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Primary Biliary Cholangitis and Superficial Pemphigus: A Case Report

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Abstract **Case Report**

Primary biliary cholangitis (PBC), a chronic autoimmune liver disease, is frequently associated with other autoimmune disorders such as Sjögren's syndrome and thyroid diseases. However, its coexistence with superficial pemphigus is extremely rare, with no prior cases reported in the literature. We present a case of a 49-year-old woman diagnosed with PBC who developed superficial pemphigus during the course of her disease, treated with corticoids followed by RITUXIMAB as background treatment with good clinical evolution. This case emphasizes the importance of considering rare autoimmune associations in PBC and highlights the need for coordinated care between hepatology and other specialists.

Keywords: Primary Biliary Cholangitis, Superficial Pemphigus, Autoimmune Disease, Rare, Case report.

INTRODUCTION

Formerly known as primary biliary cirrhosis (PBC), primary biliary cholangitis is the leading cause of intrahepatic cholestasis. This condition predominantly affects women, accounting for approximately 90% of cases [1], with a typical age of diagnosis around 55 years. It is frequently associated with various autoimmune diseases such as Sjögren's syndrome, thyroid disorders, celiac disease, or, more rarely, lupus. However, the association between primary biliary cholangitis and superficial pemphigus is extremely rare, with no cases reported in the literature to date.

PATIENT AND OBSERVATION

Patient Information: We present the case of a 49-year-old woman who sought medical attention for chronic itching, fatigue, and progressive weight loss over a period of one year.

Clinical findings: clinical examination revealed conjunctival jaundice, scratching lesions, and irregular hepatomegaly.

Diagnostic Assessment

Laboratory investigations showed pancytopenia (hemoglobin at 11 g/dL, white blood cells at 3,610, platelets at 109,000), hypoalbuminemia at 17 g/L, hepatic cytolysis (elevated transaminases five times the normal level), and icteric cholestasis (alkaline phosphatase and GGT six times the normal level). Hepatocellular insufficiency was demonstrated (prothrombin time at 50%, hypoalbuminemia, and hypocholesterolemia). Immunological testing revealed positive antinuclear antibodies (ANA), positive anti-mitochondrial antibodies of type M2. Abdominal ultrasound showed a liver with chronic liver disease signs, portal hypertension, and moderate ascites. Liver biopsy showed hepatic fibrosis estimated at F4. The diagnosis of PBC was established at the decompensated cirrhosis stage. Tests for other associated autoimmune diseases were negative (anti-transglutaminase antibodies -, normal TSH, anti-TPO antibodies -).

Therapeutic Intervention

The patient was treated with ursodeoxycholic acid (UDCA) at 13 mg/kg/day, diuretics (Lasilix 40 mg/day + Aldactone 75 mg/day), and beta-blockers. Clinical and biological follow-up showed significant improvement with a reduction in alkaline phosphatase levels to less than 3 times the upper limit of normal and normal transaminases levels according to PARIS criteria which is predictive of a good prognosis.

Follow-up and Outcomes

After eight months, the patient developed generalized bullous dermatosis with pruritic post-bullous erosions, and a diagnosis of superficial pemphigus was made based on skin biopsy. She was treated with oral corticosteroids at 80 mg/day and topical therapy for one month, followed by gradual dose reduction and initiation of Rituximab in consultation with dermatologists as a maintenance therapy. Under RITUXIMAB treatment, we observed a disappearance of dermatological lesions after a few weeks. Symptoms such as fatigue and pruritus also improved. Liver test levels remaining stable.

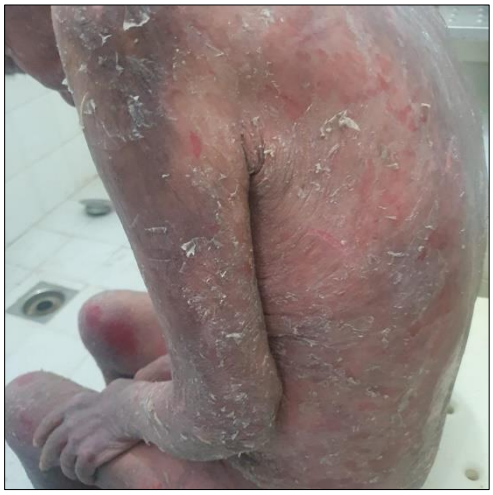

Figure 1: Generalized bullous dermatosis and post-bullous

DISCUSSION

Primary biliary cholangitis (PBC) is a chronic autoimmune disease characterized by the progressive destruction of intrahepatic bile ducts, leading to intrahepatic cholestasis and ultimately cirrhosis. The prevalence of PBC varies significantly by geographic region, with rates ranging from 10 to 40 per 100,000 inhabitants in Western countries [1]. This disease primarily affects middle-aged women, with a median age of diagnosis around 55 years [2]. The pathogenesis of PBC is strongly associated with other autoimmune diseases. About 30 to 70% of PBC patients have comorbid autoimmune conditions such as Sjögren's syndrome, autoimmune hepatitis, thyroid disorders, or celiac disease [3]. PBC is also associated with specific antibodies, particularly anti-mitochondrial antibodies of type M2, which are important diagnostic markers [4].

Superficial pemphigus, also known as pemphigus foliaceus, is a rare autoimmune disease characterized by bullous skin lesions caused by antibodies directed against desmosomes, the structures that maintain epidermal integrity [5]. This condition differs from pemphigus vulgaris by its more superficial impact on the skin and a tendency to present with less deep lesions. The autoantibodies present in superficial pemphigus target desmosomal cadherins, leading to blister formation and skin erosions.

The association between PBC and superficial pemphigus is extremely rare. No previous study has detailed this specific association. Reported cases of PBC coexisting with other autoimmune diseases primarily focus on conditions such as Sjögren's syndrome or autoimmune hepatitis [6, 7]. However, observations in case reports suggest that PBC can coexist with various rare autoimmune conditions, indicating that unexpected associations may occur. The clinical presentation of our patient highlights the importance of considering rare differential diagnoses when an autoimmune condition is diagnosed. The onset of superficial pemphigus after the diagnosis of PBC could be due to common underlying immunological mechanisms or treatment side effects [8]. It is also possible that the initiation of immunosuppressive therapy for PBC may have altered the immune balance, promoting the emergence of superficial pemphigus. Although the pathophysiological mechanisms are not yet fully elucidated, the association between primary biliary cholangitis (PBC) and superficial pemphigus can be explained by a common immune dysregulation. PBC, an autoimmune disease that leads to the destruction of the bile ducts, is often associated with the production of autoantibodies, which trigger inappropriate immune responses. Genetic and environmental factors may also increase susceptibility to other autoimmune diseases. Furthermore, the immunosuppressive treatment of PBC can disrupt immune balance, potentially allowing the emergence of manifestations such as superficial pemphigus, where autoantibodies target desmosomes, resulting in skin lesions.

The treatment of PBC primarily aims to slow disease progression, improve symptoms, and prevent complications. The first-line treatment is ursodeoxycholic acid (UDCA), recommended for its choloretic and cytoprotective effects on the liver. UDCA can improve liver tests, reduce the progression of liver fibrosis, and improve survival [9]. In cases of

progression despite UDCA therapy, immunosuppressants such as corticosteroids may be considered, although their use is generally reserved for severe cases or those associated with autoimmune hepatitis [10].

On the other hand superficial pemphigus is generally treated with systemic and topical corticosteroids to control inflammation and skin eruptions. High-dose oral corticosteroids are often required initially, followed by gradual dose reduction. In cases of inadequate response or relapse, additional immunosuppressive treatments such as rituximab, an anti-CD20 monoclonal antibody, may be used to induce prolonged remission and reduce the need for corticosteroids. Other immunosuppressive agents, such as azathioprine or mycophenolate mofetil, may also be considered based on individual treatment response**.** In our case, corticosteroid therapy was not required to treat the liver disease, but was used as a first-line treatment for the dermatological involvement, pending pathological confirmation. The result was a spectacular improvement in dermatological lesions, an improvement in asthenia and pruritus, with a stable liver test rate already obtained after initiation of AUDC.

Treatment of PBC and superficial pemphigus may sometimes overlap, particularly in the use of corticosteroids. However, corticosteroid use in PBC is limited and generally reserved for severe forms with coexisting autoimmune hepatitis, whereas in superficial pemphigus, corticosteroids play a central role in controlling symptoms. Rituximab, used for refractory superficial pemphigus, is not a standard treatment for PBC but could be considered in very specific cases requiring more aggressive immunosuppressive therapy. In our patient, the use of rituximab proved necessary as background therapy to control the dermatological disease and improve prognosis. Its introduction was decided in consultation with the gastroenterology team, after eliminating any contraindications to immunosuppressive treatment. The introduction of RITUXIMAB enabled us to maintain the remission of superficial Pemphigus already obtained on corticoids without any real change in the course of liver disease. In our case, the association of primary biliary cholangitis (PBC) and superficial pemphigus does not significantly influence the prognosis or severity of either disease. PBC, although at an advanced stage of decompensated cirrhosis, responded favorably to treatment with ursodeoxycholic acid, leading to an improvement in liver function tests. Meanwhile, superficial pemphigus, while requiring treatment with corticosteroids and Rituximab, also showed a rapid regression of skin lesions. Thus, each disease was managed independently, without mutual impact on their severity or prognosis.

The management of the patient required coordination between hepatology and dermatology specialists to optimize PBC treatment while effectively addressing superficial pemphigus, illustrating the complexity of care needed for patients with multiple and rare manifestations. This observation enhances the understanding of rare autoimmune disease associations and underscores the importance of ongoing clinical vigilance.

Patient Perspective: The patient expressed satisfaction with the care received and remained positive throughout the treatment. Despite the challenges of the condition, they stayed optimistic about their recovery.

Patient´s Consent: informed consent was obtained from the client for us to use the pictures.

CONCLUSION

The association of primary biliary cholangitis (PBC) with superficial pemphigus is extremely rare and previously unreported. This case emphasizes the need for careful monitoring of PBC patients for unusual skin conditions. Clinicians must be vigilant in identifying rare autoimmune associations to provide optimal care. Treatments for PBC and superficial pemphigus involve corticosteroids, but therapy must be personalized to each condition. Effective management requires coordination between hepatology and dermatology specialists.

Competing Interests: The authors declare no competing interests

Authors' Contributions

All authors contributed equally to the conception, design, data collection, analysis, and interpretation of the case. They all participated in the writing, revising, and final approval of the manuscript.

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