中文題目：在一位 52 歲帶有 MDA5 抗體之皮肌炎男性患者發生快速進展的間質性肺病：病例報告
英文題目：Rapid progressive interstitial lung disease in a 52-year-old man with anti-MDA5 associated dermatomyositis: A Case Report
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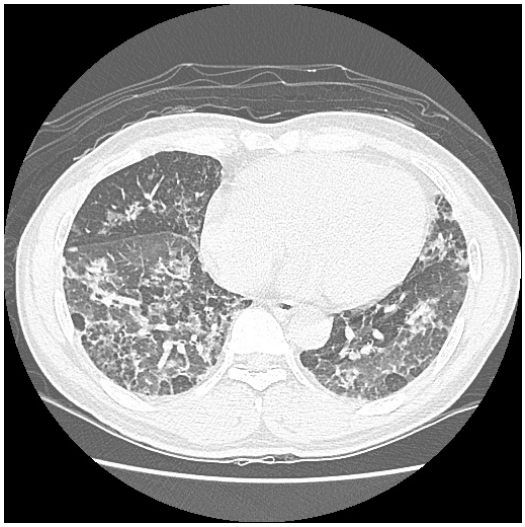
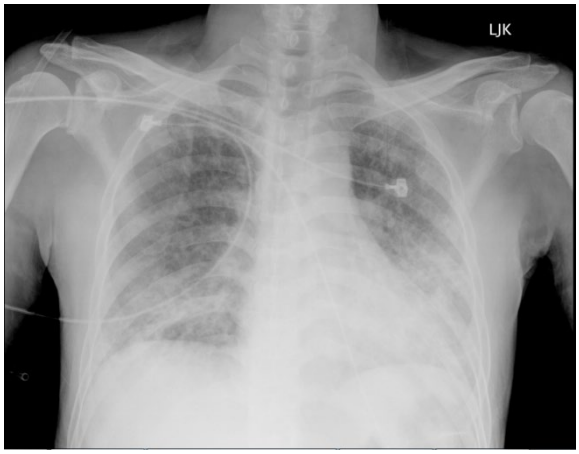
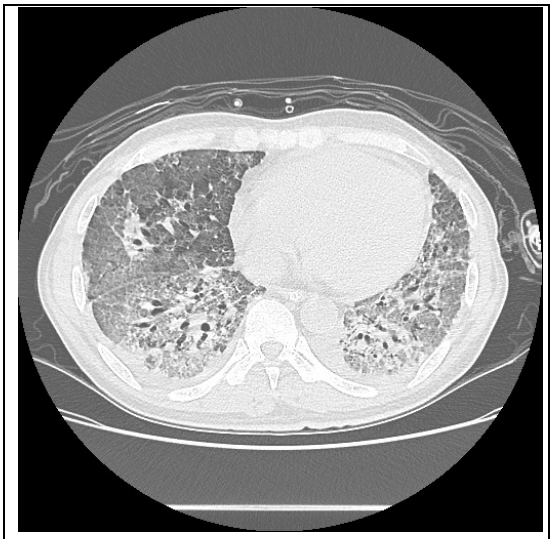
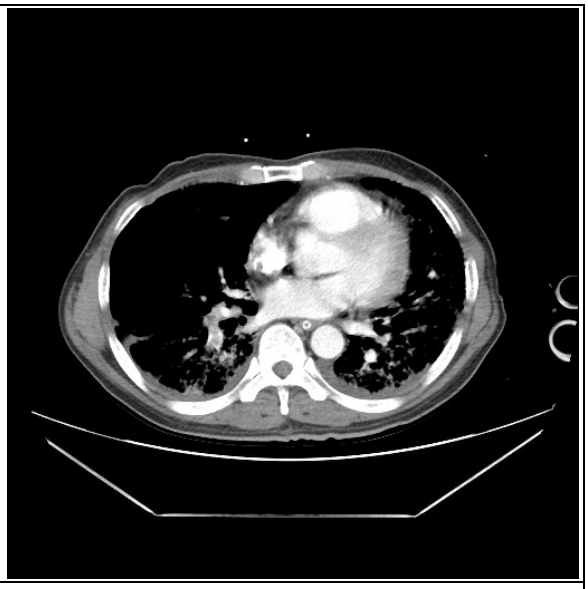
Case presentation: A 52-year-old man with chronic hepatitis B who suffered from erythematous papules over both hand for about two months. In addition, he also complained progressive proximal weakness and myalgia. Therefore, he visited our outpatient department that typical Gottron's sign and mechanic's hands were noted and dermatomyositis was impressed. Initial lab data showed elevation of creatine phosphokinase level up to 462U/dL otherwise negative finding of autoimmune profile(antinuclear antibody, anti-SS-A, anti-SS-B, anti-Jo-1, anti-Sm, anti-RNP, anti-dsDNA, Rheumatoid factor). Chest X-ray revealed mild bilateral lower lungs interstitial pattern. After glucocorticoid treatment, creatine phosphokinase level subsided to 274U/dL and his skin lesions improved with increased muscle power. Electromyography showed myopathy pattern and nailfold capillaroscopy showed 1.tortuosity, 2.loop dilatation, 3.hemorrhagic spot, 4.bush form that are all compatible with dermatomyositis. We switched medication to prednisolone 15mg twice a day, hydroxychloroquine 200mg once a day, methotrexate 7.5mg once a week, folic acid 5mg once a week. However, progressive dyspnea without obvious productive cough or fever were mentioned within 14 days, thus chest CT was arranged which showed bilateral lungs ground glass opacity and basal lung field interstitial pattern***(Picture A)***. He was then admitted due to impending respiratory failure that Chest X-ray revealed bilateral pneumonia and interstitial pattern***(Picture B)***. Empirical antibiotics with Tazocin, azithromycin and Sevatrim for covering severe pneumonia and possible PJP due to immunocompromised status. Methylprednisolone 20mg twice a day was given to control dermatomyositis with interstitial lung. High lactic dehydrogenase (374 IU/L) and ferritin(1023.7) levels were noted. Conditions downhill within 1 day that he was intubated. Poor PaO2/FiO2 ratio was noted during the next one week, thus we arranged chest high resolution computed tomography again which showed progression of interstitial lung with crazy paving pattern***(Picture C)*** and right pulmonary artery emboli***(Picture D)***. Enoxaparin was given and switched to rivaroxaban after one-week course. Antibiotics was adjusted with adding on levofloxacin for covering atypical pathogen and anidulafungin for covering fungus and treatment for Pneumocystis jiroveci pneumonia as well(Polymerase chain reaction +), then switched to Tienam plus

colistin due to sputum culture yielded multidrug-resistant Acinetobacter baumannii. After one week course, infection seems under control but still poor oxygenation status and P/F ratio that FiO2 always more than 70%. Meanwhile, anti-MDA5 antibody showed positive which reminds us that rapid progression interstitial lung disease may present. Therefore, we tried intravenous immunoglobulin and cyclosporin. Furthermore, infection was detected again with fever episodes; Methicillin-resistant Staphylococcus aureus, Cytomegalovirus were isolated from sputum culture, and we tapered glucocorticoid dose gradually. Adult respiratory distress syndrome was still noted without obvious improvement; even we administered full sedation, and prone position. Extracorporeal membrane oxygenation was suggested but family preferred conservative treatment and signed "Do not resuscitate" permit. The patient then expired on the 35th day of hospital course due to respiratory failure.

Discussion: We should always keep in mind that interstitial lung disease may occurred in rheumatology disease patient. According to the recent research and our clinical experience, positive anti-MDA5 antibody means poor prognosis and rapid progression interstitial lung disease especially when high titer. We need to balance the immunosuppressant and infection control that might be very tough for clinical physician. Early detection with symptoms, and lab data(high lactic dehydrogenase, Ferritin level are marker for poor prognosis) may help us to decide the choice of medication for these patients.

	
A. Interstitial pattern over basal lungs	B. Bilateral pneumonia with lower lungs interstitial pattern

	
C. Crazy paving pattern	D. Right pulmonary trunk emboli

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