



Figure 29-24 Retinoblastoma. **A**, Gross photograph of retinoblastoma. **B**, Tumor cells appear viable when in proximity to blood vessels, but necrosis is seen as the distance from the vessel increases. Dystrophic calcification (*dark arrow*) is present in the zones of tumor necrosis. Flexner-Wintersteiner rosettes—arrangements of a single layer of tumor cells around an apparent “lumen”—are seen throughout the tumor, and one such rosette is indicated by the *white arrow*.

KEY CONCEPTS

- **Retinal detachment**, a separation of the neurosensory retina from the retinal pigment epithelium, may be the consequence of a break in the retina (rhegmatogenous retinal detachment) or may develop without a retinal break because of pathology within or beneath the retina (non-rhegmatogenous retinal detachment).
- The clinical appearance of the retina by ophthalmoscopy can be linked to specific pathologic changes: the change in caliber and color of retinal blood vessels may reflect varying degrees of arteriosclerosis and the location of hemorrhages and exudates in the retina is related to their locations within the retinal layers.
- Several major causes of blindness result from pathologic intraocular angiogenesis including proliferative diabetic retinopathy and exudative (wet) age-related macular degeneration, among many other conditions. VEGF antagonists may prevent visual loss in many of these conditions.
- Retinoblastoma is the most common primary intraocular tumor of children.
- Primary retinal lymphoma is a aggressive tumor that often involves the brain as well.

Optic Nerve

As a sensory tract of the central nervous system, the optic nerve is surrounded by meninges, and cerebrospinal fluid circulates around the nerve. The pathology of the optic nerve is similar to the pathology of the brain. For example, the most common primary neoplasms of the optic nerve are glioma (typically *pilocytic astrocytomas*) and meningioma.

Anterior Ischemic Optic Neuropathy

There are striking similarities between stroke and a condition known in ophthalmic terminology as *anterior ischemic optic neuropathy* (AION). As used clinically, the

term AION includes a spectrum of injuries to the optic nerve varying from ischemia to infarction. Thus, transient partial interruptions in blood flow to the optic nerve can produce episodes of transient loss of vision, whereas total interruption in blood flow can give rise to an optic nerve infarct which may be segmental or total. Zones of relative ischemia may surround segmental infarcts of the optic nerve. Optic nerve function in these poorly perfused but not infarcted zones may recover. The optic nerve does not regenerate, and visual loss from infarction is permanent.

Interruption in the blood supply to the optic nerve can result from inflammation of the vessels that supply the optic nerve or from embolic or thrombotic events. Bilateral total infarcts of the optic nerve resulting in total blindness have been reported in temporal arteritis, adding urgency to the treatment of this condition with high doses of corticosteroids.

Papilledema

Edema of the head of the optic nerve may develop as a consequence of compression of the nerve (as in a primary neoplasm of the optic nerve when swelling of the nerve head produces unilateral disc edema) or from elevations of cerebrospinal fluid pressure surrounding the nerve (resulting typically in bilateral disc edema). The concentric increase in pressure encircling the nerve contributes to venous stasis and also interferes with axoplasmic transport, leading to nerve head swelling. Swelling of the optic nerve head in elevated intracranial pressure is typically bilateral and is commonly termed *papilledema*. Typically, acute papilledema from increased intracranial pressure is not associated with visual loss. Ophthalmoscopically, the optic nerve head is swollen and hyperemic; by contrast, the optic nerve head in the relatively acute phases of anterior ischemic optic neuropathy appears swollen and pale because of decreased nerve perfusion (Fig. 29-25). In papilledema secondary to increased intracranial pressure, the optic nerve may remain congested for a prolonged period of time.