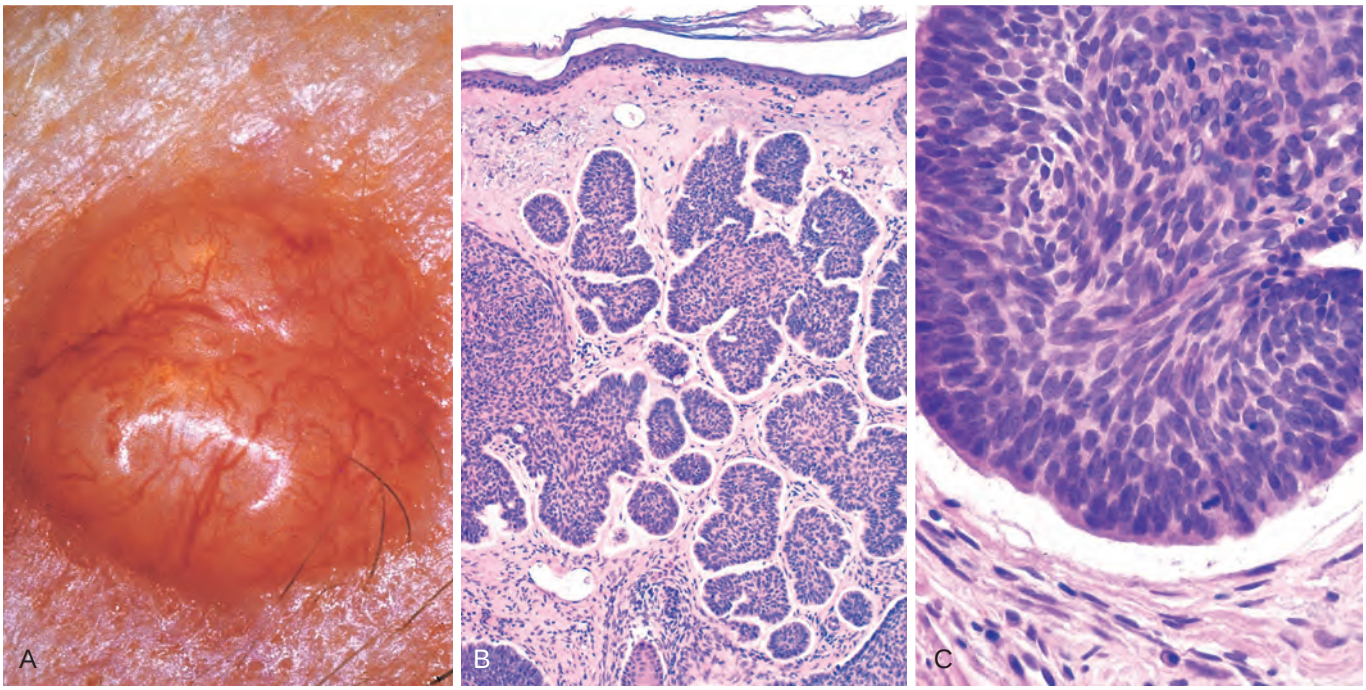

Figure 25-15 Basal cell carcinoma. Pearly, telangiectatic nodules **(A)** are composed of nests of uniform basaloid cells within the dermis **(B)** that are often separated from the adjacent stroma by thin clefts **(C),** an artifact of sectioning.

- Basal cell carcinoma, the most common malignancy worldwide, is a locally aggressive tumor associated with mutations that activate Hedgehog signaling. Metastasis is very rare.

Tumors of the Dermis

The dermis contains a variety of elements such as smooth muscle, pericytes, fibroblasts, neural tissue, and endothelium. Neoplasms comprised of cells resembling all of these elements occur in the skin, but most also involve other soft tissues and viscera and are discussed elsewhere, or are too rare to merit mention. This section discusses two dermal neoplasms—one benign, one malignant—that arise primarily in the skin.

Benign Fibrous Histiocytoma (Dermatofibroma)

Benign fibrous histiocytoma refers to a heterogeneous family of morphologically and histogenetically related benign dermal neoplasms of uncertain lineage. These tumors are usually seen in adults and often occur on the legs of young and middle-aged women. Lesions are asymptomatic or tender and may increase and decrease slightly in size over time. Their biologic behavior is indolent.

The cause of fibrous histiocytomas remains a mystery. Many cases have a history of antecedent trauma, suggesting an abnormal response to injury and inflammation, perhaps analogous to the deposition of increased amounts of altered collagen in a hypertrophic scar or keloid. These common yet curious tumors appear to be composed at least partially of factor XIIIa-positive dermal dendritic cells.

MORPHOLOGY

These neoplasms appear as firm, tan to brown papules (Fig. 25-16*A*). Most are less than 1 cm in diameter, but actively growing lesions may reach several centimeters in diameter; with time they often become flattened.

The most common form of fibrous histiocytoma is referred to as a **dermatofibroma.** These tumors consist of benign, spindle-shaped cells that are usually arranged in a well-defined, nonencapsulated mass within the mid-dermis (Fig. 25-16*B, C*). Extension of these cells into the subcutaneous fat is sometimes observed. Many cases demonstrate a peculiar form of overlying epidermal hyperplasia, characterized by downward elongation of hyperpigmented rete ridges (**pseudoepitheliomatous hyperplasia**). Numerous histologic variants are noted, such as more cellular forms or tumors with pools of extravascular blood and hemosiderin (aneurysmal).

Dermatofibrosarcoma Protuberans

Dermatofibrosarcoma protuberans is best regarded as a well-differentiated, primary fibrosarcoma of the skin. These tumors are slow growing, and although they are locally aggressive and can recur, they rarely metastasize.

Pathogenesis. **The molecular hallmark of dermatofibrosarcoma protuberans is a translocation involving the genes encoding collagen 1A1 *(COL1A1)* and platelet-derived growth factor-β *(PDGFB).*** The resulting rearrangement juxtaposes the *COL1A1* promoter sequences and the coding region of *PDGFB* and leads to overexpression and increased secretion of PDGFβ, which drives tumor cell growth through an autocrine loop. While the primary mode of treatment is wide local excision, rare cases that are