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Primary Biliary Cholangitis and Rheumatoid Arthritis: An Emerging Association

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Abbreviations: PBC: Primary Biliary Cholangitis, RA: Rheumatoid Arthritis

Introduction

Primary Biliary Cholangitis (PBC), formerly known as primary biliary cirrhosis, is an autoimmune-related cholestatic hepatopathy, representing a common cause of intrahepatic cholestasis due to progressive bile duct destruction. Often, PBC is linked with other autoimmune disorders such as Sjogren's syndrome, thyroid dysfunction, scleroderma, and systemic lupus erythematosus. However, the association of PBC with rheumatoid arthritis (RA) is rare, estimated at 1.8 to 5.6%. We present a clinical case of PBC discovered in a patient with deforming rheumatoid arthritis.

Observation

A 40-year-old female undergoing treatment for seropositive deforming rheumatoid arthritis based on ACR EULAR 2010 criteria for five years-initiated methotrexate (15mg weekly) and low-dose corticosteroids. Subsequently, she developed abdominal distension without additional clinical signs. Imaging revealed dysmorphic hepatomegaly with chronic Budd-Chiari syndrome and low-grade ascites. Viral serologies were negative. Hepatic work-up demonstrated

biological cholestasis (PAL at 375 IU/l and gamma-GT at 202 IU/l) without cytolysis. Immunological work-up revealed positive nuclear appearance anti-mitochondrial and anti-Sp100 antibodies (1280 NAA). Treatment with ursodeoxycholic acid (12mg/kg) was initiated, resulting in positive clinical and biological progress.

Discussion

The coexistence of Primary Biliary Cholangitis (PBC) and Rheumatoid Arthritis (RA) raises intriguing questions about the intricate interplay of autoimmune processes within the body. PBC, once considered a distinct liver disorder, is increasingly recognized for its association with various autoimmune conditions. While PBC is commonly linked with diseases such as Sjogren's syndrome and scleroderma, the convergence with RA remains a rare phenomenon.

The observed prevalence of PBC in RA patients, estimated between 1.8 to 5.6%, underscores the importance of considering hepatic involvement in individuals with autoimmune rheumatic diseases. In our presented case, the patient with seropositive deforming RA developed clinical

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manifestations of PBC during the course of RA management. The immunological work-up revealed the presence of antimitochondrial and anti-Sp100 antibodies, typical markers associated with PBC. The initiation of ursodeoxycholic acid treatment resulted in favorable clinical and biological responses, emphasizing the importance of early detection and intervention in managing autoimmune hepatopathies associated with RA.

It is noteworthy that the current understanding of RA-related hepatopathies is evolving. While liver involvement is not typically regarded as a frequent extra-articular manifestation of RA, this case illustrates the potential for disturbances in liver function during the course of rheumatoid arthritis. This prompts clinicians to maintain a high index of suspicion for concurrent autoimmune liver diseases, especially in the presence of unexplained hepatic abnormalities.

The debate surrounding the iatrogenicity of rheumatoid arthritis treatments, particularly methotrexate, adds complexity to the management of patients with overlapping PBC. Nonetheless, recent studies have suggested a beneficial role for methotrexate when combined with ursodeoxycholic acid in PBC, demonstrating improved outcomes compared to monotherapy.

In conclusion, this case highlights the need for heightened awareness among clinicians regarding the association of PBC with RA. A multidisciplinary approach involving rheumatologists and hepatologists is crucial for timely diagnosis and effective management, ensuring optimal outcomes for patients navigating the complexities of autoimmune liver diseases in the context of rheumatoid arthritis.

Conclusion

While hepatic involvement is not a common extra-articular manifestation of rheumatoid arthritis, liver disturbances may occur. It is crucial to consider that such disturbances might indicate an associated autoimmune hepatopathy, specifically primary biliary cholangitis. The potential iatrogenicity of rheumatoid arthritis treatments, including methotrexate, has been discussed. However, recent studies combining methotrexate with ursodeoxycholic acid in PBC patients demonstrated satisfactory results, showing improved effectiveness compared to ursodeoxycholic acid alone in terms of survival before liver transplantation.