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Letter to the Editor

# Antisynthetase syndrome and cardiac involvement: a rare association

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Antisynthetase syndrome (aSS) is an autoimmune myopathy characterized by presence of anti-tRNA synthetase antibodies [1]. Cardiac involvement is uncommon and only few cases have been previously reported. We reported here an aSS patient with myocarditis.

A 55-year-old woman was admitted in our unit for a 3-month history of polyarthritis, myalgia and dyspnea. Clinical exam revealed polyarthritis and proximal muscle weakness.

Blood tests revealed high serum creatine phosphokinase (CPK) at 6422 (20 N) U/L and T-troponin at 0.232 μg/L (5 N). Anti-Jo1 and anti-SSA antibodies were positive. Electrocardiogram was normal.

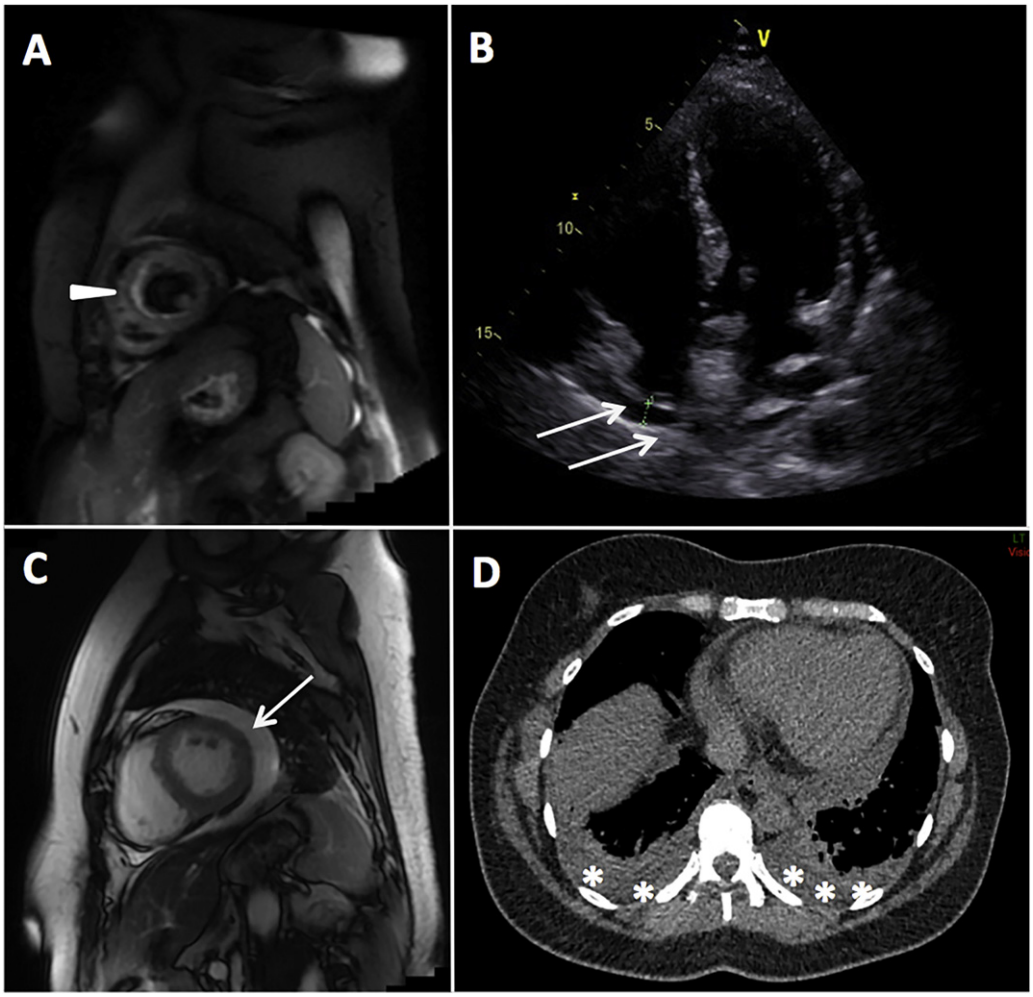


**Fig. 1.** Imaging features of the myopericarditis. A. MRI of heart showing hypersignal T2 with gadolinium-enhancement (arrowhead) of myocardium suggesting of myocarditis. B–C. Echocardiography and MRI with T1 weighted sequence-revealing pericarditis (arrows). D. CT-scan of lung in axial view showing bilateral pleural effusion (asterisk).

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Lung computed tomography scan found a mild interstitial lung disease associated with pleuropericardial effusion (Fig. 1). Echocardiography confirmed pericarditis. Magnetic resonance imaging (MRI) confirmed myocarditis with myocardium edema and diffuse gadolinium-enhancement (Fig. 1). Rituximab (1 g X 2) was started with prednisone (0.5 mg/kg/day) and methotrexate 20 mg/week. A rapid improvement of clinical symptoms and CPK and T-troponin levels were observed. One month later the patient had relapse of pericarditis confirmed by echocardiography without myocarditis (normal levels of T-troponin). Anakinra therapy was given for 7 days leading to a rapid relief of chest pain in a few days.

Myocarditis is a rare complication of aSS disease [2]. In a study of 352 aSS patients, a myocarditis was observed among only 12 (3.4%) patients [3]. Cardiac involvement was considered severe in 50% with hospitalization in intensive care unit. Similar to our patient, pericarditis is associated in 50% of patients with myocarditis and anti-Jo1 the most frequently observed antibody. Diagnosis of myocarditis is challenging, elevation of T-Troponin being non-specific and non-sensitive. Echocardiography is useful for differential diagnosis and for evaluating myocarditis complications. Endomyocardial biopsy stays the gold standard for myocarditis diagnosis but the procedure is invasive. The most reliable non-invasive procedure for the diagnosis seemed to be MRI with myocardium edema in T2-weighted sequences and early enhancement of gadolinium [4]. Thus, the association T-troponin and MRI is the most relevant non-invasive procedure to detect myocarditis. There is no consensus for the treatment of myocarditis. In our patient, the combo-therapy (corticosteroids, methotrexate and rituximab) was given without myocarditis relapse. In the literature corticosteroids, azathioprine, cyclophosphamide, rituximab, immunoglobulin and plasma exchange are the most widely used for the treatment of myocarditis associated with aSS [3,5].

Interestingly, the patient had a pericarditis successfully treated by anakinra. This is in accord with published data suggesting a role the interleukin-1 (IL-1) pathway in recurrent idiopathic pericarditis with efficacy of anakinra [6]. Furthermore, expression of interleukine-1 receptor in muscle fibers of patients with inflammatory myositis has been described [7], suggesting a role of IL-1 in myositis.

In conclusion, myocarditis in aSS is a rare condition and the diagnosis could be challenging. MRI and T-troponin are the most relevant procedure to diagnose myocarditis. The treatment is not codified but presence of myocarditis leads to intensify the treatment of such patients.

## Disclosure of interest

The authors declare that they have no competing interest.

## References

[1] Lega JC, Fabien N, Reynaud Q, et al. The clinical phenotype associated with myositis-specific and associated autoantibodies: a meta-analysis revisiting the so-called antisynthetase syndrome. Autoimmun Rev 2014;13:883–91.
[2] Imbert-Masseau A, Hamidou M, Agard C, et al. Antisynthetase syndrome. Three cases and a review of the literature. Ann Med Interne (Paris) 2003;154:483–8.
[3] Dieval C, Deligny C, Meyer A, et al. Myocarditis in patients with antisynthetase syndrome: prevalence, presentation, and outcomes. Medicine (Baltimore) 2015;94:798.
[4] Mavrogeni S, Bratis K, Karabela G, et al. Myocarditis during acute inflammatory myopathies: evaluation using clinical criteria and cardiac magnetic resonance imaging. Int J Cardiol 2013;164:3–4.
[5] Allanore Y, Vignaux O, Arnaud L, et al. Effects of corticosteroids and immunosuppressors on idiopathic inflammatory myopathy related myocarditis evaluated by magnetic resonance imaging. Ann Rheum Dis 2006;65:249–52.
[6] Jain S, Thongprayoon C, Espinosa RE, et al. Effectiveness and safety of anakinra for management of refractory pericarditis. Am J Cardiol 2015;116:1277–9.
[7] Grundtman C, Salomonsson S, Dorph C, et al. Immunolocalization of interleukin-1 receptors in the sarcolemma and nuclei of skeletal muscle in patients with idiopathic inflammatory myopathies. Arthritis Rheum 2007;56:674–87.

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