Etiopathogenesis of childhood proptosis

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Abstract

Aim: To study the etiology, presentation and management of proptosis in children less than 14 years of age

Materials and Methods: This is prospective interventional study conducted at the department of Oculoplasty and Orbital diseases of Sarojini Devi eye hospital, Hyderabad. Period of study was from August 2012 to July 2014. 120 eyes of 111 children presenting as proptosis to the department were included in the study. Adult cases of Proptosis were excluded from the study. All cases underwent a detailed proptosis evaluation by an experienced oculoplasty surgeon. CT scan of orbit, axial and coronal view, 2mm cut without contrast was modality of imaging in all cases. CT scan with contrast was done in selected cases. Complete blood picture and surgical profile investigations were done. Cases were posted for excision or incision biopsy under general anaesthesia. Histopathological diagnosis was obtained in required cases. Culture and sensitivity of pus was done in infective cases.

Results: 111(14.83%) cases of childhood proptosis were there out of 748 cases of proptosis presented during the study period to the department. The Male to Female ratio was 55:46. 102 cases were of unilateral proptosis and 9 were bilateral. Common etiology for childhood proptosis was 28 congenital, 18 infective, 20 parasitic, 36 neoplasm, 6 secondaries and 3 trauma cases. Of 111 cases 28 had acute proptosis, 25 had Sub acute presentation and 58 cases had chronic presentation.

Management of 111 cases was done as follows: 48 cases underwent surgery, 41 were managed medically, 10 cases were referred to Medical Oncologist and 12 cases of congenital crouzons disease and microphthalmos were not intervened.

Conclusion: Childhood proptosis forms 14.83% of total proptosis cases presenting at department of Oculoplasty and Orbital diseases. Etiological spectrum is different when compared to adult Proptosis.(1)

Keywords: Congenital, Infective, Neoplasm, Parasitic, Proptosis.

Introduction

There is marked variation in the incidence of various causes of childhood proptosis. Neoplastic (33-53%), congenital (34%), infection & inflammation (5-46%) in various studies conducted worldwide.(2,3) But one thing is common in all that etiology of childhood proptosis pattern is different from adult Proptosis.

Aim

To study the etiology, presentation and management of Proptosis in children less than 14 years of age.

Materials and Methods

This is prospective interventional study conducted at the Department of Oculoplasty and Orbital diseases of Sarojini Devi Eye Hospital. The study period was from August 2012 to July 2014. 120 eyes of 111 children presenting as Proptosis to the department were included in the study. Adult cases of Proptosis were excluded from the study. All cases underwent detail proptosis evaluation by senior ophthalmologist using Slit lamp, Direct and indirect Ophthalmoscope, Hertel's Exophthalmometer, Retinoscopy. Clinical diagnosis was arrived at and CT scan orbit, Axial and Coronal view 2 mm cuts without contrast was ordered in all cases. CT Scan with contrast was performed in selected cases. Complete blood picture[4], surgical profile investigations was done in all cases. Cases were posted for excision or incision biopsy under general anaesthesia. Histopathological diagnosis was obtained by an experienced pathologist. Immunohistochemistry was performed in case of lymphoma and rhabdomyosarcoma.

Results

Out of 748 cases of Proptosis 111 (14.83%) childhood proptosis were recorded during the study period. Male to female ratio was 55:46. 102 cases were unilateral and 9 were bilateral.

Congenital cause formed 34.22% (28 cases of 34 eyes).

Table 1

S. No	Etiology	No. of cases	No. of eyes
1.	Crouzon's disease	6	12
2.	Microphthalmos with cystic eye ball	6	6
3.	Dermoid Cyst	16	16
Total		28	34

Infective and inflammatory causes formed 16.21% (18 cases in 20 eyes).

Table 2

S. No	Etiology	No. of cases	No. of eyes
1.	Orbital cellulitis	13	15
2.	Pyogenic granuloma	3	3
3.	Tuberculous granuloma	1	1
4.	Dacryoadenitis	1	1
	Total	18	20

Primary vascular Neoplasia formed 13.51% (15 cases in 16 eyes).

Table 3

S. No	Etiology	No. of cases	No. of eyes
1.	Lymphangioma	7	7
2.	Cavernous hemangioma	4	4
3.	Capillary hemangioma	2	3
4.	Hemangiopericytoma	1	1
5.	Angiolipoma	1	1
	Total	15	16

Primary Neoplasia of other tissue formed 18.91% (21 cases in 21 eyes).

Table 4

S. No	Etiology	No. of cases	No. of eyes
1.	Rhabdomyosarcoma	3	3
2.	Myxoma	1	1
3.	Optic nerve glioma	1	1
4.	Lymphomas	1	1
5.	Retinoblastoma	11	11
6.	Squamous cell carcinoma	4	4
	Total	21	21

Miscellaneous etiology.

Table 5

S. No	Etiology	No. of cases	No. of eyes
1.	Secondaries in Orbit	6	6
2.	Myocysticercosis	20	20
3.	Trauma	3	3

28 cases had acute presentation of proptosis. They were 13 cases of Orbital cellulitis, 3 cases of Trauma, 3 myocysticercosis and 4 cases of lymphangioma.

25 cases had subacute presentation of Proptosis. They were 17 cases of myocysticercosis, 3 cases of lymphangioma, 3 cases of pyogenic granuloma, 1 case of tuberculous granuloma and 1 case of Dacryoadenitis.

58 cases had chronic presentation of proptosis. They were 28 cases of Congenital disorders, 11 cases of Retinoblastoma, 4 cases of Squamous cell carcinoma of conjunctiva, 4 cases of Cavernous hemangioma, 3 cases of Rhabdomyosarcoma, 2 cases of capillary hemangioma, 1 case of hemangiopericytoma, angiolipoma, myxoma, Optic nerve glioma, Lymphoma each.

Management of cases was done as follows: 48 cases underwent surgery, 41 were managed medically, 10 cases were referred to Medical Oncologist and 12 cases of congenital Crouzans disease and microphthalmos were not intervened.

Medically managed cases were, orbital cellulitis, trauma and myocysticercosis. Orbital cellulitis and trauma cases were giving broad spectrum antibiotics topically and systemically with systemic steroids. Myocysticercosis cases were given Albendazole 15-25 mg/kg body weight with Prednisolone 1mg/kg of body weight orally.

In surgical management 16 cases underwent anterior orbitotomy, 5 lateral orbitotomy, 12 enucleation, 3 exenteration, 8 excision biopsy, 4 cases had incision and drainage of Orbital Abscess.

The diagnosis of cases which underwent anterior orbitotomy was dermoid cyst of Orbit on medial side, lymphangioma, rhabdomyosarcoma, myxoma, cavernous hemangioma, hemangiopericytoma and angiolipoma. Rhabdomyosarcoma had incision biopsy to establish diagnosis all other cases had total excision of tumour.

The cases which underwent lateral orbitotomy were intra conal cavernous hemangiomas, optic nerve glioma, lymphangioma and an orbital dermoid.

Enucleation was done in cases of Retinoblastoma, Squamous cell carcinoma of conjunctiva involving sclera. Exenteration was done in cases of Retinoblastoma involving orbit by perforating sclera and Conjunctival Squamous cell carcinoma involving sclera and extra orbital muscles.

Incision biopsy was done in cases of Lymphoma, Tubercular granuloma and Pyogenic granuloma.

Incision and drainage was done in cases of Orbital abscess.

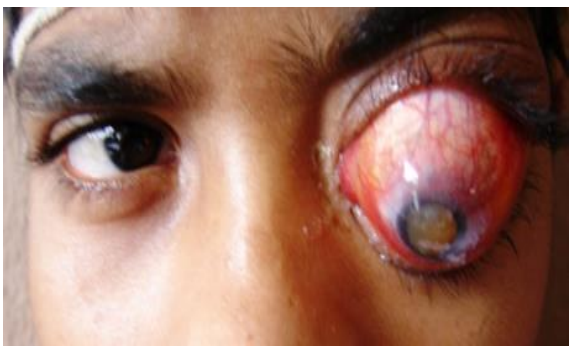

Fig. 1: Retinoblastoma presenting as left eye Proptosis

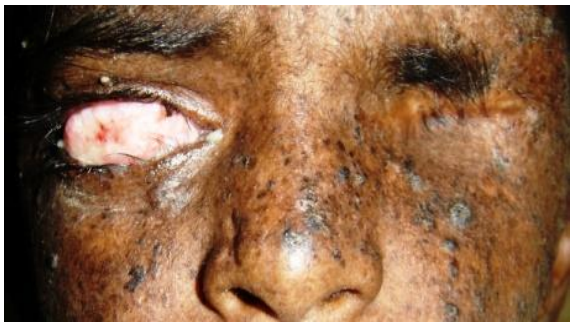

Fig. 2: Xeroderma pigmentosa left eye Exentration was done, Right eye developing Squamous cell carcinoma

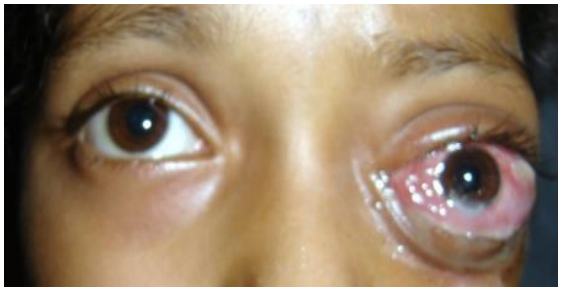

Fig. 3: Sub Periosteal Abscess presenting as left eye Proptosis

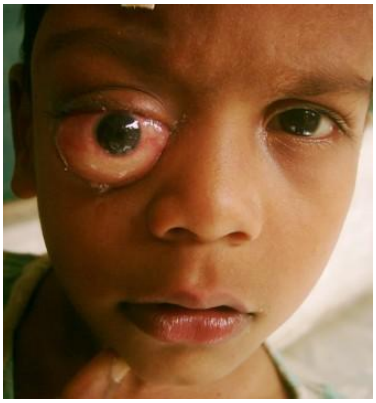

Fig. 4: Right Eye showing Salmon patch (Lymphoma)

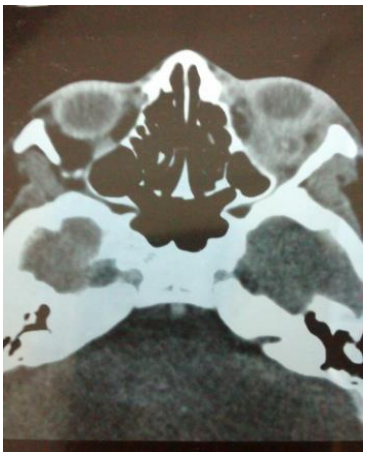

Fig. 5: CT scan orbit axial view showing Scolex in inferior rectus

Discussion

Congenital causes of Proptosis 28 cases lead the list in our study. Percentage of cases matches with Rootman J series. Myocysticercosis with 20 cases is next common cause. Inflammation and infection is third cause in our study 18 cases which matches with Otulana T ET al[5] where the inflammation is second leading cause in his series. Vascular tumours with 15 cases match with Bajaj et al study. Retinoblastoma with 11 cases is the most common childhood malignancy causing proptosis. This matches with Bakshi et al[6] study of 104 cases of childhood malignancy wherein Retinoblastoma forms 51% in their study. Secondaries in orbit with 6 cases is second malignancy to cause proptosis among children. This again matches with the Bakshi et al,[6] in whose series of 104 cases of malignant causes of childhood proptosis secondaries in orbit is second cause for proptosis. The common primary tumour causing secondaries in our series is Retinoblastoma followed by Neuroblastoma and Lymphoma.

The CT Scan Orbit, Axial and Coronal view without contrast is the most preferred imaging modality in the cases of Proptosis.[7] Final diagnosis is done by histopathological examination of the excised tumour.[7]

Following table shows comparison of our study with the study of Bajaj et al and Rootman J.

Table 6

Etiology	Our study	Bajaj et al	Rootman J et al
Congenital	28/111(25.22%)	8/119(6.72%)	70/405(17.28%)
Myocysticercosis	20/111(18.01%)	---	1/405(0.24%)
Infections & Inflammations	18/111(16.21%)	9/119(7.56%)	10/405(2.46%)
Vascular tumours	15/111(13.51%)	18/119(15.21%)	35/405(8.64%)
Retinoblastoma	11/111 (9.91%)		
Secondaries tumours	6/111(5.41%)	21/119(17.64%)	3/405(0.74%)
Trauma	3/111 (2.70%)	---	83/405(20.49%)
Rhabdomyosarcoma	3/111 (2.70%)	29/119 (24.36%)	15/405 (3.70%)
Optic nerve glioma	1/111 (0.90%)	9/119 (7.56%)	27/405 (6.66%)
Lymphoma	1/111 (0.90%)	----	11/405 (2.71%)

Conclusion

Parasitic disease like Myocysticercosis and infection and inflammation is the leading cause of Proptosis in India and other developing countries apart from congenital causes. In benign tumours dermoid cyst followed by Vascular tumours are most common. Retinoblastoma is leading childhood malignancy to cause Proptosis. Secondaries in Orbit are next common malignant cause of proptosis. We conclude that the difference between our study and Rootman J is because of geographical location. Western countries are more developed and have fewer incidences of infections and parasitic infestations.

All cases of Proptosis do not need surgical intervention. 40% of the cases can be treated by medical management.[7] Management of proptosis needs multi-disciplinary approach. The role of Medical oncologist, Pathologist, Microbiologist and Radiologist is crucial in management of cases.[7]

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