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Dermatomyositis Associated with Primary Biliary Cholangitis: A Case Report

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Abstract **Case Report**

Primary biliary cholangitis (PBC) is a chronic autoimmune cholestatic liver disease characterized by progressive destruction of the small intrahepatic bile ducts and the presence of antimitochondrial antibodies. Its association with dermatomyositis is rare. We report a case of dermatomyositis associated with PBC and briefly review the literature.

Keywords: Dermatomyositis; primary biliary cholangitis; cholestasis; autoimmunity.

INTRODUCTION

Primary biliary cholangitis (PBC) is a chronic autoimmune cholestatic liver disease characterized by progressive immune-mediated destruction of the small intrahepatic bile ducts, which may lead to fibrosis, cirrhosis, and portal hypertension [1,3,5]. PBC can be associated with other autoimmune diseases; however, its association with dermatomyositis is rare [3]. We report a case illustrating this uncommon association.

CASE REPORT

A 65-year-old woman with no significant past medical history was admitted to our department for a two-week history of facial erythema and edema associated with deterioration of her general condition and unusual asthenia.

Clinical examination revealed bilateral violaceous periorbital erythema associated with eyelid edema, consistent with a heliotrope rash, as well as Gottron's sign and elbow ulceration.

The diagnosis of dermatomyositis was supported by these clinical findings, elevated serum creatine phosphokinase (CPK) levels, a myopathic pattern on neurophysiological examination, and interface dermatitis on skin biopsy. Muscle biopsy findings were inconclusive. The paraneoplastic workup was negative.

Systemic corticosteroid therapy was initiated at a dose of 1 mg/kg/day. After one month of treatment, a marked increase in gamma-glutamyl transferase (GGT) levels was observed, reaching 27 times the upper limit of normal. Viral serologies were negative. Liver ultrasonography showed hepatic steatosis, while magnetic resonance cholangiopancreatography (MRCP) was normal. Immunological testing revealed positive M2-type antimitochondrial antibodies.

Based on the cholestatic liver abnormalities and positive antimitochondrial antibodies, a diagnosis of primary biliary cholangitis was made. Treatment with ursodeoxycholic acid (UDCA) at a dose of 600 mg/day in three divided doses was initiated.

The patient subsequently developed corticosteroid-induced diabetes. Three months later, she died following an episode of gastrointestinal bleeding.

DISCUSSION

Primary biliary cholangitis is a chronic autoimmune cholestatic liver disease that predominantly affects women [1,3]. Its pathophysiology involves a genetic predisposition combined with environmental and immunological factors [2].

The diagnosis of PBC is generally based on the presence of at least two of three criteria: biochemical evidence of cholestasis, particularly elevated alkaline phosphatase levels; the presence of antimitochondrial antibodies; and characteristic histological findings of nonsuppurative destructive cholangitis affecting the

small intrahepatic bile ducts [1,3,5]. Liver biopsy is not always required for diagnosis but may be useful in selected cases and for disease staging [1,3,5].

PBC can be associated with several autoimmune disorders, particularly Sjögren's syndrome, systemic sclerosis, rheumatoid arthritis, and other autoimmune diseases [3]. However, the association between PBC and dermatomyositis is uncommon.

Dermatomyositis is an autoimmune inflammatory myopathy characterized by muscle involvement and typical cutaneous manifestations, including heliotrope rash and Gottron's papules or signs. Liver enzyme abnormalities in patients with dermatomyositis may have several causes, including muscle involvement, drug toxicity, infection, or associated liver disease. Therefore, persistent cholestatic abnormalities should prompt further investigation.

The coexistence of dermatomyositis and PBC may reflect a shared autoimmune background. In our patient, the presence of marked cholestatic abnormalities associated with M2-type antimitochondrial antibodies led to the diagnosis of PBC. This observation highlights the importance of considering an associated autoimmune liver disease when unexplained or persistent cholestasis occurs in a patient with dermatomyositis.

Ursodeoxycholic acid remains a first-line treatment for PBC [1,4,5]. In advanced disease, management of complications of cirrhosis and portal hypertension is required, and liver transplantation may be considered in selected patients [4,5].

CONCLUSION

This case highlights the rare association between dermatomyositis and primary biliary cholangitis. In patients with dermatomyositis presenting with persistent or unexplained cholestatic liver abnormalities, an associated autoimmune liver disease should be considered. Testing for antimitochondrial antibodies may contribute to the diagnosis of PBC.

Conflicts of interest

The authors declare no conflicts of interest.

Author contributions

Abdourahman Moussa, Mohamed El Amraoui, Hamda Hassan Iltireh, and Rahma Ali Malow contributed to the preparation of the clinical case and the discussion. Rachid Frikh and Naoufal Hjira critically revised the manuscript and approved the final version.

Patient consent

Written informed consent was obtained for the publication of this case report and the accompanying clinical image.

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