- Paraganglia related to the great vessels of the head and neck, the so-called aorticopulmonary chain, including the *carotid bodies* (most common); aortic bodies; jugulotympanic ganglia; ganglion nodosum of the vagus nerve; and clusters located about the oral cavity, nose, nasopharynx, larynx, and orbit. These are innervated by the parasympathetic nervous system and infrequently release catecholamines.

MORPHOLOGY

The **carotid body tumor** is a prototype of a parasympathetic paraganglioma. It rarely exceeds 6 cm in diameter and arises close to or envelops the bifurcation of the common carotid artery. The tumor tissue is red-pink to brown. The microscopic features of all paragangliomas, wherever they arise, are remarkably uniform. They are chiefly composed of nests **(zellballen)** of round to oval chief cells (neuroectodermal in origin) that are surrounded by delicate vascular septae (Fig. 16-13). The tumor cells contain abundant, clear or granular, eosinophilic cytoplasm and uniform, round to ovoid, sometimes vesicular, nuclei. In most tumors there is little cellular pleomorphism, and mitoses are scant. The chief cells stain strongly for neuroendocrine markers such as chromogranin, synaptophysin, neuron-specific enolase, CD56, and CD57. In addition, there is a supporting network of spindle-shaped stromal cells, collectively called sustentacular cells, which are positive for S-100 protein. Electron microscopy often discloses well-demarcated neuroendocrine granules in paravertebral tumors, but their number can be highly variable and they tend to be scant in nonfunctioning tumors.

Carotid body tumors (and paragangliomas in general) are rare. They are slow-growing and painless masses that usually arise in the fifth and sixth decades of life. They commonly occur singly and sporadically but may be familial, with autosomal dominant transmission in the multiple endocrine neoplasia 2 syndrome (Chapter 24); in this setting they are often multiple and sometimes bilateral. Carotid body tumors frequently recur after incomplete resection and, despite their benign appearance, may metastasize to regional lymph nodes and distant sites. About 50% ultimately prove fatal, largely because of infiltrative growth. Unfortunately, it is almost impossible to predict the clinical course of a carotid body tumor—mitoses, pleomorphism, and even vascular invasion are not reliable indicators.

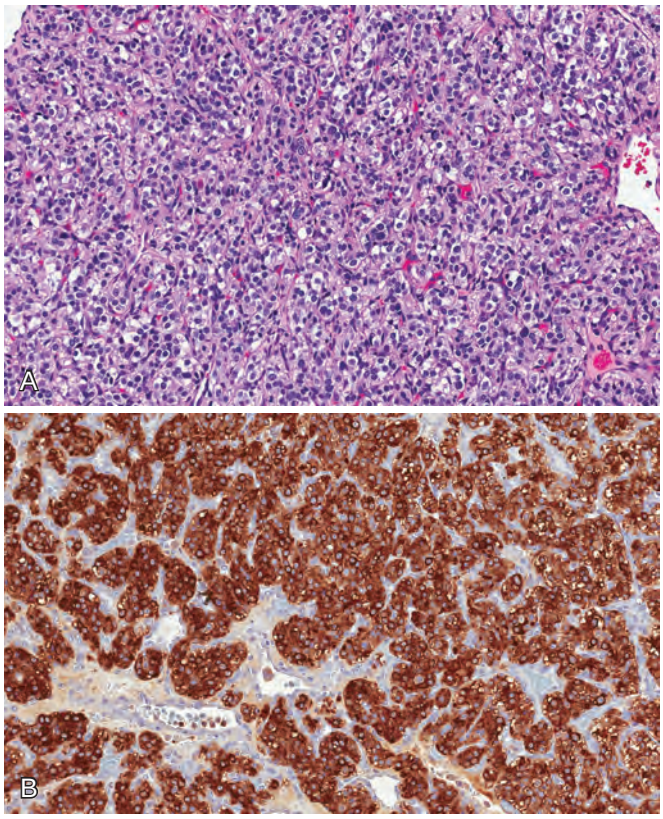

Figure 16-13 Carotid body tumor. **A,** Low-power view showing tumor clusters separated by septa (Zellballen). The septae are marked by bright red capillaries. **B,** Immunohistochemistry demonstrating positivity for chromogranin in the tumor cells.

KEY CONCEPTS

Neck

- **Branchial cysts** arise from the region of the second branchial pouch and are typically seen in young adults.
- **Thyroglossal duct cysts** arise as a result of incomplete descent of the thyroid analog from the foramen cecum in the base of the tongue.
- Seventy percent of **paragangliomas** are observed in the head and neck region that often develop in the fifth decade of life. While often sporadic in appearance, they may be associated with multiple endocrine neoplasia 2 syndrome.

SALIVARY GLANDS

There are three major salivary glands—parotid, submandibular, and sublingual—as well as innumerable minor salivary glands distributed throughout the mucosa of the oral cavity. Inflammatory or neoplastic disease may develop within any of these.

Xerostomia

Xerostomia is defined as a *dry mouth* resulting from a decrease in the production of saliva. Its incidence among various populations has been reported to be as high as 20% in individuals over the age of 70. It is a major feature of the autoimmune disorder Sjögren syndrome (Chapter 6), in which it is usually accompanied by dry eyes. A lack of salivary secretions is also a major complication of radiation therapy. However, xerostomia is most frequently a side-effect of many commonly prescribed classes of medications, including anticholinergic, antidepressant/antipsychotic, diuretic, antihypertensive, sedative, muscle relaxant, analgesic, and antihistamine drugs. Xerostomia may present as dry mucosa and/or atrophy of the papillae