Orbital Disease in Neuro-Ophthalmology



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KEYWORDS

- Orbit Orbital fracture Traumatic optic neuropathy Enophthalmos
- Idiopathic orbital inflammatory syndrome Thyroid eye disease
- IgG4-related disease

KEY POINTS

- Orbital compartment syndrome, cavernous sinus thrombosis, and orbital mucormycosis are neuro-ophthalmologic emergencies.
- Trapdoor fractures may present as acute diplopia without many stigmata of trauma, cannot be diagnosed by CT alone, and require urgent surgery.
- There is no proven effective treatment for traumatic optic neuropathy.
- Enophthalmos may be caused by orbital fracture, silent sinus syndrome, chronic intracranial hypotension, and rare fibrosing diseases.
- Infection must be excluded in cases of orbital inflammation, especially in patients who
 may be immunocompromised.

INTRODUCTION

Many abnormalities of the orbit present with neuro-ophthalmic findings, such as impaired ocular motility or alignment, and sensory changes, including optic neuropathy. Comprehensive coverage of all orbital diseases is beyond the scope of this article. This review focuses on diagnosis and management of the most common and the most vision- or life-threatening orbital conditions as well as more recently discovered entities and points of active controversy. These conditions include orbital trauma, vascular disease, inflammatory and infectious diseases, and neoplasms. Common presenting symptoms and associated neuro-orbital diseases also are summarized (Table 1).

Disclosures: None.

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Table 1 Neuro-ophthalmic manifestations of orbital disease		
Optic Disc Edema and/or Optic Neuropathy		
Compressive	Neoplastic Inflammatory	 Primary (eg, optic nerve sheath meningioma, rhabdomyosarcoma) Secondary (eg, metastasis to extraocular muscle) Vascular (eg, capillary hemangioma, cavernous hemangioblastoma) TED (Graves disease, Hashimoto thyroiditis) Orbital inflammatory disease (eg, sarcoidosis, IgG4-related
		orbital disease, idiopathic)
Infiltrative	Neoplastic Infectious	 Primary (eg, optic nerve glioma) Secondary (eg, metastatic carcinoma, lymphoma, leukemia) Bacterial
	Inflammatory	 Viral Fungal Systemic disease (eg, sarcoidosis, SLE, granulomatosis with polyangiitis) Idiopathic optic perineuritis
Traumatic	Direct	Impingement or transection from bone fragment or foreign body
	Indirect	Avulsion injuryBlunt trauma to the forehead or orbit
Diplopia and/or Motility Abnormality		
Mechanical		 Orbital fracture (with or without enophthalmos) Silent sinus syndrome Sagging eye, sunken brain syndrome Globe dystopia from mass lesion (eg, vascular tumor)
Inflammatory		 Myositis TED Orbital inflammation (eg, IOIS, IgG4-RD, orbital cellulitis)
Neuropathic		Infiltrative (inflammatory or neoplastic) Compressive (eg, cavernous sinus or orbital apex syndrome)
Myopathic		Chronic progressive external ophthalmoplegia Congenital fibrosis syndrome

ORBITAL TRAUMA

Orbital fractures are common and may present with periocular swelling, proptosis, enophthalmos, ecchymosis, chemosis, subconjunctival hemorrhage, infraorbital nerve hypoesthesia, diplopia, and decreased vision. One should first assess for globe rupture and orbital compartment syndrome, because emergent intervention for these conditions carries the potential to reverse or prevent permanent vision loss. The treatment of traumatic optic neuropathy (TON) is more controversial. Trapdoor fractures constitute another urgent and sometimes challenging diagnosis; comminuted fractures may also present with acute and/or delayed dysmotility and diplopia. 1–4 Ischemic extraocular muscle entrapment requires urgent surgical intervention to reduce the risk of chronic dysmotility. 4 Enophthalmos often results from orbital trauma, but other mechanisms are also described in later discussion.

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