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CASE REPORT

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The Disappearance of a Hepatic Mass in Anti-Synthetase Syndrome

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Abstract

Anti-Synthetase Syndrome (ASyS) is a rare chronic autoimmune disorder characterized by myositis, interstitial lung disease (ILD), polyarthralgia, "mechanic's hands" and Raynaud's phenomenon. Liver lesions are quite rare in ASyS. In our ASyS case, we will discuss a 58-year-old man presenting with muscle weakness, arthralgia, and ILD. He was positive for anti-Jo-1 antibodies, substantiating the diagnosis, and was started on treatment. This was followed by the appearance of a liver mass that disappeared when the patient achieved remission.**(International Journal of Biomedicine. 2017;7(2):141-143.)**

Key Words: Anti-Synthetase Syndrome • anti-Jo-1 antibody • nodular regenerative hyperplasia • treatment

Abbreviations

ASyS, Anti-Synthetase Syndrome; **CT,** computed tomography; **ILD**, interstitial lung disease; **ILM**, idiopathic inflammatory myopathies (IIM); **MSA**, myositis-specific autoantibodies; **NRH**, nodular regenerative hyperplasia.

Introduction

Autoantibodies are a hallmark in the diagnosis of many systemic autoimmune rheumatic diseases, including idiopathic inflammatory myopathies (IIM).[1-3] A number of autoantibodies, called myositis-specific autoantibodies (MSA), have been described in IIM.[4,5] The presence of MSA is a key feature for diagnosis of clinically distinguishable IIM subsets (polymyositis (PM), dermatomyositis (DM), inclusion body myositis, and myositis associated with malignancy and other connective tissue diseases).

Among MSA, autoantibodies against aminoacyl-tRNA synthetases (ARS) were detected in 25%–35% of IIM patients.[6] The anti-Jo-1 antibody directed against the antihistidyl–tRNA synthetase is the most common of anti-ARS autoantibodies, predominantly found in 15%–30% of patients with polymyositis and in 60%–70% of those with ILD.[7] Anti-ARS autoantibodies, especially the anti-Jo-1 antibody, characterize their own clinical IIM phenotype, which has become known as Anti-Synthetase Syndrome (ASyS) and which is characterized by multiple organ involvement, primarily ILD, and is often accompanied by myositis, non-erosive arthritis, Raynaud's phenomenon, "mechanic's hands," skin rashes, and constitutional symptoms, such as fever.[8,9] With proper diagnosis and early initiation of therapy, ASyS is potentially treatable. In this text, we report the rare appearance and disappearance of a liver mass in ASyS as the patient's condition improved.

Case report

A 58-year-old male with a history of diabetes mellitus and asthma presents to the emergency room complaining of swelling and pain of the lower extremities, generalized weakness, and weight loss for about 2 months' duration. Upon further questioning, he agrees that he has joint pain, stiffness, shortness of breath, and decreased exercise tolerance. He has also noticed a bluish discoloration of the fingers in cold weather. His weakness was throughout his body, but worse in the arms, and was associated with numbness and tingling, which had prompted a work-up for cervical radiculopathy the month prior at the neurosurgeon's office, whereby electromyography showed left ulnar neuropathy. A physical exam found loss of grip strength, "mechanic's hands," lungs

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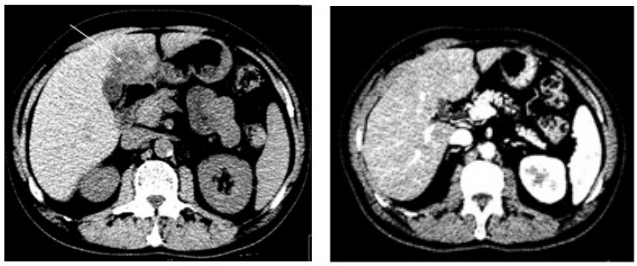
that are clear to auscultation, synovitis of the wrists bilaterally, tenderness and bony enlargement of the first, second, and third metacarpophalangeal joints, lower extremity edema, and proximal muscle weakness of three-fifths strength. Labs noted to be elevated are creatinine kinase, aldolase, and a positive result for anti-Jo-1 antibodies. Rheumatoid factor, anti-CCP, and ANA were negative, and liver function test unremarkable. A chest X-Ray revealed increased interstitial marking, warranting a chest CT, which showed patchy ground-glass areas in the lung bases suggestive of possible ILD. The patient was started on prednisone 60 mg and azathioprine 50 mg. A repeat chest CT 4 months after initiating medication showed resolution of ground-glass opacities, but noted a hypodense hepatic mass. A biopsy was offered but declined by the patient. Gradually, prednisone was tapered and azathioprine up titrated to 150 mg daily. Five months later, a follow-up abdominal CT showed resolution of the hepatic mass (Fig.1). The patient's disease is currently in remission, with resolution of symptoms at 2.5 mg prednisone daily and 150 mg of azathioprine.



a) b)

Fig. 1. *Abdominal CT scans before (a) and after (b) treatment*

Case discussion

The true population prevalence of ASyS is unknown. Several retrospective studies have found that the reported annual incidence of IIM has been 2 to 10 new cases per million adults per year and that ASS antibodies were detected in 20% to 40% of such cases. Due to the high association with malignancies, patients should be screened for age- and gender-appropriate malignancies.[10] Diagnosis is based on the constellation of symptoms, autoantibodies, and muscle histopathology. The use of electromyography and magnetic resonance imaging are controversial as they are costly and lack sensitivity and specificity.[1] Respiratory symptoms are by far the most commonly seen within the disease, where at least 60% of patients have such symptoms on presentation. Therefore, it is common that those patients presenting with unexplained ILD, should have ASyS in the differential diagnosis.[1,10] Those not presenting with respiratory symptoms should have pulmonary function tests, and a thoracic high-resolution CT. High-dose corticosteroids are the first-line treatment for ASyS.[11] Although corticosteroids are considered the mainstay of treatment, additional immunosuppressive agents, such as azathioprine and methotrexate, are often used, both as corticosteroid sparing agents and to achieve disease control. In those with severe or refractory disease, rituximab may be used. The long-term prognosis for people with ASyS varies based on the severity of the condition and symptoms present; although, with new therapy available, more cases are becoming chronic and requiring life-long therapy.[12]

In our case, we illustrate ASyS with liver involvement. Liver lesions are quite rare in ASyS. Upon review of the literature, we found that the most common liver abnormalities were abnormal liver function, chronic active hepatitis, and hepatomegaly. Few cases of liver conditions, such as nodular regenerative hyperplasia (NRH) or Budd-Chiari in ASyS, are reported. Our patient's lesion is likely NRH; however, it remains unconfirmed by biopsy. NRH of the liver is characterized by diffuse nodularity of the liver with little or no fibrosis.[13] It has been associated with autoimmune disease especially after drug treatment, notably azathioprine. NRH is one of the known causes of non-cirrhotic portal hypertension. Patients have presented with variceal bleeding and ascites. The natural history is unknown as there are limited data on the long-term prognosis and outcomes of patients with NRH. Experts believe that if NRH is identified, treatment should be directed at identifying an etiologic agent and removing it, if possible. Due to increased longevity of patients with ASyS, the physician should be aware of rare presentations, such as that of liver lesions and complications to monitor and follow.

In conclusion, ASyS is characterized by a variety of signs and symptoms leading to an array of presentations: myositis, ILD, polyarthralgia and mechanic's hands associated with anti-Jo1 antibodies. The physician should be vigilant when the patient is presenting with weakness, arthralgia, and shortness of breath. As more patients are achieving chronicity with the disease in remission, the physician should be aware of possible associated liver lesions and closely follow their evolution.

Competing interests

The authors declare that they have no competing interests.

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