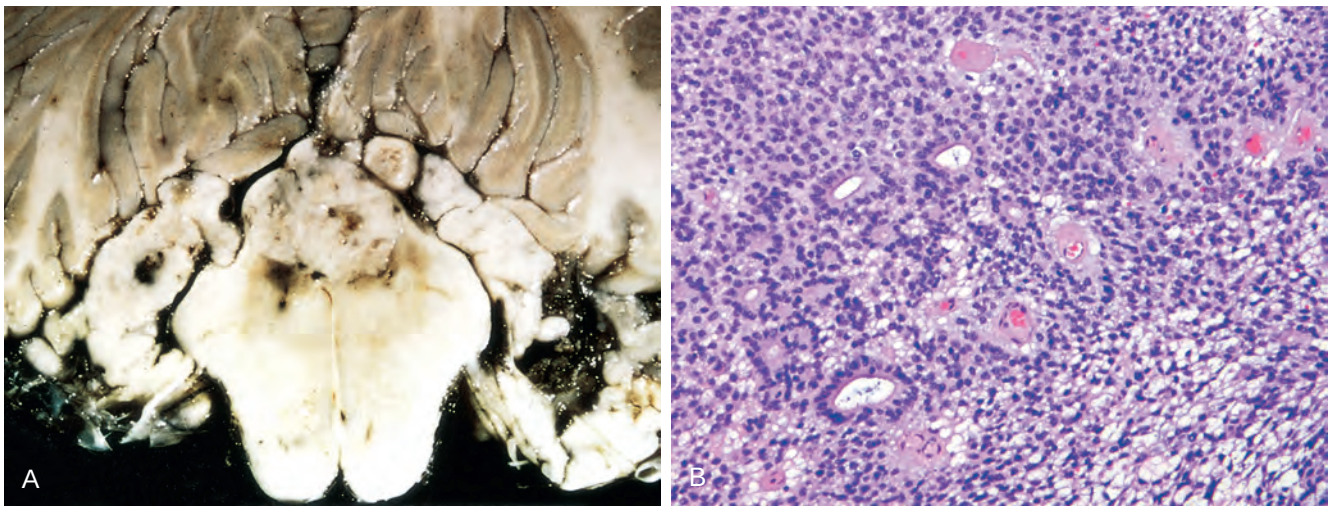

Figure 28-51 Ependymoma. **A,** Tumor growing into the fourth ventricle, distorting, compressing, and infiltrating surrounding structures. **B,** Microscopic appearance of ependymoma.

Myxopapillary ependymomas are distinct but related lesions that occur in the filum terminale of the spinal cord and contain papillary elements in a myxoid background, admixed with ependymoma-like cells. Cuboidal cells, sometimes with clear cytoplasm, are arranged around papillary cores containing connective tissue and blood vessels. The myxoid areas contain neutral and acidic mucopolysaccharides. Prognosis depends on completeness of surgical resection; if the tumor has extended into the subarachnoid space and surrounded the roots of the cauda equina, recurrence is likely.

Clinical Features. Posterior fossa ependymomas often manifest with hydrocephalus secondary to progressive obstruction of the fourth ventricle. Because of the relationship of ependymomas to the ventricular system, CSF dissemination is a common occurrence and portends a poor prognosis. Posterior fossa lesions have the worst overall outcome, particularly in younger children, in whom the 5-year survival is roughly 50%. The clinical outcome for completely resected supratentorial and spinal ependymomas is better.

Several other tumors occur either immediately below the ependymal lining of the ventricle or in association with the choroid plexus, which sits in continuity with the ependyma. With the exception of the rare choroid plexus carcinoma, these are benign; however, because of location they may cause clinical problems.

- *Subependymomas* are solid, sometimes calcified, slow-growing nodules attached to the ventricular lining and protruding into the ventricle. They are usually asymptomatic and are incidental findings at autopsy or imaging; if they are sufficiently large or strategically located, they may cause hydrocephalus. They are most often found in the lateral and fourth ventricles and have a characteristic microscopic appearance, with clusters of ependymal-appearing nuclei scattered in a dense, fine, glial fibrillar background.
- *Choroid plexus papillomas* may occur anywhere along the choroid plexus and are most common in children, in whom they are usually found in the lateral ventricles. In adults, they more often involve the fourth ventricle. These papillary growths almost exactly recapitulate the structure of the normal choroid plexus. The papillae have connective tissue stalks covered with a cuboidal or columnar epithelium. Clinically, choroid plexus papillomas usually present with hydrocephalus due to obstruction of the ventricular system by tumor or overproduction of CSF. The far rarer *choroid plexus carcinomas* resemble adenocarcinoma. These tumors are usually found in children; in adults, they must be differentiated from metastatic carcinoma, which is much more common than primary carcinomas of the choroid.
- *Colloid cyst of the third ventricle* is a non-neoplastic enlarging cyst that most often occurs in young adults. The cyst is attached to the roof of the third ventricle, where it can obstruct one or both of the foramina of Monro and, as a result, causes noncommunicating hydrocephalus which may be rapidly fatal. Headache, sometimes positional, is an important clinical symptom. The cyst has a thin, fibrous capsule and a lining of low to flat cuboidal epithelium; it contains gelatinous, proteinaceous material.

Neuronal Tumors

Far less common than glial tumors are those that exhibit neuronal differentiation. In general, neuronal tumors are more often seen in younger adults and often present with seizures.

- *Gangliogliomas* are tumors comprised of a mixture of mature neuronal and glial cells. They are typically superficial lesions that present with seizures. These are the most common of the neuronal tumors of the CNS. Most of these tumors are slow growing, but the glial component occasionally becomes anaplastic, and the disease then progresses rapidly. When gangliogliomas present because of a seizure disorder, surgical resection of the tumor is usually effective in controlling the seizures. Approximately 20% of these tumors have an