PROPTOSIS; CAUSES AND DIAGNOSTIC ROLE OF CT

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ABSTRACT

Proptosis of the eye is an important clinical manifestation of orbital diseases. Proptosis due to any cause can compromise visual function and the integrity of the eye. Delayed diagnosis and improper treatment can lead to unintended sequelae.

The aim of this study was to retrospectively analyze the causes of adult proptosis and was to present the diagnostic role of CT in evaluation of proptosis.

We reviewed 39 adult patients, over 15 years of age. They presented to out-patient of Kamphaengphet Hospital with clinical proptosis. The period of collections extended from January, 2006 to February, 2008.

There are numerous causes of proptosis. In order of frequency, the causes of proptosis is trauma[15] following by tumors[9] and inflammatory diseases including thyroid opthalmopathy and orbital pseudotumor.[7] The most common cause of adult proptosis in Kamphaenphet hospital is trauma, which is different from those reported in other literatures.

In conclusion, with recent improvements in scanning techniques along with the wider availability of the current CT scanner, it has been proven to be excellent in identifying orbital pathology responsible for proptosis especially in places where MRI is not available.

Key words: CT, proptosis

INTRODUCTION

Proptosis is forward projection or protrusion of one or both eyeballs. Oweing to the rigid bony structure of the orbit with only anterior opening for expansion and seeing the objects by the eye, any increase in orbital contents taking place from the sides or from behind will displace the eyeball forward.

The definition of exophthalmos and proptosis are similar but in common usage, the latter tends to be more severe and gross. Some authors reserve the term exophthalmos as protrusion secondary to endocrine dysfunction and proptosis as any non -endocrine-mediated protrusion.[2,3,5]

Proptosis can be the sign of myriad disease processes including tumor, infection, inflammation, trauma, metastasis, endocrine dysfunction and vascular diseases.[1,2,3,5]

Whatever the underlying pathology, the orbit is basically conical in shape and so if something increases the volume within the orbit, the eye will be pushed outward.

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The patient who presents with proptosis must be evaluated. Imaging studies and corresponding laboratory investigations to ascertain the causes of the diseases must be obtained. The failure to address these findings can lead to unexpected sequelae. Certain symptoms, such as pain or a sudden diminution of vision, indicate a more rapid course and impose a more rapid investigation to obtain the diagnosis and treatment. Preservation of vision and function of the globe and orbit is of paramount importance.

We describe the variety of causes of proptosis and present the diagnostic role of CT in evaluation of the precise location and extent of the lesions.

MATERIALS AND METHODS

In clinical, the definition of proptosis is anterior displacement of one or both globes. The usual amount of ocular protrusion as measured from the lateral orbital rim to the corneal apex is 14 to 21 mm in adults. Protrusion greater than 21 mm or a 2 mm difference between the two sides is generally abnormal.[2,3,5]

Pseudoproptosis is either the simulation of abnormal prominence or a true asymmetry that is not the result of increased orbital contents.[5]

All of our patients, clinical proptosis were determined by ophthalmologist. 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.[5]

We retrospectively reviewed all 39 patients who came to out patient department of Kamphaengphet Hospital with proptosis, from January, 2006 to February, 2008. All of them underwent to perform CT by double slices spiral CT, (HiSpeed CT/e Dual CT scanner system, GE). Before commencing for CT examination, the evaluation including all the preceding history, complete ocular examination and laboratory investigation were recorded.

Various techniques of CT were applied to solve the different problems. The CT technique was to obtain a lateral scannogram with supine position and contiguous axial sections with slice thickness of 3-5 mm. Coronal CT with 3 mm. thickness in prone position were obtained in selected cases when required. The scans were obtained both prior to and after administration of intravenous contrast media, except in cases of trauma that were obtained only in plain study.

Final diagnosis of our patients was made by the CT findings correlated with clinical details, laboratory data, response to treatment and histopathological findings.

RESULT

39 patients with various age groups from over 15 to 68 years years, both sexes, (male 23 and female 16) with unilateral (36) and bilateral (3) proptosis were recorded.

39 patients were studied with proptosis, apparently resulting from multiple etiologies. In order of frequency, the causes of proptosis were: 15 orbital and facial trauma, 9 tumors including orbital tumors (metastasis and lymphoma) and paraorbital tumors (meningioma, sella mass, local spread of nasopharyngeal and paranasal sinus CA), 4 thyroid eye disease, 3 orbital pseudotumor, 3 orbital cellulitis, 1 ethmoidal mucocele, 1 fibrous dysplasia, 1 post traumatic carotid-cavernous sinus fistula, 1 cavernous sinus thrombosis, 1 sphenoid dysplasia in neurofibromatosis type I.

Various lesions causing proptosis are grouped in the table 1.

TRAUMA (15 patients)

Trauma is the most common cause. Most of them were young men with motorcycle accident. All 15 patients of traumatic proptosis have various degrees of intraorbital hemorrhage and soft tissue injury including preseptal and retro-bulbar regions (Figure 1). One or more fractures of orbital osseous wall and periorbital soft tissue contusion were seen

in all cases. 7 patients had orbital soft tissue emphysema. Associated facial bony fractures are seen for 5 cases. There were 8 patients who had evidence of directed or indirected sign of skull base fracture. 6 patients had intracranial injury, 2 of 6 were acute subdural hematoma and the rest 4 patients were localized frontal and or temporal brain contusions. One patient developed orbital cellulitis.

TABLE 1

CAUSES OF PROPTOSIS OF 39 PATIENTS

CAUSES OF PROPTOSIS	NO. OF PATIENTS	PERCENTAGE
TRAUMA	**15**	**39%**
TUMORS	**9**	**23%**
1. Orbital tumor		
- Metastasis	1	
- Lymphoma	1	
2. Paraorbital tumors		
- Meningioma	2	
- Maxillary CA	2	
- Nasopharyngeal CA	2	
- Sella mass	1	
INFLAMATION	**7**	**18%**
1. Thyroid ophthalmopathy	4	
2. Pseudotumor	3	
INFECTION	**4**	**10%**
1. Orbital cellulitis	3	
2. Ethmoidal mucocele	1	
VASCULAR	**2**	**5%**
1. Post traumatic CCF	1	
2. Cavernous sinus thrombosis	1	
OTHERS	**2**	**5%**
1. Fibrous dysplasia	1	
2. Sphenoidal, NF type1	1	

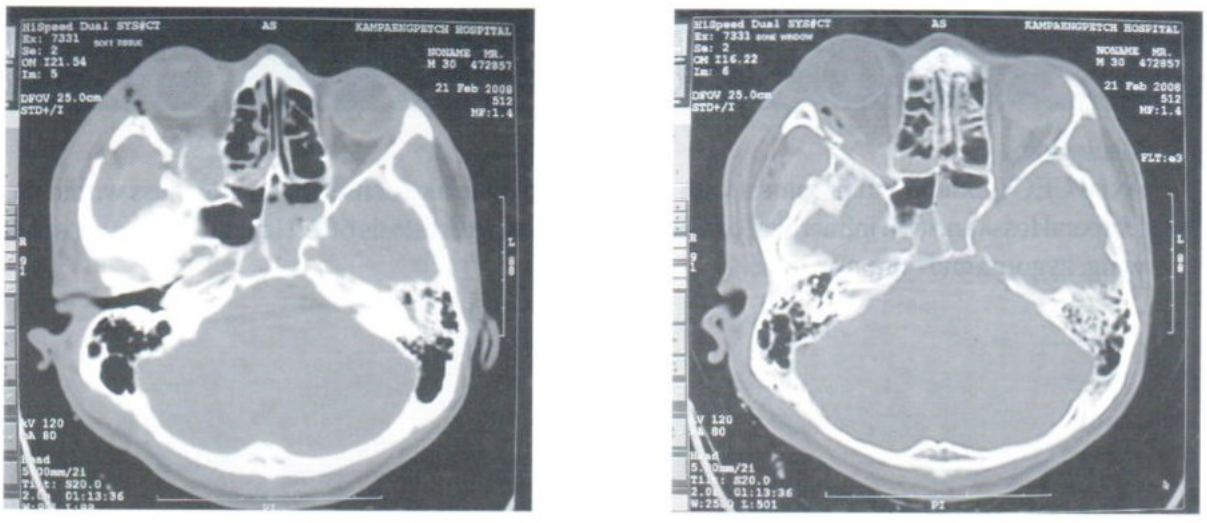

Fig.1 Axial CT with bone window setting shows right proptosis, comminuted fractures of right lateral orbital wall, retrobulbar hemorrhage, soft tissue swelling and emphysema. Skull base fracture with blood within the sphenoid and ethmoid sinuses are observed.

TUMOR (9 patients)

Second most common causes is tumor, there are 9 cases, including orbital and paraorbital tumors, as described.

Two patients were meningioma, CT reveal dense enhancing enplaque mass and dural enhancement within the temporal fossa and bony hyperostosis of sphenoid wing, zygoma, lateral orbital wall and temporal bone resulting in proptosis (Figure 2).

Four patients were primary paranasal sinus CA[2] and nasopharyngeal CA.[2] CT showed aggressive inhomogeneous enhancing primary soft tissue mass, adjacent bony destruction with orbital and or cavernous sinus invasion.

One patient was found to have a huge sella mass with gross cavernous sinus and orbital cone involvement. It was not known the exact pathology. This patient was referred.

One patient was probably orbital lymphoma; the CT shows homogeneous enhancing mass with extraconal and intraconal components. The tumor involved adjacent lacrimal gland and outward displacement of the globe. No bony destruction or optic nerve invasion is seen.

Orbital metastasis was presumed for one case, she had underlying primary CA of the breast. She developed unilateral proptosis. CT of orbit was performed. It reveals large soft tissue mass at the lateral extraconal retrobulbar space with blastic and lytic destruction of sphenoid bone (Figure 3).

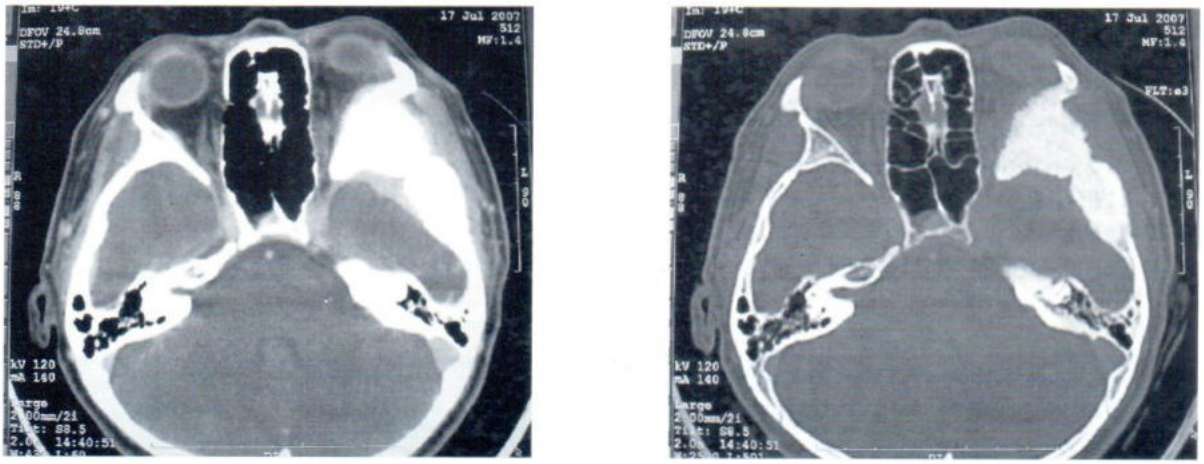

Fig. 2 Meningioma
Axial CECT exhibit left proptosis with extra-axial plaque like dense enhancing mass within the left temporal fossa and thin intraorbital component. There is hyperostosis of left lateral orbital wall, sphenoid wing, zygoma and temporal bone.

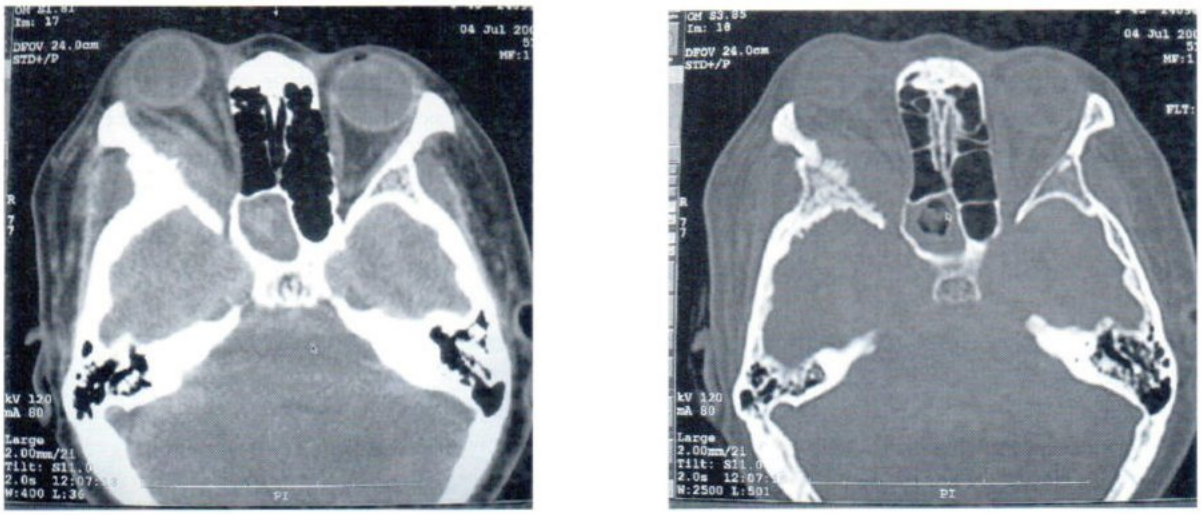

Fig. 3 Metastasis
Axial plain CT demonstrate right proptosis with large right lateral extraconal retrobular mass extending into sphenoid sinus. There is mixed sclerotic and lytic destruction of the right sphenoid wing, mimic meningioma.

INFLAMMATION (7 patients)

Thyroid ophthalmopathy with proptosis were found in 4 patients. All cases have history of hyperthyroid. 3 of them have abnormal thyroid function test. Bilateral multiple extraoccular muscles involvement were seen in 3 cases, one of them was not symmetry. Unilateral involvement was seen in one case. CT were reviewed, the contents of the orbits swelling due to inflammation that affects primarily the muscles. Various degrees of enlarged extraoccular muscles with almost tapering insertions were observed (Figure 4). The optic nerve at the orbital apex did not be grossly affected in all cases.

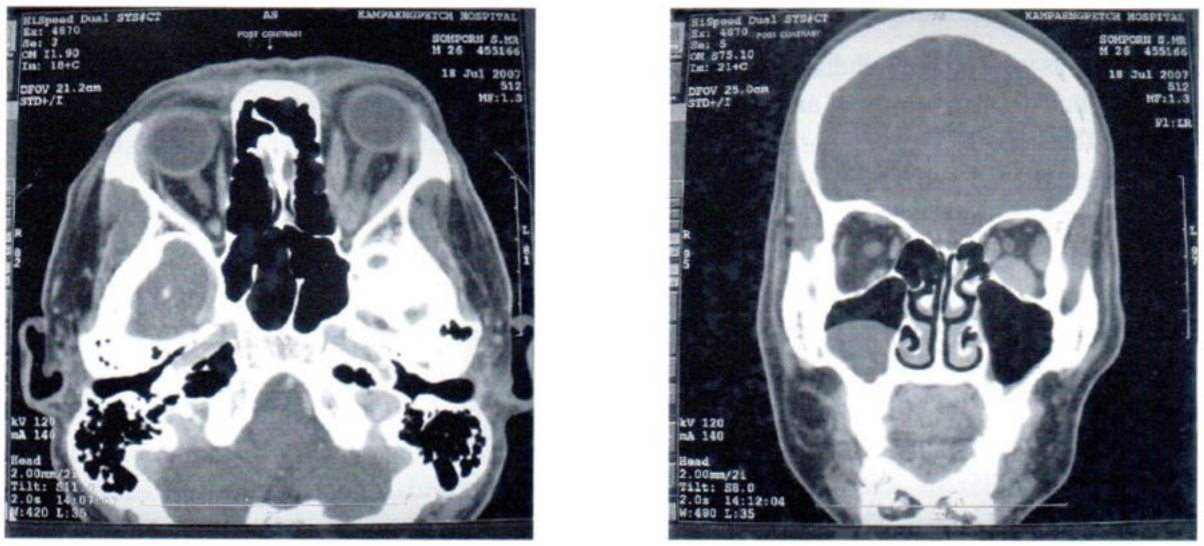

Fig.4 Thyroid Ophthalmopathy
Axial and coronal CT demonstrate unilateral left proptosis, enlarged left extraoccular muscles, maximally in the mid part with typical tapering at the insertions, involving inferior, lateral and medial rectus muscles.

There were 3 cases of pseudotumors, all of them are unilateral involvement. CT showed inflamed intraorbital structures including globe, lacrimal gland, extraocular muscle and orbital fat. There was no optic nerve involvement of our collected cases. One of them was a myositic type with isolated enlarged inferior rectus muscle and tendon insertion, it may be named pseudotumor variant (Figure 5). The rest two patients are diffuse lesions, their CT show diffuse enhancing uveal-scleral thickening, slight enlarged rectus muscles, retrobulbar fat infiltration and preseptal soft tissue swelling. Lacrimal gland involvement is seen in one case. Clinical details, CT features, prompt clinical response to steroid treatment and dramatic improvement on CT support the diagnosis of pseudotumor.

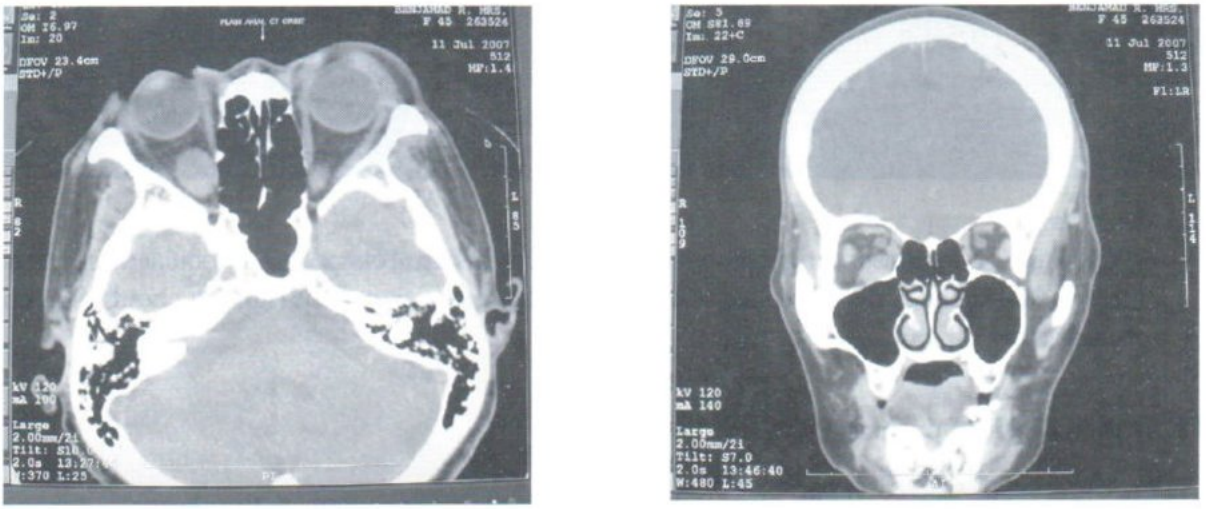

Fig.5 Pseudotumors
Axial and coronal CT reveal myositic type of pseudotumor, there is isolated enlarged right inferior rectus muscle belly and tendon insertion

INFECTION (4 patients)

3 patients were orbital cellulitis with proptosis. First patient had diffuse preseptal and retrobulbar cellulitis with eyeball involvement secondary to foreign bodies; the CT shows proptosis, preseptal soft tissue swelling, uveoscleral thickening and some increase enhancing retrobulbar fat. The second patient developed small subperiosteal involvement secondary to head and facial trauma. Formation of small subperiosteal abscess with medial enhancing rim along the lateral orbital wall, swollen enlarged preseptal soft tissue, retrobulbar fat infiltration, some enlarged rectus muscles and loss of soft tissue plain in the infratemporal fossa. The last patient was diffuse orbital cellulitis secondary to sinusitis, the CT features reveal increase density of soft tissue and inhomogeneous contrast enhancement in all compartments including preseptal and retrobular spaces.

One patient was diagnosed as ethmoidal mucocele. CT showed a large nonenhancing cystic mass arising from right ethmoid air cell. Expansion and remodeling of the bony wall, some erosion of medial wall of the right orbit were found. It was situated along the posteromedial aspect of right globe. Proptosis and optic nerve compression were seen (Figure 6).

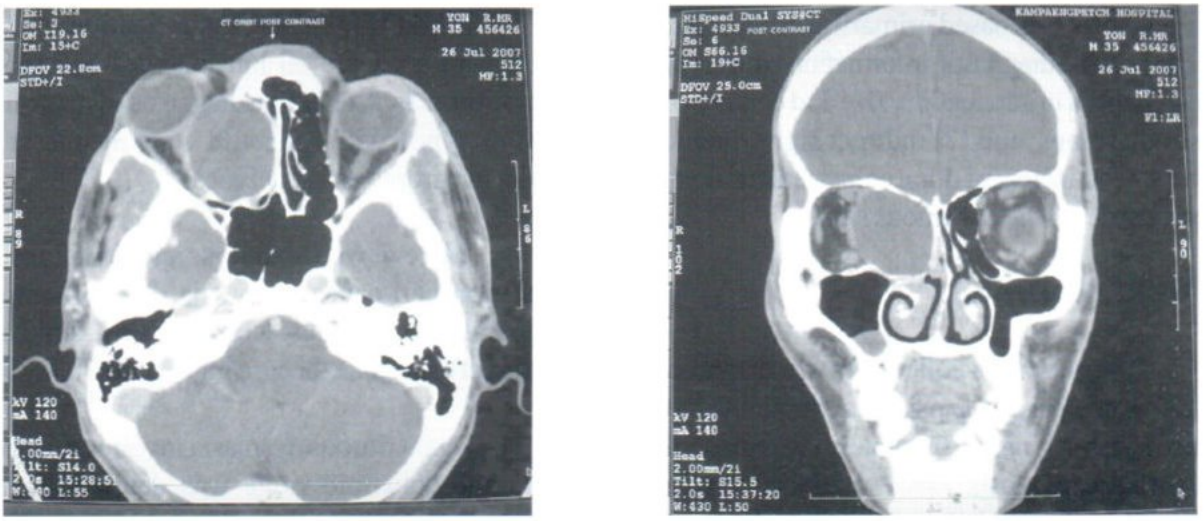

Fig.6 Ethmoidal Mucocele
Axial and coronal CT demonstrate right proptosis with a large expansile nonenhancing cystic mass arising from right ethmoid sinus with bony remodeling and some erosion of right medial orbital wall.

VASCULAR (2 patients)

In our result, there was one patient who was diagnostic as carotid cavernous fistula after head trauma. He presented as palsatile proptosis. CT reveal unilateral proptosis, associated enlargement and tortuous of the superior ophthalmic vein, swollen dense enhancing cavernous sinus with enlarged contour, some thickened extraocular muscles and notable ipsilateral dilated intracranial cortical venous drainage (Figure 7).

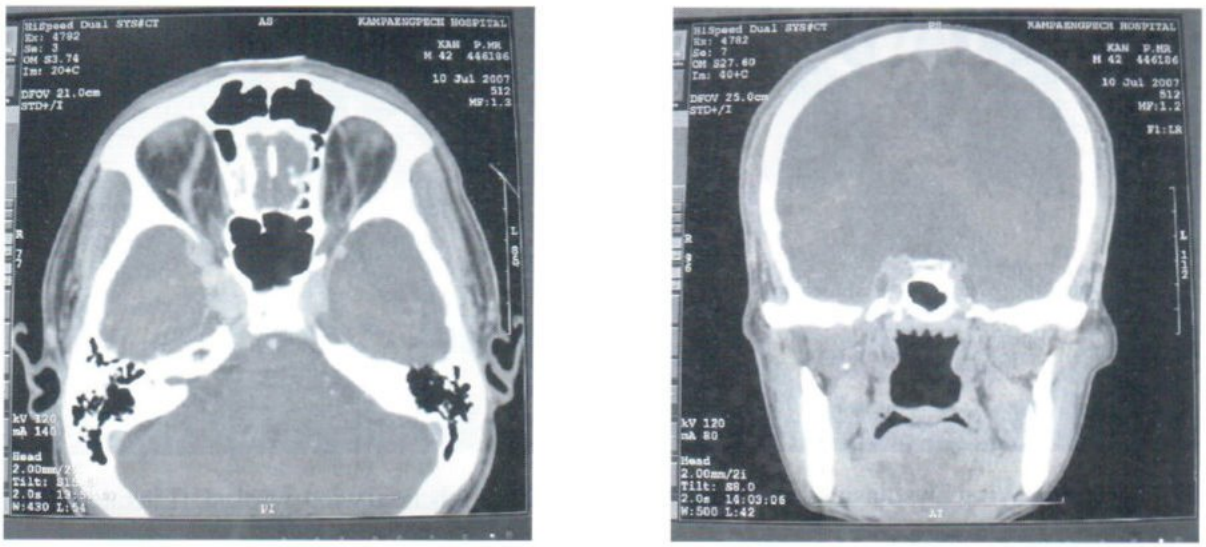

Fig.7 Carotid Cavernous Fistula
Axial and coronal contrast enhanced CT images show right proptosis with enlargement and tortuous of the right superior ophthalmic vein, swollen enhancing right cavernous sinus.

Another patient had cavernous sinus thrombophlebitis caused by paranasal sinusitis in 58-year-old woman, underlying poor controlled DM. She presented with right ocular pain and diplopia.

The diagnosis was based primarily on clinical data. CT can provide diagnostic information with (1) direct signs, changes in attenuation, size and contour of the cavernous sinus, and (2) indirect signs, including proptosis and increased dural enhancement along the lateral border of the cavernous sinus

OTHERS (2 patients)

The one of our collections was diagnosed as neurofibromatosis, proptosis was complained by the owner. Eye examination by clinician was made, there is no definite proptosis. However, she underwent an outpatient CT. CT demonstrate significant right sphenoid wing abnormality. Much of greater wing was absent. Slight herniation of the temporal lobe into the orbit and a convex bulging temporal fossa were found. Direct contact of the temporal lobe with the orbital soft tissue was noticed (Figure 8). Proptosis was measured on CT; there is no definite proptosis, too.

In addition general examination revealed more than six Café au lait macules and several neurofibromas. A tentative diagnosis of neurofibromatosis type1 with sphenoid wing dysplasia was made, but proptosis is equivocal.

We reviewed textbooks and literatures, this condition can cause proptosis due to herniated temporal lobe and bulgy temporal fossa, simulating an intraorbital space-occupying lesion (Figure 9).

Facial fibrous dysplasia involving temporal bone and sphenoid ridge was detected for one patient, the CT show some proptosis with enlarged expansion of involved bones.

Pseudoproptosis was not found in our series, it means that there was no negative CT finding in patients coming with proptosis whom CT were taken.

Fig.8 Sphenoid Wing Dysplasia (Case Study)

Fig.9 Sphenoid wing dysplasia (Literature Review)

Fig.8 An axial CT scan demonstrates dysplasia and absence of the right sphenoid bone and slight bulging of right temporal lobe, no definite proptosis.

Fig.9 An axial CT shows much absence of left sphenoid, enlarged middle cranial fossa, herniated temporal lobe and accompanying CSF sleeve anteriorly, simulating and intraorbital space occupying lesion, resulting in proptosis.[9]

DISCUSSION

Proptosis is a sign of an underlying disorder, and not a diagnosis itself. There are numerous causes. Due to the lack of direct visualization of the pathology in orbital disease, a thorough history and radiographic investigations are extremely helpful in arriving at a differential diagnosis and appropriate treatment. Actually, the diagnosis of proptosis can be made by multiple radiographic studies including ultrasonography (US), Magnetic Resonance Imaging (MRI) and computed tomography (CT).

Plain radiograph; there are a variety of views of the orbit that can be requested such as Caldwell, Water's, Rhese, lateral and axial basal views, however the findings are not pathognomonic of most of the proptosis.[1,2,5]

US has the advantage of ease to use, no ionizing radiation, excellent tissue differentiation and cost effectiveness. Additionally its safety, noninvasive nature and low cost makes it an attractive first line imaging modality and as a screening method before undertaken CT and MRI in some places. However, US is inferior to CT and MRI in depicting the bony wall and orbital apex, and of limited value in assessing lesions in the sinus and intracranial spaces. Other disadvantages of US are that, one can not take image of both orbits simultaneously and it is difficult for a nonspecialist to interpret. For these reason, US remains unpopular.[1,2,5]

MRI with its superb soft tissue contrast and multiplanar ability provides excellent rendering of orbital anatomy and pathology but is limited by lack of wider availability and high cost.[1,2,3,5]

CT is the most useful initial investigation, due to easy availability and operability, good maintenance and high speed. CT is currently the most valuable technique for delineating the shape, location, extent and character of lesion of the orbit. Furthermore, current CT scanner administers a very low dose of radiation.[2,3,5]

In our work up of proptosis, we found that plain CT was usually sufficient and most lesions were already well visualized especially in cases of orbital trauma, foreign body, orbital lesion associated with paranasal sinus diseases and thyroid ophthalmopathy. While contrast CT was generally used to assess tumor, vascular disorder and inflammation.

The common causes of proptosis in adult and children are different. A retrospective analysis by Sindhu et al examined 57 children with proptosis. Orbital cellulitis was the most common cause (22 cases), followed by thyroid eye disease (8 cases), optic nerve glioma (8 cases), orbital rhabdomyosarcoma (7 cases).[1] KK. Sabharwal et al reported 50 patients, the most common cause is tumor (23 cases) and infection (14 cases), followed by inflammatory disease (9 cases), and trauma (3 cases).[2]

Of our report, 39 cases of adult proptosis can be the result of a myriad of diseases. The most common cause is trauma (15 cases), following by tumor (9 cases), inflammatory disease (7 cases), infection (3 cases). The most common condition causing proptosis of our study was trauma that does not correlate with prior literatures. The explanation is that motorcycle accident without wearing helmet has been a big problem in our province. With the exception of traumatic cause, our result correlates well with the findings of KK. Sabharwal et al and Masud MZ et al. Both of them also described tumors as the most common causes in their studies.[1,2]

CONCLUSION

The most common cause of proptosis in adult presenting to Kamphaengphet Hospital was trauma, following by tumors and thyroid ophthalmopathy. Our result is not comprehensive but can help in forming a differential diagnosis of proptosis.

Proptosis is just a sign, it is usually necessary to undertaken specific radiographic investigation to further narrow down the different causes of proptosis.

We thought that CT is the main tool and most useful investigation. It is useful to characterize the precise location and extension. Furthermore, CT is more precise in demonstrating the bony changes.

CT features, in relation with age, clinical pictures, laboratory investigation, and response to treatment are able to give the correct diagnosis in almost cases. Once the etiology of proptosis is established, the appropriate treatment was done, the undesirable sequelae is diminished.

Although, MRI is the most excellent modality, we presumed that, CT scan can be considered as a cost effective, non invasive and reliable diagnostic modality for evaluation of proptosis. This is particularly true in the places where MRI and specialist for orbital US are unavailable.

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