Severe Aortic Regurgitation Secondary to Antisynthetase Syndrome

Kerunne S. Ketlogetswe, MD; Joseph Aoki, MD; Thomas A. Traill, MD; Oscar H. Cingolani, MD

A 47-year-old woman with hypertension and a history of antisynthetase syndrome (ASS) diagnosed in 2002 (positive anti-PL-7 antibodies) and on long-term immunosuppressive therapy was admitted to the hospital in 2008 for worsening chest pain, shortness of breath, hypoxemia, and diffuse muscle weakness. Plain chest radiograph showed bilateral lower-lobe infiltrates and computed tomography confirmed bilateral patchy infiltrates consistent with interstitial lung disease. After being treated aggressively with high-dose immunosuppressive therapy, the patient improved and was discharged home. She was subsequently readmitted a few days later with progressive dyspnea. A transthoracic echocardiogram (TTE) showed moderate to severe aortic regurgitation (Figure 1A; online-only Data Supplement Movie IA and IB) that was not present in a prior TTE performed 4 months before admission (requested to screen for left ventricular hypertrophy; patient was asymptomatic at that time; Figure 1B and online-only Data Supplement Movie IIA and IIB). During the next 4 weeks, the regurgitation became more severe. Systolic pulmonary pressure was estimated at 70 mm Hg (Doppler). Her progressive dyspnea, together with increased left ventricular dimensions in a follow-up TTE prompted aortic valve replacement.

The native aortic valve cusps were described as diffusely edematous and buttery, with no vegetations (Figure 2A). Histopathology showed a mixed inflammatory infiltrate, with lymphocytes and neutrophils, and fragments with extensive acute and chronic inflammation with microabscesses (Figure 2B and C). Tissue culture for microorganisms as well as specific stains for fungi (Grocott Methenamine Silver), bacterial (Gram-Weigert), and acid-fast bacilli were all negative.

The patient was discharged home after recovering from surgery and was doing well 1 year after the operation on continuing immunosuppressive therapy. Attempts to reduce the immunosuppressive therapy (lowest tolerable dose being prednisone, 8 mg/d) resulted in muscle weakness.

As far as we know, there are no reported cases of ASS compromising cardiac valves. Because patients with ASS are prone to developing recurrent pulmonary infiltrates and hemorrhages and sometimes require invasive bronchoscopic procedures, it was decided to replace the native aortic valve with a bioprosthesis. The patient is being followed up...
periodically (every 6 months) at the Johns Hopkins cardiology outpatient center. So far, she has been asymptomatic, without dyspnea.

Antisynthetase syndrome is a connective tissue autoimmune disorder, classically characterized by interstitial lung disease, myositis, polyarthritis, fever, Raynaud phenomenon, and thick cracked skin on the fingers (mechanic’s hands) in the presence of an antisynthetase autoantibody, most commonly anti-Jo-1.\textsuperscript{1,2}

Other, less common antibodies in the antiaminoacyl-tRNA synthetase group include anti-PL-7 (the present case), anti-PL-12, anti-EJ, anti-OJ, anti-KS, anti-YRS, and anti-Zo.\textsuperscript{1} The aminoacyl-tRNA synthetases are cytoplasmic enzymes that catalyze the binding of amino acids to their corresponding tRNAs, and thus play a role in protein synthesis by recognizing a specific amino acid and enabling correct sequencing within the polypeptide chain. Autoantibodies against the various synthetases lead to idiopathic inflammatory myopathies. Each anti-aminoacyl-tRNA synthetase antibody is associated with a unique clinical syndrome and falls under the umbrella characterization of ASS.

The most common manifestation of ASS is interstitial lung disease, which occurs in >70\% of patients in most series.\textsuperscript{1,2} Approximately 5\% to 8\% of cases of ASS manifest as overlap syndromes with other connective tissue diseases, including rheumatoid arthritis, lupus, scleroderma, and Sjögren syndrome.\textsuperscript{1} Cardiac involvement in the idiopathic inflammatory myopathies includes isolated electrocardiographic changes such as nonspecific ST changes and conduction abnormalities, coronary vasculitis, ischemic abnormalities, heart failure, myocarditis, and pericarditis.\textsuperscript{1,2} Cardiac involvement in ASS is extremely rare. We present a case of ASS involving a cardiac valve during a disease flare in a patient with anti-PL 7 antibodies. To our knowledge, this represents the first reported case of severe aortic regurgitation occurring in the setting of an ASS flare.

Disclosures

None.

References


Figure 2. A, Macroscopic appearance of the native aortic cusps. The valvular apparatus was diffusely edematous and buttery without the presence of vegetations. B, Microscopic appearance of one of the aortic cusps showing acute and chronic inflammation (blue box) and necrosis (red box). C, Higher magnification. Accumulation of neutrophils and lymphocytes with necrosis, consistent with microabscsses formation.
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