



## IRA E-BULLETIN : ISSUE 1 | JANUARY 2020

### CASE SOLVING

#### ROWELL SYNDROME WITH MYOSITIS

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This 12-year-old patient presented with low grade fever, erythematous rash and bilaterally symmetrical progressive proximal weakness for 1 month. She was found to have oral ulcer over hard palate, papulo-squamous and erythema multiforme-like rash involving face, trunk, upper and lower limbs (figure below), chilblains in extremities, power of 3/5 in proximal muscles of b/l upper and lower limb. Her blood tests showed pancytopenia (Hb- 6.5g/dl, TLC- 1600, Plt- 125,000), ESR- 86, raised CKNAC (787 U/L) and LDH (1765 U/L), low C3 and normal C4, ANA 2+ (speckled). Patient satisfied the classification criteria for systemic lupus erythematosus and the revised criteria proposed by Zeitouni et al for Rowell syndrome. She was started on wysolone 2mg/kg/day, hydroxychloroquine and fluticasone ointment for skin lesions. Her skin lesions and muscle power improved, and was started on methotrexate weekly. She's doing well on follow-up.



Figure showing erythema-multiforme like lesions over face (left) and multiple papulo-squamous to discoid lesions resembling subacute lupus over back of trunk (right)

#### Discussion

Lupus is an autoimmune disorder with diverse clinical manifestations and virtual involvement of any organ. Skin lesions are heterogenous which can be Lupus-specific or non-specific as given by Gilliam et al. Rowell syndrome occurs rarely in patients with lupus, wherein they develop erythema-multiforme like lesions. It occurs more commonly in middle-aged and young females. The most consistent feature is speckled pattern of ANA which occurs in 88% of cases. Zeitouni et al proposed modified diagnostic criteria for Rowell's syndrome which has 3 major- lupus erythematosus (LE)– systemic, discoid or subacute LE, EM-like lesions, ANA positivity (speckled pattern), and 3 minor- chilblains, anti-Ro/La positivity or rheumatoid factor positivity. All 3 major and minor criteria are required for diagnosis(1). To the best of our knowledge, this is the first case reported to be of lupus presenting initially as myositis with EM- like skin lesions, Rowell syndrome (RS). Physicians should be aware of this as index presentation of lupus. Azathioprine, antimalarials, cyclosporine and prednisolone have good response. The index case responded well to steroids and hydroxychloroquine.

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