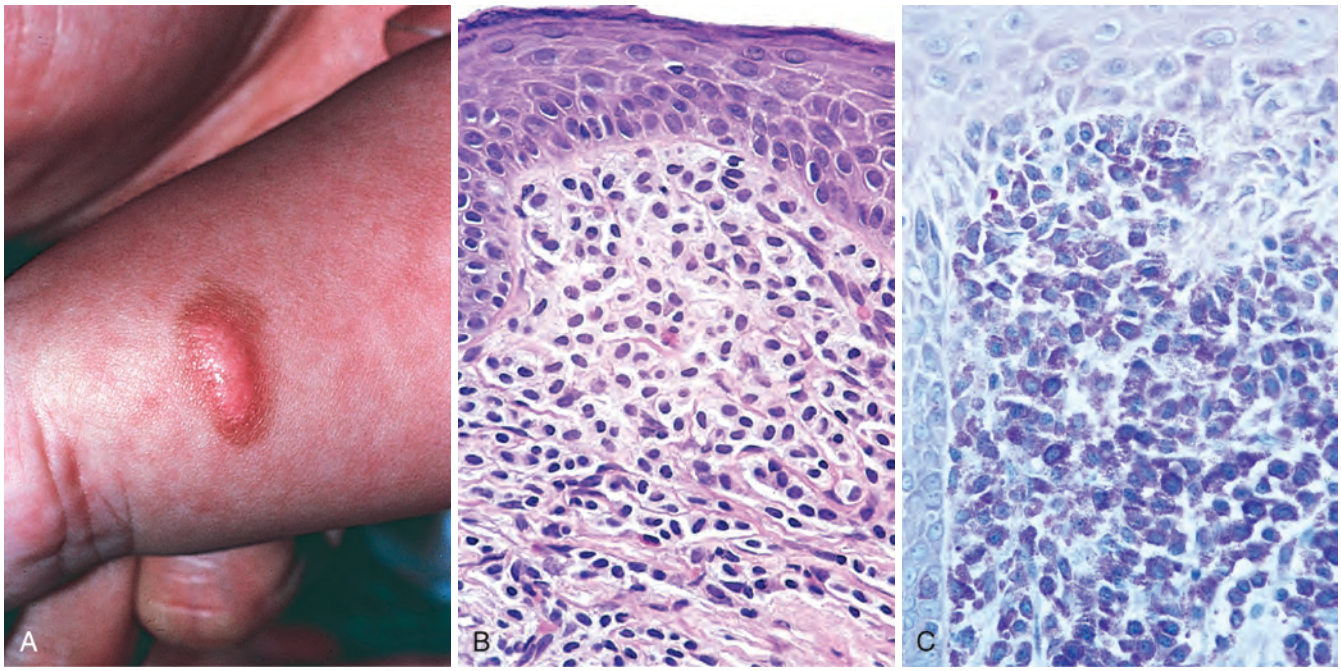


**Figure 25-19** Mastocytosis. **A,** Solitary mastocytoma in a 1-year-old child. **B,** By histology, numerous ovoid cells with uniform, centrally located nuclei are observed in the dermis. **C,** Giemsa staining reveals purple "metachromatic" granules within the cytoplasm of the mast cells.

### MORPHOLOGY

The pathologic findings are highly variable. In **urticaria pigmentosa**, lesions are multiple and widely distributed, consisting of round to oval, red-brown, nonscaling papules and small plaques. Solitary **mastocytoma** presents as a pink to tan-brown nodule that may be pruritic or show blister formation (Fig. 25-19*A*). The histologic picture in urticaria pigmentosa or solitary mastocytoma varies from a subtle increase in the numbers of spindle-shaped and stellate mast cells around superficial dermal blood vessels, to large numbers of tightly packed, round to oval mast cells in the upper to mid-dermis (Fig. 25-19*B*). Fibrosis, edema, and small numbers of eosinophils may also be present. Mast cells may be difficult to differentiate from lymphocytes in routine, hematoxylin and eosin–stained sections, and special metachromatic stains (toluidine blue or Giemsa) must be used to visualize their granules (Fig. 25-19*C*). Even with these stains, extensive degranulation may result in failure to recognize these cells by light microscopy, but their identity can be readily confirmed with immunohistochemical stains for mast cell markers, such as mast cell tryptase and KIT.

## Disorders of Epidermal Maturation

### Ichthyosis

Of the numerous disorders that impair epidermal maturation, ichthyosis is perhaps one of the most striking. The term is derived from the Greek root *ichthy,* meaning "fishy," and accordingly, this group of inherited disorders is associated with chronic, excessive keratin buildup (hyperkeratosis) that results clinically in fishlike scales (Fig. 25-20*A*). The clinical types of ichthyosis vary according to the mode of inheritance, histology, and clinical features; the primary categories include *ichthyosis vulgaris* (autosomal dominant or acquired), *congenital ichthyosiform erythroderma* (autosomal recessive), *lamellar ichthyosis* (autosomal recessive), and *X-linked ichthyosis*. Most ichthyoses become apparent either at or around the time of birth. Acquired (noninherited) variants also exist; one such variant, *ichthyosis vulgaris,* may be associated with lymphoid and visceral malignancies.

***Pathogenesis.*** The primary abnormality in some forms of ichthyosis is defective desquamation, leading to retention of abnormally formed scale. For example, X-linked ichthyosis is caused by a deficiency of steroid sulfatase, an enzyme helps to remove proadhesive cholesterol sulfate from intercellular spaces. In its absence cholesterol sulfate accumulates, resulting in persistent cell-to-cell

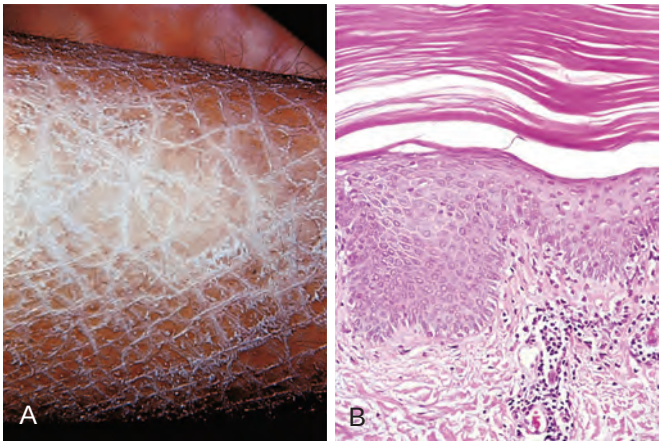


**Figure 25-20** Ichthyosis. Note prominent fishlike scales (**A**) and compacted, thickened stratum corneum (**B**).