Autoantibody to PL-12 (Anti-Alanyl-tRNA Synthetase) in an African American Girl with Juvenile Dermatomyositis and Resolution of Interstitial Lung Disease

To the Editor:

Juvenile dermatomyositis (JDM) is an immune-mediated pediatric idiopathic inflammatory myopathy (IIM) characterized by muscle and skin inflammation. Anti-PL-12 is a myositis-specific antibody (MSA), directed against alanine-tRNA synthetase, often with debilitating interstitial lung disease (ILD), that is sporadically reported in childhood. We describe a child with anti-PL-12 antibody who recovered lung function after therapy.

A 14-year-old African American girl developed bilateral ankle stiffness associated with swelling and redness of the upper eyelid. One month later, she noticed proximal weakness of shoulder girdle and pelvic girdle. Four months later, she had fever to 102°F; an erythematous rash over upper chest, arms, and upper eyelids; difficulty walking and getting up from bed, with shortness of breath during exertion. After admission to intensive care unit for suspected aspiration pneumonia, she was diagnosed with JDM on the basis of a muscle biopsy. She received intravenous (IV) steroid (30 mg/kg/dose) for 5 days, followed by oral prednisone (1 mg/kg/day), and was lost to followup.

Six months later, she was admitted to Children’s Memorial Hospital with fever, malaise, weakness, worsening rash, and bilateral knee arthritis. Laboratory data were elevated: creatine kinase 12,464 IU/l (normal 29–165 IU/l), aldolase 150 IU/l (normal 3.4–8.6 IU/l), alanine transaminase 156 IU/l (normal 2–30 IU/l), aspartate transaminase 257 IU/l (normal 16–52 IU/l), and lactate dehydrogenase 1496 IU/l (normal 126–289 IU/l). Markers of inflammation were increased: erythrocyte sedimentation rate (ESR) 37 mm/h (normal < 20 mm/h), C-reactive protein (CRP) 4.41 mg/dl (normal < 0.8 mg/dl), von Willebrand factor antigen (vWF:Ag) 420% (blood group B normal = 57%–241%); and elevated neopterin level 30.4 nm/l (normal < 10 nm/l). Periungual capillaroscopy showed dilated capillaries, and moderately severe dropout of the nailfold capillary end-row loop (ERL): 4.28 (normal value 7–10%). Tests for anti-dsDNA, Scl-70, and rheumatoid factor were negative, but her sera were positive for anti-Ro and anti-PL-12 (laboratory of Dr. I. Targoff). On pulmonary function testing, she had decreased forced vital capacity, 73%, and diffusion capacity (DLCO/VA) of 66%, consistent with restrictive lung disease (Table 1). Computed tomography (CT) scan of the chest showed peripheral intralobular and interlobular septal thickening, early parenchymal cystic changes, and micronodules consistent with ILD (Figure 1).

Our patient’s pulmonary function tests showed a pattern of restrictive lung disease, commonly associated with diminished core strength as well as a diffusion defect. The chest CT identified interlobular septal thickening, one of the most common radiological findings in PL-12-positive adults. Although myositis is usually mild in patients with anti-PL-12, our patient had severe myositis with markedly elevated muscle enzymes, and her score on the Childhood Myositis Assessment Scale was markedly decreased at 21/52. She was negative for antibody to ribonuclear protein, which is present in 36%–65% of patients who are anti-PL-12-positive. VWF:Ag is often an indicator of a disease flare in IIM, and our patient exhibited high VWF:Ag levels at presentation, persisting for 9 months. Her disease course was similar to those of adults with both anti-PL-12 and anti-Ro antibody, in whom ILD is difficult to control. In contrast to reported cases, however, her pulmonary function tests normalized and the chest CT scan showed significant resolution of ILD. To our knowledge this is the first complete report of a child with IIM with both anti-PL-12 and anti-Ro autoantibodies and improvement of lung function in response to intensive immunosuppressive therapy.

Table 1. Lung function data over time (dates shown). All data are percentages.

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<tr>
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<th>Pretreatment</th>
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<tbody>
<tr>
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<tr>
<td>TLC</td>
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<td>90</td>
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<tr>
<td>DLCO/VA</td>
<td>66</td>
<td>88</td>
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FEV: forced expiratory volume; FVC: forced vital capacity; TLC: total lung capacity.
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REFERENCES


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Figure 1. CT scans before treatment showing peripheral intralobular interstitial thickening at the lung bases, left greater than right (A, B, C); and after treatment showing resolution of previous changes (D, E, F).