

# Malar Rash in Systemic Juvenile Idiopathic Arthritis

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We describe malar rash in a patient presenting with systemic juvenile idiopathic arthritis (sJIA).

Midsummer, a 9-year-old girl developed an evanescent rash on the legs, which generalized within days. Fever began on the third day, then arthritis. These improved with corticosteroids, but reappeared on tapering. She had one oral ulcer, but no hair loss or Raynaud's phenomenon. A photograph shows her facial rash during the seventh week (Figure 1).

Medications included acetaminophen, ibuprofen, and prednisone. She had not taken sulfa-containing medications<sup>1</sup>. Family history revealed no autoimmune diseases. Muscle strength and nailfold microscopy were normal. She had no Gottron's papules. Laboratory studies demonstrated mild normocytic anemia, and elevated C-reactive protein (71 mg/dl), erythrocyte sedimentation rate (50 mm/h), ferritin (564 ng/ml), aldolase (10.3 U/l), and lactate dehydrogenase (796 U/l). Twice, antinuclear antibodies were negative. Additional normal or negative investigations included anti-double-stranded DNA and extractable nuclear antibodies (including anti-Ro), complement C3 and C4, total hemolytic complement, IgG, creatine kinase, alanine and

aspartate aminotransferases, urinalysis, and magnetic resonance imaging of the proximal thighs. During the ninth week, she demonstrated quotidian fevers, 39°–40°C. White blood cell count was 15,400 with 12,300 neutrophils; platelets 364,000; and ferritin 5,278, suggesting occult macrophage activation syndrome<sup>2</sup>.

Treatment for sJIA was instituted with corticosteroids and methotrexate. The malar rash resolved. Adalimumab was added. Complete remission was achieved after substituting anakinra for adalimumab. This is the first report of a malar rash in a patient presenting with sJIA. This case highlights the need to maintain an open differential diagnosis, even in the presence of a highly characteristic examination finding.

## REFERENCES

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2. Behrens EM, Beukelman T, Paessler M, Cron RQ. Occult macrophage activation syndrome in patients with systemic juvenile idiopathic arthritis. *J Rheumatol* 2007;34:1133-8.



Figure 1. Facial rash at 7 weeks after onset of symptoms.