7-45 U/L
CK (creatinine kinase)	549 U/L	40-200 U/L
CK-MB (creatinine kinase-MB)	13.25 ng/mL	0-5.00 ng/mL
ANA (antinuclear antibody)	1 : 1000 (cytoplasmic granular)	Negative
AMA (antimitochondrial antibody)	Positive	Negative
Anti-M2-3E (anti-mitochondrial antibody M2-3E subtype)	Positive	Negative
Anti-SP-100 (anti-Sp100 antibody)	Positive	Negative
Anti-GP-210 (anti-gp210 antibody)	Positive	Negative

staining (40 $\times$ ), consistent with Sjögren's disease (SJD).

**Liver biopsy** (Figure 4): Chronic hepatitis with features of PBC (stage 1).

**Right medial thigh muscle biopsy** (Figure 5): Scattered necrosis, regenerating fibers with phagocytosis; upregulation of MHC-I expression in muscle fibers; complement deposition on membranes of some non-necrotic fibers, consistent with IMNM.

**Immunofluorescence** (Figure 6): Anti-GP210 antibody immunofluorescence staining (40 $\times$ , 100 $\times$ ) in muscle and liver tissues.

#### Diagnosis and Treatment

According to the diagnostic criteria for PBC established by the Asia-Pacific Association for

the Study of the Liver,<sup>1</sup> the patient met the criteria for PBC. Based on the 1975 Bohan/Peter criteria for polymyositis (PM)/dermatomyositis,<sup>2</sup> she fulfilled the diagnostic criteria for PM. Furthermore, according to the 2017 European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for IIM,<sup>3</sup> her score (6.7-8.7) fell within the "probable IIM" range. Combined with pathological findings of scattered necrosis and membrane attack complex (MAC) deposition, the diagnosis was refined to IMNM, representing a more precise myositis subtype classification.

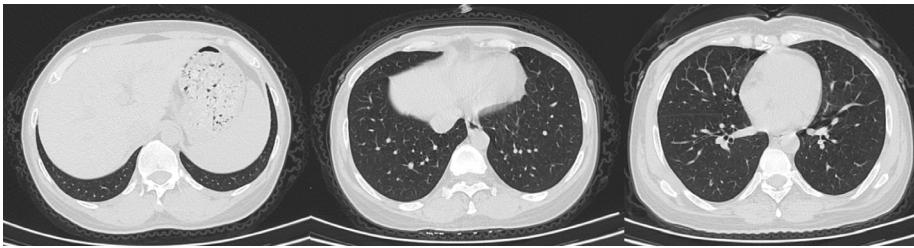
In addition, labial gland biopsy (Figure 3) revealed lymphocytic foci (>1 focus/4 mm<sup>2</sup>), consistent with SJD, along with anti-SSA antibody positivity. However, according to the 2016 ACR/EULAR classification criteria for SJD,<sup>4</sup>

**Table 2. Tumor Markers**

Parameters	Result	Reference Range
Alpha-fetoprotein (AFP)	2.73	0.00-20.00 ng/mL
Carcinoembryonic Antigen (CEA)	0.79	0-5.00 ng/mL
Ferritin	166.5	4.6-204.0 ng/mL
Squamous Cell Carcinoma Antigen (SCC)	0.45	0-1.50 ng/mL
Carbohydrate Antigen 199 (CA19-9)	10.29	0-37.00 U/mL
Carbohydrate Antigen 125 (CA125)	11.54	0-35.00 U/mL
Carbohydrate Antigen 153 (CA15-3)	11.55	0-31.30 U/mL
Carbohydrate Antigen 50 (CA50)	12.52	0-30.00 IU/mL
Carbohydrate Antigen 242 (CA242)	7.55	0-20.00 U/mL
Carbohydrate Antigen 72-4 (CA72-4)	0.23	0-6.90 U/mL
Cytokeratin 19 Fragment (CYFRA 21-1)	0.53	0-3.30 ng/mL
Euron-Specific Enolase (NSE)	15.85	0-17.77 ng/mL

#### Highlights

- Rare case of primary biliary cholangitis (PBC) presenting with low back pain (LBP) and complicated by immune-mediated necrotizing myositis (IMNM).
- Anti-mitochondrial antibody subtype M2 positivity detected, with paraspinal muscle involvement confirmed by magnetic resonance imaging and biopsy.
- Anti-GP210 expression in muscle and liver suggests a potential role in multisystem autoimmune damage.
- Clinicians should consider systemic autoimmune diseases in atypical LBP presentations.



**Figure 1.** Chest CT showed no interstitial changes or other abnormalities.

she did not meet the inclusion threshold, as she reported only mild xerostomia and xerophthalmia without objective evidence (Schirmer's test was not performed, as symptoms were self-reported).

The final diagnosis, integrating clinical presentation, laboratory findings, and histopathology, was PBC complicated by IMNM.

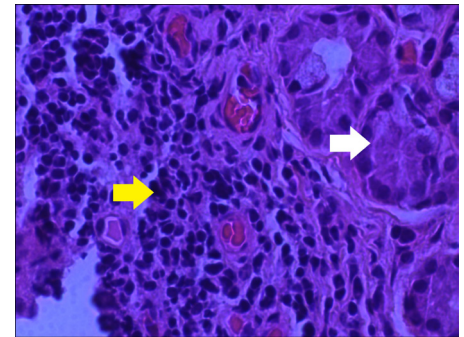
To alleviate potential SS-related sicca symptoms and systemic inflammation, hydroxychloroquine (100 mg twice daily) was initiated. Due to poor tolerance of higher doses and the patient's relatively low body weight (50 kg), along with a favorable therapeutic response, a reduced dose of mycophenolate mofetil (500 mg twice daily) was used. The treatment regimen comprised ursodeoxycholic acid (250 mg twice daily), methylprednisolone (40 mg once daily), hydroxychloroquine (100 mg twice daily), and mycophenolate mofetil (500 mg twice daily). After two weeks, her symptoms improved, and she was discharged on

prednisone (30 mg once daily), hydroxychloroquine (100 mg twice daily), and mycophenolate mofetil (500 mg twice daily).

The corticosteroid dose was gradually tapered, with regular follow-up. By April 2025 (15 months), liver enzyme levels had nearly normalized, creatine kinase-MB (CK-MB) and LDH levels were within normal limits, and low back pain (LBP) had resolved. She reported no chest discomfort, fatigue, myalgia, or muscle tenderness. Muscle strength was grade 5 in all limbs and neck flexors, with no muscle atrophy. Cardiac rhythm was regular, lung auscultation was clear, and neurological examination was unremarkable. The patient had resumed normal daily activities and work.

### Discussion

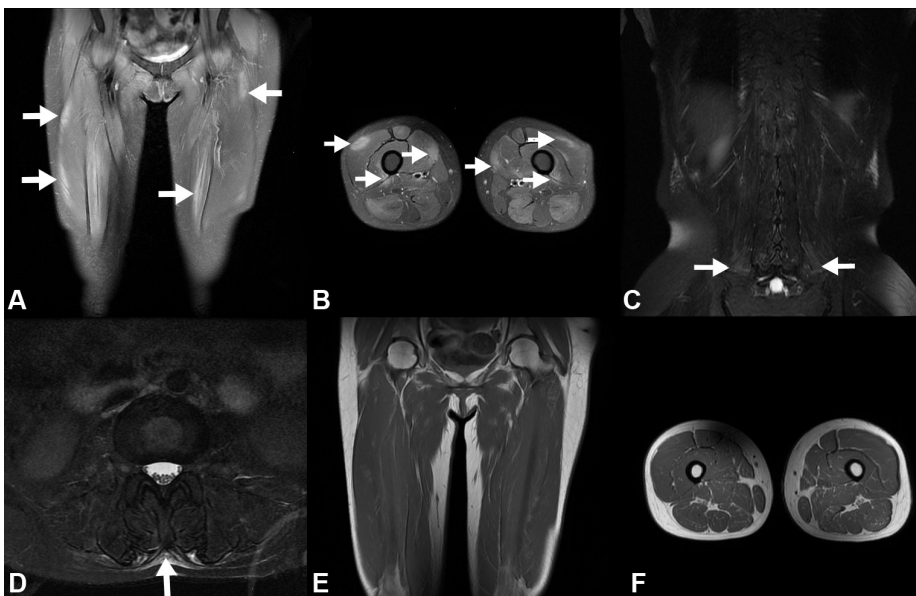
Low back pain (LBP) is most commonly caused by mechanical factors (e.g., lumbar strain, intervertebral disc herniation) or degenerative conditions (e.g., osteoarthritis) and can usually be identified readily through history-taking



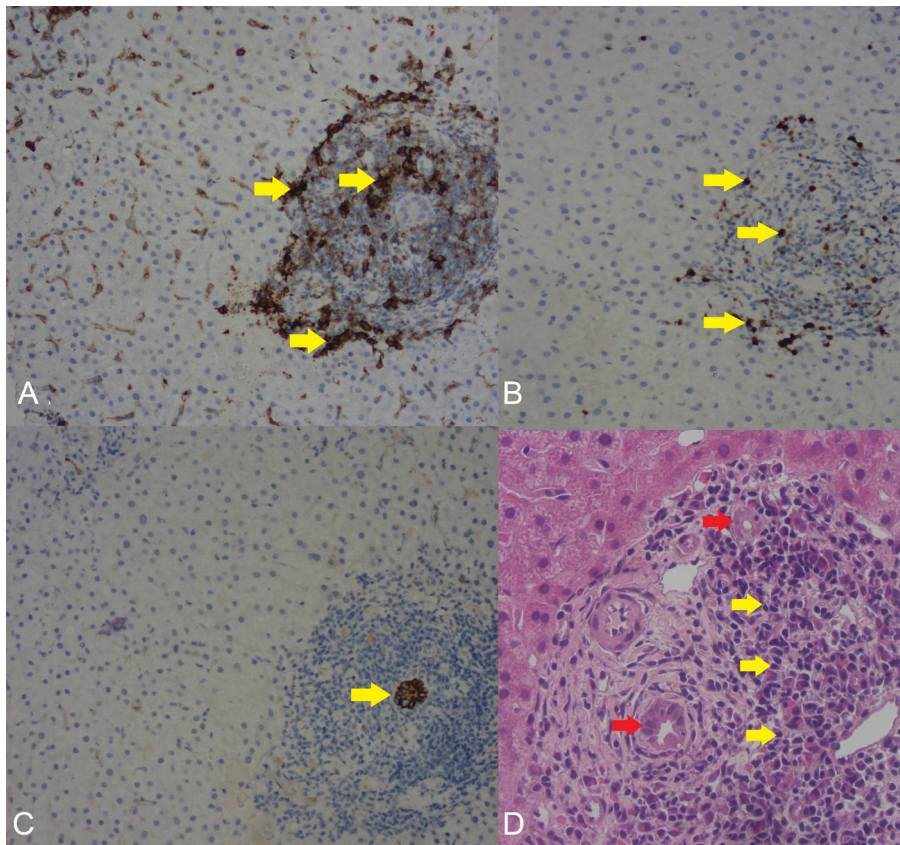
**Figure 3.** HE of labial gland biopsy (40X magnification): Interstitial lymphocytic and plasma cell infiltration with lymphoid foci (>1 focus/4 mm<sup>2</sup>, yellow arrows); normal acinar structures (white arrows). Note: HE, hematoxylin-eosin staining.

and imaging. Rare diseases presenting initially with LBP are easily overlooked. In the present case, PBC complicated by IMNM manifested with LBP as the first symptom. The reported prevalence of IIM in PBC patients is approximately 1.4%–7.4%,<sup>5</sup> with proximal muscle weakness being the most common initial symptom (60%–80%).<sup>6,7</sup> Paraspinal muscle involvement is relatively uncommon. In this patient, MRI demonstrated hyperintense signals in the erector spinae fascia, suggesting that paraspinal myositis was the cause of LBP. Uenaka et al. reported that in anti-mitochondrial antibody subtype M2 (AMA-M2)-positive myositis, the quadriceps muscles may be preserved while the paraspinal muscles exhibit severe atrophy,<sup>8</sup> indicating that paraspinal involvement may be a distinguishing feature. The present case showed similar characteristics, supporting the possibility that AMA-M2 positivity may serve as a pathogenic antibody in IMNM.

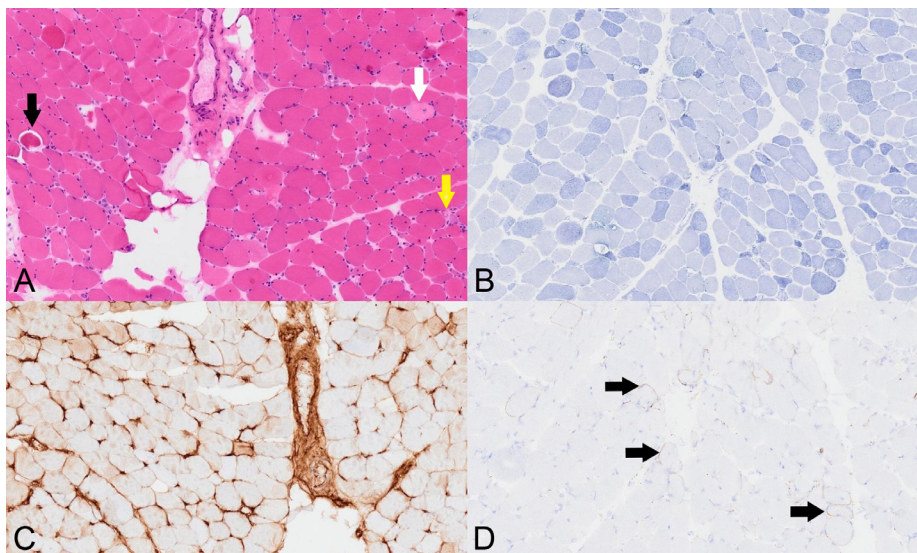
Cardiac involvement is another recognized feature of AMA-M2-positive myositis, with a reported prevalence of 28%–71%.<sup>9</sup> The patterns of involvement are diverse, including arrhythmias and conduction blocks. Studies have suggested that myocardial involvement often precedes muscle weakness, is associated with a prolonged disease course (average >5 years), and is more frequently observed in patients with PBC overlap syndromes.<sup>10–12</sup> In the patient, despite the coexistence of PBC, muscle weakness was mild, and there were no symptoms of chest tightness, dyspnea, or palpitations. Both echocardiography and electrocardiography were normal. Although CK-MB and LDH levels were elevated, troponin I remained normal, and the CK-MB/CK ratio was 0.024, indicating that the elevated creatine kinase likely originated from skeletal muscle



**Figure 2.** (A, B) Coronal and axial fat-suppressed T2-weighted (short-tau inversion recovery, STIR) images showing multifocal patchy hyperintense signals in bilateral thigh muscles (arrows), with partial distribution along tendons; (C, D) Coronal and axial fat-suppressed T2-weighted images revealing hyperintense signals in paraspinal muscle fascia (arrows); (E, F) T1-weighted images showing no significant abnormalities.



**Figure 4.** (A) CD38 staining (40x magnification) showing positive plasma cells (arrows); (B) MUM1 staining (20x magnification) indicating positive plasma cells (arrows); (C) CK7 staining (40x magnification) demonstrating bile duct injury (arrows); (D) Hematoxylin-eosin (HE) staining (40x magnification) revealing lymphocytic and plasma cell infiltration in the portal area (yellow arrows) and bile duct epithelial degeneration (red arrows). CD38, ADP-ribosyl cyclase; CK7, cytokeratin 7; MUM1, multiple myeloma oncogene 1.

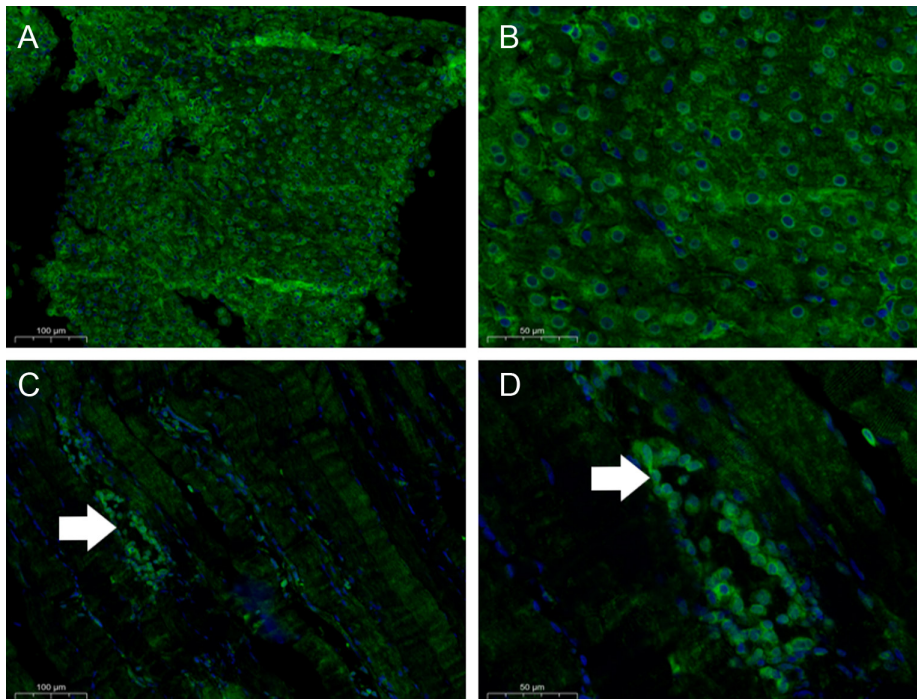


**Figure 5.** (A) Hematoxylin-eosin (HE) staining (40x magnification) of muscle fiber cross-sections showing mild variation in fiber size and irregular morphology (black arrows), scattered necrotic and regenerating fibers with phagocytosis (yellow arrows), and mild increase in internalized nuclei (white arrows); (B) NADH staining (40x magnification) indicating normal myofibrillar architecture except in necrotic fibers; (C) Major histocompatibility complex class I (MHC-I) immunohistochemistry (40x magnification) demonstrating diffuse upregulation of MHC-I expression in muscle fibers; (D) Immunohistochemistry (40x magnification) showing membrane attack complex (MAC) deposition on non-necrotic fiber membranes (arrows). MAC, membrane attack complex; MHC-I, major histocompatibility complex class I; NADH, nicotinamide adenine dinucleotide staining.

rather than myocardium. At present, there is no evidence of myocardial involvement.

Unlike previously reported IMNM cases that frequently involve pulmonary complications such as interstitial lung disease,<sup>13</sup> this patient's chest CT was unremarkable (Figure 1), indicating that the presentation was dominated by musculoskeletal symptoms. Previous studies have noted that seronegative IMNM is associated with an increased risk of malignancy.<sup>13,14</sup> In the present case, both anti-HMGCR and anti-SRP antibodies were negative, and comprehensive tumor marker screening (Table 2) was also negative, thereby ruling out malignancy. This may be related to the relatively short disease duration (~24 months) and reinforces the marked heterogeneity of AMA-positive myositis.

Anti-AMA-M2 is a specific antibody for PBC, with a detection rate of 70%-85% in PBC-IIM overlap.<sup>15</sup> It serves as a serological marker for both diagnosis and assessment of disease activity in PBC.<sup>16</sup> Recently, AMA has been recognized as a novel myositis-associated autoantibody, with AMA-M2 positivity reported



**Figure 6.** (A, B) Liver tissue (40x and 100x magnification) showing diffuse anti-GP210 antibody expression; (C, D) Muscle tissue (40x and 100x magnification) demonstrating focal anti-GP210 antibody infiltration (arrows).

in approximately 2.5%-19.5% of autoimmune myopathy patients.<sup>6</sup> The AMA-M2–positive myopathies are predominantly IMNM and PM, with IMNM accounting for 50%-60%,<sup>15</sup> consistent with the present case. Notably, immunofluorescence staining revealed anti-GP210 expression not only in the liver but also in muscle tissue. The GP210, a nuclear envelope antibody, is positive in 20%-30% of PBC patients and is associated with disease progression and liver failure. Anoun et al<sup>5</sup> reported that GP210 antibodies may contribute to multisystem immune damage in complex autoimmune syndromes. Whether GP210 antibodies mediate or participate in the pathogenesis of IMNM remains speculative and warrants further investigation.

The PBC is a chronic autoimmune liver disease in which the central mechanism involves AMA-M2–mediated small bile duct epithelial injury, leading to dysregulated Th1/Th17 immune responses and release of inflammatory cytokines (e.g., IL-6, TNF- $\alpha$ ).<sup>17</sup> This immune dysregulation can extend systemically, affecting other organs.<sup>18</sup> The IMNM is pathologically characterized by muscle fiber necrosis and complement activation, with the MAC, C5b-9 playing a pivotal role in muscle injury.<sup>13</sup> It is plausible that systemic inflammation in PBC may amplify the pathological processes of IMNM via complement activation, although the precise mechanisms require further study.

The LBP is a common clinical symptom, but approximately 5% of cases are attributable to systemic diseases. When LBP presents in association with rare disorders, clinicians should maintain a high index of suspicion to ensure timely diagnosis.

**Data Availability Statement:** The data that support the findings of this study are available on request from the corresponding author.

**Ethics Committee Approval:** Ethical committee approval was received from the Ethics Committee of Jiaying Second Hospital (Approval No.: 2024-CA-65; Date: January 10, 2025).

**Informed Consent:** Written informed consent was obtained from the patient who agreed to take part in the study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept – L.Z., Q.Y.; Design – L.Z., T.Y., Q.Y.; Supervision – Q.Y.; Resources – L.Z., Q.Y.; Materials – L.Z., T.Y.; Data Collection and/or Processing – L.Z., T.Y.; Analysis and/or Interpretation – L.Z., Q.Y.; Literature Search – L.Z., T.Y.; Writing – L.Z.; Critical Review – T.Y., Q.Y.

**Declaration of Interests:** The authors have no conflicts of interest to declare.

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