BMJ Case Reports

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Dramatic heliotrope dermatitis and uncommon pulmonary involvement in juvenile dermatomyositis

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DESCRIPTION

Juvenile dermatomyositis (JDM) is a systemic autoimmune myopathy of childhood. It is characterised by pathognomonic skin rashes and proximal muscle weakness, which are the most useful diagnostic clues.[1] Pulmonary involvement in JDM has been less reported than in adult patients[2] and interstitial lung disease is frequently identified abnormality by CT.[3] Here we present a JDM case with severe heliotrope rash and uncommon manifestation in CT image of the lung. A 15-year-old girl presented with a 3-month history of progressive periorbital swelling and a reddish-purple rash on the upper and lower eyelids (figure 1A). Her vision acuity decreased and MRI showed only thickening of soft tissue in bilateral eyelids. Three weeks before admission, she started to present with middle grade fever, leukocytopenia, anorexia, polyarthralgia and mild symmetrical proximal muscle weakness. She had a skin rash in the V neck region and periungual vascular dilatation, but neither Gottron's papules or the shoulder shawl cutaneous signs were identified. Her lung fields were clear to auscultation. Laboratory findings demonstrated that serum levels of muscle enzymes were mildly elevated. Antinuclear antibodies and anti–Jo–1 were negative. Electromyography demonstrated myopathic changes. Histologic findings of quadriceps biopsy were compatible with JDM. CT of the chest showed multiple patchy opacities (figure 1C). The patient underwent CT-guided needle biopsy. Pathological analysis showed no signs of lung malignancy. Cultures for

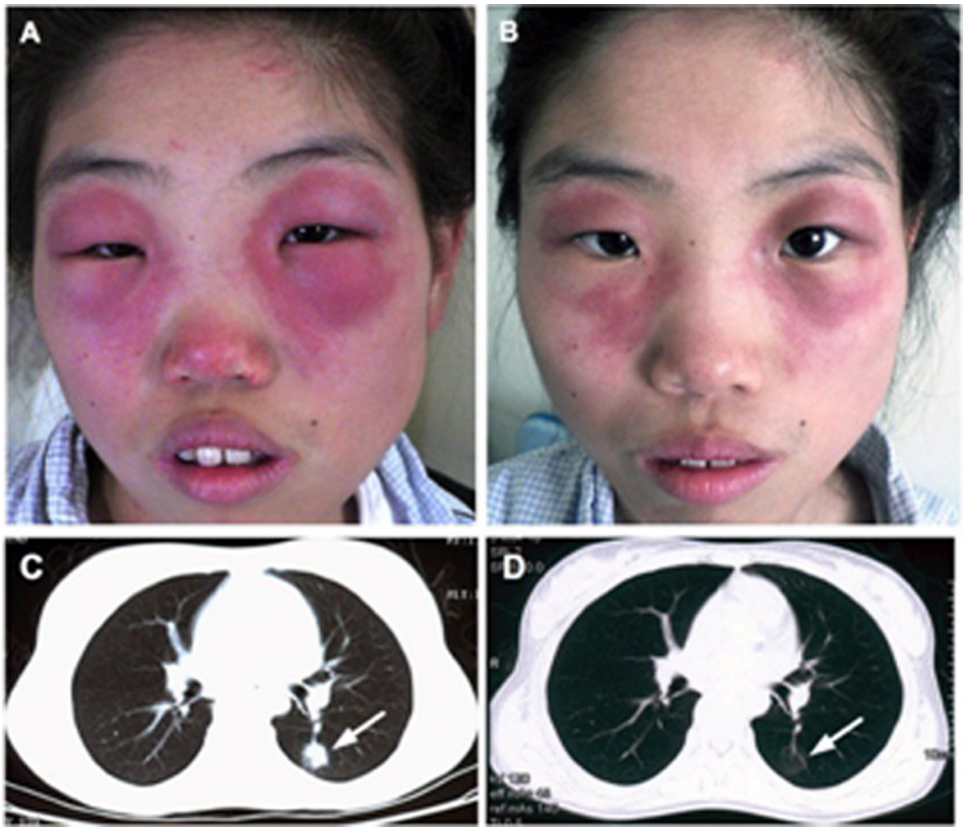

Figure 1 (A) Severe reddish-purple rash around eyelids. (B) The heliotrope rash was resolved significantly 1 month after prednisone treatment. (C) Pulmonary involvement shown in CT of the chest. (D) Repeated CT scan result showed improvement after 1 month treatment.

bacterium, fungus and acid-fast bacillus were negative. She received prednisone therapy (1 mg/kg per day) and intravenous cyclophosphamide. One month later, the patient was free of symptoms. Her periorbital rash was significantly resolved. A repeated CT scan displayed demonstrable improvement of the pulmonary involvement.

Acknowledgements This study was funded by Beijing Nova Program (No 2008B49).

Competing interests None.

Patient consent Obtained.

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