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Orbits, Vision, and Visual Loss

F.J. Wippold II and for the Expert Panel on Neurologic Imaging

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Orbital Disorders

Imaging analysis of orbital diseases is facilitated by a compartmental approach that establishes differential diagnoses on the basis of the location of the process within the orbit.[1,2] General visual disturbances and/or ophthalmoplegia may be caused by such conditions as tumors, infections, inflammations, and vascular disorders, which may be found at any location along the visual pathway extending from the globe to the occipital lobes. Therefore, the choice of appropriate imaging technique and focus depends on the specific clinical condition and may include portions of the orbits, anterior skull base, and/or brain.

CT and MR imaging are complementary diagnostic procedures and may be used together in some circumstances (Table).[3] Dedicated thin-section multiplanar orbital imaging is necessary for detecting orbital abnormalities.[4] The intrinsic contrast provided by orbital fat allows for excellent anatomic visualization with either technique. Contrast enhancement is important in assessing most orbital disorders. CT is useful in evaluating bony structures, and MR imaging excels in evaluating soft tissues.[5-10] Because of the absence of radiation and the utility of fat-suppressed contrast-enhanced images, MR imaging has emerged as the procedure of choice for orbital disorders, with the exception of trauma and assessment for foreign bodies.[9,11-13] Moreover, specialized surface coils have expanded the utility of MR imaging.[14] Sonography and fluorescein angiography are also important modalities; however, these special procedures are usually performed by the ophthalmologist.

Optic Nerve and Sheath Disorders

Primary disorders of the optic nerve and optic nerve sheath include optic nerve tumors such as gliomas, astrocytomas, hamartomas, and meningiomas.[15] Extension of these tumors into the optic chiasm, optic tracts, and lateral geniculate bodies of the thalami is more accurately depicted on MR imaging than on CT. The size and shape of the optic canals are best assessed in the axial projection, while the size and shape of the optic nerves are best appreciated on coronal and oblique sagittal images. Many optic nerve tumors exhibit fusiform homogeneous enhancement.

Meningiomas may be cufflike, surrounding the optic nerves, or eccentrically located on 1 side of the nerve. CT scans will often demonstrate calcifications. Enhancement parallel to the length of the optic nerves with the intact nerve seen within the mass ("tram-tracking") is seen on both CT and MR imaging. MR imaging scans also readily depict the spread into adjacent meninges.

The papilledema associated with pseudotumor cerebri or intracranial mass may enlarge the optic nerve as detected on CT or MR imaging. If severe, there is a reversal of the optic nerve head, with bulging forward into the posterior wall of the globe. While dilatation of the perioptic subarachnoid space is best appreciated on fat-suppressed T2-weighted images, reversal of the nerve head may be more readily detected on CT than on MR imaging because of the chemical shift artifacts inherent to the MR imaging studies. MR imaging may also monitor optic nerve damage in other disorders such as glaucoma.[16]

Other Orbital Neoplasms

Imaging with MR imaging also demonstrates other orbital tumors such as benign cavernous hemangiomas, complex vascular lesions such as lymphangiomas and capillary hemangiomas, schwannomas, metastases, and lymphomas. CT, as a complementary method, may reveal calcifications and bone destruction.

Optic Nerve Neuropathies

Optic neuritis is best seen on MR imaging as focal or diffuse enlargement of the optic nerve, abnormal hyperintensity on T2-weighted images, and/or enhancement. These features are best appreciated on fat-suppressed T2-weighted and contrast-enhanced T1-weighted images. MR imaging has become an essential study for evaluating patients with optic neuropathy and suspected multiple sclerosis and supplements other clinical studies.[17] Even when MR imaging findings of the orbit are normal, imaging of the brain may reveal foci of demyelination.

Radiation-induced optic neuropathy may manifest after a latency period of 6–36 months following treatment. Clinically, the nerve head may appear normal, but gadolinium-enhanced fat-suppressed MR imaging will show patchy, linear, or confluent enhancement along the portions of the optic nerve, chiasm, or optic tract.

Vascular Disorders

In patients with cavernous carotid fistulas, arteriovenous malformations, or orbital varices, MR imaging or CT will demonstrate the dilated ophthalmic veins, facial veins, and other regional venous structures along with enlargement of the cavernous sinus. Large edematous extraocular muscles and periorbital structures may be identified. Proptosis is typically

This article is a summary of the complete version of this topic, which is available on the ACR Website at www.acr.org/ac. Practitioners are encouraged to refer to the complete version.

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Clinical Condition: Orbits, Vision and Visual Loss	MR Imaging Head and/or Orbit Without and With Contrast	MR Imaging Head and/or Orbit, Without Contrast	CT Head and/or Orbit With Contrast	CT Head and/or Orbit Without Contrast	MRA Head and Neck With or Without Contrast	CTA Head and Neck	X-ray Orbit
Infant or child with orbital asymmetry, proptosis and visual loss	8	7	6	5	4[a]	2[b]	1
Child with slowly progressing visual loss	8	7	6	5	4[a]	2[b]	1
Adult with sudden onset of painless or painful visual loss	8	7	6	5	5[a]	5[b]	1
Adult patient with proptosis and/or painful visual loss	8	7	6	5	4[a]	4[b]	1
Adult patient with uveitis, scleritis, and visual loss	8	7	5	4	4[a]	4[b]	1
Adult patient with ophthalmoplegia	9	6	6	5	6[a]	6[b]	1
Head injury with visual loss	5[d]	7[d]	3	9[c]	4[a]	4[b]	2

Note:—Appropriateness criteria scale from 1 to 9, 1 = least appropriate, 9 = most appropriate.
[a] Selected cases when vascular disease is suspected. See statement regarding contrast in text under "Anticipated Exceptions."
[b] If vascular disease suspected.
[c] In acute trauma, a scan without contrast is usually sufficient; in subacute trauma, contrast may be useful.
[d] If MR imaging safe.

present. The addition of MR angiography or CT angiography allows for flow assessments along with the static morphologic changes.[18,19] In some cases, conventional angiography may be required to make the definitive diagnosis, though it is most commonly used in conjunction with therapeutic interventional procedures.

Thin-section CT scans with multiplanar reconstruction are the most useful in traumatic optic neuropathy. Such images provide accurate identification of indirect signs of injury to the optic nerve, such as dehiscence, or bony fragments within the orbit or optic nerve canal; narrowing of the optic canal; or significant bony separations, which indicate likely optic nerve injury. MR images have been shown to be more sensitive for detecting optic nerve edema or avulsion.

Inflammatory Orbital Syndrome

Inflammatory orbital syndrome (IOS) (orbital pseudotumor, inflammatory fibromyotendinitis) may appear on CT and MR imaging as a diffuse infiltrate or focal mass. In the diffuse form, there is inflammatory infiltrate of the orbital fat, extraocular muscles, and adnexal structures, particularly in the orbital apex. The more focal forms of IOS commonly involve the tendinous insertions of the extraocular muscles (myositic form), the uveal structures (anterior form), the scleral region (posterior form), or the lacrimal gland (lacrimal form). Commonly, the retrobulbar fat has a "dirty" appearance. Occasionally, there may be a well-defined mass lesion that mimics a neoplasm. In virtually all cases, there is prominent enhancement on postcontrast CT or MR imaging scans. In the chronic form of the disease, there is increased fibrosis in the lesions, resulting in decreased signal intensity on T2-weighted images. When there is secondary thrombosis of the sphenoidal veins or cavernous sinus, a painful ophthalmoplegia results, with the presumptive diagnosis of Tolosa-Hunt syndrome. Although usually confined to the orbital soft tissues, IOS can produce bone destruction or extraorbital extension. Posterior scleritis shows inflammatory signs in the coat of the eye sclera with thickening of the posterior coat of the eye sclera that may be identified as areas of enhancement on CT or MR imaging. The thickened sclera, enhanced by contrast, presents as a so-called "ring sign." CT or MR imaging scans may be used to follow the course of the illness until it resolves or recurs in the chronic form of the disease.

Endocrine Disorders

CT and especially MR imaging studies demonstrate enlargement of 1 or more of the extraocular rectus muscles in thyroid ophthalmopathy (Grave disease) with relative sparing of tendinous insertions. The ability to measure the T2 signal intensity on MR imaging helps both in determining which patients may benefit from corticosteroid therapy (those with high T2 values) and/or which patients require combined therapies including cyclosporin (based on a measurable response on serial MR images).

Retinal, Choroidal, and Subhyaloid Detachments

MR imaging may differentiate choroidal effusion from choroidal hemorrhage. In acute hemorrhages, CT may be more specific, showing the increased attenuation of subchoroidal hemorrhage.

Retinal detachments may also occur with ocular neoplasms such as retinoblastomas in children and uveal malignant melanomas in adults. Ocular sonography may be more accurate in detecting small tumors; however, enhanced MR images are useful in determining the true extent of lesions beyond the ocular structures and also in demonstrating associated retinal detachments. Postcontrast T1-weighted images are most helpful in detecting uveal melanomas and in differentiating melanomas from subretinal fluid collections. CT scanning reveals small punctuate calcifications in retinoblastoma.

The differentiation of an amelanotic melanoma from a subretinal hemorrhage is based on both the precontrast and postcontrast T1-weighted images. Of note are metastatic lesions to the retina or certain inflammatory conditions that cannot be consistently differentiated from primary uveal melanomas. Doppler sonography may help detect vascularity within an intraocular tumor and help differentiate such entities from nonvascular choroidal, subretinal, or subhyaloid effusions or from hematomas.

Summary

- Imaging analysis of the orbit is facilitated by a compartmental approach.
- CT and MR imaging are complementary diagnostic procedures for suspected orbital pathology.
- CT is useful in evaluating bony structures.
- MR imaging is useful in evaluating soft-tissue structures such as the globe and optic nerves and intraconal and extraconal spaces.
- CTA, MR angiography, and conventional angiography may be useful in vascular conditions.
- Conventional angiography may be useful in delivering therapeutic intervention.

Anticipated Exceptions

Nephrogenic systemic fibrosis (NSF) is a disorder with a scleroderma-like presentation and a spectrum of manifestations that can range from limited clinical sequelae to fatality. It appears to be related to both underlying severe renal dysfunction and the administration of gadolinium-based contrast agents. It has occurred primarily in patients on dialysis, rarely in patients with very limited glomerular filtration rate (GFR) (ie, <30 mL/min/1.73 m^2), and almost never in other patients. There is growing literature regarding NSF. Although some controversy and lack of clarity remain, there is a consensus that it is advisable to avoid all gadolinium-based contrast agents in dialysis-dependent patients unless the possible benefits clearly outweigh the risk and to limit the type and amount in patients with estimated GFR rates <30 mL/min/1.73 m^2. For more information, please see the ACR *Manual on Contrast Media*.[20]

Review Information

This guideline was originally developed in 1999. The last review and update were completed in 2009.

Appendix

Expert Panel on Neurologic Imaging: Franz J. Wippold II, MD, Principal Author and Panel Chair; Rebecca S. Cornelius, MD; Daniel F. Broderick, MD; Douglas C. Brown, MD; James A. Brunberg, MD; Patricia C. Davis, MD; Robert L. De La Paz, MD; Charles F. Garvin, MD; Isabelle Germano, MD, American Association of Neurologic Surgeons/Congress of Neurologic Surgeons; Charles T. McConnell Jr, MD; Michael W. McDermott, MD, American Association of Neurologic Surgeons/Congress of Neurologic Surgeons; Suresh Kumar Mukherji, MD; David J. Seidenwurm, MD; Michael A. Sloan, MD, MS, American Academy of Neurology; James G. Smirniotopoulos, MD.

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