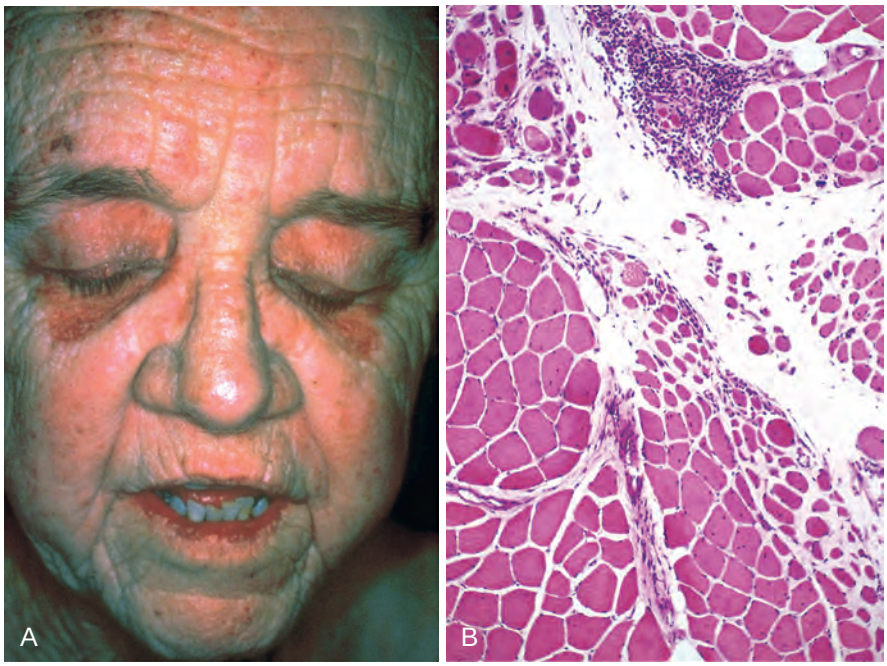

Figure 27-8 **A,** Dermatomyositis. Note the heliotrope rash affecting the eyelids. **B,** Dermatomyositis. The histologic appearance of muscle shows perifascicular atrophy of muscle fibers and inflammation. (Courtesy Dr. Dennis Burns, Department of Pathology, University of Texas Southwestern Medical School, Dallas, Texas.)

studies may identify an infiltrate rich in CD4+ T-helper cells and the deposition of C5b-9 in capillary vessels. Electron microscopic studies may show tubuloreticular endothelial cell inclusions, a feature of a number of inflammatory disorders that are linked to a type I interferon response.

Clinical Features. Muscle weakness is slow in onset, symmetric, and often accompanied by myalgias. It typically affects the proximal muscles first. As a result, tasks such as getting up from a chair and climbing steps become increasingly difficult. Fine movements controlled by distal muscles are affected only late in the disease. Associated myopathic changes on electrophysiologic studies and elevation in serum creatine kinase levels are reflective of muscle damage. Various rashes are described in dermatomyositis, but the most characteristic ones are a lilac colored discoloration of the upper eyelids *(heliotrope rash)* associated with periorbital edema (Fig. 27-8*A*) and a scaling erythematous eruption or dusky red patches over the knuckles, elbows, and knees (*Gottron papules*). *Dysphagia* resulting from involvement of oropharyngeal and esophageal muscles occurs in one third of the affected individuals, and another 10% of patients have *interstitial lung disease*, which can sometimes be rapidly progressive and lead to death. Cardiac involvement is common, but rarely leads to cardiac failure.

Juvenile and adult forms are recognized. The average age of onset of juvenile dermatomyositis is 7 years, whereas adult cases tend to present from the fourth to sixth decade of life. Dermatomyositis is the most common inflammatory myopathy in children. Compared to adult disease, childhood disease is more likely to be associated with calcinosis and lipodystrophy and less likely to be associated myositis-specific antibodies, cardiac involvement, interstitial lung disease, or an underlying malignancy. As might be expected based on these differences, the overall prognosis is better in children than in adults. From 15% to 24% of adult patients have an associated malignancy, and in such patients dermatomyositis may be viewed as a paraneoplastic disorder.

Polymyositis

Polymyositis is an adult-onset inflammatory myopathy that shares myalgia and weakness with dermatomyositis but lacks its distinctive cutaneous features and is therefore to some degree a diagnosis of exclusion. As in dermatomyositis, patients typically develop symmetric proximal muscle involvement, and there may be inflammatory involvement of the heart and the lungs, as well as similar autoantibodies.

Pathogenesis. The pathogenesis of polymyositis is uncertain, but it is believed to have an immunologic basis. CD8-positive cytotoxic T cells are a prominent part of the inflammatory infiltrate in affected muscle, and it is hypothesized that these cells are the mediators of tissue damage. Unlike dermatomyositis, vascular injury is not believed to have a major role in polymyositis.

MORPHOLOGY

Mononuclear inflammatory cell infiltrates are present, but in contrast to dermatomyositis, these are usually endomysial in location. Sometimes myofibers with otherwise normal morphology appear to be invaded by mononuclear inflammatory cells. Degenerating necrotic, regenerating, and atrophic myofibers are typically found in a random or patchy distribution. The perifascicular pattern of atrophy that is characteristic of dermatomyositis is absent.