中文題目：帶有 MDA-5 抗體之皮肌炎患者之間質性肺炎表現與治療後產生之非分支結核桿菌伺機性感染

英文題目：A case report of anti-MDA-5 positive dermatomyositis patient diagnosed with secondary hemophagocytic lymphohistiocytosis due to opportunistic NTM infection after systemic steroid therapy for interstitial lung disease

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Background: Dermatomyositis(DM) is a inflammatory myopathy with characteristic skin manifestations accompanying or preceding muscle weakness. Pulmonary complications, due primarily to interstitial lung disease, are reported in 10 to 40% of patients. anti-MDA-5 is associated primarily with interstitial lung disease.

Method: Here, we presented a case report of anti MDA5 positive DM patient, who was admitted on July, 2018 due to fever, and later diagnosed with hemophagocytic lymphohistiocytosis (HLH).

Clinical case: This 50-year-old male had history of dermatomyositis and had initial presentation of exertional dyspnea and proximal muscle weakness without typical skin manifestations in August, 2017. he underwent muscle biopsy and at first polymyositis was impressed and treated with steroid pulse therapy for his interstitial lung disease. In March, 2018, bilateral digital pulp ulcer developed and his serum was positive for anti-MDA-5. He was thereafter diagnosed as dermatomyositis.

He was admitted in July, 2018 due to fever and general malaise. No definite infectious focus was identified after serial blood tests, culture and diagnostic imaging modalities. Bicytopenia developed and bone marrow biopsy was arranged as hematologist suggestion. Histiocytosis was found. Bone marrow acid fast stain was 1+ and TB-qPCR yielded negative result. Elevated level of ferritin (2632ng/mL) and splenomegaly revealed by sonography promoted diagnosis of secondary HLH due to NTM infection.

Conclusion: Anti-MDA5 were originally identified in clinically amyopathic DM patients. This case was notable for its initial presentation of proximal muscle weakness and late manifestation of acral ulcerations. Anti-MDA positive DM patient was vulnerable to fatal rapidly progressive interstitial lung disease and requires aggressive systemic immunosuppressive therapy, and predisposed them at higher risk of opportunistic infection.