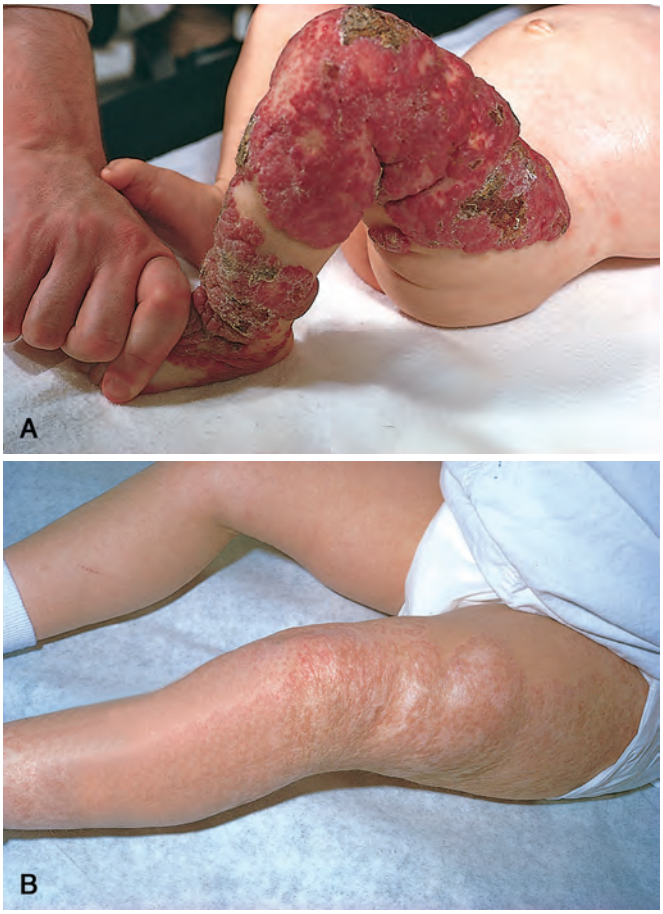

Figure 10-22 Congenital capillary hemangioma at birth (**A**) and at age 2 years (**B**) after spontaneous regression. (Courtesy Dr. Eduardo Yunis, Children's Hospital of Pittsburgh, Pittsburgh, Pa.)

greater detail in appropriate chapters; here a few comments are made about their special features in childhood.

Hemangioma. *Hemangiomas are the most common tumors of infancy* (Chapter 11). Architecturally, they do not differ from those occurring in adults. Both cavernous and capillary hemangiomas may be encountered, although the latter are often more cellular than in adults, a feature that is deceptively worrisome. In children, most are located in the skin, particularly on the face and scalp, where they produce flat to elevated, irregular, red-blue masses; some of the flat, larger lesions (considered by some to represent vascular ectasias) are referred to as *port-wine stains*. Hemangiomas may enlarge along with the growth of the child, but in many instances they spontaneously regress (Fig. 10-22). In addition to their cosmetic significance, they can represent one facet of the hereditary disorder von Hippel-Lindau disease (Chapter 28). A subset of CNS cavernous hemangiomas can occur in the familial setting; these families harbor mutations in one of three *cerebral cavernous malformation* (CCM) genes.

Lymphatic Tumors. A wide variety of lesions are of lymphatic origin. Some of them—*lymphangiomas*—are hamartomatous or neoplastic, whereas others seem to represent abnormal dilations of preexisting lymph channels known as *lymphangiectasis.* The *lymphangiomas* are usually characterized by cystic and cavernous spaces. Lesions of this nature may occur in the skin but are more often encountered in the deeper regions of the neck, axilla, mediastinum, retroperitoneal tissue, and elsewhere. Although histologically benign, they tend to increase in size after birth, owing to the accumulation of fluid and the budding of preexisting spaces. In this manner they may encroach on vital structures, such as those in the mediastinum or nerve trunks in the axilla, and give rise to clinical problems. *Lymphangiectasis*, in contrast, usually presents as a diffuse swelling of part or all of an extremity; considerable distortion and deformation may occur as a consequence of the spongy, dilated subcutaneous and deeper lymphatics. The lesion is not progressive, however, and does not extend beyond its original location. Nonetheless, it creates cosmetic problems that are often difficult to correct surgically.

Fibrous Tumors. Fibrous tumors occurring in infants and children range from sparsely cellular proliferations of spindle-shaped cells (designated as *fibromatosis*) to richly cellular lesions indistinguishable from fibrosarcomas occurring in adults (designated as *congenital-infantile fibrosarcomas*). Biologic behavior cannot be predicted based on histology alone, however, in that despite their histologic similarities with adult fibrosarcomas, the congenital-infantile variants have an excellent prognosis. *A characteristic chromosomal translocation, t(12;15)(p13;q25), has been described in congenital-infantile fibrosarcomas, which results in generation of an ETV6-NTRK3 fusion transcript.* The normal *ETV6* gene product is a transcription factor, while the *NTRK3* gene product (also known as TRKC) is a tyrosine kinase. Like other tyrosine kinase fusion proteins found in human neoplasms, ETV6-TRKC is constitutively active and stimulates signaling through the oncogenic RAS and PI-3K/AKT pathways (Chapter 7). Among soft-tissue tumors, the ETV6-NTRK3 fusion transcript is unique to infantile fibrosarcomas, making it a useful diagnostic marker.

Teratomas. Teratomas illustrate the relationship of histologic maturity to biologic behavior. Teratomas may occur as benign, well-differentiated cystic lesions (mature teratomas), as lesions of indeterminate potential (immature teratomas), or as unequivocally malignant teratomas (usually admixed with another germ cell tumor component such as endodermal sinus tumor) (Chapter 21). They exhibit two peaks in incidence: the first at approximately 2 years of age and the second in late adolescence or early adulthood. The first peak is congenital neoplasms; the later occurring lesions may also be of prenatal origin but are more slowly growing. *Sacrococcygeal teratomas* are the most common teratomas of childhood, accounting for 40% or more of cases (Fig. 10-23). They occur with a frequency of 1 in 20,000 to 40,000 live births, and are four times more common in girls than boys. In view of the overlap in the mechanisms underlying teratogenesis and oncogenesis, it is interesting that approximately 10% of sacrococcygeal teratomas are associated with congenital anomalies, primarily defects of the hindgut and cloacal region and other midline defects (e.g., meningocele, spina bifida) not believed to result from local effects of the tumor.