Role of MRI and CT in Ocular and Orbital Diseases

Dr. Tejas Kalyanpur MS, Dr. Mathew Cheriyan MS

Abstract

CT and MRI have made significant contributions to the field of ophthalmology. Familiarity with salient imaging features of conditions affecting the eye and orbit will help the ophthalmologist to better understand disease process and evaluate response to therapy. The article outlines important diagnostic features with an algorithmic approach which would help in making a diagnosis.

Introduction

The eye and the orbit constitute a unique and complex region of the nervous system which poses a challenge to the ophthalmologist and neuroradiologist. While history and clinical examination are invaluable, CT and MR provide an insight into the retrobulbar area, the orbital apex and the brain. CT was the preferred investigative modality with its faster acquisition times and lack of sensitivity to eyeball motion which were seen as major problem areas in MR imaging. With the advent high-tesla MR systems and small diameter dedicated surface coils, the skepticism against MR has receded and despite the limitations, MRI is now the modality of choice and can provide information that is unavailable on CT because of superior soft tissue resolution. The article focuses on salient MRI and CT findings of few commonly encountered conditions in the eye and orbit, and also aims to provide a systematic approach to these conditions.

Techniques

MRI Technique:

MRI is performed using a head or preferably a surface coil with the patient in supine position. The common pulse sequences used for orbit are Spin Echo T1 and T2, Fat suppression and Short Tau Inversion recovery. These help in tissue characterization. Post contrast T1 sequences help in assessing the vascularity of lesions, the enhancement patterns of some of which may help in reaching a definitive diagnosis.

CT Technique:

With the advent of Multi Detector row CT, the images obtained with the patient scanned in supine position and can be reformatted to coronal and sagittal planes in slice thickness of as low as 0.6 mm.

Radiological Anatomy of The Globe and Orbit:

To identify and characterize the lesions of orbit and globe one should be aware of normal structures and their signal intensities on various MRI sequences. Representative image with anatomical structures is shown below. (Figs 1a, 1b)
Lesions In The Globe

The lesions in globe and orbit can be summarized in Table 1 and 2. The classification system followed is based on pathology. The imaging approach to each disease entity is dealt with at the end of the discussion.

Table 1. Approach to lesions in Eye

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Microphthalmia</th>
<th>Macrophthalmia</th>
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<tbody>
<tr>
<td>Staphyloma</td>
<td>Coloboma</td>
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<tr>
<td>Ocular Detachments</td>
<td></td>
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<tr>
<td>Retinal</td>
<td>Posterior subhyaloid</td>
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<tr>
<td>Vitreous</td>
<td>Choroidal Detachment</td>
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<td>Inflammation</td>
<td>Scleritis</td>
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<tr>
<td>Uveitis</td>
<td>Laukocoria</td>
<td></td>
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<tr>
<td>Retinoblastoma</td>
<td>Persistent primary hyperplastic</td>
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<tr>
<td>vitreous</td>
<td>Coats Disease</td>
<td></td>
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<tr>
<td>Retinopathy of prematurity</td>
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<td></td>
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<tr>
<td>Others</td>
<td>Tumours</td>
<td></td>
</tr>
<tr>
<td>Melanoma</td>
<td>Lymphoma</td>
<td></td>
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<tr>
<td>Lymphoma</td>
<td>Metastasis</td>
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</tbody>
</table>

Congenital
- Microphthalmia
- Macrophthalmia
- Staphyloma
- Coloboma
- Ocular Detachments
- Posterior subhyaloid
- Retinal
- Posterior Vitreous
- Choroidal Detachment
- Inflammation
- Scleritis
- Uveitis
- Laukocoria
- Retinoblastoma
- Persistent primary hyperplastic vitreous
- Coats Disease
- Retinopathy of prematurity
- Others
- Tumours
- Melanoma
- Lymphoma

Table 2. Approach to Orbit

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Dermoid/epidermoid</th>
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</thead>
<tbody>
<tr>
<td>Inflammatory</td>
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<tr>
<td>Orbital cellulitis</td>
<td>Subperiosteal abscess</td>
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<tr>
<td>Cavernous sinus thrombosis</td>
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<td>Fungal Simmsitis</td>
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<td>Pseudotumor</td>
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<td>Graves</td>
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<td>Sarcoidosis</td>
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<td>Histiocytosis</td>
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<td>Tunors</td>
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<td>Lymphoma</td>
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<td>Metastasis</td>
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<tr>
<td>Vascular</td>
<td>Capillary Hemangioma</td>
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<tr>
<td>Lip angioma</td>
<td>Cavernous Hemangioma</td>
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<tr>
<td>Lymphangioma</td>
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<td>Orbital varix</td>
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<td>Carotico cavernous fistula</td>
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<tr>
<td>Neural</td>
<td>Schwannoma</td>
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<tr>
<td>Neurofibromatosis</td>
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<td>Optic nerve sheath</td>
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<td>Optic neuritis</td>
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<td>Optic N glioma</td>
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<td>Optic N meningioma</td>
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based on pathology. The imaging approach to each disease entity is dealt with at the end of the discussion.

Congenital Microphthalmia

It is defined as a globe with AP diameter of less than 21 mm in adult or less than 19 mm in an infant. The condition may be isolated or may present with other ocular and craniofacial anomalies, congenital rubella,
leukokorias. If the embryological insult occurs before the complete invagination of the optic vesicle then an orbital cyst is formed and is referred to as microphthalmos with orbital cyst. (Fig 2).

**Staphyloma and Coloboma**

Staphyloma results due to diffuse thinning and stretching of the sclera-uveal coats of the eyeball. The causes include progressive myopia, glaucoma, post infectious and inflammatory conditions. Coloboma on the other hand is a notch, gap or a fissure in which a tissue or portion of tissue is lacking. The cleft appears in the inferonasal quadrant of the globe with optic disc excavation in a typical Coloboma. Retinal detachments are well documented with Colobomas. Ocular colobomas may involve iris, lens, ciliary body, retina, choroid, optic nerve or sclera.

**Ocular Inflammatory Disorders**

Majority of the inflammatory disorders of the globe such as infectious conditions, papilledema, episcleritis and uveitis are usually a clinical diagnosis. Imaging does not play a significant role except when there is an abscess or space occupying lesion like a cysticercus which is causing the symptoms.

**Leucokoria**

Leucokoria is a white, pink white or a yellow white pupillary reflex and results from any intraocular abnormality that reflects the incident light back to the observer. Causes of Leucoria are Retinoblastoma, Persistent fetal vasculature, Retinopathy of prematurity, Cataract, Coloboma (fissure or cleft) of choroid or optic disc.
disc, Uveitis, Toxocariasis, Coats’ disease, Vitreous hemorrhage, Retinal dysplasia

The radiological approach to common causes of leucokoria can be summarized as in Table 3.

**Retinoblastoma**

It is the most common intraocular tumor of childhood. Imaging plays a crucial role in differentiating it from benign mimics and evaluating the extent of retrobulbar spread of the lesion. It rules out tri/tetralateral lesions in the pineal gland or suprasellar region. Punctate or finely speckled calcification is seen in 90-95 % cases on CT. MR is useful for assessing extraocular and intracranial disease as extension beyond the eye signifies close to 100 % mortality. (Fig5.)

![Retinoblastoma CT image](image1)

**Fig. 5.** Retinoblastoma. CT image reveals a hyperdense calcified mass in right globe of a 2 year old child.

**Coats’ disease**

Coats’ disease also known as primary retinal telangiectasias, is seen as subretinal exudates. Calcification is not a feature. It is important to differentiate this from a non-calcified retinoblastoma which is not possible on CT. On MRI Coat’s disease shows hyperintense signal on T1, T2 and proton density images whereas retinoblastoma appears hyper on T1 and proton density, and hypointense on T2W images. Enhancement may be seen which is attributed to idiopathic intraretinal telangiectasia and microaneurysms.

**Persistent hyperplastic primary vitreous (PHPV)**

PHPV is caused by failure of embryonic hyaloid vascular system to regress normally. On CT the globe is small,

![PHPV CT image](image2)

**Fig. 6.** a, b: PHPV. CT shows hyperdense retrolental soft tissue in posterior chamber of right eye. Colour doppler demonstrates the persistent hyaloid artery in the right globe coursing anteriorly in the vitreous chamber.
calcification is absent and a generalized increased density of vitreous chamber may be visible. The enhancement of abnormal intravitreal tissue (the retrolental tissue stalk) may be seen. There may be an associated small and irregular lens with a small and shallow anterior chamber. (Fig. 6a, b).

**Retinopathy of prematurity (ROP)**

ROP is seen in premature low birth weight babies due to prolonged exposure to oxygen therapy. Spontaneous regression is seen in 85-95% with the disappearance of neovascularisation and formation of dense membrane or a vascularised mass that is left back as a permanent evidence of active phase. Calcification is rare. Involvement is bilateral and often asymmetric. There may be associated massive persistent hyaloid vascular system. Clinical history plays a crucial role in differentiating these from other causes of bilateral leucokoria.

**Uveal Masses: Uveal melanoma**

Malignant uveal melanomas are more frequent in whites than blacks. Less than 2% of patients affected are under 20 years. Dynamic CT may help to differentiate it from a choroidal hemangioma. They appear as elevated hyperdense sharply marginated lenticular or mushroom shaped lesions and show a moderately high signal on T1W and PD images and low on T2W sequences. Lesions smaller than 3 mm are better detected on ultrasound. They show moderate post contrast enhancement which is important in defining the extraocular extent of the disease. Other mass lesions which are difficult to differentiate from each other are uveal metastasis, choroidal hemangioma and retinal astrocytoma.

**Orbital Diseases**

**Congenital Dermoid/Epidermoid**

These are choristomas which are among the most common orbital tumor in childhood and are seen superior or temporal in location. Both appear as well circumscribed smoothly marginated low density masses. Fatty tissue or calcifications are a feature of dermoids. On MR both are seen as hyperintense on T1 and T2 depending upon the content of fat and are of negative HU values on CT. Minimal enhancement of capsule may be seen. On CT they show negative Hounsfield values typical of fat (Fig. 7).

**Inflammatory**

The majority of acute inflammatory disorders are of paranasal sinus origin. Inflammatory disorders can be classified as inflammatory edema, subperiostal phlegmon and abscess, orbital cellulitis, orbital abscess and ophthalmic vein and cavernous sinus thrombosis. Imaging in inflammation is aimed at detecting the underlying cause if any, intraocular and intracranial extension and bony involvement. In post septal orbital cellulitis, orbital imaging with contrast enhanced study is indicated to differentiate inflammatory edema, cellulitis, phlegmon and orbital abscess. Subperiostal phlegmons (Fig. 8) may result from collection of inflammatory tissue and edema beneath the periosteum seen as diffusely enhancing soft tissue and this may progress to abscess (Fig. 9a, b) formation when...
it shows peripheral enhancement with a non-enhancing centre.

Complications such as cavernous sinus thrombosis is seen as low attenuation non enhancing structure on contrast scans. Engorgement of cavernous sinus ophthalmic veins and extraocular muscles is seen. MR Venography is more sensitive than CT and MR angiography reveals deformity of cavernous portion of internal carotid artery.

**Mycotic infections**

Mucor and aspergillus are the most common fungal organisms incriminated and usually affect diabetics and immunocompromised. Spread to orbit occurs usually from paranasal sinuses. Hypointensity of the mycetoma on T2W images due to the paramagnetic materials produced by the fungi is an important finding and helps to distinguish from other lesions which are frequently iso to hyperintense on T2. (Fig. 10a, b)

**Pseudotumors**

Orbital pseudotumours are part of a non-granulomatous inflammatory process in the orbit or eye and are without known local or systemic causes. Patients present with acute onset orbital pain, restricted eye movement, diplopia, proptosis or impaired vision. This acute presentation helps to differentiate other lesions that are similar in appearance but have a chronic presentation. Pseudotumours can be classified as follows:

1. Acute and subacute idiopathic orbital inflammation seen as thickening of uveal-scleral rim with obscuration of optic nerve junction and post contrast enhancement.

2. Acute and subacute idiopathic diffuse orbital inflammation seen as a diffuse lesion which fills up the retrobulbar space and moulds itself around the globe while respecting its shape. No bony erosions are seen even in large masses. (Fig. 11a, b)

3. Acute and subacute idiopathic myositis orbital inflammation which present as enlargement of muscles of superior complex and medial rectus. The enlargement extends anteriorly to involve tendon insertion. The ragged fluffy borders of involved muscles with infiltration of the fat with an inward bowing of the medial contour of the muscle belly help in diagnosis. (Fig. 12a, b)

4. Acute and subacute idiopathic apical orbital inflammation is seen as irregular infiltrative process at the orbital apex that may extend anteriorly along the posterior aspect of extraocular muscles (EOM's) or optic nerve.
5. Idiopathic dacryoadenitis is seen as enlargement of the gland. A viral etiology, sarcoidosis, Sjogren's disease, lymphoproliferative disorders, cysts, neoplasia has to be ruled out by biopsy if required. (Fig. 13a, b).

6. Perineuritis seen as ragged edematous enlargement of the optic nerve sheath complex.

**Tolosa-Hunt Syndrome (THS)**

THS is unilateral recurrent painful ophthalmoplegia involving 3rd, 4th, 5th and 6th cranial nerves. It is considered as a regional variant of idiopathic orbital tumor and is located in the superior orbital fissure or cavernous sinus. The cavernous ICA may show adventitial thickening with a cuff of inflammatory tissue surrounding it on MR and the superior ophthalmic vein may be occluded.

**Thyroid Orbitopathy**

Grave's dysthyroid ophthalmopathy is the most common cause of uni- and bi-lateral exophthalmos in the adult population. Generally more common in women and orbital manifestations appear approximately 2 to 5 years after onset of thyroid disease. Thyroid orbitopathy is presumed to be an autoimmune disease. Initially the extraocular muscles are infiltrated by lymphocytes, and later they undergo fibrosis resulting in restrictive myopathy. Exophthalmos and limitation of ocular motility are the most common presenting symptoms, and exophthalmos results mainly from enlargement of the EOMs and/or increased orbital fat volume. Inferior rectus muscle is involved most commonly, and the exophthalmos is almost always bilateral, usually being relatively symmetric. Imaging in the early stage may reveal bilateral proptosis with markedly swollen retrobulbar contents. Later on the muscle bellies start enlarging, with coronal sections being especially valuable in their evaluation. However, axial views are best to see the strangulation of the optic nerve. On CT or MRI, the characteristic finding is enlarged muscle belly with normal anterior tendinous insertion. (Fig. 14a,b). Another helpful finding is the presence of hypodense areas within the muscle bellies. Additional findings are excessive orbital fat, enlargement (engorgement) of lacrimal glands, lid edema, anterior displacement of orbital septum, and stretching of the optic nerve with or without associated “tenting” of the posterior globe. In chronic stages, when fibrosis of the EOMs occur, CT and MRI may show fatty replacement or a string-like appearance of the EOMs.

**Sarcoidosis**

Granulomatous systemic disease affecting both sexes and all ages. Ophthalmic lesions are seen in approximately one fourth of the cases, and any part of the globe may be involved. The most common form of orbital involvement is chronic dacryoadenitis, which may mimic a lacrimal gland tumor. MRI is the modality of choice, with post contrast images being essential for the diagnosis. Multiple Enhancing granulomatous lesions which can involve any part of optic tract and

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Fig. 12. a, b: Patient with Myositic Pseudotumor. Coronal IR and T1 sequences display uniformly enlarged EOM on right side.

Fig. 13.a,b. Lacrimal Gland Pseudotumor. Patient presented with an acute history of orbital swelling. MRI shows an enlarged left lacrimal gland iso on T1 and hyper to EOM’s on T2W sequences with involvement of musculotendinous junctions.

Fig. 14.a,b: Grave’s Ophthalmopathy. Axial MRI showing fusiform enlargement of EOM’s hyperintense on T2 axial Fat sat images with sparing of musculotendinous junctions.
globe along with pituitary stalk involvement is known. Chest x-ray may be helpful in diagnosis especially in the active stages of the disease. 18.

**TUMOURS**

**Orbital Lymphoma**

Lymphoid tumours account for approximately 10% to 15% of orbital masses. Imaging cannot reliably differentiate benign from malignant lymphoid tumours. The CT and MRI findings are usually nonspecific, and based solely on imaging, it may be impossible to diagnose lymphoma confidently. Orbital lymphomas are well defined relatively high density homogenous masses and are often seen in the anterior portion of orbit, the retrobulbar area, or in the superior orbital compartment. Mild to moderate enhancement is usually present. All orbital lymphoid tumours tend to shape themselves around the orbital structures with no associated bony erosions. However, frank destruction of bone may be seen in aggressive malignant lymphomas. (Fig. 15a, b, c). Deformity of the globe shape is rare. Both pseudotumours and lymphoma may have intermediate to low signal intensity on T1-weighted and proton density MR images, and appear iso to hypointense to fat on T2-weighted images.

**Secondary Orbital Tumours**

Tumours may invade the orbit from its vicinity such as those arising from sinonasal cavities, skin of the face etc. The orbit may also be involved by metastases, especially from breast carcinoma in women, and carcinoma of the lung, kidney, or prostate in men. These may involve any of the orbital contents. (Fig. 16a, b)

**Orbital Vascular Conditions**

**Capillary hemangioma**

They are tumours of infancy show a rapid growth followed by gradual decrease in size 21. They derive their blood supply from either external or internal carotid arteries and are capable of bleeding profusely. These tumours can extend intracranially through the superior orbital fissure, optic canal and orbital roof. On CT they appear as fairly well marginated, irregular intensely enhancing lesions most of which are extracanal. On MR, they appear hypointense on T1 and hyperintense on T2 with intense enhancement on contrast injection. (Fig. 17a, b)

**Cavernous hemangioma**

It is the most common orbital vascular tumor of the orbit in adults and tends to occur in second to fourth
decade and progressively increase in size. They have a fibrous pseudocapsule and are well defined masses majority of which are intraconal. On CT they are smoothly marginated homogenous masses that respect the contour of the globe. (Fig. 18a, b, c) On MR they are hypo on T1 and hyper on T2. Dynamic MRI can help differentiate cavernous hemangioma from intraconal schwannoma. Hemangiomas show typical pattern of enhancement that starts from a point and then spreads to the entire mass whereas schwannomas show homogenous enhancement that involves a large area of the mass in the early phase. Prominent arterial supply is usually absent which is in contrast to capillary hemangiomas.

Lymphangioma

Lymphangiomas are tumours of children and young adults and also progressively enlarge during the first two decades. On CT they are poorly circumscribed heterogeneous masses often of increased density in intra or extracanal space with varying degree of enhancement. Bony remodeling may be present. On MRI fluid-fluid levels related to hemorrhages are characteristic. (Fig. 19a, b)

Orbital varix

These are congenital venous malformations characterized by proliferation of venous elements and massive dilatation of one or more orbital veins. A tortuous enhancing structure which changes form and gets distended on valsalva is confirmatory for orbital varix. (Fig. 20a, b) MRI can be performed with patient in supine and then prone to increase the venous pressure. They can present as thrombosis and hemorrhage.

Caroticocavernous fistula

Caroticocavernous fistula can be post traumatic or spontaneous secondary to osteogenesis imperfecta, Ehlers-Danlos syndrome and pseudoxanthoma elasticum. CT and MR imaging helps in identifying engorgement of superior ophthalmic vein. Angiography is the gold standard and helps in categorizing the fistula and deciding further management.

Neurogenic Tumors

Schwannomas and Neurofibromas together form approx 4% of all the orbital tumors. Schwannomas in the orbit arise from 3rd, 4th, 5th, 6th, 7th and the autonomic nerves and appear similar to cavernous hemangioma differing.
its enlargement. Imaging should include evaluation of the entire visual pathway. On MRI they are isointense to cortex and hypointense to white matter on T1 and hyperintense on T2.

**Perioptic meningiomas**

These benign tumors arise from the meningoendothelial cells of the arachnoid. They occur in the 4th and 5th decades with a female predominance. They present as an eccentric mass at the orbital apex or as a well defined tubular thickening or fusiform enlargement of the optic nerve sheath. Moderate to marked enhancement of the tumor gives a tram track appearance due to circumferential enhancement of the meningioma around a optic nerve and may simulate a pseudotumor. MRI displays the tumor as an abnormally enlarged optic nerve sheath and appear hypo to grey matter on both T1 and T2. Calcification may be seen.

**Other rare tumours**

Fibrous histiocytoma is a mesenchymal tumor and are seen as well circumscribed masses that may be intraconal and appear moderate to markedly hypointense on T2 images. These may not be able to differentiate from neurofibromas and other fibrous tumours. Rhabdomyosarcomas, although rare still remain the most common primary orbital malignancy of childhood. CT and MRI play an important role in staging. Imaging should include the possible metastatic sites. On CT they appear as enhancing tumours with bone destruction and invasion of surrounding structures. They can appear heterogeneous due to hemorrhage within the tumor. Findings are nonspecific and tissue diagnosis is required to differentiate it from other aggressive lesions.

**Lacrimal Gland Lesions**

Lacrimal gland lesions may be broadly classified as inflammatory or neoplastic. Inflammatory lesions cause diffuse enlargement of the gland. There may be associated features of myositis and scleritis. Inflammatory processes tend to involve all parts of the gland including the palpebral lobe. Pleomorphic adenoma account for 50 % of the tumors and appear as well defined and rounded (Fig. 23a, b, c) whereas...
Fig. 23.a,b,c: Lacrimal gland mass lesion: Coronal and Axial T2 weighted fat sat images (a,b) show hyperintense mass lesion involving orbital lobe of lacrimal gland. The lesion is enhancing on post-contast fat sat T1-weighted image (c). Histopathological correlation revealed a pleomorphic adenoma.

Adenoid cystic carcinoma is the commonest malignant tumor to affect the orbit tend to have microserrations. Table 4a,b.

Conclusion

Cross sectional imaging plays an important role in identifying and characterization of lesions in eye and orbit. Awareness of the usual location of the lesions, their density and signal intensities frequently help in clinching the diagnosis. The algorithmic approach provided in the article may help reach a diagnosis in most situations.

References


