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

To the Editor:

Extra- and intermusculocutaneous 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.6,9 Recently, anti-CADM-140 autoantibody was discovered in amyopathic dermatomyositis and was associated with rapidly progressive ILD.7,8 We describe a fatal case of JDM complicated by ILD and cardiac involvement in which serum measured 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 clawed fingers indicative of low-limb muscle weakness. He had Gottron’s papules but no heliotrope rash. Cardiac sounds revealed a gallop rhythm and he had fine crackles over both lung fields. His erythrocyte sedimentation rate was 38 mm/h, white cell count 1800/μl, hemoglobin 9.3 g/dl, platelets 115,000/μl, aspartate aminotransferase 207 U/l (normal 11–39), alanine aminotransferase 101 U/l (normal 5–40), lactate dehydrogenase 564 U/l (normal 119–229), aldolase 16.7 U/l (normal 2.1–6.1), creatine kinase (CK) 104 U/l (normal 45–160), C-reactive protein (CRP) 0.24 mg/dl (normal 0.0–0.3), antinuclear antibody 80 (normal < 40), anti-Jo-1 antibody negative, brain natriuretic peptide 55.2 pg/ml (normal 0.0–19.5), and Krebs von den Lungen-6 (KL-6) 1275 U/ml (normal 0–500). His electrocardiogram revealed sinus tachycardia at 150 beats/min and T wave flattening. Echocardiography demonstrated a low ejection fraction of 44%. Chest high-resolution computed tomography (HRCT) revealed bilateral pleural effusions. Whole-body scintigraphy with 67Ga-citrate showed increased uptake in the lower lobes of the lungs. Electromyography showed low amplitude and short durations compatible with myopathy. Magnetic resonance imaging showed increased signals at bilateral adductor and pectoral muscles on T2-weighted images. He was diagnosed with JDM and initially treated with oral methylpredonisolone (mPSL). However, his CK increased to 315 U/l two weeks later and two courses of mPSL pulse therapy were added. After each mPSL pulse therapy, CK elevation was 5138 U/ml. His respiratory condition rapidly deteriorated and pneumocystis jiroveci pneumonia was diagnosed. Repeat HRCT demonstrated progressive bilateral infiltration in the lower lobes, then intravenous cyclosporin A was administered. KL-6 was elevated to 5138 U/ml. His respiratory condition rapidly deteriorated and pneumomediastinum, subcutaneous emphysema, and left pneumothorax developed (Figure 1). He was intubated and mechanically ventilated and underwent plasmapheresis. Despite intensive treatment, he died 1 week after intubation, which was 3 months after the initial hospitalization.

With his guardian’s consent, an autopsy was performed. The lungs were entirely firm and showed remarkable congestion, and the microscopie examination revealed intense alveolar hemorrhage, formation of hyaline membrane, and infiltration of inflammatory cells suggesting diffuse alveolar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2). Neither interstitial changes nor infection of the lungs were present. The heart demonstrated hypertrophy and histologic lar damage (Figure 2).
**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).