A STUDY ON CHILDHOOD PROPTOSIS

Malarvizhi Raman[1], Anuradha[2], Keerthana[3]

[1]Professor, Department of Ophthalmology, Regional Institute of Ophthalmology, Egmore, Chennai.
[2]Assisatnt Professor, Department of Ophthalmology, Regional Institute of Ophthalmology, Egmore, Chennai.
[3]Junior Resident, Department of Ophthalmology, Regional Institute of Ophthalmology, Egmore, Chennai.

ABSTRACT

BACKGROUND

Proptosis in children usually presents as a diagnostic dilemma. This study is to evaluate the incidence, mode of onset, various clinical presentations in children up to 14 years. The various treatment modalities and the final outcome of treatment were also assessed.

MATERIALS AND METHODS

This prospective study was conducted at Orbit and Oculoplasty Services, RIOGOH, Egmore, Chennai for a period of 26 months from April 2014 to May 2016. Thirty patients presenting with proptosis were examined with detailed history taking, complete general examination and ocular examination, slit lamp biomicroscopy, ophthalmoscopy, Hertel's exophthalmometry, colour vision, refraction, intra ocular pressure, perimetry, laboratory investigations, radiological investigations and histopathological evaluation were done to aid in the etiological diagnosis and to plan the management.

RESULTS

Of the total 90 patients of proptosis who attended the hospital during the study period, 30 were children upto 14 years of age (33.33%). 18 cases (60%) of patients presented with axial proptosis and 12 cases (40%) were eccentric. 25 cases (83%) presented with unilateral proptosis and 5 cases (17%) with bilateral proptosis. The common aetiologies for proptosis in children were inflammatory (33%) followed by neoplastic (20%) commonly secondaries. Among the inflammatory causes orbital cellulitis was the most common cause (30%). Leukemic infiltration was the commonest cause of proptosis due to secondary orbital infiltration. Among all cases inflammatory lesions had better outcome. Out of 30 cases majority (40%) got cured and showed clinical improvement by medical and surgical methods of treatment.

CONCLUSION

This study reveals that in children inflammatory condition is the predominant cause. Most of the cases were unilateral and axial in presentation. Overall the most common aetiology of childhood proptosis is orbital cellulitis. Among the malignancies, secondaries were more common than the primary orbital tumours. Haematological malignancy accounts for the majority of cases of orbital secondaries in children. Imaging techniques are the best modality of investigation in all cases of proptosis to detect the early lesions within the orbit.

KEYWORDS

Childhood Proptosis, Axial Proptosis, Eccentric Proptosis, Exophthalmometer.

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BACKGROUND

The orbits are a pair of bony cavities in the skull situated on either side.

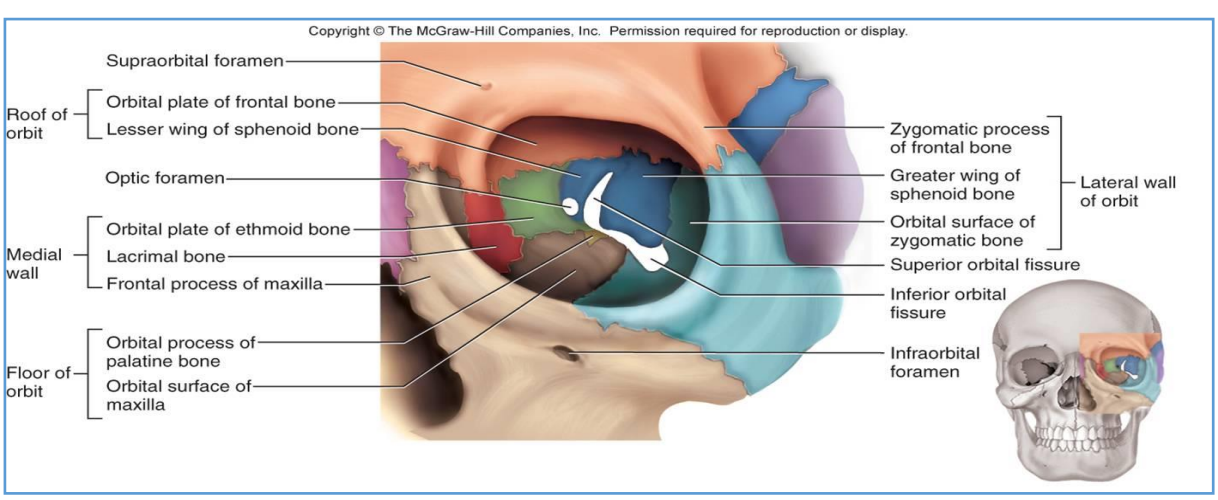

Figure 1. Anatomy of Orbit

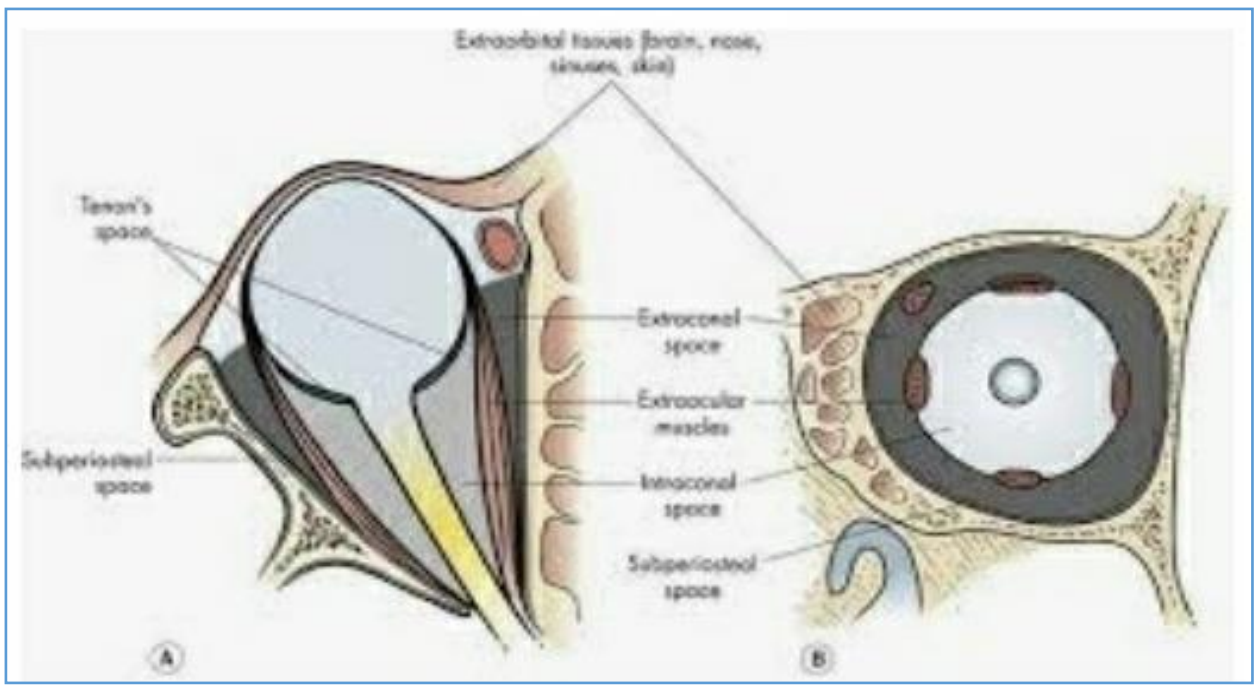

Figure 2. Surgical Spaces of Orbit

Classification of Proptosis

1. Proptosis may be acute.[1] or chronic.
2. It may be unilateral or bilateral.
3. It may be axial or eccentric.[2]
4. It may be classified based on the aetiology.

Causes of Childhood Proptosis

1. Congenital and developmental Anomalies

a. Craniofacial Dysostosis

Proptosis in this condition is the result of shallow orbit due to anterior displacement of greater wing of sphenoid.[3] e.g. Crouzon's (Figure 12) and Apert's syndrome.[4]

Figure 3. Pseudoproptosis in Crouzon's Syndrome

b. Encephalocele and Meningocoele

Due to congenital dehiscence in the bones the meninges herniate causing a cystic tumour with cerebrospinal fluid, it is a meningocoele. If there is brain tissue it is an encephalocele.

c. Teratoma

Arises from two or more germ layers, including ectoderm and endoderm or mesoderm or both.[5]

d. Dermoid Tumours

Most common cystic tumours occurring in the orbital and periorbital region and most common orbital neoplasm in the paediatric age group Result of sequestration of surface ectoderm at suture lines or lines of embryonic closure.

Fronto zygomatic suture is the most common site.

2. Inflammation

a. Orbital Cellulitis

This is one of the most common.[1] cause of proptosis in childhood.

Figure 4. Orbital Cellulitis

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Corresponding Author:
Dr. Malarvizhi Raman,
No. 9, Golden Castle Apartment,
24/16, J Block, 2nd Street,
Anna Nagar, Chennai.
E-mail: docmalar@gmail.com
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Figure 5. Orbital Cellulitis

b. Orbital Abscess

Orbital Abscess presents with acute proptosis, ophthalmoplegia, palpable fluctuant mass and papilledema.

Complications

- Infective /toxic neuropathy
- Cavernous sinus thrombosis
- Intracranial complications.[6]

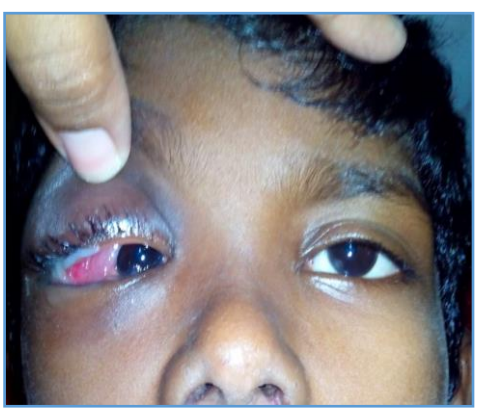

Figure 6. Orbital Abscess

c. Idiopathic Orbital Inflammatory Disease

Nonspecific inflammatory conditions of the orbit without identifiable local or systemic causes. Group of inflammatory disorders that produce a pseudo neoplastic orbital mass.[7] This causes pain, proptosis, chemosis and diplopia with visual loss.

d. Cysticercosis

Presents as cystic lesion in the conjunctiva, muscles or intraocular structures.

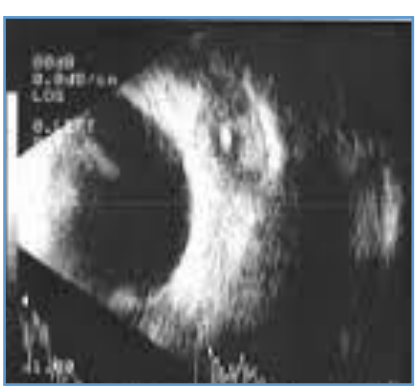

Figure 7. B scan-cystic Lesion with Scolex

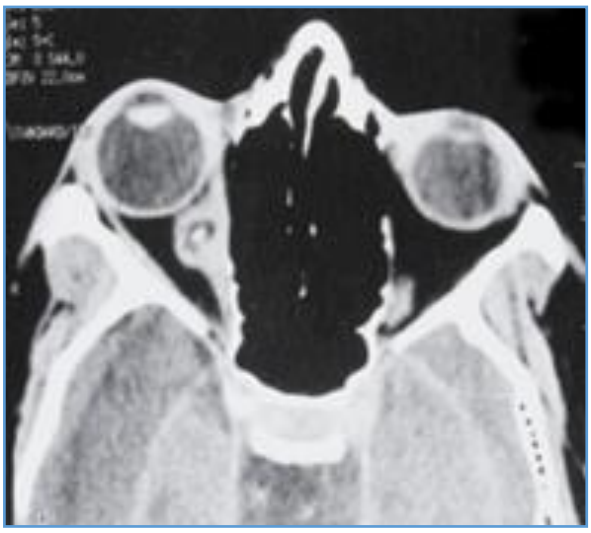

Figure 8. CT scan-cystic Lesion with Scolex

3. Vascular Lesions

a. Haemangioma

Capillary Haemangioma**-** It is the most common orbital lesion of infancy. It appears as an ill-defined, compressible bluish mass that has a predilection for the upper nasal quadrant of the orbit.[8]

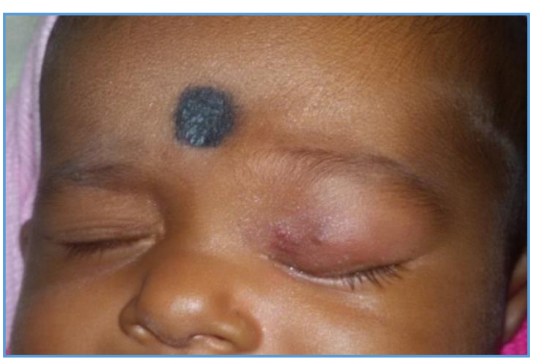

Figure 9. Capillary Haemangioma

Cavernous Haemangioma- The most common benign intraorbital tumor. Causing axial proptosis. Microscopically, the lesion is composed of dilated vascular channels.

b. Lymphangioma

It is a rare congenital tumour of the orbit, becoming classically apparent in the early years of childhood.[9] Spontaneous haemorrhage into the cyst leads to sudden increase in size of proptosis and formation of "chocolate cysts".

c. Orbital Varices

Orbital varices may be congenital weakness in the venous wall or acquired weakness because of AV shunts.[10] Onset usually during first five years of life.

d. A – V Malformations

AV malformations may be high-flow or low-flow lesions.[11] A high flow malformation demonstrates pulsating exophthalmos, bruit, marked orbital swelling clinically due to retrograde flow into the venous system.

Low flow lesions shows lesser degree of Proptosis, absent or minimal bruit, milder elevation of venous pressure leading to dilated episcleral, orbital, intraocular veins and raised intraocular pressure.

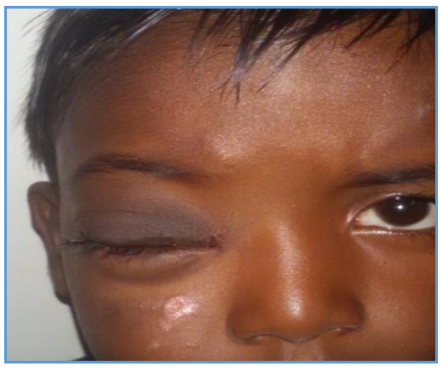

Figure 10. Vascular Malformation Showing Proptosis

Figure 11. Showing Vascular Malformation

4. Neurogenic Lesions

a. Glioma- Optic nerve gliomas are more common than meningiomas. The peak incidence is 2-6 years of age.[12]

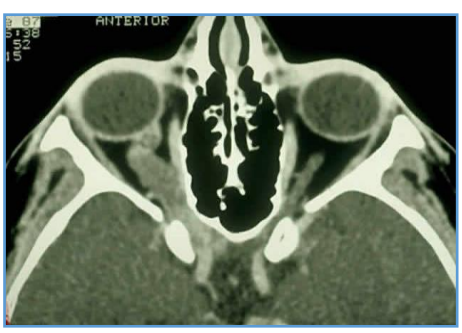

Figure 12. CT scan Optic Nerve Glioma

b. Meningioma- Meningioma although rare, bilateral cases may present much earlier approximately by 13 years of age.

c. Peripheral Nerve Sheath Tumours
Neurofibromatosis
Due to developmental absence of orbital bones posteriorly presenting as pulsatile proptosis.

5. Mesenchymal Tumours-
Striated Muscle Tumours
Rhabdomyosarcoma- Rhabdomyosarcoma is the most common primary malignant orbital tumour of childhood. Orbit the second most common site in the head and neck (the most common being the parameninges) accounting for about 10% of all rhabdomyosarcomas.

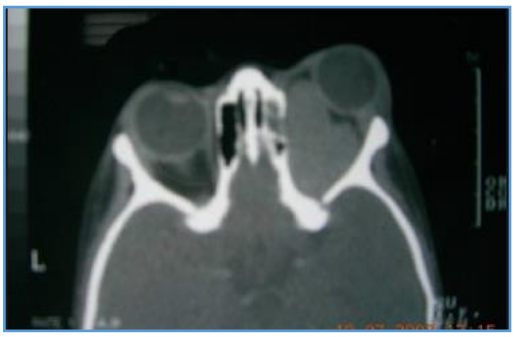

Figure 13. Rhabdomyosarcoma

6. Fibro osseous Lesion

a. Osteoma- Osteoma is a common benign tumour of the paranasal sinuses. It usually arises within the frontal sinus. They usually present in the fourth and fifth decades and occur equally in males and females.

b. Fibrous Dysplasia- It is a non-neoplastic disorder of the childhood that frequently involve the orbital bones. Most patients present with facial asymmetry, proptosis and globe displacement. Associated symptoms include diplopia, anosmia, hearing defects, nasal obstruction and epiphora. Increased intracranial pressure and cranial nerve palsies can also occur. Extensive disease results in deformed facies known as 'leontiasis ossea'. Progressive disease can result in optic nerve compression.

7. Secondary Tumours of the Orbit

a. Ocular:
Retinoblastoma- Retinoblastoma orbital extension may present with proptosis and most likely to occur at the site of scleral emissary veins.[13]

b. Orbital Metastasis- Metastasis to the orbit occurs secondary to haematological spread of a primary tumor. Of all the orbital cases 1.5 to 3.3 % are metastatic of which 7 % cases are bilateral. In the paediatric age group, the common primaries include neuroblastoma, Wilm's tumor and Ewing's sarcoma

Figure 14. B/L Severe Proptosis with Ecchymosis Due to Secondaries from Neuroblastoma

8. Endocrine

Thyroid Orbitopathy- Thyroid dysfunction is the most common cause of unilateral proptosis.[14] Hertel's exophthalmometer is most commonly used exophthalmometer to measure proptosis.[15]

Aim of the Study- To evaluate the various causes of Proptosis in children up to 14 years. To analyse the incidence, mode of onset, various clinical presentations in patients with childhood Proptosis. To assess the various treatment modalities and the final outcome of treatment of childhood Proptosis.

MATERIALS AND METHODS

This prospective study was conducted at Orbit and Oculoplasty Services, RIOGOH, Egmore, Chennai for a period of 26 months from April 2014 to May 2016. 30 Patient presenting to Orbit and Oculoplasty Services with proptosis were registered, evaluated and followed up during the study period. A detailed history of the patient, complete general examination and ocular examination, slit lamp biomicroscopy, ophthalmoscopy, Hertel's exophthalmometry, fields, colour vision, refraction, intra ocular pressure and examination of proptosis, laboratory investigations, radiological investigations and tissue examination were done to aid in the etiological diagnosis and to plan the management.

Inclusion Criteria

- Children up to 14 yrs. presenting with proptosis were enrolled.

Exclusion Criteria

- Children with severe debilitation were excluded.

RESULTS

Total number of proptosis cases reported in RIOGOH during the study period between from April 2014 to May 2016 were 90 cases, including both children and adults.

1. Distribution of Proptosis among 90 Cases

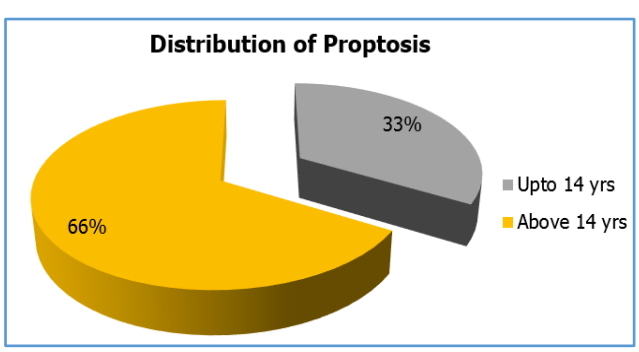

Chart 1. Pie chart Showing Distribution of Proptosis

The incidence of proptosis was highest in patients above the age group of 14 yrs. 60 cases (66%). Only 30 cases (33%) were under the age group of 14 yrs.

2. Age Distribution among Children with Proptosis

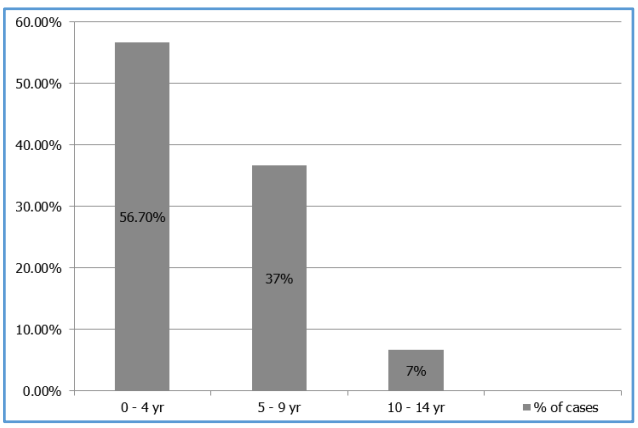

Chart 2. Histogram Showing Age Distribution among Children with Proptosis

The highest incidence was seen between the age group of 0-4 years 17 cases (56.7%) 11 cases (37%) were between 5-9 yrs. and 2 cases (7%) were 10-14 yrs.

3. Sex Distribution among Children with Proptosis

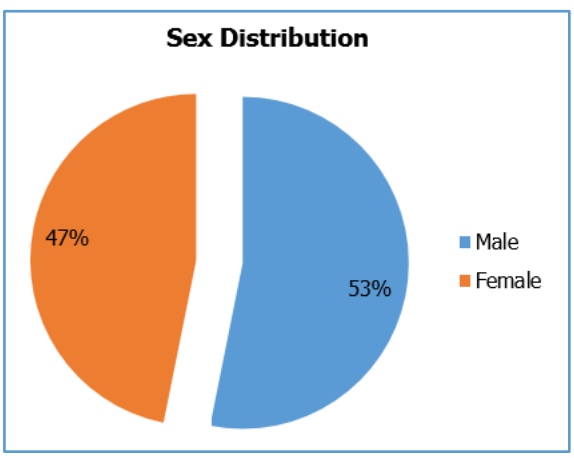

Chart 3. Pie Chart showing Sex Distribution among Children

Of the reported childhood proptosis in this study 16 cases (53.3%) were males and 14 cases (46.7%) were females.

4. Direction of Proptosis

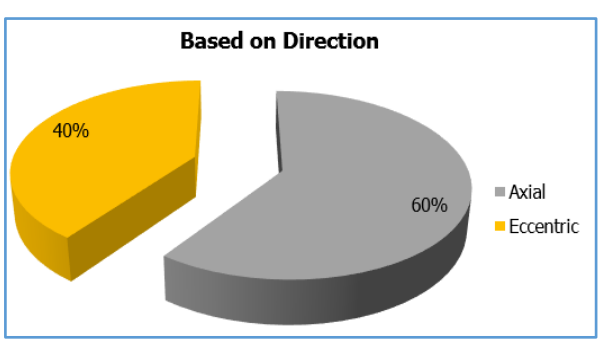

Chart 4. Pie Chart Showing Direction of Proptosis in Children

Axial proptosis was more common than the eccentric proptosis. Of the total 30 cases 18 cases (60%) were axial and 12 cases (40%) were eccentric proptosis

5. Laterality of Proptosis

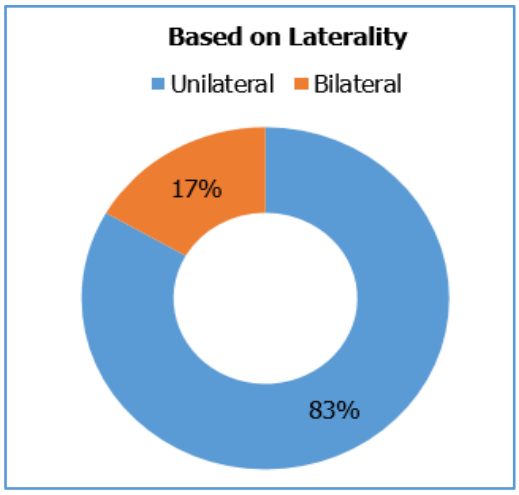

Chart 5. Doughnut Chart Showing Laterality of Proptosis in Children

Incidence of unilateral proptosis was more common 25 cases (83.3%) than the bilateral proptosis which is 5 cases (16.67%)

6. Onset of Proptosis

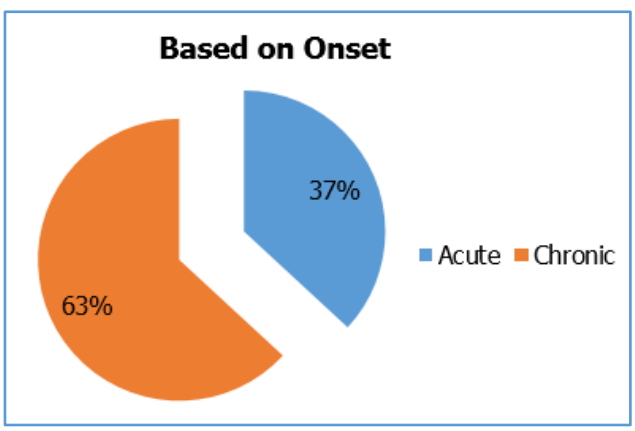

Chart 6. Pie Chart Showing Onset of Proptosis in Children

Acute onset 11 cases (37%) of proptosis is less common than the chronic cases 19 cases (63%).

7. Causes of Proptosis in our Study Population

Aetiology	No. of Cases	Percentage
Inflammatory	12	40.00 %
Congenital	9	30.00 %
Malignant	6	20.00 %
Grave's eye disease	2	06.67 %
Traumatic	1	03.33 %

Table 1. Various Causes of Proptosis in Our Study Population

Inflammatory causes ranks the first, 12 cases (40%) followed by congenital conditions 9 cases (30%) and neoplastic conditions 6 cases (20%). Among the inflammatory conditions orbital cellulitis 9 cases (30%) is the most common cause.

8. Proptosis due to Various Tumours

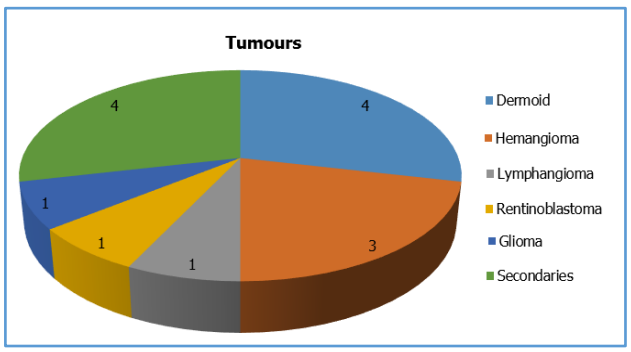

Chart 7. Pie chart showing Proptosis Due to Various Tumours

Among tumours benign tumours are more common than the malignant tumours.

Among benign tumours dermoid 4 cases (13.33%) is the most common cause followed by haemangioma 3 cases (10%). Among the malignant causes secondaries 4 cases (13.33%) are more common than the primary 2 cases (6.67%). Among the secondaries haematological malignancies 3 cases (10%) are common followed by neuroblastoma 1 case (3.3%).

9. The various Clinical Presentations of Proptosis

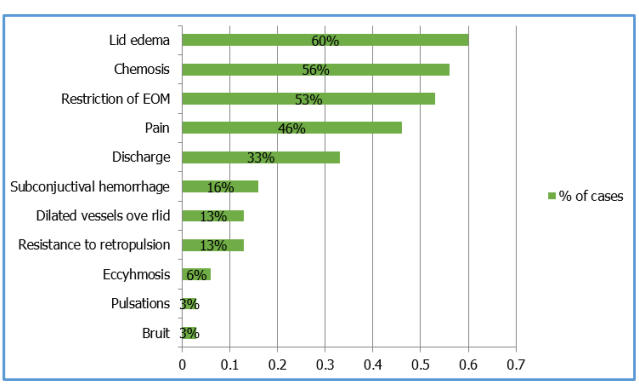

Chart 8. Various clinical presentations of Proptosis

Lid oedema 18 cases (60%) was the most common presentation of proptosis followed by chemosis in 17 cases (57%) and restriction of extra ocular movements were 16 cases (53%). Proptosis without pain were more common among total cases than the painful one. Among the painless conditions dermoid was more common followed by capillary haemangioma. Among the painful conditions orbital cellulitis was the most common cause followed by orbital secondaries due to haematological malignancies.

10. Analysis of Various Investigational Procedures

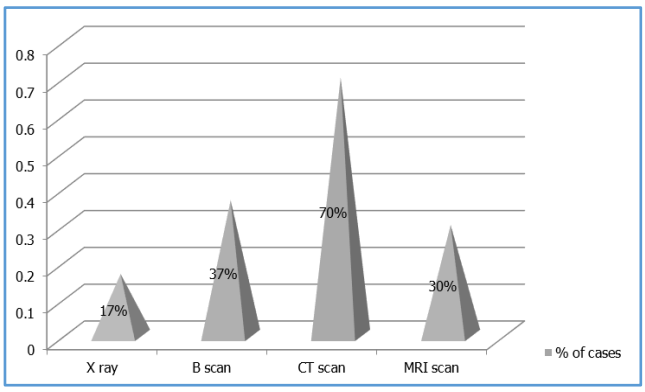

Chart 9. Various Radiological Investigations Done for Proptosis

X-ray- 5 cases (17%) underwent x ray orbit /PNS/skull.

- 3 cases of orbital cellulitis revealed sinus haziness.
- 1 case of dermoid revealed bony defect.
- 1 case of trauma revealed haziness in extraconal space suggestive of hematoma which was later confirmed with CT scan.
- Also skeletal survey in neuroblastoma patient revealed widening of skull sutures and multiple lytic lesions in the long bones and skull suggestive of secondaries.

USG (B Scan)- 11 cases (37%) underwent B scan

- 3 Patients with orbital cellulitis were subjected to USG and they showed low reflective echoes of the soft tissues, with mottled appearance of orbital fat "dirty fat sign".
- 2 cases of thyroid ophthalmopathy showed a low to medium reflectivity echoes and enlargement of extraocular muscles with tendon sparing.
- 3 cases of capillary Haemangioma showed a typical well demarcated cystic lesion with high reflectivity and strong transmission.
- 2 patients of cysticercosis showed well circumscribed cyst with high reflective echoes inside the cyst wall suggestive of scolex.
- In case of Retinoblastoma diffuse mass filling the globe with areas of high echo reflectivity suggestive of calcification was noted.
- The safety and relative low cost of ultrasound on comparison to CT Scan and MRI gives it a distinct and practical advantage.

CT Scan- 21 cases (70%) underwent CT –orbit

- Among 30 cases of childhood Proptosis taken for study, in 21 cases diagnosis was confirmed with CT scan.
- Of all the orbital cellulitis, 6 cases showed ethmoidal sinusitis, 2 cases showed sphenoidal sinusitis, 1 case showed both sphenoidal and ethmoidal sinusitis. All the cases of orbital cellulitis showed hyperdense lesions involving the soft tissues of the orbit.
- All 4 cases of dermoid showed non-enhancing cystic lesion with smooth margins. Bony defect was noted in 2 cases.
- Orbital abscess showed a ring lesion with wall enhancement with central hypodense region.
- Orbital hematoma revealed an isodense lesion laterally along lateral rectus muscle compressing the globe medially.
- Optic nerve glioma showed an isodense, fusiform mass lesion within the muscle cone arising from the optic nerve.
- CT scan of retinoblastoma showed calcification.
- Most of the secondary malignant orbital tumours showed hyperdense, diffuse lesions with some orbital enlargement and bony involvement.
- 128 slice CT angiogram was done in child with suspected AV malformations which revealed low flow vascular malformation.

MRI Scan- 9 cases (30%) underwent MRI orbit.

- In 2 cases of dermoid, MRI done showed hyperintensity consistent with fat was seen in both T1W and T2W sequences.
- In 3 cases of capillary haemangioma, hypointensity in T1W images and iso to hyperintensity in T2W images were seen.
- In case of lymphangioma variable intensity was noted in T1W sequence.
- In 2 cases of cysticercosis both T1W and T2W revealed isointense lesion with eccentric scolex.
- In case of neuroblastoma heterogenous soft tissue mass displacing the globe both in right and left eye was noted along with retroperitoneal hypointense mass from left adrenal gland consistent with primary neuroblastoma.

11. Analysis of the Various Treatment Modalities

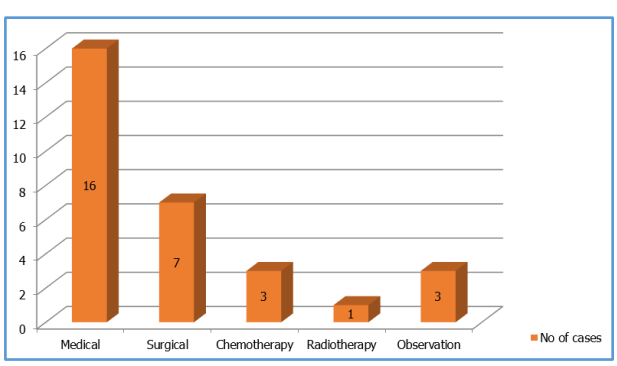

Chart 10. Analysis of the Various Treatment Modalities in Proptosis

16 cases (53%) were treated by medical management, 7 cases (23%) by surgical management, 3 cases (10%) by chemotherapy, 1 (3%) by radiotherapy, 3 (10%) by observation.

12. Analysis of Outcome of Various Types of Proptosis

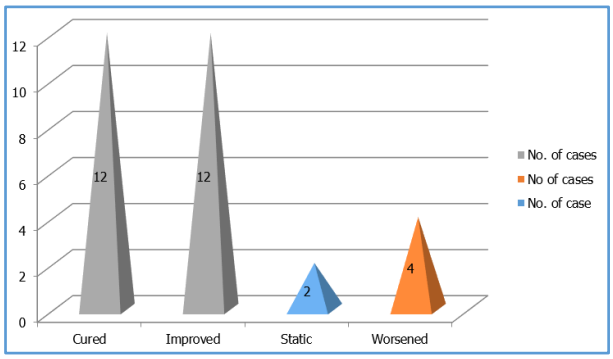

Chart 11. Analysis of Outcome of Various Types of Proptosis

12 cases (40%) cured, 12 cases (40%) improved, 2 cases (6.6 %) didn't improve while 4 cases (13.2%) worsened.

Out of the 9 cases of orbital cellulitis 7 cases got resolved with parenteral broad-spectrum antibiotics and 2 patients had improvement.

A case of orbital abscess underwent surgical drainage and with broad spectrum iv antibiotics. Out of the 2 cases of orbital cysticercosis 1 got completely cured and 1 child improved clinically with residual lesion in the imaging. All the dermoid, dermolipoma patients underwent surgical removal with good postoperative vision. Of the 3 cases of capillary haemangioma, 2 had marked reduction in size of proptosis after topical timolol gel forming solution and oral propranolol and one child had no change in the size of proptosis. 2 cases of Grave's eye disease was treated medically with antithyroid drugs. Both of them showed good improvement. 1 case of orbital hematoma improved after three weeks of steroid therapy. 1 case of optic nerve glioma was sent for neurosurgical intervention. 1 case of neuroblastoma was referred for chemotherapy and radiotherapy. Among 3 cases of haematological malignancies which were treated with chemotherapy 1 improved and 2 worsened.

DISCUSSION

The findings of the analysis are as follows-

Of the total 90 cases of proptosis attended the hospital during the study period, 30 were children upto 14 years of age (33.33%). 18 cases (60%) of patients presented with axial proptosis and 12 cases (40%) were eccentric. 25 cases (83%) presented with unilateral proptosis and 5 cases (17%) with bilateral proptosis. Of the 30 cases, 11 (37%) were acute in onset and 19 (63%) were chronic. M Loganathan et al[1] in 2014 conducted a study on childhood Proptosis among 50 cases which revealed orbital cellulitis was the most common cause of childhood Proptosis which is similar to our study. This study also showed that axial Proptosis were more common than eccentric. Proptosis among malignancies, secondaries were more common than primary. These results were comparable to our study. In our study, the common aetiologies for proptosis in children were inflammatory (33%) followed by neoplastic (20%) esp. secondaries. Among the inflammatory causes orbital cellulitis was the most common cause (30%) and all cases of inflammatory proptosis were acute in onset. According to the study conducted by Bakshi et al[16] in 2008 among 104 cases of malignant childhood Proptosis secondary tumours (59.6%) were more common than primary tumours (51%) which is similar to my study. In our study, leukemic infiltration was the commonest cause of proptosis due to secondary orbital infiltration. A study done by Thakre Snehalr.[17] et al in 2016 revealed malignant ocular and extraocular tumours being the most common cause of childhood proptosis. But in OUR study inflammatory lesions were the most common cause of childhood Proptosis. In a study done by Belmekki M.[18] et al in 1999 among 54 cases of Morocan children with proptosis retinoblastoma was found to be the leading cause of proptosis whereas in OUR study inflammatory lesions was found to be the leading cause of proptosis.

In a study conducted by Ganessan K et al[19] among 45 children with proptosis, 80% were unilateral and 17% were bilateral which is similar to my study. This study also shows that AML was the most common cause of bilateral proptosis and this supported our data wherein haematological malignancies leads the cause of bilateral proptosis. Plain X ray and CT scan were useful tool in the diagnosis of proptosis due to bony lesions. Ultrasonography was very useful in diagnosing soft tissue and cystic lesions. CT scan is the essential investigation in proptosis. It helps in locating the intracranial extension of the orbital lesions. It is also very helpful to detect early soft tissue and bony lesions which cannot be picked up by the USG. Most of the soft tissue inflammatory lesions were managed by medical treatment. The mass lesions (tumours and secondaries orbit) causing the proptosis has to be tackled by combined modalities viz. surgery, chemotherapy and or radiotherapy in association with various specialities like neurosurgery, ENT, oncologist.

Among all cases inflammatory lesions had better outcome. Out of 30 cases majority got cured and showed clinical improvement by medical and surgical methods of treatment.

CONCLUSION

The suspicion of orbital disease in a child produces anxiety and concern in the patient, the family, and the treating physician. Although proptosis is uncommon complaint in children, its presence usually reflects a serious threat and needs urgent intervention. Risk of malignancies should be considered especially in subacute and chronic cases. Thorough examination of an orbital problem in children is a challenge. But Complete general and regional examination with special importance to abdominal examination is mandatory as they may give clue to the diagnosis.

This study reveals the aetiology of proptosis in children up to 14 years of age which is definitely different from that in adults. In adults, thyroid disease is the most common cause wherein children inflammatory (orbital cellulitis) is the predominant cause. There was not much gender difference

in the incidence of childhood proptosis. Most of the cases were axial and unilateral.

Among the malignant neoplasms, secondaries were more common than the primary orbital tumours and haematological malignancy accounts for majority of cases of secondaries in the orbit in children. X-ray and USG are initial investigation in diagnosing the proptosis. CT and MRI scans are the better modality of investigation in all cases of proptosis to detect the early lesions within the orbit and to identify the extent of lesion in planning further management.

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