

中文題目：一位 23 歲的年輕男性伴隨有 NXP2 陽性的皮膚炎—從診斷到疾病進展與治療

英文題目：A 23-year-old man with NXP2 Ab positive Dermatomyositis-from initial diagnosis, disease progression to treatment

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### **Abstract:**

Myositis-specific autoantibodies (MSAs) are useful tools for detected the idiopathic inflammatory myopathies (IIM) including polymyositis (PM) and dermatomyositis (DM). Anti-NXP2 Ab (nuclear matrix protein antibodies ) is a one of the MSAs but more often found in juvenile dermatomyositis (JDM). Anti-NXP2 Ab has higher association with male gender, dysphagia, myalgia, peripheral edema and calcinosis.

A 23-year-old man has no systemic disease before. His initial presentation was puffy eyes and erythematous change of upper bilateral eyelid, calcinosis over forehead and back at 2018/04. On 2018/06/18, he had myalgia, general weakness, poor appetite and abdominal discomfort. Due to elevated GOT/GPT level, then he was admitted under the impression of acute hepatitis. Progressive four limbs weakness, dysphagia, and prominent erythema and swelling over upper eyelids and bilateral lateral necks, some calcinosis skin lesion over neck, chest, arms, back and abdomen were noticed. Lab data showed elevated CPK. Typical dermatomyositis findings: heliotrope sign (+), Gottrons signs (+), V sign(+), Shawl sign(+), muscle power decreased were noted. Blood examination showed: NXP2 Ab = Strong positive. For dermatomyositis, pulse steroid therapy with methylprednisolone 1000mg used. However, his clinical condition deteriorated in days, he had respiratory failure and then was intubated. CXR showed bilateral pneumonia, but no interstitial lung pattern. So, in addition to methylprednisolone 40mg q12h, he received plasma exchange five times, and had Cyclophosphamide 500mg on 2018/7/26. Also, he had monoclonal anti-CD20 antibody Rituximab twice. The pathologic report of muscle biopsy was inflammatory myopathy. After treatment, his muscle power improved, but still poor airway protection and difficult weaning from ventilator. Finally, tracheostomy was done, and the patient had successful weaning, and now is under oral form dexametasone 4mg QD and cyclophosphamide 100mg QD.

### **Conclusions:**

1. Novel myositic-specific antibodies including anti-NXP2 were developing and helps physician to earlier diagnose dermatomyositis.