Evaluation of etiopathogenesis of proptosis

Mohammed Ather1,*, Navneet Servey2, Rahmathunissa3, T. Sai Shreya4, Pallavi Gupta5, S. Saudamini6, Mohammed Arshad Ali7

1Professor, 2, 5Senior Resident, 3Assistant Professor, 5, 7Junior Resident, Dept. of Ophthalmology, Gandhi Medical College, Hyderabad, Telangana

*Corresponding Author:
Email: ather11258@gmail.com

Abstract
Aim: To study etiopathogenesis of Proptosis by clinical evaluation and confirming it by histopathological examination.

Materials and Methods: This is a prospective, interventional study carried out on 200 patients of proptosis attending Department of Oculoplasty and Orbital Disorders, of Sarojini Devi Eye Hospital, Hyderabad, which is attached to Gandhi Medical College. The study was conducted during the period of August 2011 to September 2013. All cases of proptosis which presented to our eye Hospital during the study period were included in study. The cases of proptosis that treated elsewhere were excluded from the study. All cases included were clinically evaluated using slit lamp, Hertel’s exophthalmometer, direct and indirect Ophthalmoscopy. Basic surgical profile was ordered in all cases who were suppose to undergo surgical intervention. Radiologically cases were evaluated by B scan ultrasonography, CT scan and / or MRI. In endocrine causes of proptosis T3, T4 and TSH was estimated Cases were subjected to orbitotomy to do incisional or excisional biopsy. Specimen were send for histopathological examination to come to final diagnosis. The cases of proptosis which require medical management were started on requisite medical treatment after confirming diagnosis.

Results: Out of 200 cases 108 (54%) were males and 92 (46%) were females. 176 (88%) of the cases had unilateral proptosis and 24 (12%) had bilateral proptosis. 112 (56%) cases had axial proptosis and 88 (44%) had eccentric proptosis. Out of 200 cases, 148 cases (74%) had chronic onset, 40 cases (20%) had subacute onset and 12 cases (6%) had acute onset of presentation. Etiology varied from infection, Inflammatory conditions like TRO, NSOIS, benign neoplasias like Hemangiomias, Dermoid, Pleomorphic adenoma of lacrimal gland to malignant neoaplasias like Lymphomas, Retinoblastoma, Rhabdomyosarcoma and miscellaneous condition like Carotico cavernous fistulas.

Keywords: Axial, CT scan, Eccentric, HPE(Histopathological exam), IHC Immuno histo chemistry, Proptosis

Introduction
The orbits are a pair of closed and guarded cavities. The volume of the globe is only 25% (7cc) of the volume of the orbit (30cc). The orbit is in intimate relationships with the cranial cavity, nasal cavity, and the para nasal air sinuses. This makes it vulnerable to many disorders that might involve the orbit by contiguity or through the venous drainage. For clinical and pathological purposes the orbits are divided into the intra-conal (surrounded by the extra-ocular muscles and their investing sheath) and the extra conal (outside the muscle cone) compartments. This again is subdivided into a superior, and an inferior extra conal orbital spaces. The important structures that occupy the intra conal space are the optic nerve with its meningeal coverings and the subarachnoid space, the ophthalmic artery and the sympathetic plexus. A lesion in this location e.g.: an intra-conal tumour produces noticeable axial proptosis. Lesion in the extra conal compartments viz., superior and inferior tend to push the globe not only forward but also in an opposite direction thus making the proptosis eccentric. Proptosis may be seen from infancy to very old age. Both sexes are involved. It may be unilateral or bilateral. Course of disease may be acute, sub-acute and chronic.

Most common symptom is protrusion of eyeball forwards. Other symptoms will be diminution of vision, watering, redness. Signs will be axial or eccentric proptosis, chemosis of conjunctiva, Vision will be effected by exposure keratopathy or Hyperopic shift by pressure of tumor on globe and Optic nerve compression by tumor itself or enlarged extra ocular muscles at the apex of the orbit.

Aim
To evaluate etiopathogenesis of cases of proptosis clinically and radiologically and confirm by histopathological examination after surgical intervention.

Materials and Methods
This is a prospective and interventional study carried out on 200 patients of proptosis attending Department of Oculoplasty and Orbital Disorders, of Sarojini Devi Eye Hospital, Hyderabad. The study was conducted from August 2011 to September 2013. All cases of unilateral and bilateral proptosis who had given informed consent and agreed to come for follow up were included in the study. Recurrent cases and the patient who were not sure of coming for follow up were excluded from the study.

A detailed clinical examination of orbit according to protocol and examination of nasal cavity, oral cavity, abdominal examination and CNS examination was carried out. Measurement of axial proptosis was done.
using Hertel’s Exophthalmometer. Eccentric proptosis was documented by horizontal and vertical displacement of globe. Clinical diagnosis was made.

Routine hematological investigations and surgical profile was done in all cases. B-scan ultrasound and CT scan of orbits axial and coronal view 2 mm cut was done in all cases. MRI of orbits and brain was done in cases who had intracranial extension of orbital tumor.

Cases which can be managed medically without further investigations like cases of TRO, NSIOS, Orbital cellulitis and Mycysticercosis were given required medical treatment.

The cases of Lymphomas, secondaries in orbit and sub peri-ostal abscess were subjected to biopsy or drainage of pus. Pus drained was sent for culture and sensitivity. Cases of lymphomas and secondaries were confirmed by HPE and IHC and referred to medical oncologist or Radiation oncologist for further management.

Cases who had tumor in intra conal or extra conal space were subjected to Orbitotomy under GA. Cases of Sinuorbital masses and intra cranial extension cases were operated along with ENT surgeon and Neuro surgeon. Mass excised was sent for HPE and IHC if needed.

**Results and Observations**

Out of 200 cases 108 (54%) were males and 92 (46%) were females. 176 (88%) cases had unilateral proptosis and 24 (12%) had bilateral proptosis. 112 (56%) cases had axial proptosis and 88 (44%) had eccentric proptosis. 148 cases (74%) had chronic onset, 40 cases (20%) had sub acute onset and 12 cases (6%) had acute onset of presentation.

44 cases had Thyroid related orbitopathy which was most common cause of Proptosis (22%) followed by Non specific orbital inflammatory syndromes 28 (14%), 36 had orbital cellulitis (18%) and 24 had pleomorphic adenoma of Lacrimal gland (12%). 16 had Dermoid cyst (8%), 12 had Non Hodgkin’s lymphoma (6%), 12 had Retinoblastoma with extra ocular extension (6%), 12 had cavernous hemangioma (6%), 4 had Rhabdomyosarcoma (2%), 4 had secondaries in orbit (2%), 6 had mycysticercosis (3%) 1 had fungal granuloma (0.5%) and 1 had carotico cavernous fistula (0.5%).

Microscopic examination of pus was done in 36 cases of orbital cellulitis, Staph. aureus was the most common pathogen.

**Discussion**

Out of 200 cases of proptosis in our study, majority were in the age group of 20-29 years that accounted for 30% of total cases. 16% each was from the age group of 0-9 years and 10-19 years. Male to female ratio in our study was 1.17:1 which correlated with the study of Khalid Farooq et al[4] where male to female ratio was 1.23:1. This in contrast to study by Rituraj et al[6] were the male to female ratio was 0.69:1. Unilateral proptosis was common and accounted for 88% of the total cases. Bilateral proptosis was seen in only 12% of cases, implying that systemic causes of proptosis are rarer than local causes. This correlated with study of Khalid Farooq et al[4] where unilateral cases were 88.5%. Among 200 cases of proptosis majority were axial proptosis accounting for 56% of cases and eccentric proptosis were about 44%. This correlated with study of Rao. K.P. et al[5] where axial proptosis were majority accounting for 68% of cases. Majority of cases in our study were of chronic onset which accounted to 74% of cases. Only 6% cases had an acute onset. The most common etiology was thyroid related orbitopathy which accounted to 22% of cases followed by pseudotumour attributing to 14%, Lacrimal gland tumours forming 12%, orbital cellulitis attributing to 18% and other causes contributing to 40% of cases. This correlated with study of Rituraj et al[6] where thyroid eye disease was the commonest etiology contributing to 33% followed by pseudo tumour forming 14% and Lacrimal gland tumours attributing to 12%.

Majority of cases were managed surgically which included orbitotomy and exenteration, surgical drainage of pus and Incision biopsy that accounted for 52% while 26% of cases were managed medically. Orbital exenteration was done in all the cases of retinoblastoma with extra ocular extension implying that the tumour is the most common intra-ocular cause of proptosis. These figures are comparable to study carried out by Asif et al[7].

**Conclusion**

The most common cause of proptosis in our study is thyroid related orbitopathy followed by pseudotumor (NSOIS), Lacrimal gland tumors and orbital cellulitis. Proptosis was the presenting complaint in all the cases which was associated with symptoms like visual deterioration, restricted extra ocular movements and diplopia. 60% of proptosis require interventions, the remaining 40% will be medically managed. This is in accordance to proptosis studies all over the world. CT scan is invaluable in evaluating a case of proptosis, but histopathological examination provides definitive diagnosis of the exact etiology.
Table 1: Showing incidence of various conditions causing Proptosis

<table>
<thead>
<tr>
<th>Condition</th>
<th>Cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thyroid related orbitopathy</td>
<td>44</td>
<td>22%</td>
</tr>
<tr>
<td>Lacrimal gland tumours</td>
<td>24</td>
<td>12%</td>
</tr>
<tr>
<td>NSOIS</td>
<td>28</td>
<td>14%</td>
</tr>
<tr>
<td>Orbital cellulitis</td>
<td>36</td>
<td>12%</td>
</tr>
<tr>
<td>Lymphomas</td>
<td>12</td>
<td>6%</td>
</tr>
<tr>
<td>Retinoblastoma with extraocular extension</td>
<td>12</td>
<td>6%</td>
</tr>
<tr>
<td>Dermoid cyst</td>
<td>16</td>
<td>8%</td>
</tr>
<tr>
<td>Carotid cavernous fistula</td>
<td>1</td>
<td>0.5%</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>12</td>
<td>6%</td>
</tr>
<tr>
<td>Embryonal Rhabdomyosarcoma</td>
<td>4</td>
<td>2%</td>
</tr>
<tr>
<td>Fungal granuloma</td>
<td>1</td>
<td>0.5%</td>
</tr>
<tr>
<td>Secondaries in orbit</td>
<td>4</td>
<td>2%</td>
</tr>
<tr>
<td>Myocysticercosis</td>
<td>6</td>
<td>2%</td>
</tr>
</tbody>
</table>

Fig. 1: Pie diagram showing male to female ratio

Fig. 2: A case of TRO

Fig. 3, 4: CT Scan Orbit Coronal & Axial view of same patient

Fig. 5: A case of Lacrimal gland tumour
Fig. 6: CT scan of same patient

Fig. 7a, b: A case of Orbital cellulitis before & after drainage of pus

Fig. 8: CT scan showing sub periosteal abscess involving roof of orbit

Fig. 9a

Fig. 9b
Fig. 9c

Fig. 9a, b, c: A case of NSOIS before & after treatment with steroid

Fig. 10: CT scan showing enlargement of medial rectus involving belly & tendon

References
6. Dr. Rituraja Baruah, Dr. Ashok Kumar Grover, Dr. Shalu Bageja. Proptosis: A Retrospective analysis of presentation, management and outcome.