

Primary pulmonary diffuse large B-cell lymphoma with anti-NXP2-positive dermatomyositis: a case report

Sirs,

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterised by cutaneous involvement. It was reported that DM patients had an increased risk for malignancy, especially anti-Tif- γ , anti-NXP2, and anti-SAE positive DM patients (1).

Lymphoma was the most common haematological malignancy (2), but primary pulmonary lymphoma was seldom reported. Here, we present a case of primary pulmonary diffuse large B-cell lymphoma (PP-DLBCL) with anti-NXP2-positive DM that presented as a lung mass and cyst in CT findings.

A 73-year-old man presented with a 3-month history of progressive symmetric proximal limb weakness, mild myalgia, facial erythema, and shortness of breath. He recently lost 8KG in weight. He was a smoker and had a history of chronic bronchitis. On physical examination, facial erythema (Fig. 1A) and weakness of neck flexors were noted. The muscle strength of the proximal limbs was level III, and the distal limbs was level IV. Laboratory tests showed erythrocyte sedimentation rate of 67 mm/h (normal range 0–15 mm/h), C-reactive protein of 12.3 mg/L (normal range 0–10 mg/L), serum lactate dehydrogenase of 781 U/L (normal range 120–250 U/L), creatine kinase of 7608 U/L (normal range 40–200 U/L), creatine kinase isoenzyme of 125 U/L (normal range 0–25 U/L), myoglobin of 765 ng/ml (normal range 0–70 ng/ml), alanine aminotransferase of 75 U/L (normal range 0–40 U/L), aspartate aminotransferase of 310 U/L (normal range

0–40 U/L), serum albumin of 25.8 g/L (normal range 35–55 g/L), ferritin of 1756 ng/ml (normal range 80–130 ng/ml). The complete blood count, urine analysis, serum IgG, IgM, and IgA were all within normal ranges. Negative for rheumatoid factor, anti-CCP antibodies, antinuclear antibodies and HIV antibodies. The blood autoantibody test for idiopathic inflammatory myopathy (IIM) showed anti-NXP2 antibody was positive. Electrophysiological tests of the extremities demonstrated a manifestation of myogenic damage. Positron emission tomography-computed tomography (PET-CT) showed a 6.4×4.7 cm mass in the middle lobe of the right lung with SUVmax of 15.78, a 4.7×4.4 cm lung cyst in the lower lobe of the right lung with variable wall thickness, SUVmax of 4.2 (Fig. 1B and 1C). He underwent ultrasound-guided percutaneous needle biopsy and immunohistochemistry of the lung mass showed the proliferation of large cells positive for CD20, CD79a, PAX-5, Bcl-2, Mum-1, C-myc (about 10%), and Ki-67 (about 70%). Bone marrow aspirate revealed no lymphoma involvement. Based on the above examination results, he was finally diagnosed as PP-DLBCL with DM. R-CHOP (rituximab, cyclophosphamide, liposomal doxorubicin, vincristine, prednisolone) chemotherapy was given once every 3 weeks. Enhanced computed tomography after the 5th cycle showed lung mass diminished and the solid component of the cyst wall disappeared (Fig. 1D). As well as the creatine kinase returned to normal.

Primary lung lymphoma is a relatively rare disease, which is defined as clonal lymphoid infiltration of the lung with or without hilar or mediastinal lymph node and with no evidence of extra thoracic disease up to 3 months after the time of diagnosis (3). PP-DLBCL was reported as the second most common sub-type of

primary pulmonary lymphoma, mostly affecting individuals with acquired immune deficiency. At imaging, PP-DLBCL can present as different findings, such as areas of consolidation, single/multiple nodules, a reticular/interstitial infiltrate or ground-glass appearance, with or without mediastinal lymphadenopathy. Lesions are mostly bilateral and tend to be located peripherally in the lower lobe (4).

A lung cyst can be a rare imaging finding of a tumour, mainly primary lung cancer which is now called lung cancer associated with cystic airspaces (5), rarely lymphoma. Our patient's lung mass diminished and the solid component of the cyst wall disappeared after R-CHOP chemotherapy treatment. Based on the changes, we considered the lung cyst was associated with PP-DLBCL. To our knowledge, this is the first case referring to lung cyst associated with PP-DLBCL, as well as the first case referring to anti-NXP2-positive DM associated with PP-DLBCL. When one anti-NXP2-positive DM patient had atypical lung lesions, we should pay attention due to the association between anti-NXP2 and malignancy.

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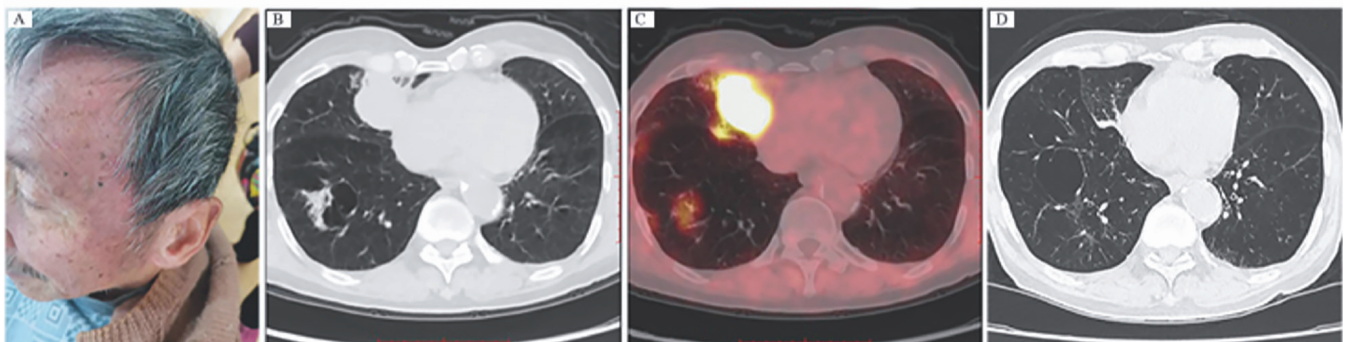


Fig. 1. Physical examination, CT evolution before and after R-CHOP chemotherapy. **A:** Facial erythema. **B, C:** Lung mass with SUVmax of 15.78 and lung cyst with variable wall thickness, SUVmax of 4.2 before treatment. **D:** Lung mass diminished and the solid component of the cyst wall disappeared after treatment.

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