The sunken eye: An educational review of imaging findings in enophthalmos

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Learning objectives

To review the various techniques and indices that establish the presence of enophthalmos on cross-sectional images. To offer a systematic approach to the imaging manifestations of traumatic and non-traumatic etiologies of enophthalmos. To present common, uncommon, and rare etiologies of enophthalmos.
Enophthalmos, as measured on CT and MRI on the axial plane, is defined as asymmetric posterior displacement of the globe of at least 2 mm of the affected side. Specifically, the displacement on the axial plane must be measured at the interzygomatic line where a transversely oriented imaginary line is drawn from one bony zygomatic arch to the other (Fig. 1). The radiographic measurements for bilateral enophthalmos are still debated in the literature. However, ophthalmologic exam defines enophthalmos, unilateral or bilateral, as exophthalmometric values of less than 14 mm. Exophthalmometry is an instrument used to measure displacement of the globes both on a relative and absolute scale. Although enophthalmos most commonly follows traumatic facial injuries, in the absence of trauma it can be concerning for an underlying systemic disease. Clinically, patients with enophthalmos can present with diplopia, ptosis, and apparent contralateral exophthalmos. In this educational exhibit, we highlight the many CT and MRI manifestations of this condition.
Findings and procedure details

TABLE OF CONTENTS/OVERVIEW:

1. Traumatic
   - Orbital/Facial Fracture
2. Nontraumatic
   - Structural Changes to Orbital Contents
     a. Syndromic
     b. Vascular
     c. Infectious
   - Structural Changes to the Globe
     a. Phthisis Bulbi (can also be post-traumatic)
     b. Anophthalmic
   - Globe Retraction
     a. Metastatic
     b. Congenital

We consider the varied etiologies of enophthalmos in a systematic approach. Enophthalmos can be traumatic or non-traumatic. The non-traumatic subset can be further stratified into entities that result in: (1) structural changes to orbital contents, such as in silent sinus syndrome, silent brain syndrome, orbital varix, tuberculosis, neurofibromatosis; (2) structural changes to the globe itself, such as phthisis bulbi and anophthalmic enophthalmos; and (3) globe retraction, occurring with scirrhous orbital metastases or congenital fibrosis.

Traumatic - Orbital/Facial Fracture:

The most common cause of enophthalmos overall is post traumatic, generally secondary to fractures of the orbital floor or medial orbital wall. It is postulated that in the event of an orbital floor fracture, the direct trauma to the inferior orbital rim results in buckling of the orbital contents and subsequent postero-inferior displacement. With medial orbital
wall fractures, hydraulic forces from the increased intraorbital pressure lead to rupture of the orbital wall.

Early radiographic diagnosis of post-traumatic enophthalmos is challenging, often detected on follow up imaging weeks to months after the initial insult. Imaging, however, does play a pivotal role in such trauma by delineating the extent of the fractures, involvement of the adjacent soft tissues including potential optic nerve injury, and by possibly predicting the extent of enophthalmos based off of the severity of the orbital fractures (Fig. 2). Subsequent surgical repair is generally determined based on presence of symptoms.

Nontraumatic - Syndromic - Silent Sinus Syndrome:

Silent sinus syndrome (SSS) is a sequela of chronic sinusitis resulting in gradual atelectasis of an opacified sinus. This sinus wall collapse then causes negative intraorbital pressure that leads to enophthalmos. It is generally unilateral, occurs equally in males and females, and is most commonly seen in the third to fifth decades.

SSS demonstrates characteristic imaging findings on CT and MRI such as opacification and decreased volume of the affected sinus, and downward bowing of the orbital floor (Fig. 3). Treatment is generally surgical with repair of the orbital wall defect.

Nontraumatic - Syndromic - Silent Brain Syndrome:

Silent Brain Syndrome (SBS) is a rare form of nontraumatic acquired bilateral enophthalmos secondary to VP shunt placement for hydrocephalus. It is characterized by symmetric, severe, bilateral enophthalmos with gradual progression years after VP shunt placement with associated retraction of the eyelids in the absence of systemic intracranial disease. It is postulated that sudden reduction in the intraorbital pressures result in this posterior displacement. Patients often develop dryness and irritation within their eyes, a foreign body sensation, and even blurry vision.

SBS is unique as a rare cause of bilateral enophthalmos. Imaging shows symmetric posterior displacement of the globes (Fig. 4) with possible air entrapment between the globes and eyelids in addition to severe upward bowing of the orbital roof into the anterior cranial fossa. Treatment consists of orbital implants and upper eyelid repair.

Nontraumatic - Syndromic - Parry Romberg Syndrome:

Parry Romberg Syndrome (PRS) is a form of progressive self-limiting enophthalmos secondary to hemifacial atrophy involving the skin and forehead, most commonly unilaterally involving the periorbital soft tissues. It often occurs in children and young adults with an increased incidence in females. It has been associated with scleroderma.
It is thought that the intraorbital fat in PRS is gradual replaced with collagen leading to enophthalmos.

Characteristic imaging findings include asymmetric of the facial bones on the affect side with deviation of the nose and decreased volume of the ipsilateral ethmoid sinuses (Fig. 5). Absence of subcutaneous fat in the inframaxillary region of the affect side has been reported. Additional non-orbital imaging findings include linear calcifications of the ipsilateral subcortical frontal lobe with associated hypoattenuation/ T2 hyperintensity and atrophy of the white matter on CT and MRI respectively. Surgical repair can be performed with autologous fat and muscle grafts.

Nontraumatic - Syndromic - Neurofibromatosis 1:

Neurofibromatosis 1 (NF1) is an autosomal dominant syndrome characterized by café au lait spots, neurofibromas, optic nerve gliomas, sphenoid wing dysplasia, foci of abnormal parenchymal signal intensity, and vascular anomalies. It has been shown to cause pulsatile enophthalmos.

Imaging findings of NF1 enophthalmos include absence of the sphenoid wing of the affect side (Fig. 6), prolapse of the ipsilateral temporal lobe, and kinking of the ipsilateral optic nerve. It can lead to progressive vision loss, which dictates ophthalmological management.

Nontraumatic - Vascular - Orbital Varix:

Orbital varices are vascular low flow, low pressure hamartomas leading to markedly distended vessels, more commonly associated with exophthalmos but also seen with enophthalmos. While initially pushing the globe forward due to mass effect from distention, over time, the anomaly can cause atrophy of the orbital fat resulting in posterior displacement of the globe.

Imaging findings include lobulated homogenously enhancing structure commonly within the intraconal space with associated bony defects and atrophy of the surrounding fat (Fig. 7). Treatment is conservative but embolization can be performed as clinically indicated.

Nontraumatic - Infectious - Tuberculosis:

Mycobacterium Tuberculosis (TB) is one of the most common mimickers of systemic disease. One such manifestation that can cause diagnostic dilemmas is TB related enophthalmos. This entity can present as a hard mass and is therefore challenging to differentiate from metastasis or orbital inflammatory disease. TB related enophthalmos is thought to manifest due to granulomatous reactions leading to fibrosis.
Imaging findings of TB enophthalmos include an ill-defined enhancing intraorbital mass causing retraction of the globe (Fig. 8). Secondary imaging findings can include an underlying abscess or osteomyelitis of the adjacent bony structures. Biopsy is generally performed for definitive diagnosis and treatment consistent of anti-TB medications for at least six months, after which the condition commonly resolves.

Nontraumatic/Traumatic - Structural - Phthisis Bulbi

Phthisis bulbi (PB) is a severely atrophied globe with a disorganized appearance, deemed nonfunctioning end stage eye disease. PB can be either traumatic or nontraumatic with nontraumatic causes consisting of infection, radiation, malignancy (retinoblastoma), or post inflammatory conditions.

Imaging findings of PB include marked reduction in volume of the affected globe with dystrophic calcifications and thickening sclera (Fig. 9). Enucleation can be performed for cosmetic or symptomatic reasons.

Nontraumatic - Structural - Anophthalmic

Anophthalmic Enophthalmos is a form of enophthalmos due to an anophthalmic socket. It is also termed post enucleation/ evisceration socket syndrome (PESS) consisting of ptosis, enophthalmos, superior sulcus deformity, and eyelid laxity.

PESS can be seen radiographically after treatment with polyacrylamide gel with a soft tissue density in the intraconal space (Fig. 10). PESS can be prevented surgically by achieving adequate orbital post implantation volume at the initial oculoplastic surgery.

Nontraumatic - Globe Retraction - Metastatic

Metastases represent up to 4% of orbital tumors with enophthalmos seen in nearly a quarter of such cases. Scirrhous breast carcinoma metastases are by far the most common cause of metastasis related enophthalmos with lung and stomach also reported in exceedingly rare cases. Symptoms can manifest years after the initial primary malignancy has been treated and include diplopia, pain, and gradual vision loss. Patients may initially present with exophthalmos with gradually developing enophthalmos on clinical exam over multiple years.

Characteristic imaging findings of scirrhous breast metastatic enophthalmos include an ill-defined enhancing mass with fibrosis, retraction of the globe, and potentially invasion of the adjacent extraocular muscles (Fig. 11). Biopsy is confirmatory and prognosis is poor. Palliative care is generally necessary for improved survival.

Nontraumatic - Globe Retraction - Congenital
Congenital fibrosis is a rare form of enophthalmos secondary to extraocular muscle fibrosis. It is most commonly bilateral with unilateral muscle fibrosis. It can present in infancy with restricted eye movements, progression decreased vision, diplopia, and strabismus.

Neuroimaging findings include retraction of the globe secondary into an intraorbital mass with thickening of the extraocular muscles (Fig. 12). Management is commonly surgical to relieve the restricted movement and fix the strabismus.
Fig. 1

Fig. 2: Enophthalmos secondary to Facial Trauma. Post-traumatic left enophthalmos related to complex facial fractures.

Fig. 2

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Fig. 3: Enophthalmos secondary to Silent Sinus Syndrome. Coronal bone CT in a patient with silent sinus syndrome shows an opacified right maxillary sinus with inferior position of the roof of the sinus (white solid arrow) with increased volume in the right orbit. The uncinate process (white open arrow) is lateralized, narrowing the infundibulum.

Citation: StatDx
Fig. 4

Fig. 5: Enophthalmos secondary to Parry Romberg Syndrome. Axial CT images of a 44-year-old woman with Parry Romberg Syndrome obtained for cosmetic surgical planning. (A) There is prominent hemiatrophy of the skin, subcutaneous fat, and masseter muscle. (B) The right maxillary sinus is considerably smaller than the left. (C) Marked right enophthalmos is present.

Citation: M. Wong et al. AJNR Am J Neuroradiol 2015;36:1355-1361

Fig. 5

Fig. 6

Fig. 7

Fig. 8: Enophthalmos secondary to Orbital Tuberculosis. Left, External photograph shows mild enophthalmos OS. Middle, Coronal CT shows an ill-defined anterior orbital mass located inferotemporally in the left orbit, with minimal contrast enhancement. Right, Axial CT shows an ill-defined anterior orbital mass with enophthalmos of the left globe.

Citation: Ophthalmic Plastic & Reconstructive Surgery. 22(3):219-221, May/June 2006.
DOI: 10.1097/01.iop.0000214528.96858.ad
Fig. 9: Enophthalmos secondary to Phthisis Bulbi. Atrophied, collapsed left globe resulting in left enophthalmos which is compatible with phthisis bulbi.

Citation: https://radiopaedia.org/articles/phthisis-bulbi

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Fig. 10

Fig. 11: Enophthalmos secondary to Metastatic Breast Cancer. (A) Clinical photograph of a 59-year-old female with enophthalmos of the left eye secondary to metastatic scirrhou carcinoma of the breast. (B) The CT scan demonstrates irregular opacification (metastatic disease with fibrosis) in the left posterior orbit (arrow) with resulting retraction of the globe.

Fig. 12

Conclusion

Enophthalmos is most commonly associated with trauma. Nonetheless, in the absence of trauma it can be a marker of underlying serious systemic diseases. In this pictorial and educational review we present the many etiologies that can result in enophthalmos. Recognition of enophthalmos clinically and with the aid of cross-sectional imaging can potentially reveal indolent diseases, leading to timely diagnosis for afflicted patients.
References


