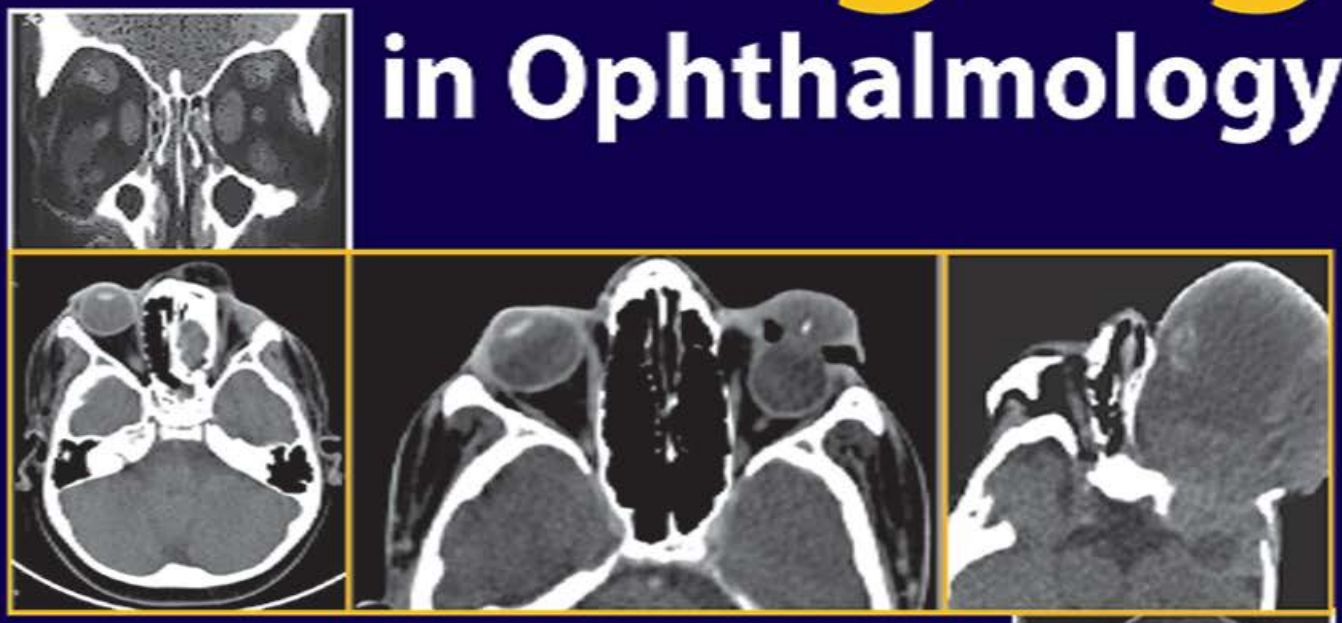
Sankara Nethralaya's

Atlas of Imaging in Ophthalmology



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Foreword
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Chapter 3

Congenital Anomalies of the Globe

CONGENITAL CYSTIC EYE

- Congenital cystic eye results due to failure of the optic vesicle to invaginate during the 4th week of embryogenesis
- It can be associated with other nonocular abnormalities such as facial clefts, choanal atresia, malformation of the sphenoid bone, agenesis of corpus callosum and midbrain deformities.

IMAGING

Imaging is done to confirm the diagnosis and evaluate any associated intracranial anomalies.

CT Orbit

- The normal globe is not visualized
- Cystic structure of CSF density not resembling the normal globe is seen in the orbit. If large can cause remodeling of the bony orbit
- A rudimentary connection to a thinned optic nerve may be seen
- The cyst may have an attached stalk. If the stalk is patent, then size of the cyst remains small due to communication of the cyst with the cranial cavity
- The ipsilateral superior orbital fissure may be widened
- Extraocular muscles and optic nerve are usually absent.

MRI Orbit

- On MR, the signal intensity of the cyst fluid may not be equal to that of normal vitreous because the cyst is normally filled with serum
- A nodular focus may be seen adjacent to the cyst wall.

DIFFERENTIAL DIAGNOSIS

Microphthalmia with colobomatous cyst.

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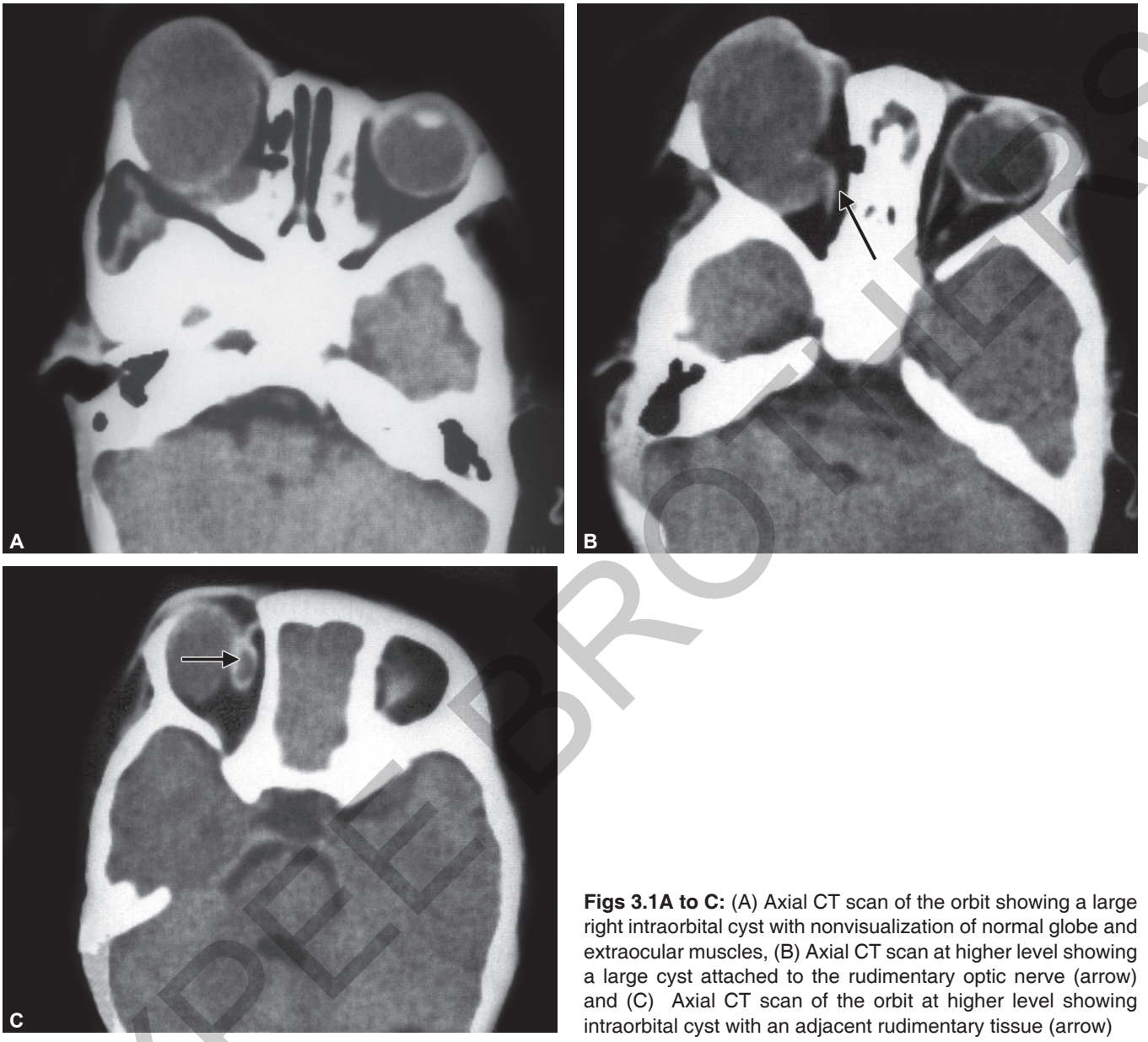

Figs 3.1A to C: (A) Axial CT scan of the orbit showing a large right intraorbital cyst with nonvisualization of normal globe and extraocular muscles, (B) Axial CT scan at higher level showing a large cyst attached to the rudimentary optic nerve (arrow) and (C) Axial CT scan of the orbit at higher level showing intraorbital cyst with an adjacent rudimentary tissue (arrow)

ANOPHTHALMIA

- Anophthalmia is complete absence of an eye by birth due to a developmental defect
- Structures not derived from the neuroectoderm such as extraocular muscles, eyelids, conjunctiva, lacrimal apparatus and bony orbit are relatively preserved
- Bilateral anophthalmia is rare.

CT/MRI FINDINGS

- Small rudimentary tissue present
- Dystrophic calcification may be present within the rudimentary tissue
- Bony orbit is smaller in size
- The optic nerve and extraocular muscles are hypoplastic
- In true anophthalmia, optic nerve and optic chiasm may be absent. Optic tracts may be rudimentary and lateral geniculate body may be gliotic.

RECOMMENDED IMAGING

CT or MRI orbit with brain to look for intracranial abnormalities especially midline defects, hydrocephalus and intracerebral cysts.

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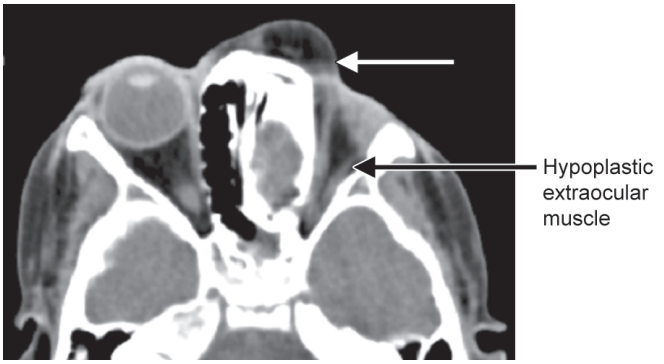

Fig. 3.2: Axial CT scan of the orbit showing absent left globe with hypoplastic extraocular muscles (long arrow) and small bony orbit. Also noted is a nasal dermoid (white arrow)

Fig. 3.3: Three-dimensional CT of the skull of the same patient showing an extremely small left bony orbit

MICROPHTHALMIA

- Microphthalmia is defined as an eye that has an axial length of less than 21 mm in an adult or less than 19 mm in a one-year-old child
- It can occur as an isolated disorder or with other ocular and craniofacial anomalies such as Hallermann-Streiff syndrome or systemic abnormalities such as microphthalmos, dermal aplasia and sclerocornea (MIDS)
- Other causes of microphthalmia include congenital rubella syndrome, persistent hyperplastic primary vitreous and retinopathy of prematurity.
- Microphthalmia is of two types:
  1. Small anatomically correct eye—simple
  2. Small malformed eye complex.

It may be unilateral or bilateral. Bilateral microphthalmia can be seen in Lowe's syndrome (oculocerebral renal disease).

It may or may not be associated with colobomatous cysts. Those with colobomatous cysts result from failure of fusion of the fetal optic fissure.

CT/MR FINDINGS

- A small globe with small bony orbit may be seen
- Extraocular muscles and optic nerve are hypoplastic
- Colobomatous cyst may vary from small to large almost occupying the orbit
- Colobomatous cyst may cause a conical deformity on the posterior globe
- Cysts can be very large and at times larger than the microphthalmic eye resulting in bony remodeling
- Often intraocular calcification may be seen which may represent calcified lens.

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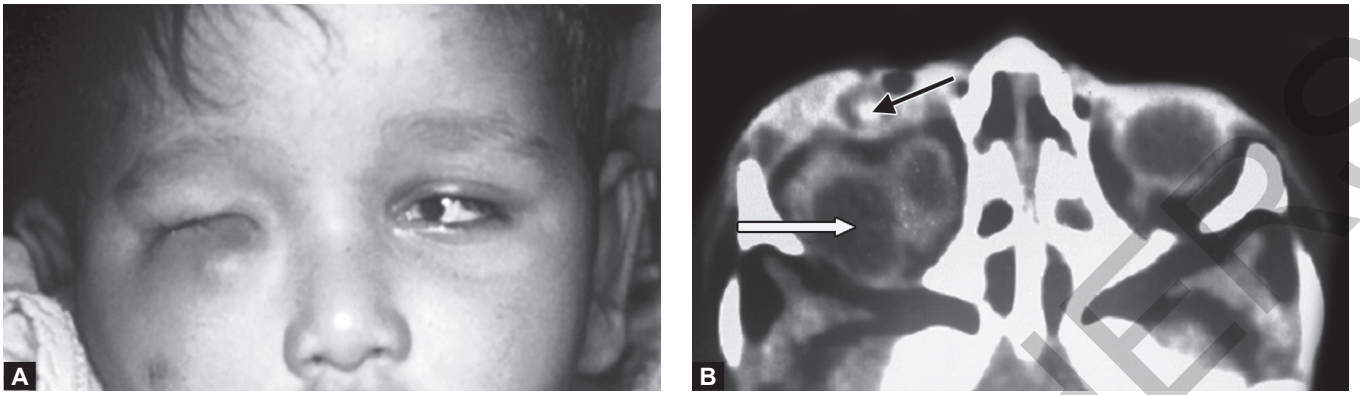
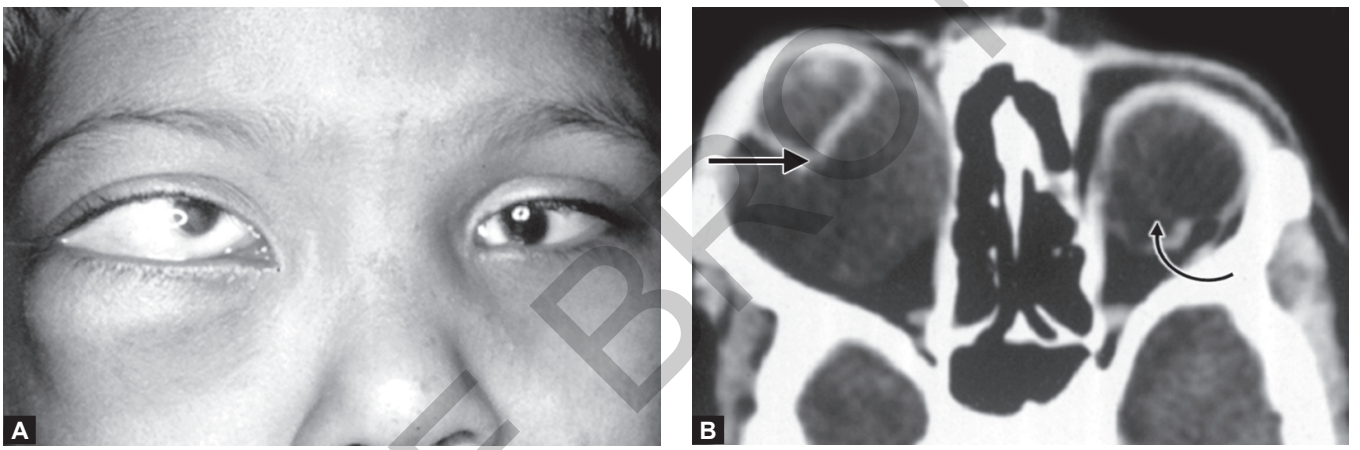


Figs 3.4A and B: (A) Clinical photograph of a child showing microphthalmic right eye and (B) Axial CT scan of the orbit of the same patient showing a small right globe (arrow) with multiple retro-ocular cysts (block arrow) resulting in expansion of the bony orbit



Figs 3.5A and B: (A) Clinical photograph showing a microphthalmic right eye, which is displaced upwards and forwards. Fullness seen in right lower lid suggestive of inferior periocular cyst and (B) Axial CT orbit showing a small right globe with a large solitary retro-ocular cyst causing posterior conical defect of the globe (arrow). The left eye shows a large disc coloboma (curved arrow)

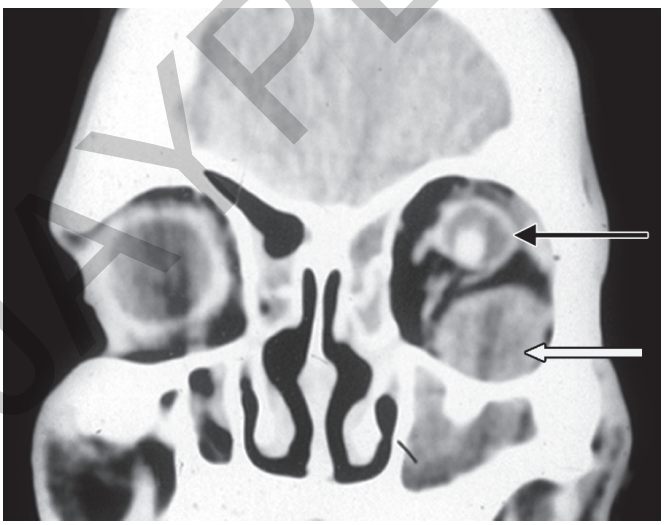


Fig. 3.6: Coronal CT orbit showing small left globe (arrow) with calcified lens and an inferior orbital colobomatous cyst (block arrow) displacing the globe superiorly

CRYPTOPHTHALMIA

- Cryptophthalmia consists of partial or complete failure of development of the eyelids, eyebrow, palpebral fissure, eyelashes and conjunctiva
- Patients will have hidden eyes because the skin of the eyelids is partially or fully sealed
- It is classified into three types:
  1. Complete—eyelid is completely fused over existing eye
  2. Incomplete—eyelid is partially fused over existing eye
  3. Abortive—eyelid is completely fused and underlying eye does not form.
- It may be unilateral or bilateral
- It may be associated with skin like cornea, an incompletely developed anterior segment or a rudimentary cyst-like globe
- Cryptophthalmos may be associated with systemic anomalies
- Fraser's syndrome consists of cryptophthalmos associated with other malformations such as abnormalities of genitals, kidneys, larynx, and ear and abnormal or fused digits (syndactyly)
- There may be abnormal or absent punctums/canaliculi and associated hypertelorism.

IMAGING (CT/MRI)

Incompletely developed anterior segment, or a rudimentary cyst-like globe.

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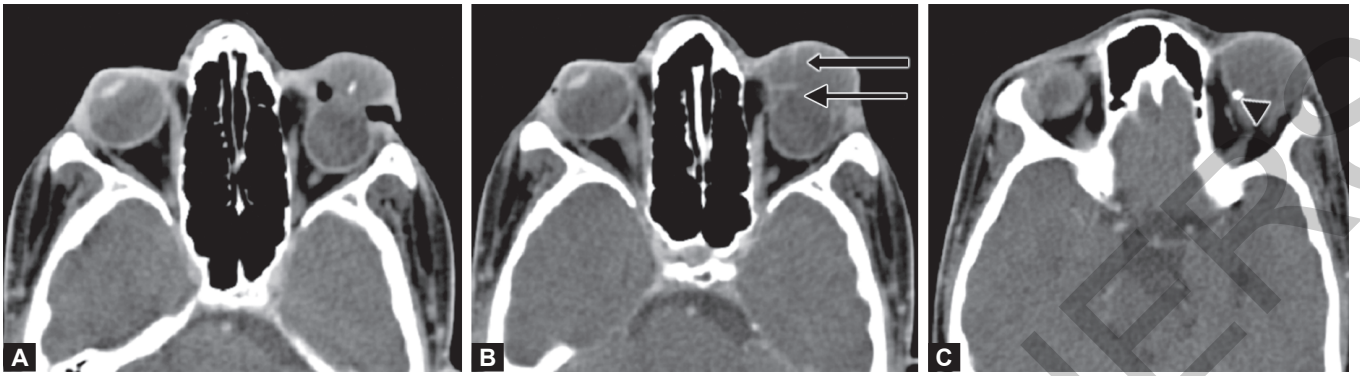

Figs 3.7A to C: Axial CT scan of the orbit in a case of cryptophthalmos: (A and B) Showing a malformed left globe (thin arrow) with malformed anterior chamber, absent lens and an anterior cystic component (thick arrow) and (C) Showing intraocular calcification (arrowhead). The posterior half of the globe and optic nerve are well preserved

STAPHYLOMA

- Staphyloma is defined as areas of scleral ectasia that are lined by uveal tissue. It can be anteior or posterior
- High myopia causes posterior staphyloma. Less prominently, one can see equatorial staphylomata at sites of scleral thinning
- Staphyloma involving the ciliary body is called ciliary or intercalary staphyloma depending on its location. It is caused by glaucoma and scleritis
- Anterior staphyloma develops secondary to infection, trauma or radiotherapy. It consists of ectatic pseudocornea (formed by fibrous tissue) lined by uveal tissue.

CT/MRI FINDINGS

- It is seen as a focal bulge of the ocular coats
- Posterior staphyloma is seen as a focal bulge in the ocular coats temporal to the optic disc
- In anterior staphyloma, as the name suggest the bulge is on the anterior ocular surface.

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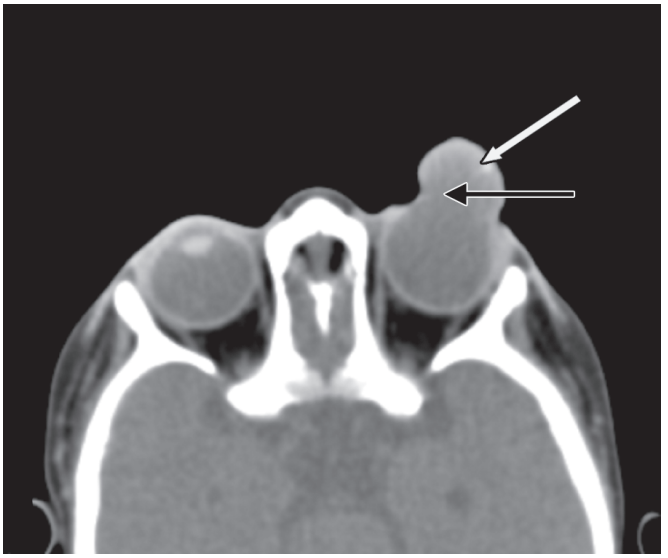

Fig. 3.8: Axial CT orbit showing a focal bulge of the anterior ocular coats (anterior staphyloma) (arrow) with eccentric calcification (block arrow)

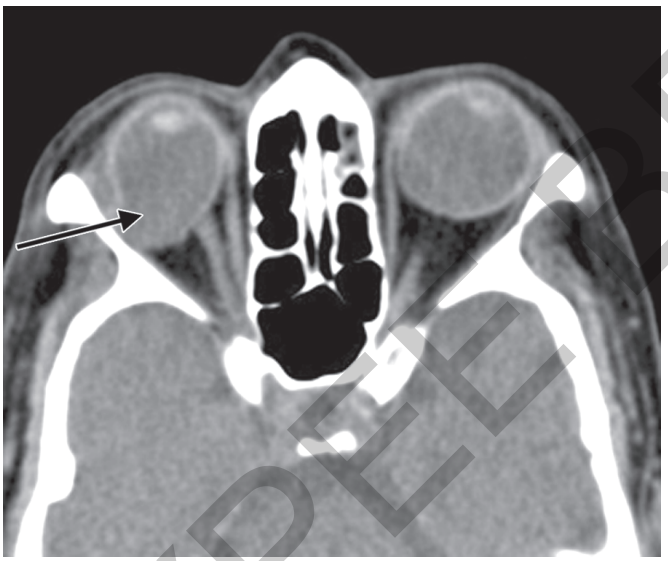

Fig. 3.9: Axial CT orbit of a myopic patient showing a focal bulge temporal to optic disc with scleral thinning—posterior staphyloma (arrow)

Atlas of Imaging in Ophthalmology

Salient Features

- Consists of extensive collections of computed tomography (CT) and magnetic resonance imaging (MRI) of various ocular, orbital and neuro-ophthalmic disorders
- Deals with most of the neurological diseases with ophthalmic manifestations with vivid portrayal of their imaging characteristics
- Aims to cover the important role of computed tomography and magnetic resonance imaging in the diagnosis of various ophthalmic diseases and thereby enhancing patient management
- Deals also with imaging aspects in ophthalmology
- Includes enormous data and collection of images of various common and relatively rare clinical conditions
- A good guide to all practicing ophthalmologists, neurologists and radiologists in their day-to-day practice
- Illustrated all clinical images used in this atlas belong to patients who were seen in our organization (Sankara Nethralaya—a Unit of Medical Research Foundation) which is a tertiary referral ophthalmic institution.
- It can be used as a reference book for undergraduates, ophthalmology, radiology and neurology postgraduates.

Ambika Selvakumar is a Senior Consultant and Director, Department of Neuro-ophthalmology, Sankara Nethralaya (a Unit of Medical Research Foundation), Chennai, Tamil Nadu, India. She has many publications in the field of Neuro-ophthalmology and participated as a presenter in numerous national and international ophthalmic and Neuro-ophthalmic conferences. She has given various invited lectures in Neurology and Neurosurgical Society Conferences, apart from Ophthalmic conferences. She has authored the book *The Atlas of Neuro-ophthalmology* (2002), which was the first Indian atlas in this specialty. She has released the second edition (2007) of the *Atlas of Neuro-ophthalmology* along with her colleagues. She has done observership in Neuro-ophthalmology with legends such as Dr Andrew Lee, University of Iowa, Dr James Goodwin, University of Illinois, Dr Averatna Noronha, University of Chicago, and Dr Joel Glaser, Bascom Palmer Eye Institute, Miami, USA.

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