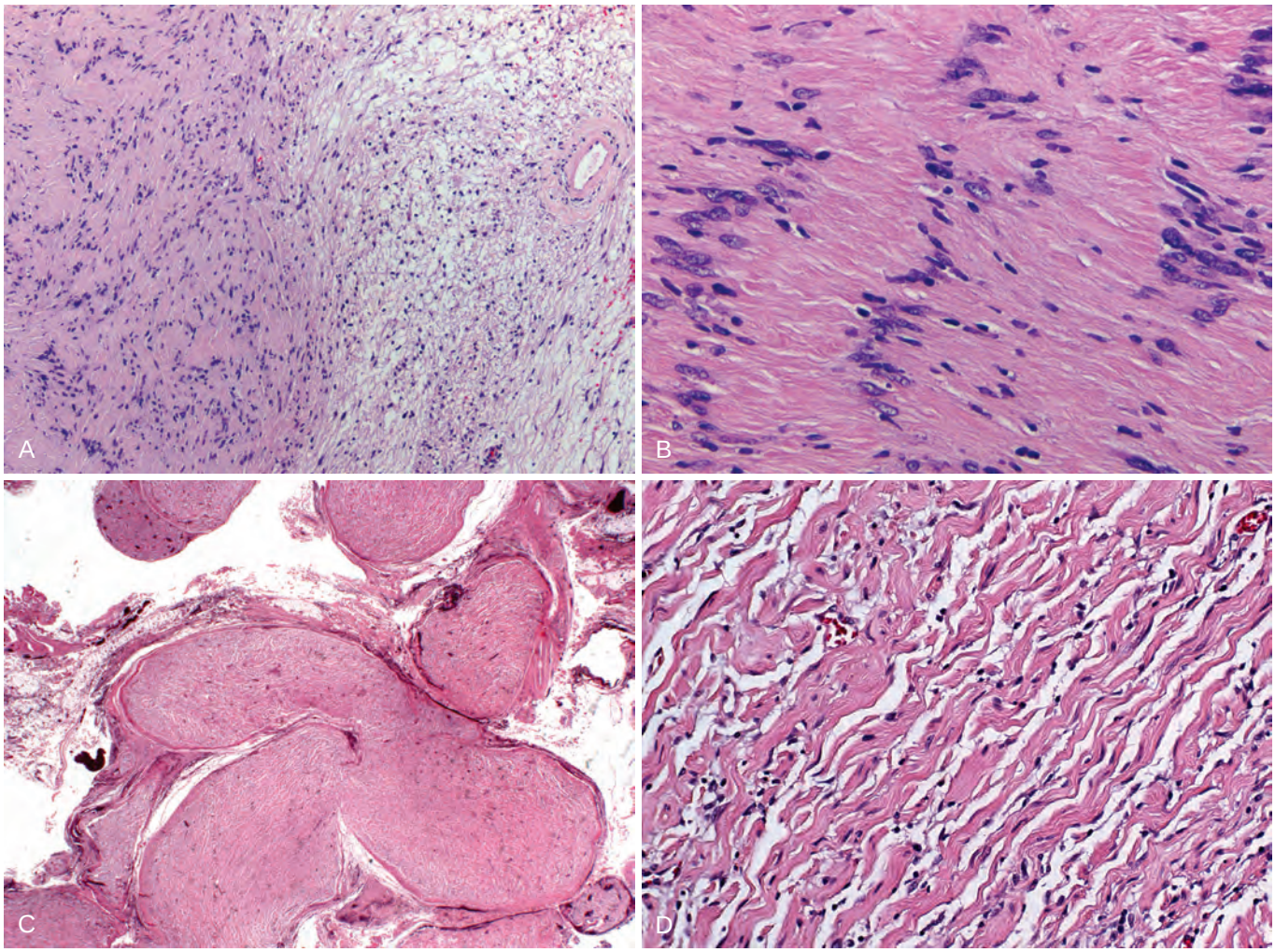

Figure 27-14 Schwannoma and plexiform neurofibroma. **A** and **B,** Schwannoma. As seen in **A,** schwannomas often contain dense eosinophilic Antoni A areas (*left*) and loose, pale Antoni B areas (*right*), as well as hyalinized blood vessels (*right*). **B,** Antoni A area with the tumor cell nuclei aligned in palisading rows leaving anuclear zones and resulting in the formation of structures termed *Verocay bodies*. **C** and **D,** Plexiform neurofibroma. **C,** Multiple nerve fascicles are expanded by infiltrating tumor cells. **D,** At high power bland spindle cells are admixed with wavy collagen bundles resembling carrot shavings.

Diffuse neurofibroma. This tumor has morphologic features similar to those seen in localized cutaneous neurofibromas, but exhibits a distinctly different growth pattern. The tumor diffusely infiltrates the dermis and subcutaneous connective tissue, entrapping fat and appendage structures and producing a plaque-like appearance. Some of these neurofibromas can grow to large sizes. Focal collections of cells mimicking the appearance of Meissner corpuscles (so-called **pseudo-Meissner corpuscles** or **tactile-like bodies**) are an associated feature.

Plexiform neurofibroma. These tumors grow within and expand nerve fascicles (Fig. 27-14*C*), entrapping associated axons. The external perineurial layer of the nerve is preserved, giving individual nodules an encapsulated appearance. The expanded, ropy thickening of multiple nerve fascicles results in what is sometimes referred to as a "bag of worms" appearance. The tumor has cellular composition similar to that of other neurofibromas. The extracellular matrix varies from loose and myxoid to more collagenous and fibrous. Often the collagen is seen in bundles likened to "shredded carrot" (Fig. 27-14*D*).

Malignant Peripheral Nerve Sheath Tumors (MPNST)

Most MPNSTs (approximately 85%) are high-grade tumors, but low-grade variants are recognized. About half arise in NF1 patients and are assumed to result from malignant transformation of a plexiform neurofibroma. Sporadic cases may arise de novo. Most are associated with larger peripheral nerves in the chest, abdomen, pelvis, neck or limb-girdle. MPNSTs exhibit complex chromosomal aberrations, including chromosome gains, losses, and rearrangements. The molecular alterations driving malignant transformation of a neurofibroma to MPNST are still poorly understood.

MORPHOLOGY

The lesions are poorly defined tumor masses that frequently infiltrate along the axis of the parent nerve and invade adjacent soft tissues. A wide range of histologic appearance can be encountered. Typical cases show a fasciculated arrangement