Anti-CADM-140 Antibody-positive Juvenile Dermatomyositis with Rapidly Progressive Interstitial Lung Disease and Cardiac Involvement

To the Editor:

Extramusculocutaneous manifestations in juvenile dermatomyositis (JDM) may lead to life-threatening consequences. Interstitial lung disease (ILD) has been reported as one such serious complication in JDM. However, cardiac involvement in JDM is a rare complication and is seldom reported. Recently, anti-CADM-140 autoantibody was discovered in amyopathic dermatomyositis and was associated with rapidly progressive ILD. We describe a fatal case of JDM complicated by ILD and cardiac involvement in which serum preserved at admission was shown to contain anti-CADM-140 antibody.

A 9-year-old boy was admitted to our hospital with a 4-month history of low-grade fever and erythematous rashes on his face, hands, elbows, and knees. He was developmentally delayed from an unknown cause. He could not describe muscle weakness or tenderness but showed claudication indicating lower-limb muscle weakness. He had Gottron’s papules but no掌纹变化.

The patient had weak lower-limb muscles, decreased active and passive range of motion of the fingers, and weakness of the lower limbs. His lower limbs were elevated (25.256 units, normal below 8). Cardiac sounds revealed a gallop rhythm and he had fine crackles over both lung fields. His respiratory condition rapidly deteriorated and pneumothorax developed. Whole-body scintigraphy with 67Ga-citrate showed increased uptake in the lower lobes of the lungs. The patient had a history of recurrent bacterial pneumonias.

Serum ß-D-glucan and cytomegalovirus antigen were also negative. With his guardian’s consent, an autopsy was performed. The lungs were entirely firm and showed remarkable congestion, and the histologic examination revealed intense alveolar hemorrhage, formation of hyaline membranes, and infiltration of inflammatory cells suggesting diffuse alveolar damage. The heart demonstrated hypertrophy and histologic examination detected no specific changes of myocarditis, pericarditis, or fibrosis except for partial degeneration of the papillary muscle. The presence of anti-CADM-140 antibody in our patient may suggest that this antibody is a predictor of rapidly progressive ILD and poor prognosis even in JDM. We describe a case of fatal JDM with rapidly progressive ILD and cardiac involvement. Positive anti-CADM-140 antibody may predict a poor prognosis for JDM with ILD.

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Figure 1. A. Chest radiography showing diffuse bilateral consolidation of the lungs with pneumomediastinum and subcutaneous emphysema of thoracic walls. B. Chest HRCT shows extensive bilateral lobar consolidation, pneumomediastinum, subcutaneous emphysema of the thoracic walls, and left pneumothorax.

Figure 2. Autopsy findings of the lungs. A. Severe congestion of the lungs. B. The lung histology shows alveolar hemorrhage, formation of hyaline membrane, and infiltration of inflammatory cells compatible with diffuse alveolar damage (H&E stain, original magnification ×200).