Functions

• Special afferent (SA) for vision.

Anatomy (Figs. 2.1, 2.2)

Retina

• Divided into four quadrants by horizontal and vertical meridians (superior/inferior and nasal/temporal)
• Phototransduction occurs by photoreceptors (rods and cones), which contain light-sensitive pigments. Rods are absent from macula and optic disc (blind spot). Cones (color vision, three types: R, G, B) (~7 million cones) are tightly packed in the macula (100,000), which is at the posterior pole 4 mm lateral to optic disc and accounts for most visual acuity.
• Flow of information in retina is from photoreceptors to bipolar cells to ganglion cells. Other cell types in retina include horizontal cells (neurons), amacrine cells (neurons) and Müller cells (glial cells) that are thought to modulate the signal (e.g., surround inhibition).
• The axons of retinal ganglion cells form the optic nerve at the optic disc. Note that, like the olfactory nerve and unlike other cranial nerves, the optic nerve is really a CNS tract.
Optic Nerves and Optic Chiasm

- Optic nerve (Figs. 2.3, 2.4) contains 1 million fibers (comparison: cochlear nerve 50,000 fibers). Macular fibers are on the temporal side of the optic disc and the adjacent optic nerve and move to the central part of the nerve as the papillomacular bundle for most of the distal pathway. The larger nonmacular fibers are on the periphery.
- Optic nerve is 50 mm long and has four portions:
  1. Intraocular (1 mm) (also called optic nerve head). Axons become myelinated.
  2. Intraorbital (25 mm). From back of globe to optic canal.
  3. Intracanalicular (9 mm). Traverses optic canal with ophthalmic artery and sympathetic plexus.
  4. Intracranial (4 to 16 mm). Lies superior to the internal carotid artery (ICA) as ICA exits cavernous sinus and gives off ophthalmic artery. The sphenoid sinus is inferomedial, whereas the anterior cerebral artery (ACA) (A1 segment), gyrus rectus, olfactory tract, and anterior perforated substance lie superior.
- See Table 2.1 for a summary of vascular supply of the visual pathway.
- The two optic nerves converge at the optic chiasm where the nasal axons from the nasal retina decussate and the axons from the temporal retina remain ipsilateral.

Fig. 2.2 Slightly more detailed view of visual pathway demonstrates the retinocollicular and retinopretectal tracts as well as the retinogeniculate pathway. Not shown is the retinohypothalamic tract (to suprachiasmatic nucleus (SCN) of hypothalamus) (see text).

Fig. 2.3 Axial T2-weighted image demonstrates high signal intensity fluid within the ocular globes. The right lens is well seen (black straight arrow). Normal optic nerve (white concave arrow) is seen surrounded by perioptic fluid in the optic sheath. More posteriorly, the nerves traverse the optic canal (white arrowhead) to meet at the optic chiasm (black arrowhead) in the midline. Note that the pituitary stalk (small black curved arrow) is located just posterior to the optic chiasm.

Fig. 2.4 Coronal T2-weighted image shows normal optic nerves entering the optic canals. The right optic nerve is indicated (arrow). The nerves lie medial to the anterior clinoid processes (arrowhead) and just above the flow void of the internal carotid artery.
Positioning of the optic chiasm: The prefixed chiasm (9%) lies over the tuberculum sellae, 80% over the sella turcica (L., “Turkish saddle”); the postfixed chiasm (11%) lies over the dorsum sellae.

Retinal Targets
- There are four brain regions that directly receive visual information from the retina:
  1. Retinogeniculate pathway. Primary pathway for visual information; to lateral geniculate nucleus (LGN) of thalamus for conscious vision
  2. Retinopretectal tract. Retina to pretectal area of mid-brain for pupillary light reflex (see Appendix B)
  3. Retinocollicular tract. To superior colliculus for eye movement reflexes (e.g., conjugate eye movements in response to head movements)
  4. Retinohypothalamic tract. To bilateral suprachiasmatic nuclei (SCN) of hypothalamus for circadian rhythms and neuroendocrine function

Optic Tracts
- Extend from chiasm posterolaterally around hypothalamus, around cerebral peduncles to lateral geniculate nuclei

Vascular relationships: Posterior communicating artery (PCommA) located inferior to optic tracts in suprasellar cistern, posterior cerebral artery (PCA) and basal vein of Rosenthal apposed to optic tracts in perimesencephalic cistern

Table 2.1 Vascular Supply of Visual Pathway

<table>
<thead>
<tr>
<th>Structure</th>
<th>Vascular Supply</th>
</tr>
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<tbody>
<tr>
<td>Retina</td>
<td>• The ophthalmic artery branches from the supraclinoid ICA and lies below and lateral to CN II in the optic canal. Five to 15 mm from the globe it gives off central retinal artery (CRA) which pierces CN II and continues forward in its core to divide into superior and inferior branches at the optic disc. Second-order nasal and temporal branches supply nerve fiber layer and inner retina (including ganglion cells). Infarction in territory of CRA may be caused by emboli, thrombi, hypercoagulable states, migraine, and arteritis. • Ophthalmic artery also gives off dural branches (anterior falcine and recurrent meningeal arteries), orbital branches, short posterior ciliary arteries (outer retinal layers, sclera, rods, cones) and long posterior ciliary arteries (ciliary body and iris), which form the anastomotic network, supply some of the optic disc, and 50% supply the macula.</td>
</tr>
<tr>
<td>Optic nerve</td>
<td>• Proximal part supplied by small branches of ophthalmic artery (pial plexus)</td>
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<tr>
<td>Optic chiasm</td>
<td>• Superior part supplied by perforators from ACommA</td>
</tr>
<tr>
<td>Optic tract</td>
<td>• Inferior part supplied by perforators from ICA, PCommA, PCA</td>
</tr>
<tr>
<td>Lateral geniculate nucleus</td>
<td>• Lateral part supplied by anterior choroidal artery</td>
</tr>
<tr>
<td>Optic radiations</td>
<td>• Medial part supplied by lateral posterior choroidal artery</td>
</tr>
<tr>
<td>Visual cortex</td>
<td>• Upper (parietal) supplied by MCA branches</td>
</tr>
<tr>
<td></td>
<td>• Lower (temporal) supplied by PCA branches</td>
</tr>
</tbody>
</table>

Abbreviations: ACA, anterior cerebral artery; ACommA, anterior communicating artery; CN, cranial nerve; CRA, central retinal artery; ICA, internal carotid artery; MCA, middle cerebral artery; PCA, posterior cerebral artery; PCommA, posterior communicating artery.
Lateral Geniculate Nucleus (Fig. 2.6)
- In the inferolateral thalamus lateral to the upper midbrain
- Contains tertiary neurons that form the optic radiations to the primary visual cortex surrounding the calcarine fissure
- Contains six laminae (I–VI). I, IV, VI from contralateral eye; II, III, V from ipsilateral eye

Optic Radiations (Geniculocalcarine Tracts) (Figs. 2.1, 2.2, 2.7)
- Formed by axons of LGN neurons projecting to calcarine cortex of occipital lobe.
- Sweep posteriorly around lateral and posterior portion of lateral ventricles.
- Parietal lobe optic radiations start in the medial LGN and take a direct, nonlooping course over the top of the inferior horn of the lateral ventricle and then travel posteriorly through the parietal white matter to the superior calcarine cortex (subserve inferior visual fields).
- Temporal lobe optic radiations start in the lateral LGN, course anteriorly toward the temporal pole before turning posteriorly in a sharp loop (Meyer’s loop) to course around the lateral wall of the inferior horn of the lateral
ventricle then posteriorly to the inferior calcarine cortex (subserve superior visual fields). They account for contralateral superior quadrantanopia following temporal lobectomy.
- Ventral optic radiations are closely associated with the posterior limb of the internal capsule.

Visual Cortex and Visual Association Areas
- Primary visual cortex (area 17) in walls and floor of calcarine sulcus in medial occipital lobe.
- Geniculocalcarine afferent input to layer IV of six-layered neocortex.
- On cross-section, white matter stria (stria of Gennari, thick white matter band in layer IV) can be seen with naked eye (thus striate cortex).
- Macula projects to posterior one third of the calcarine cortex (occipital pole). Macula represented posteriorly, peripheral retina anteriorly (fovea at occipital pole), upper-field inferior, right field on left.
- Primary visual cortex projects to secondary visual areas (peristriate cortex) V2 (area 18) and V3 (area 19) in adjacent occipital lobe cortex.

Optic Nerve: Normal Images (Figs. 2.3, 2.4, 2.5, 2.6)

Optic Pathway Lesions
- The most common visual disorders are pathologies of the globe and vary by age:
  - Children: nearsightedness (myopia)
  - Adults: farsightedness (hyperopia)
  - Elderly: cataracts, glaucoma, retinal hemorrhages/detachment, macular degeneration
- Episodic visual loss in early adulthood is usually caused by migraines and in late adulthood by transient ischemic attacks (TIAs).

Nonneurologic Causes of Visual Loss (related to abnormal light transmission through the eye)
- Cornea. Scarring, deposits, infection, ulceration, and trauma.
- Anterior chamber. Hemorrhage, infection, open-angle glaucoma (90% of glaucoma cases; drainage pathway is partially open, there is gradual visual loss, and the eye looks normal), closed-angle glaucoma (increased intraocular pressure, red and painful eye).
- Vitreous humor. Hemorrhage from retinal or ciliary vessels due to trauma, arteriovenous malformation (AVM), or aneurysm (Terson syndrome, usually from ruptured anterior communicating artery (ACommA) aneurysm). “Floaters” are opacities in the vitreous humor. A sudden increase in floaters with a flash of light occurs with retinal detachment.
- Uveitis. Inflammation of the uvea (consists of iris, ciliary body, and choroid layer); accounts for ~10 to 15% of all cases of total blindness in the United States and has many causes, including inflammatory disease (e.g., systemic lupus erythematosus, sarcoidosis, rheumatoid arthritis, ulcerative colitis, Behçet disease), infection (e.g., cytomegalovirus [CMV], tuberculosis [TB], Toxoplasma, Histoplasma), and autoimmune disease (e.g., multiple sclerosis [MS]).

Types

Retinal Pathologies
- Hypertensive retinopathy. Associated with arteriolar narrowing, A-V nicking, hemorrhage and “cotton wool” exudates, and copper- or silver-wiring.
- Senile macular degeneration. Leading cause of blindness in the elderly. The hereditary form may also occur in children and young adults. Usually spares the peripheral retina.
- Retinal detachment. Fluid collects between the sensory retina and the retinal pigment epithelium.
- Amaurosis fugax. Sudden onset of transient monocular blindness usually from a small fibrin embolus.
- Retinitis pigmentosa. Begins in childhood and adolescence, male preponderance. Autosomal recessive (AR) or autosomal dominant (AD) inheritance on chromosome 3. Degeneration of all retinal layers (neuroepithelium and pigment epithelium) with foveal sparing. Starts with impairment of twilight vision (nyctalopia) progressing to blindness.
  - Usher syndrome. AR, retinitis pigmentosa with congenital hearing loss as well.
  - Lawrence-Moon-Biedl syndrome. AR, retinitis pigmentosa, obesity, congenital heart disease, developmental delay, polydactyly, renal disease, diabetes insipidus.
  - Cockayne syndrome. AR, retinitis pigmentosa, cataracts, photosensitivity, pendular nystagmus, ataxia, severe cerebellar atrophy, striatocerebellar calcifications, proagnathism, anhidrosis, poor lacrimation.
  - Stargardt disease. Onset at 6 to 20 years of age; causes slow macular degeneration, especially of the central cones (opposite of retinitis pigmentosa).
Optic Nerve Pathologies

- **Retinopathy of prematurity (retrolental fibroplasia).** Vascular proliferative disease of the retina originally related to oxygen therapy to premature infants. Risk factors are low birth weight and prematurity.
- **Retinoblastoma.** Congenital malignant retinal tumor. Presents with leukocoria (white pupil).
- **Subarachnoid hemorrhage (SAH).** Associated with hemorrhage between the internal limiting membrane and the vitreous fibers (subhyaloid or preretinal hemorrhages).
- **Roth spot.** A pale spot in the retina from the accumulation of white blood cells (WBC) and fibrin, associated with subacute bacterial endocarditis (SBE) or embolic plaques.

**Central retinal artery.** Produces an altitudinal field deficit, flame hemorrhage, and edema with disc atrophy. One third of cases are bilateral, and are usually associated with diabetes or hypertension.

- **Toxic and nutritional optic neuropathies.** Cause bilateral, symmetric central or centrocecal scotomas (unlike demyelinating disease) with normal peripheral fields. Caused by B vitamin deficiencies, ethanol (EtOH), methanol, and other toxins. Patients do well if treated early.

- **Traumatic optic neuropathy.** Can occur at any point from posterior globe to optic chiasm, but usually occurs at the level of the optic canal. Can result from direct nerve injury or vascular compromise. Optic canal fractures are frequently associated with concomitant ICA injury with the risk of developing a carotid-cavernous fistula (CCF).

- **Optic nerve glioma.** Presents in children (usually <10 years of age) with visual dysfunction, hypothalamic dysfunction, and potentially obstructive hydrocephalus; associated with neurofibromatosis type 1 (NF-1) (see Case 2.5 below).

Visual Field Deficits (Fig. 2.7)

- **Hemianopia.** Blindness in half the visual field.
- **Homonymous.** Same field is involved in each eye (left or right).
- **Concentric visual field constriction** (tunnel vision). May be psychogenic or caused by glaucoma, papilledema, and retinitis pigmentosa. If psychogenic, the field does not change with distance.

- **Prechiasmatic lesion deficits.**
  - Monocular blindness (if entire nerve involved).
  - Scotoma. An island of decreased vision surrounded by normal vision. Central scotoma involves a fixation point and cecocentral is a fixation point connected to the blind spot.
  - Visual loss extending out to the periphery.

- **Chiasmatic lesion deficits.**
  - Junctional scotoma. Caused by a lesion at the optic nerve/chiasm border, resulting in ipsilateral central scotoma and a contralateral superior temporal quadrant defect (“pie in the sky”).
  - Bitemporal hemianopsia. Due to sellar and suprasellar masses, sarcoidosis, aneurysms, and Langerhans cell histiocytosis.

- **Retrochiasmatic lesion deficits.**
  - Homonymous deficits.
  - Congruous deficits. Means an identical field defect in each eye. Lesions closer to the cortex create more congruous deficits.
  - **Meyer’s loop.** Temporal lobe lesion involving fibers from the superior field causes a pie in the sky deficit (contralateral superior quadrant defect).
  - **Macular sparing.** Caused by a PCA stroke with the occipital pole being supplied by middle cerebral artery (MCA) collaterals.

- **Visual Field De-**
  - **Central disc edema.**
    - Bilateral. Papilledema (90% of cases), papillitis, optic neuritis, ischemic optic neuropathy, neuroretinitis (disc edema with an inflammatory macular star), inflammatory disease, infiltrative disease.
  - Unilateral. Optic neuritis, asymmetric papilledema, ischemic optic neuropathy, inflammatory disease, infiltrative disease, Foster-Kennedy syndrome (ipsilateral disc edema and contralateral disc atrophy caused by frontal masses), neuroretinitis.
  - Papillitis is optic disc edema associated with inflammatory or demyelinating diseases.
  - Papilledema is optic disc edema associated with increased intracranial pressure. Causes enlarged visual blind spot and constricted visual field without visual acuity change. Develops as increased cerebrospinal fluid (CSF) pressure in the optic sheath compresses nerve fibers, resulting in axonal swelling and leaking. Unilateral papilledema can occur with optic nerve tumors. Visual acuity and pupillary light reflexes are usually normal (especially early).

- **Optic neuritis.** Inflammation, infection, and/or demyelination of the optic nerve. Presents with rapid partial or total loss of vision in one or both eyes. It usually occurs in young adults with scotoma in macular area and blind spot (cecocentral scotoma), may be associated with retrobulbar neuritis or papillitis, and local tenderness or pain is present with eye movements. Magnetic resonance imaging (MRI) may reveal T2 prolongation and gadolinium enhancement associated with the involved nerve(s). It is the first symptom of MS in 15% of cases. Vision returns to normal within a few weeks in two thirds of cases. Steroids speed the recovery (intravenous methylprednisolone followed by oral prednisone). Dyschromatopsia is a common persistent problem. In children, it is more frequently bilateral and of a viral or post-viral etiology. Differential diagnosis includes MS, acute disseminated encephalomyelitis (ADEM), syphilis, Lyme disease, vasculitis, neurosarcoidosis, and prior radiation.

- **Ischemic optic neuropathy.** Most common cause of painless monocular blindness in patients older than 50 years. It has abrupt onset and is caused by occlusion of the
- **Bilateral central scotomas.** Caused by occipital pole strokes.
- **Homonymous altitudinal hemianopsias.** Caused by bilateral occipital strokes. Monocular altitudinal deficits are usually caused by ischemic optic neuropathies.
- See Table 2.2 for differential diagnosis of lesions affecting the visual pathway.

### Optic Nerve: Pathologic Images

#### Case 2.1

A 3-month-old girl was found to have leukocoria in the left eye (Figs. 2.8, 2.9).

**Diagnosis**

Retinoblastoma

#### Table 2.2 Imaging Differential Diagnosis of Visual Pathway Lesions

<table>
<thead>
<tr>
<th>Location</th>
<th>Lesions</th>
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<tbody>
<tr>
<td>Prechiasmatic</td>
<td>Retina</td>
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<td></td>
<td>Retinoblastoma</td>
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<tr>
<td></td>
<td>Retinal detachment</td>
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<tr>
<td></td>
<td>Optic nerve/sheath tumors</td>
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<tr>
<td></td>
<td>Optic glioma</td>
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<tr>
<td></td>
<td>Optic nerve sheath meningioma</td>
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<tr>
<td></td>
<td>Inflammatory lesions of optic nerve/sheath</td>
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<tr>
<td></td>
<td>Optic neuritis</td>
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<td></td>
<td>Orbital pseudotumor</td>
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<tr>
<td></td>
<td>Sarcoidosis</td>
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<tr>
<td></td>
<td>Orbital masses extrinsic to optic nerve</td>
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<tr>
<td></td>
<td>Hemangioma</td>
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<tr>
<td></td>
<td>Lymphangioma</td>
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<tr>
<td></td>
<td>Graves disease (enlarged extraocular muscles)</td>
</tr>
<tr>
<td></td>
<td>Pseudotumor (may or may not involve muscles)</td>
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<tr>
<td></td>
<td>Malignant tumor</td>
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<tr>
<td></td>
<td>Metastatic tumor</td>
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<tr>
<td></td>
<td>Lymphoma</td>
</tr>
<tr>
<td></td>
<td>Rhabdomyosarcoma</td>
</tr>
<tr>
<td>Chiasmatic</td>
<td>Pituitary adenoma</td>
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<td>Rathke cleft cyst</td>
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<td>Optic glioma</td>
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<td></td>
<td>Craniohypophyseoma</td>
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<tr>
<td></td>
<td>Parasellar meningioma</td>
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<tr>
<td></td>
<td>Aneurysm (ICA, ophthalmic, ACA, ACommA)</td>
</tr>
<tr>
<td>Retrochiasmatic</td>
<td>Infarction (e.g. anterior choroidal artery, PCA)</td>
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<tr>
<td></td>
<td>Neoplasm, either primary glial or metastatic</td>
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<tr>
<td></td>
<td>Multiple sclerosis/ADEM (demyelinating disease)</td>
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<td></td>
<td>AVM</td>
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<tr>
<td></td>
<td>Temporal lobectomy</td>
</tr>
</tbody>
</table>

**Abbreviations:** ACA, anterior cerebral artery; ACommA, anterior communicating artery; ADEM, acute disseminated encephalomyelitis; ICA, internal carotid artery; PCA, posterior cerebral artery.

#### Fig. 2.8 Axial 3-D fast spin echo T2-weighted image shows a large mass within the left globe extending to the level of the optic disc and nerve (arrowhead). Note the smaller retinal-based mass (arrow) within the right globe near the macula in this patient with bilateral retinoblastoma.

#### Fig. 2.9 Postcontrast fat-saturated T1-weighted image of the same patient as in Fig. 2.8 shows homogeneous enhancement of both large and small (arrow) masses.

#### Retinoblastoma

- **Epidemiology.** Most common malignant extracranial solid tumor in children. Ninety percent occur before 5 years of age. Genetic predisposition due to loss of the Rb tumor suppressor gene (chromosome 13q), then a spontaneous mutation of Rb on the other (normal) chromosome (“two-hit” hypothesis).
**Acute Disseminated Encephalomyelitis**

- **Epidemiology.** Monophasic demyelination that typically occurs after a viral illness or vaccination (smallpox, rabies, varicella, rubella).
- **Clinical presentation.** Acute focal neurologic deficits related to CNS demyelination, often including headache, meningeal signs, and seizures. Unilateral or bilateral optic neuritis (painful change/loss in vision) is common.
- **Imaging.** Orbital imaging may demonstrate edema and/or enhancement of the optic nerve(s) or chiasm. Abnormal brain/spinal cord findings include multifocal areas of T2 hyperintensity in the white matter, with variable enhancement. Imaging findings may mimic MS and clinical correlation is required for diagnosis.
- **Pathology.** Perivenous demyelination with axonal sparing and mononuclear infiltration. Thought to be an autoimmune response to a CNS antigen possibly similar to a viral epitope (molecular mimicry).
- **Treatment.** Steroids. In isolated cases intravenous immunoglobulin (IVIG) and plasmapheresis have been used. Most recover without persistent neurologic deficits, though deficits may persist. In some cases the process may be relapsing.

**Case 2.3**

A 41-year-old male had presented previously with complaints of sudden onset, red proptotic eye for 1 to 2 weeks.
CT scan showed intraorbital mass, hyperdense on precontrast images and homogeneously enhancing. The patient then developed significant loss of visual acuity over a 24-hour period, which prompted emergent biopsy (Figs. 2.12, 2.13).

**Diagnosis**

Unilateral (left) optic nerve sheath meningioma

**Meningioma**

- **Epidemiology.** Fifteen percent of primary intracranial tumors; peak at age 40 to 60 years, and females are more commonly affected. Incidence increased by prior radiation and the presence of genetic syndromes such as neurofibromatosis type 2 (NF-2).
- **Clinical presentation.** Headache, seizures, and/or focal neurologic deficit related to tumor location.
- **Imaging.** MR typically demonstrates a well-circumscribed extraaxial mass that is isointense on T1-weighted imaging (T1WI), iso/hyperintense on T2WI, and shows intense and homogeneous enhancement and often a dural tail. Associated vasogenic edema (best seen on T2 or fluid-attenuated inversion recovery [FLAIR]) in the brain parenchyma may be present, and bony hyperostosis may be appreciated. CT often best demonstrates associated calcifications or adjacent bony hyperostosis. Catheter angiography is often performed for preoperative embolization.
- **Pathology.** Originate from arachnoid cap cells. Blood supply is typically from carotid artery (ECA) branches, but meningiomas may parasitize pial vessels from the internal carotid circulation, especially around the skull base. Often involve adjacent dura and bone. Rarely metastasize. Several different pathologic types (three basic types: meningothelial, fibroblastic, and transitional) have been described. Grading: Grade I, benign; Grade II, atypical; Grade III, anaplastic/malignant.
- **Treatment.** Surgery with or without preoperative embolization. Radiation therapy may be indicated for residual and/or recurrent disease. Prognosis is generally good. Recurrence is dependent on grade and postoperative tumor residual volume.

**Imaging Pearl**

- Meningiomas of the optic nerve sheath may show peripheral linear enhancement along the sheath known as tram-tracking.

**Case 2.4**

A 57-year-old female presented with progressive left vision loss and no specific diagnosis was made. She then re-presented nine years later with progressive right vision loss (Figs. 2.14, 2.15).

**Diagnosis**

Bilateral optic nerve sheath meningiomas