Cutaneous Polyarteritis Nodosa

A 12-year-old girl presented with digital ulcer, palpable purpura and subcutaneous nodules involving the extremities (Fig. 1). Systemic examination was normal. Investigations showed a raised erythrocyte sedimentation rate and C-reactive protein. Liver function tests, renal function tests, and complement levels were normal. Rheumatoid factor, anti-nuclear antibody, anti-double stranded DNA, anti-neutrophil cytoplasmic antibody, anti-phospholipid antibody, lupus anti-coagulant and serum cryoglobulins were all negative. ASO titers were normal. Mantoux test and serologies for hepatitis B and C infections were negative. Deep incisional skin biopsy from the foot lesion showed leucocytoclastic small vessel vasculitis of the deep dermis. A diagnosis of Cutaneous Polyarteritis Nodosa (CPAN) was made. The patient responded to high dose pulse methyl prednisolone therapy and is at present on low dose steroid for last 6 months without any relapse (Fig. 2).

CPAN is a rare vasculitis affecting the small and medium vessels of the dermis without any systemic involvement of the liver, kidney or heart. CPAN should be differentiated from systemic PAN which may present with similar skin involvement but has florid systemic manifestations. Unlike other vasculitis, CPAN lacks immunological markers. It may be associated with streptococcal sore throat, hepatitis B, C infection or tuberculosis. Though affecting skin and having a good prognosis, CPAN may be resistant to low dose steroids and may require aggressive therapy with a relapsing course.

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