

IMAGES IN EMERGENCY MEDICINE

Pediatrics, dermatology

Child with fever, rash, and abnormal gait

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PRESENTATION

A 21-month-old Caucasian male presented to the emergency department with 3 months of fever, rash, and abnormal gait. He first developed intermittent fever that progressed to daily high-spiking fevers for a 1-month duration before presentation. Erythematous maculopapular lesions progressed from his nose, forehead, cheeks, to his hands. He had difficulty walking up stairs and moved "like an old man" when sitting and getting up from the floor. Examination revealed erythematous papules over the metacarpophalangeal (MCP) and proximal interphalangeal joints (PIP) with nailbed erythema and diffuse non-pitting edema in his hands (Figure 1). Gait was stiff and antalgic without limping. Laboratory studies showed mild leukopenia (white blood count 4×10^9 cells/L), anemia (hemoglobin 10.2 g/dL), and an elevated erythro-



FIGURE 1 Erythema and papules on dorsal MCP and PIP joints of right hand (Gottron's papules). Mild, diffuse soft tissue swelling of the hands and fingers and nailbed erythema is also seen

cyte sedimentation rate (25 mm/h). C-Reactive protein and creatine kinase were normal.

DIAGNOSIS

Juvenile dermatomyositis

Additional evaluation by pediatric rheumatology showed myositis (aspartate aminotransferase 863 units/L, alanine aminotransferase 338 units/L, lactate dehydrogenase 731 units/L, and aldolase 26.3 units/L). Diagnosis of juvenile dermatomyositis was made with evidence of Gottron's papules and symmetric proximal muscle weakness. Myositis antibody testing later revealed positive antibodies against melanoma differentiation-associated protein 5 (MDA-5). Recognition of characteristic skin changes can direct appropriate evaluation to confirm diagnosis and screen for morbid complications.

Possible emergent complications include intestinal perforation,^{1,2} or hemorrhage from vasculitis,³ severe diaphragmatic muscle weakness, dysphagia, and dysphonia leading to respiratory failure or aspiration.⁴ Interstitial lung disease can occur and potentially be rapidly progressive, with anti-MDA-5-positive patients at increased risk.⁵ Macrophage activation syndrome secondary to juvenile dermatomyositis can also occur. Unlike adults with dermatomyositis, children with juvenile dermatomyositis do not have increased malignancy rates.⁶⁻⁹ Immunosuppressed patients, and/or those with ulcerative calcinosis are at risk for systemic and opportunistic infection.¹⁰⁻¹²

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