A COMPREHENSIVE ANALYSIS OF AETIOLOGY AND CLINICAL PRESENTATION OF UNILATERAL PROPTOSIS PRESENTING TO TERTIARY EYE CARE CENTRE

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ABSTRACT

BACKGROUND

Proptosis is the hall mark of orbital diseases. Prompt and early diagnosis become essential as this socket small space in between orbital wall will lead to increased damage than one expects, and removal of masses will become difficult in later stages. It causes cosmetic disfigurement which becomes alarming to patients.

Aim- A comprehensive analysis of various aetiology and clinical profile of unilateral proptosis in tertiary eye care center.

MATERIALS AND METHODS

A hospital based retrospective case study of all the patients attending the orbit clinic of Department of Ophthalmology Each patient was thoroughly examined. After recording the detailed history including past history, general and ocular examination were carried out. All the findings were entered in a proforma for further analysis.

Inclusion Criteria- All cases of unilateral proptosis.

Exclusion Criteria- All cases of bilateral proptosis, enophthalmos, pseudoproptosis were excluded from the study.

RESULTS

Bilateral proptosis- 12 (6.41%), Unilateral proptosis- 60 (51.28%), the ratio of bilateral and unilateral proptosis is 1:5. Inflammation and neoplasms constitute more than 55% of the cause for unilateral proptosis in children under 15 years of age.

CONCLUSION

Unilateral proptosis was the presenting feature in the orbit clinic patients. Unilateral proptosis was eight times more common than bilateral proptosis. Defective vision was the commonest associated symptom. Inflammation was the commonest etiology found in adults, which leads to defective reversible defective vision.

KEYWORDS

Orbit, Unilateral Proptosis, Idiopathic Inflammatory Orbital Disorders.

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BACKGROUND

The orbit houses the eyeball and subserves the major functions of protection of the eye and facilitates extreme degree of motility possible to the eye. The orbit is pear shaped cavity, the stalk of which is the optic canal. The orbital diseases gain importance due to involvement of optic nerve[1] hence causing decreased vision and disturbance in ocular movements. The intraorbital portion of optic nerve is longer (25 mm) than the distance between, the back of the globe and optic canal (18 mm). This allows for significant forward displacement of the globe called proptosis.[2]

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Clinically a case of unilateral proptosis is challenging even for experienced ophthalmologist with special interest in orbital diseases. Since orbit is in close relation to paranasal sinus, maxilla and cranium, primary diseases of these areas may initially present as proptosis. Hence a detailed study of age of occurrence of disease, finding of examination, progression of disease, available incidence of such disease, investigation results become essential to diagnose a case of proptosis.

Proptosis is the hall mark of orbital diseases. Prompt and early diagnosis become essential as this socket small space in between orbital wall will lead to increased damage than one expects, and removal of masses will become difficult in later stages. It causes cosmetic disfigurement which becomes alarming to patients.

MATERIALS AND METHODS

A hospital based retrospective case study of all the patients attending the orbit clinic of Department of Ophthalmology. Each patient was thoroughly examined. After recording the detailed history including past history, general and ocular examination were carried out.

Examination of the proptosis was done to know whether it is axial or eccentric, reducibility, pulsatility, tenderness, thrill, bruit, presence of visible mass and change associated with cough impulse and Valsalva manoeuvre. Exophthalmometry was done for all cases. Investigations like total count, differential count, urine sugar, peripheral smear, VDRL & Mantoux test, were done in our department laboratory. X-rays, CT scan and were done in the radiology department T3, T4 and TSH estimation were done in the endocrinology department. Cases were followed up in the eye department. All the findings were entered in a proforma for further analysis.

Inclusion Criteria

All cases of unilateral proptosis

Exclusion Criteria

All cases of bilateral proptosis, enophthalmos, pseudo proptosis were excluded from the study.

RESULTS

During the period of study, the total number of patients with unilateral proptosis examined was sixty. The number of males was 29. The number of females was 31. The numbers of children (under 15 years) in this study were 28. Most of the children were below six years[3]. The numbers of adults were 32.

Incidence

No of cases referred to orbit clinic – 78

I. Bilateral proptosis - 12 (6.41%)
   - Unilateral proptosis - 60 (51.28%). The ratio of bilateral and unilateral proptosis is 1:5.

II. The side involved was
   - Right eye- 31
   - Left eye- 29

III. The type of proptosis encountered[4]
   - Axial- 28
   - Eccentric- 32

IV. Painful proptosis was seen in 25 patients

V. The onset of proptosis was sudden in 25 cases, Gradual in 30 cases, since birth in 5 cases.

Sex

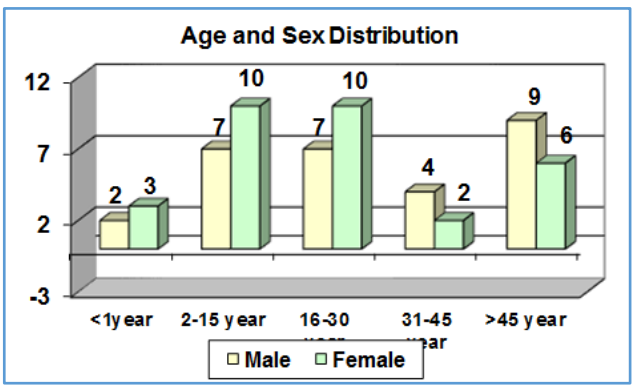
A condition with a 4:1 female prevalence[5] we still found 57% of the patients to be female. Female patients account for 57% of the subjects in Silva's orbital tumor survey. Whereas Henderson's review of 764 tumours that did not include Graves' disease lists a female:male ratio of 12:13.

Age (yrs.)	Percentage
0-10	8
11-20	7
21-30	10
31-40	11
41-50	16
51-60	19
61-70	18
71-80	9
81-90	2

Table 1. Distribution of Orbital Disease by Decade

No.	Age	Male	Female	Total
1.	≤ 1 year	2	3	**5**
2.	2-15 years	7	10	**17**
3.	16-30 years	7	10	**17**
4.	31-45 ears	4	2	**6**
5.	>45 years	9	6	**15**
Total		29	31	**60**

Table 2. Age and Sex Distribution

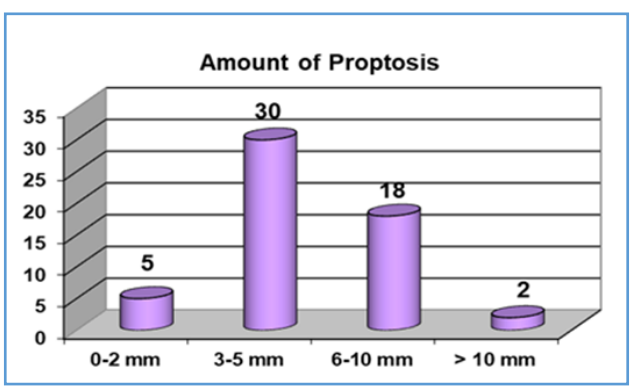


Since birth in 5 cases

The amount of proptosis encountered in this study is as follows:

No.	Proptosis	Number
1.	0-2 mm	5
2.	3-5 mm	30
3.	6-10 mm	18
4.	>10 mm	2
Total		**55**

Table 3. Amount of Proptosis



In 5 children proptosis could not be measured.

Diplopia was observed in 6 cases. (10%). It totally subsided in 4 patients. In one patient with osteomyelitis it was present till the end of this study. Fundus examination revealed leucocoria in 3 patients.

Primary optic atrophy was present in 7 patients. Two patients with carotid cavernous fistula presented with dilated, tortuous retinal veins which totally subsided following carotid ligation. Rest of the patients had a normal fundus. Of the 60 cases, 44 patients presented with primary orbital pathology while in the remaining 16 patients the orbit was involved secondary to a systemic cause.

Primary orbital involvement	44	72.5%
Secondary orbital involvement	16	27.5%

Table 4. Primary and Secondary Orbital Lesions – Incidence

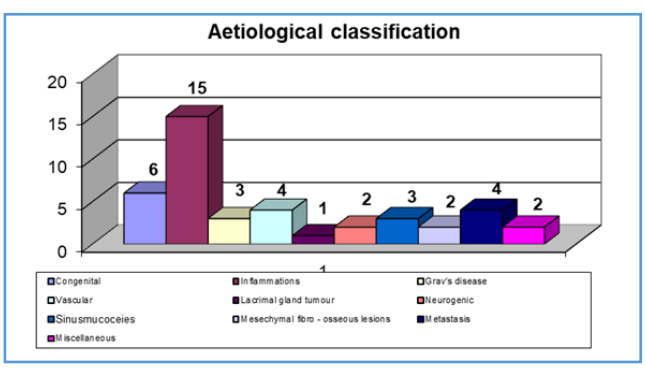
Lesion	Children	Adults	Total
Primary orbital involvement	19	25	44
Secondary orbital involvement	7	9	16

Table 5. Primary and Secondary Orbital Lesions Children and Adult Ratio

In 18 cases diagnosis was confirmed by histopathological examination. In 2 cases of carotid cavernous fistula diagnosis was confirmed by carotid angiography and Doppler studies. In the remaining 40 cases diagnosis was arrived at after therapeutic trial and observation. There was one case of pseudo proptosis due to hemi facial atrophy. Rare cases like one case of malignant medulloepithelioma arising from the optic disc and two cases of carotid cavernous fistula were also included in the study. In a 3 years old child with proptosis of left eye due to orbital secondaries from adrenal neuroblastoma the right eye presented with orbital deposits but without proptosis.

Aetiology	No. of cases	Percentage
Congenital	6	10.0%
Inflammatory	15	37.5%
Grave's disease	3	7.5%
Vascular	4	10.0%
Lacrimal gland tumour	11	2.5%
Neurogenic	2	5.0%
Sinus mucoceles	3	7.5%
Mesenchymal fibro osseous lesions	2	12.5%
Metastasis	4	10.0%
Miscellaneous	2	5.0%

Table 6. Aetiological Classification[6]



Inflammation and neoplasms constitute more than 55% of the cause for unilateral proptosis in children under 15 years of age.

DISCUSSION

The literature on unilateral proptosis is reviewed. Henderson's review of 764 tumours that did not include Graves' disease lists a female: male ratio of 12:13. As Root man's series and ours both included Grave's orbitopathy, these series may reflect a closer approximation to the incidence of the various orbital diseases encountered by most clinicians. Nonetheless, it is instructive to observe the complications of the other larger series that are principally based on tumours, the five-series complication done by Wilson and Grossniklaus is Henderson's orbital tumor series from the Mayo clinic and with Romans series from British Columbia inflammatory orbital disease, 1965 (31.0%) had systemic conditions involving the orbit, 1277 (20.1%) had neoplasm, 600 (9.4%) had congenital lesions, 308 (4.8%) had trauma, and 17 (0.2%) had vascular disease. Of the 2161 patients presenting with inflammatory disease, 1473 (68.1%) had idiopathic orbital inflammation, 270 (12.5%) had infection, 126 (5.8%) had dacryoadenitis, and 292 (13.5%) had other aetiologies. Among the 1965 patients presenting with systemic disease involving the orbit, 1938 (98.6%) were diagnosed with thyroid orbitopathy, 22 (1.1%) with amyloidosis, and 5 (0.2%) with sarcoidosis. Of the 1277 patients with neoplasm, the tumor was vascular in 369 (28.8%), neural in 336 (26.3%), lymphoid or leukemic in 131 (10.2%), secondary neoplasm in 82 (6.4%), epithelial in 68 (5.3%), adipose in 53 (4.1%), metastatic in 39 (3.0%), and fibrous, fibro-osseous, striated muscle, histiocytic, and other cellular origin in 40 (3.1%), 37 (2.8%), 23 (1.8%), 21 (1.6%), and 78 (6.1%), respectively. Of the 600 patients with congenital lesions, 427 (71.1%) had dermoid and 170 (28.3%) had dermolipoma, followed by meningoencephalocele.

	Cases	Percentage
Graves' disease	590	32
Pseudo tumours	107	6
Pseud proptosis	107	6
Vascular	101	6
Orbital cellulites	91	5
Undetermined	117	6
Meningioma (primary – secondary)	52	3
Non- Hodgkin's lymphoma	50	3
Metastatic	50	3
Dermoid – epidermoid	42	2
Sinus mucocele pyocele	33	2
Traumatic mass lesions	33	2
All others	452	24
Total	1825	100

Table 7. Common Orbital Disorders[7]

Benign and malignant lesions

70% of the orbital disease in our overall series was benign,[8] although one must use the term with cation was benign, although one must use the term with caution. For instance, benign mixed tumour of the lacrimal gland and sclerosing pseudotumour are benign histologically, but their clinical

behaviour can be quite destructive. 58% of the tumours in his series to be benign histologically, three of the orbital disorders in table are in inflammatory in nature. i. e. Grave's ophthalmopathy, idiopathic inflammatory pseudotumour, and orbital cellulitis. Jones and Jakobiec[9] lend support to our findings by stating, non-organismal inflammations and infections collectively cause well over 50 per cent of cases of orbital disease and proptosis. Two- thirds dural cavernous sinus fistula, aneurysm, intracranial arteriovenous malformations, cavernous sinus syndrome, and carotid cavernous sinus fistula all have been attended by pathologic orbital findings. vascular lesions 4.5% of the primary vascular neoplasm 6.3% arteriovenous malformations, orbital varices, and organizing haemorrhage accounted for the largest number of vascular lesions, whereas cavernous haemangioma and capillary haemangioma accounted for the largest number of primary vascular neoplasms, closely followed by lymphangioma decreasing order of frequency. Cavernous haemangiomas most prevalent haemangiomas in adults, capillary haemangioma was the most common childhood haemangioma. Globe asymmetry, cranial nerve paralysis, and congenital asymmetry to be the three most common causes of pseudo proptosis. Six most common categories of mass lesions neurogenic, lymph reticular, congenital cystic, vascular, inflammatory, and metastatic.

Fifty-eight primary 424 were secondary 68% of Illiff and grebe and Illiff and ossofsky reviews and 48 percent of Hou and Garg's series. Combined five series compilation tabulated by Wilson and Grossniklaus, 55% of the orbital mass lesions were primary and 45% were secondary lesions.

	Cases	Percentage
Neurogenic	97	18
Cysts	96	17
Vascular	68	12
Lymphoreticular	66	12
Inflammatory	60	11
Metastatic	50	9
Secondary epithelial	28	5
Lacrimal intrinsic Neoplasms	16	3
Osseous cartilage	10	2
Miscellaneous	24	4
Undetermined	35	7
Total	**549**	**100**

Table 8. Orbital Mass Lesions [1]

Cystic lesions mucocele 4.1% metastatic tumours 9% of the orbital tumours 4.5% Wilsom Grossniklaus the most common tumour to metastasize to the orbit in adults is breast carcinoma, followed in frequency by carcinoma of the lung. Carcinoma of the prostate is another common source of orbital metastases in adult men. Neuroblastoma[10] is the most common childhood entity to metastasize to the orbit.

Secondary neoplasms are those cancers that have invaded the orbit form adjacent structures, periorbital skin conjunctiva nasopharynx, and nasal cavity. Paranasal sinus neoplasms[2] accounted for 54% of the previously mentioned subdivision. 50% of Henderson's secondary epithelial neoplasms, whereas nasal sinus neoplasms accounted for 9% of his secondary epithelial neoplasms.

Secondary neoplasia accounted for 11.5% of the combined five series compilation tabulated by Wilson and Grossniklaus. A full 2.2% of the total series was secondary to orbital invasion of uveal melanoma.

Wilson and Grossniklaus inflammatory 37% and mass lesion 37% categories are the most common, followed by trauma 6% vascular (2%) bony development (2%), general diseases 1%, and pseudo proptosis.

CONCLUSION

The literature on unilateral proptosis is reviewed. Materials and methods employed are stated. Sixty cases were studied out of whom 19 were males and 21 were females. Unilateral proptosis was the presenting feature in 51.28% of all the orbit clinic patients. Unilateral proptosis was eight times more common than bilateral proptosis. Defective vision was the commonest associated symptom which was present in 82% of the patients with unilateral proptosis. Pain was associated with 40% of cases and Diplopia was present is only 10% of cases. Right eye was more commonly involved than the left eye. Eccentric proptosis was more common than axial proptosis. Irreversible optic nerve damage resulted in 12.5% patients. Inflammation was the commonest aetiology found in adults (50%). Vascular causes come next with 18%. Neoplasm was the commonest cause in children (33.3%). Inflammatory lesions come second with 22.2%.

REFERENCES

[1] Modi PJ, Shah NA, Bhalodia JN, et al. Orbital tumors in children: a descriptive study at tertiary care centre. National Journal of Medical Research 2013;3(4):362-366.

[2] Sultana A. Analysis of prevalence and etiology of proptosis at a tertiary care centre of south, India. India Ann Int Med Den Res 2017;3(2):7.

[3] Amudhavadivu S, Shanmugam UV. An aetio-epidemiological inquiry of proptosis. Int J Intg Med Sci 2016;3(10):439-442.

[4] Rao KP, Rao KV, Rao MR, et al. Clinical study of proptosis. Indian Journal of Ophthalmol 1982;30(5):489-490.

[5] Kennedy RE. An evalution of 820 orbital cases. Trans Am Ophthalmol Soc 1984;82:134-157.

[6] Henderson J. Orbital tumors. 3rd edn. New York, NY: Brian C. Decker 1994:43-51.

[7] Jakobiec FA, Jones IS. Orbital inflammations. In: Duane TD, ed. Clinical ophthalmology. 2nd edn. New York: Harper & Row 1980:1-75.

[8] Kim UR, Khazaei H, Stewart WB, et al. Spectrum of orbital disease in south India: an aravind study of 6328 consecutive patients. Ophthal Plast Reconstr Surg 2010;26(5):315-322.

[9] Wilson MW, Buggage RR, Grossniklaus HE. Orbital lesions in the south eastern United States. Orbit 1996;15(1):17-24.

[10] Tang W, Hei Y, Xiao L. Recurrent orbital space-occupying lesions: a clinicopathologic study of 253 cases. Chin J Cancer Res 2013;25(4):423-429.