



ISSN: 0976-3031

Available Online at <http://www.recentscientific.com>

International Journal of Recent Scientific Research
Vol. 8, Issue, 3, pp. 15802-15808, March, 2017

**International Journal of
Recent Scientific
Research**

DOI: 10.24327/IJRSR

Research Article

ROLE OF CT IN EVALUATION OF PROPTOSIS

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DOI: <http://dx.doi.org/10.24327/ijrsr.2017.0803.0007>

ARTICLE INFO

Article History:

Received 15th December, 2016

Received in revised form 25th

January, 2017

Accepted 23rd February, 2017

Published online 28th March, 2017

Key Words:

Proptosis, MDCT, Orbital Cellulitis, Optic Nerve Meningioma, Orbital Pseudotumor.

ABSTRACT

Proptosis is a common problem, encountered in ophthalmic outpatient department (OPD) requiring radiological evaluation for early diagnosis and to know the extent of involvement. CT is crucial not only in the diagnostic evaluation but also in treatment planning and follow up monitoring of diseases. Multidetector CT (MDCT) has the advantage of rapid acquisition of volumetric dataset (isotropic imaging) allowing excellent multiplanar reconstruction (MPR) images. We reviewed 3 cases of orbital cellulitis, 2 cases of idiopathic orbital pseudotumor, 2 cases of optic nerve meningioma, 2 cases of lymphoma, 2 cases of hemangioma, 1 case ethmoidal mucocele, 1 case of maxillary carcinoma and 1 case of tolosa hunt syndrome.

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INTRODUCTION

Proptosis is defined as an abnormal protrusion of the ocular globe. There are various lesions of the orbit causing proptosis which may be unilateral or bilateral. These include Idiopathic Inflammatory pseudotumor, orbital tumors (benign or malignant), orbital myositis, graves ophthalmopathy. The most common benign orbital tumor in adults is cavernous hemangioma. The most frequent cause of bilateral proptosis is Graves ophthalmopathy. Contrast Enhanced Computerized Tomography with axial and coronal views may be considered as a single, non-invasive diagnostic tool, which not only localizes and characterizes the lesion but also, will show calcification, cystic changes and extent of disease process.¹

Non-ionic iodinated contrast is used in appropriate doses to assess the tissue enhancement patterns in many orbital conditions. The orbit is the site of a large number of pathologies of diverse etiologies, and imaging has to be tailored to the symptoms and clinical findings.²

CT can distinguish normal and abnormal structures of different tissue density on the basis of differential X-ray absorption. The presence of orbital fat allows high spatial and density resolution of orbital structures. In contrast to direct coronal scans, sagittal and coronal reformations avoid high spatial frequency artefacts from dental appliances and other metal implants. Multiplanar reformations enable a lesion to be viewed in the optimum anatomic plane and its location to be

assessed relative to contiguous orbital, bone, sinus, and central nervous system structures. Modern CT equipment enables application of spiral technique of 2/3 mm slice-thickness and distance respectively.³

Diagnostic criteria for orbital proptosis; plane of the scan or axial sections must be parallel to the plane passing through the optic nerve head and lens.⁴

On CT scans, proptosis is defined as globe protrusion > 21 mm anterior to the inter-zygomatic line on axial scans at the level of the lens. Interzygomatic line is drawn first, a straight line connecting the anterior margins of zygomatic processes, at the level of median portion of the globe or orbit. The distance from posterior sclera margin to Interzygomatic line (IZL), Normal is 9.9mm ± 1.7mm. Computed tomography is of utmost importance in evaluating the orbital diseases especially when performed in orthogonal planes. It is also of great importance in the diagnosis and defining fields of surgery and radiotherapy. CT has become imaging modality for evaluating trauma, tumors, endocrinal and inflammatory lesion.⁵

Contrast enhancement may add definition and specificity in setting of orbital tumours, vascular malformations and inflammations. The strength of CT includes exquisite bony details, speed of examinations and excellent spatial resolutions. Rapid post processing of CT image provides 3D picture of lesions and helps surgeons in salvaging the normal structures during surgery. It is also possible to obtain appropriate tissue for biopsy under CT guidance and place the radioisotope in

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malignant tissues where other treatment modalities are not effective.⁵

Objective

To study the role of multi detector computed tomography in the evaluation of proptosis. To study various CT appearances of orbital lesions causing proptosis and to assess CT accuracy in the diagnosis of orbital space occupying lesions causing proptosis.

MATERIALS AND METHODS

Prospective study of 14 patients who were referred to the department of Radiodiagnosis in teaching hospitals attached to Bapuji Hospital and Chigateri General hospital, Davangere, Karnataka. Referred patients with clinically, fundoscopically or sonologically suspected of having orbital space occupying lesions causing proptosis CT was carried out using TOSHIBA Activion 16 slice MDCT machine. Imaging technique will include plain CT and IV contrast CT.

RESULTS

Idiopathic Orbital Pseudotumor

In the present study 2 patients were diagnosed with pseudo tumour, both the patients presented with painful proptosis and conjunctival infection with unilateral eye involvement in 1 cases, CT revealed enlarged and enhancing extraocular muscles. Attenuation was isodense, the muscles involved were thickened involving both the muscle bulk and tendon. Evidence of fat stranding in retrobulbar compartment was noted in almost all the cases, there was no bony remodeling or extra orbital extension.

Tolosa Hunt Syndrome

In the present study 1 patient was diagnosed with Tolosa hunt syndrome was seen. A 70 year old male patient who presented with decreased vision, diplopia and pain on left side. CT showed abnormal ill-defined isodense soft tissue with mild to moderate homogeneous enhancement in region of left orbital apex, and proximal left cavernous sinus globes and the right cavernous sinus were normal, there was No evidence of bony involvement.

Orbital Cellulitis

In the present study 3 patients were studied who presented with rapidly developing proptosis. On CT examination there was diffuse soft tissue thickening of eyelid [preseptal compartment] with extension of inflammation into postseptal space, orbital apex was abnormal in all the cases.

Optic Nerve Meningioma

In the present study they were 2 cases of optic nerve meningioma, On CT examination evidence of dilated optic nerve which was enhancing on contrast and showed homogeneously enhancing intraconal mass with stippled calcification was noted along the optic nerve, there was no E/O bony erosion / intracranial extension.

Ethmoidal mucocele

In the present study one case of ethmoidal mucocele was diagnosed. On CT examination there was evidence of

homogenous low density soft tissue mass noted in the ethmoidal sinus causing erosion of adjacent bone and protruding into orbit.

Lymphoma

In the present study two cases of orbital lymphoma were diagnosed. In one cases CECT showed Bilaterally enhancing cone shaped mass lesion in the superolateral compartment of the orbits, predominantly extraconal with mild intraconal extension.

Maxillary Carcinoma

In the present study there was 1 case of maxillary carcinoma, heterogeneously attenuated Lesion was seen extending from maxillary sinus into orbit destructing the orbital apex and was also seen extending into ethmoid sinus, sphenoid sinus and cheek.

Hemangioma

In the present study there were 2 cases of intraconal hemangioma, CECT showed intensely enhancing mass superior to retro orbital part of optic nerve compressing optic nerve inferiorly.

DISCUSSION

Idiopathic Orbital Inflammation Syndrome (IOIS)

Idiopathic orbital inflammation syndrome (IOIS) also known as idiopathic orbital pseudotumor (first described by Birch-Hirschfield⁶ in 1905), is a benign, non-infective, inflammatory condition of the orbit with no identifiable local or systemic causes.⁶ It is the third most common orbital disease after thyroid orbitopathy and lymphoproliferative disorder.⁷ Lesions are commonly restricted to the orbit. However, nonspecific inflammatory tissue of the orbit may extend into the adjacent retro-orbital structures through one or more foramina, including the superior orbital fissure, optic canal, and inferior orbital fissure.

In a study conducted by Yumei Li *et al*⁸ 8 of 73 patients (11%) had CT evidence of retro-orbital involvement. These 8 patients were all diagnosed with Idiopathic orbital pseudotumour following biopsy of the orbital lesion that was associated with abnormal retro-orbital soft tissue. All 8 patients had a unilateral orbital lesion

Sheedy-Forbes GS *et al*,⁹ reported the attenuation of pseudotumors on CT isodense to increased density. In our series, the attenuation was isodense. Tier they stated that the recti muscles were thickened in almost all cases and sometimes associated with a mass and the mass was well-enhancing. This forcing pattern was also seen in our series of orbital pseudotumor studied however the enhancement was mild.

Enzmann *et al*¹⁰ noted that sparing of the muscle insertion in Graves disease and bilateral Involvement helps to distinguish this condition from a pseudotumor. We also applied the same criteria in distinguishing the two lesions.

Tolosa-Hunt syndrome (THS)

Is a rare disorder indicated by recurrent painful ophthalmoplegia caused by non-specific inflammation of the cavernous sinus or superior orbital fissure (SOF). The disease

shares histopathological features with idiopathic orbital pseudotumour; however, owing to its anatomical location, it produces characteristic clinical manifestations.¹¹ Recurrent retro-orbital pain, with palsies of the third, fourth or sixth cranial nerves as well as the first and second divisions of the trigeminal nerve, are typical. Clinically, immediate response to steroid therapy is a hallmark of the condition.¹¹ Tolosa first described the condition in 1954, in a patient with unilateral recurrent painful ophthalmoplegia involving cranial nerves III, IV, VI and VII. The patient was imaged using carotid angiography, and segmental narrowing of the carotid siphon was seen.

Orbital Cellulitis

Orbital cellulitis is a postseptal infectious process most commonly caused by paranasal sinusitis, which spreads to the orbit via a perivascular pathway.¹² Thus, bone destruction is not usually seen. The symptoms include swelling and erythema of the eyelids, chemosis and proptosis. Visual acuity is usually maintained.¹³ Treatment of orbital cellulitis typically requires the intravenous administration of antibiotics. Complications of orbital cellulitis include thrombosis of the superior ophthalmic vein, the cavernous sinuses, or both; bacterial meningitis; epidural and subdural abscess; and parenchymal brain abscess.¹³

In a study conducted by KK. Sabharwal¹⁴ subperiosteal involvement was the most common with preseptal extension seen in all showing increased density of the soft tissues. Formation of subperiosteal abscess with medial enhancing rim was seen. Diffuse orbital involvement with increased density of soft tissues in both intraconal and extraconal spaces was also seen. Concurrent ethmoid and maxillary sinusitis was seen.¹⁴

Optic Nerve Meningioma

Optic nerve meningiomas are benign tumours arising from the arachnoid cap cells of the optic nerve sheath, and represent ~20% of all orbital meningiomas, the majority of which are direct extensions from intracranial meningiomas.

Frederick A Jacobeik,¹⁵ conducted a study combining the clinical and computed tomography diagnosis of optic nerve glioma and meningioma, in their study. Patients with gliomas generally manifested massively swollen fusiform optic nerves with clear-cut margins due to circumscription by an intact dura. Kinks and bucklings of the optic nerve as well as infarctive cysts distinguished the glioma CT-scan patterns from the meningiomas. CT-scan features of the meningiomas were narrowly and diffusely enlarged nerves, calcification; irregular excrescent margins signifying extradural invasion into the orbital soft tissues; and often revealed irregular margins signifying transgression of the dura.

Ethmoid Mucocele

A mucocele is an epithelial-lined, mucus-containing sac that is the most common cause of paranasal sinus expansion. Accumulation of mucoid secretions behind an obstructed paranasal sinus ostium is the primary etiology. This accumulation expands the sinus cavity and produces thinning or erosion and remodeling of the bony wall. Occur most frequently in the fronto-ethmoidal region (90%). The frontal sinuses are involved approximately in 60% of cases, the

ethmoidal complex in 30%, the maxillary in 10%, and the sphenoid sinuses are involved only rarely. There is an increased incidence in patients with cystic fibrosis.¹⁶

Mucoceles are usually observed during adulthood. In most cases, patients have a clinical history of chronic nasal polyposis or pansinusitis. The leading symptoms are unilateral proptosis with ophthalmalgia, double vision or decreased acuity, a palpable mass in the superior medial aspect of the orbit or medial canthus with the fronto-ethmoidal mucoceles, and headaches.

On CT examination, a soft-tissue density mass may be seen obliterating the sinus with associated expansion of the involved sinus. Bony changes, such as erosion or thinning, may also be seen, especially with frontal mucoceles. Surrounding zone of bone thickening due to chronic infection is another manifestation. After intravenous contrast administration, there is uniform lack of enhancement with only a rim of enhancement from the infected mucosal membrane.

Lymphoma

Orbital lymphoma represents a small fraction of all systemic lymphomas that account for approximately 1–2 % of non-Hodgkin lymphomas. Lymphoma is reported to constitute between 6 and 8 % of orbital tumours, however, the incidence is estimated to be rising and has doubled in the last two decades.^{17,18}

Lymphoproliferative disease includes a spectrum of disorders ranging from benign (lymphoid hyperplasia) to malignant disease (lymphoma), passing by atypical lymphoid hyperplasia. Orbital lymphomas are a heterogeneous group of malignancies, most of them are primary extra nodal lymphoma of the marginal zone of mucosa associated with lymphoid tissue (MALT type lymphoma). These lymphomas arise in lymphoid tissue acquired in certain extranodal sites as a result of chronic inflammation or autoimmune disorders.

This tumour is seen more commonly in the 5th–7th decades of life with a slight female predominance. The orbital infiltration by lymphoma is characterised by a palpable, firm or rubbery mass. Other symptoms are progressive proptosis, decreased visual acuity, motility disturbances and diplopia. Occasionally, periorbital edema is seen.^{19,20,21}

In a study conducted by Sharma P *et al*,²² Lymphoma was seen in 6.7% of patients of proptosis, as compared to our study of 2(6.7%) cases, lymphoma appeared as a homogeneously enhancing soft tissue density mass lesion in the extraconal compartment. Margo CE *et al*,²³ reported lymphoma as the most common malignant orbital tumor. Similar to this, Sabharwal KK *et al*¹⁴ reported lymphoma as the most common cause of proptosis.

Maxillary Carcinoma

Maxillary sinus carcinomas are rare, comprising 0.2–0.8% of neoplasms, 3% of head and neck carcinomas, and 80% of all cases of paranasal sinus tumors.²⁴

The majority of tumors occurring in the maxillary antrum are of epithelial origin and epidermoid carcinomas correspond to more than 80% of all cases of malignant neoplasms, the

adenocystic carcinoma being the second more frequent of them.

Computed tomography (CT) and magnetic resonance imaging (MRI) are well-established and useful techniques for evaluating the tumor extension to adjacent areas.

Because of the tumor localization and absence of early symptoms, the patients usually present with advanced tumors at the moment of diagnosis and when the tumors are small sized, they are misdiagnosed as chronic sinusitis, nasal polyp, lacrimal duct obstruction, or even cranial arteritis. In 40% to 60% of cases there are facial asymmetry, oral cavity swelling and tumor extension to the nasal cavity. These lesions extend medially towards the nasal cavity; superiorly they may invade the orbit and ethmoid sinus; anterolaterally, they may reach soft tissues and cheek; and, inferiorly, the maxillary sinus floor, dental alveolus and palate. Posteriorly, they may reach the pterygopalatine fossa and pterygoid muscles.

Hemangioma

Cavernous malformations, also known as cavernous hemangiomas, are the most common vascular lesions in adults. Cavernous malformations occur most often in women (60%–70%) between the ages of 18 and 72 years (mean age, 43–48 years), slowly and progressively enlarge, and do not involute.²⁵ Cavernous malformations usually are solitary and most often occur in the lateral aspect of the retrobulbar intraconal space. Bone remodeling is not uncommon, and intralesional calcification occurs occasionally.²⁵ CT demonstrates the cavernous haemangiomas as a homogeneously dense mass with smooth margins which shows uniform contrast enhancement. Most are located within the muscle cone. They do not deform the globe. It can be easily demarcated from the adjacent optic nerve and the muscles.²⁶



A 56 years old patient complaining of slowly progressive proptosis of right eye.



A 56 years old patient complaining of slowly progressive proptosis of right eye.

Case – 1 A Case of Optic Nerve Meningioma



Axial CECT section shows large heterogeneously enhancing mass in the right ethmoid sinus eroding medial wall of orbit.



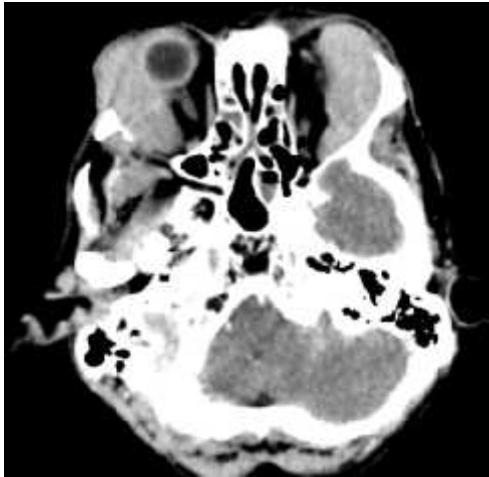
Coronal CECT section of same patient-shows evidence of heterogeneously enhancing mass in the right ethmoid sinus with erosion of medial wall and roof of maxillary sinus.

Case – 2 A Case of Ethmoidal Mucocele

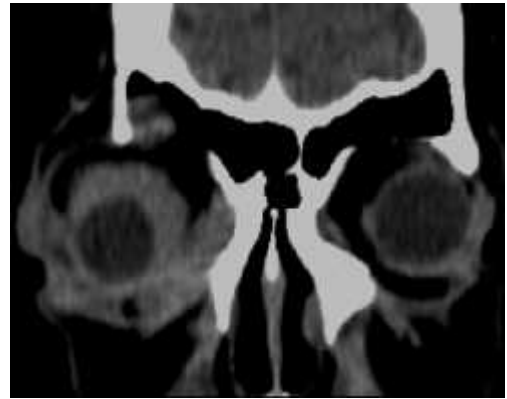


CECT axial section shows intensely enhancing mass superior to retro orbital part of optic nerve compressing optic nerve inferiorly

Case – 3 A Case of Right Orbital Intraconal Hemangioma



CECT axial sections shows Bilaterally enhancing cone shaped mass lesion in the superolateral compartment of the orbits, predominantly extraconal with mild intraconal extension.



CECT coronal section of same patient shows lesion extending anteriorly upto lacrimal fossa.

Case – 5 a case of orbital pseudo tumor

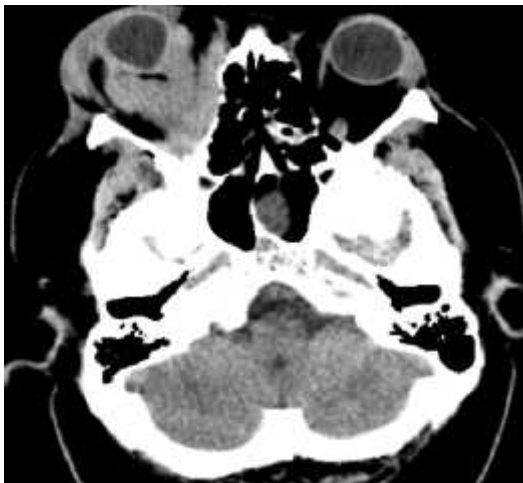


A 62 years old patient with previous paranasal sinus infection.



Coronal CECT section of same patient shows the mass in close relation with the supero lateral wall of orbit.

Case – 4 A Case of Lacrimal Gland Lymphoma



CECT axial section shows Homogeneously enhancing ill defined mass in the right orbital region involving both intra/extraconal compartments with encasement of the optic nerve and extraocular muscles.

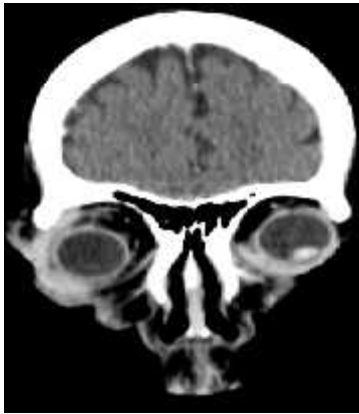


CECT axial section shows minimally enhancing soft tissue density in the superomedial aspect of right intraconal compartment with optic nerve involvement.

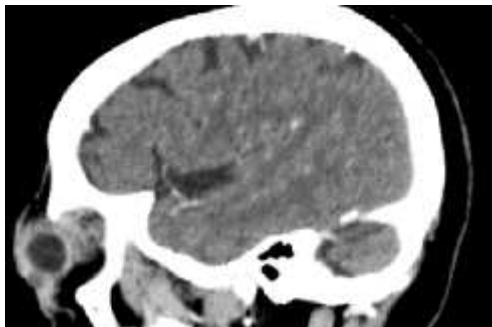
Case 6 A Case of Orbital Cellulitis



CECT axial section shows Soft tissue thickening with minimal enhancement in the right periorbital region with extension into preseptal space.



Coronal CECT section of same patient shows periorbital soft tissue thickening.



Sagittal CECT section of same patient shows periorbital soft tissue swelling extending into lateral rectus muscle

Case- 7 Another Case of Orbital Cellulitis



CECT axial section shows mass extending upto left infratemporal lobe

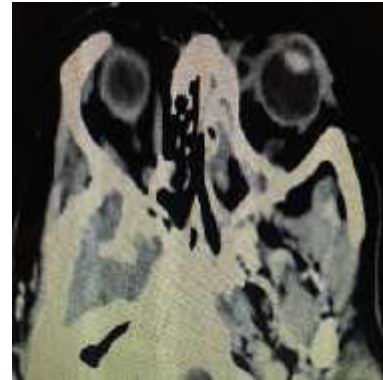


CECT coronal section of same patient shows mass anteriorly invading the orbit and posteriorly base of skull at left petrous bone

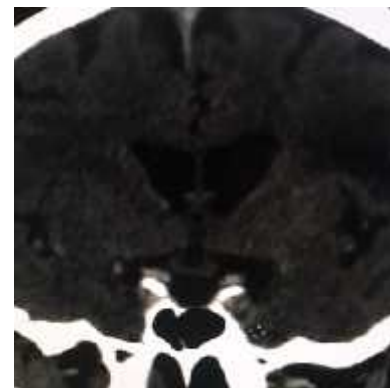
Case 8 A Case of Left Maxillary Carcinoma



70 years old patient with painful ophthalmoplegia



CECT axial section shows soft tissue density lesion in the left intraconal compartment extending to orbital apex



Same patient CECT coronal sections shows prominent left cavernous sinus in comparison with right side

CASE 9 A Case of Tolosa Hunt Syndrome

CONCLUSION

- Computed tomography (CT) has revolutionized the diagnosis and management of ocular and orbital diseases. The use of thin sections with multiplanar scanning (axial, coronal and sagittal planes) and the possibility of three-dimensional reconstruction permits thorough evaluation
- CT is modality of choice in assessing orbital lesions. Compartmentalization helps in arriving at a diagnosis. CT is useful to characterize: the precise location of the lesion - the intraconal space (including muscles & Optic nerve), the extraconal space (associated or not to an extra orbital lesion), or the eyeball; the features of the lesion (density, calcification, enhancement).

These findings are helpful to generate a differential diagnosis.

- CT is also useful to demonstrate the precise extension of the orbital lesion, the involvement of adjacent paranasal sinuses & nasal cavity, the evidence of bone erosion and intracranial extension which helps in pre treatment evaluation & post treatment follow up.
- CT also helps monitoring response to therapy. CT was fairly accurate in narrowing differential diagnosis.
- To conclude CT scan can be considered as a cost effective, non invasive, reliable diagnostic tool for evaluation of proptosis.

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How to cite this article:

Kishore Chautakuri *et al.* 2017, Role of Ct In Evaluation of Proptosis. *Int J Recent Sci Res.* 8(3), pp. 15802-15808.