# **CLINICAL CASE VIGNETTES**

# Rowell's syndrome: A rare presentation of childhood lupus

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A 11-year-old girl presented to the clinic with intermittent fever, multiple erythematous maculopapular lesions (erythema multiforme (EM) like lesions) of targetoid appearance over face, trunk, and limbs (Fig. 1), and a large palatal ulcer (Fig. 2). She was tested positive for ANA by Hep-2 (speckled pattern) and for anti-Ro, anti-nucleosome, anti-histone, and anti-dsDNA antibodies by immunoblot. She had reduced complement levels (C3, C4) and tested negative for rheumatoid factor and viral markers. Further evaluation has concluded that the patient met the diagnostic criteria for Rowell's syndrome.

In 1963, Rowell *et al.* described the syndrome as the presence of discoid lupus erythematosus with erythema multiforme and with a positive serology for rheumatoid factor, speckled ANA, and precipitating antibodies to saline extract of human tissue (anti-SjT identical to anti-Ro and anti-La antibodies).<sup>1</sup> Lee *et al.* (1995) have included chilblains as one of the characteristic features of Rowell's syndrome.<sup>2</sup> In 1997, Zeitouni *et al.* redefined the major and minor diagnostic criteria for Rowell's syndrome as given below:<sup>3</sup>

The major criteria include

- Systemic lupus erythematosus (SLE), discoid LE (DLE) or subacute cutaneous LE (SCLE)
- EM-like lesions with or without mucosal involvement
- Speckled pattern of ANA

The minor criteria are:

- Chilblains
- Positive anti-Ro or anti-La antibodies
- A reactive rheumatoid factor

## Fig 1:Targetoid lesion over right cheek



#### Fig 2: Palatal ulcer



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