

Management of Vascular Malformation of Orbit in a Tertiary Eye Hospital

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Abstract

Background: Orbit, having a volume of 30cc, contains neurosensory, vascular, motor and secretory structures; is surrounded by facial structures. Visual impairment and restricted ocular motility may be caused by diseases of these structures. Inflammation, neoplasia, structural abnormalities, vascular lesion, and degeneration and deposition can occur within or around the orbit. **Objective:** To evaluate the outcome of the management of vascular lesions of orbit and ocular adnexa attending at a tertiary eye hospital. **Method:** A prospective interventional study was conducted in a tertiary eye hospital, during the period of July, 2008 to June, 2010. A total of 60 patients with proptosis/lid growth irrespective of age and sex were selected for this study according to selection criteria. Method of medical and surgical treatment were recorded and postoperative details examination including visual acuity, amount of proptosis, cosmetic outcome had been done. Patients were followed after first day, first week, six weeks and six months of postoperative period. **Result:** In vascular lesion of the orbit, 53.33 % patients were cavernous haemangioma, 26.67 % patients were capillary haemangioma, 8.33% patients were orbital varices, 6.67 % patients were lymphangioma and 5 % were haemangiopericytoma. Only 10 (16.67%) cases managed by medical treatment and bulk of the patients about 50 (83.33%) patients surgically managed. 10 (62.5%) patients of capillary haemangioma were managed by medically. Among the surgical complications stitch granuloma was higher (10) followed by recurrence (6). There were no complications occurred in 30 (50%) cases. **Conclusion:** Cavernous haemangioma was higher among vascular lesions of orbit and adnexa treated surgically.

Keywords: Orbit, Cavernous haemangioma, Capillary haemangioma, Lymphangioma, Haemangiopericytoma, Granuloma.

Introduction: The orbits are bilateral structures in the upper half of the face below the anterior cranial fossa and anterior to the middle cranial fossa that contain the eyeball, the optic nerve, the extra-ocular muscles, the lacrimal apparatus, adipose tissue, and the nerve and vessels that supply these structures.¹ Orbit is commonly affected with inflammation, neoplasia, structural abnormalities, vascular lesion and

degeneration and deposition. Among these, vascular lesion is the fourth most common orbital process which is 4.6%.² Among the vascular lesions capillary hemangioma, cavernous hemangioma, lymphangioma, carotid-cavernous fistula and varices are common; that can be classified by malformations, shunts and newgrowths. Malformations include capillary hemangioma, cavernous hemangioma,

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lymphangioma, varices, shunts include carotid-cavernous fistula and new growths include hemangiopericytoma, angiosarcoma etc. Capillary hemangioma is the most common tumour of the orbit and adnexa in childhood. It usually appears in first or second week after birth and grows up to first year of life then starts to regress. About 70% regress by 7 years of age. Superficial cutaneous lesion presents with bright red color and deep orbital tumour presents with unilateral proptosis but no skin discoloration. Hemangioma causing amblyopia, anisometropia, optic nerve compression and exposure keratopathy need treatment. There are different methods of treatments which includes laser therapy, intralesional steroid injection, systemic steroids, subcutaneous injection of interferon alpha-2b and local resection with cutting cautery. Most common benign orbital tumour in adult is cavernous hemangioma. Surgical excision of it is easier due to well encapsulated.³ Lymphangiomas are abortive vascular malformations which occur in early childhood which usually increase in size with upper respiratory tract infection. Superficial lymphangiomas of lid and conjunctiva, multiple cysts like structures are excised easily. But deep lymphangiomas has venous connection and is friable with non-capsulated. So surgical excision, of it is difficult. It excises sub totally by carbon dioxide and contact no: YAG laser. Other method of treatment is by sclerosing agents.⁴ Orbital varices are weakened, dilated segments of orbital venous system. Usually presents from childhood to middle ages. Which causes non-pulsatile proptosis, increase with straining. It is treated usually conservatively but surgical intervention is done when optic nerve compression present. Complete surgical removal is usually not possible due to friability and not capsulated.⁴ Capillary hemangiomas are the most common orbital tumors found in children. Lined by vascular endothelium and pericytes, these histologic benign lesions manifest at birth or within the first 3 months of life, enlarge rapidly, and begin to commence contracting around age 1 year. Other benign orbital lesions include dermoids, lymphangiomas, and histiocytic tumors. In adults, cavernous hemangiomas are the most common de novo orbital tumefaction. CT scan reveals a round, encapsulated, well-defined orbital lesion. Histologically, large blood-filled, endothelial-lined spaces with fibrous interstitial tissue and smooth muscle are discerned. These lesions usually are well tolerated by the patient and managed by conservative

therapy and reassurance, unless visual acuity or field loss is found.⁵ Carotid-cavernous fistula, abnormal communication between carotid artery and cavernous sinus. Surgery is needed if spontaneous closer secondary to thrombosis of the cavernous sinus does not occur. Interventional radiology involves detachable balloon occlusion of the fistula. Treatment is indicated with secondary-glaucoma, diplopia, intolerable bruit, severe proptosis causing exposure keratopathy.³ Hemangiopericytoma is a uncommon vascular tumour of orbit which occurs in middle ages. It causes proptosis, conjunctival prolapse and restricted ocular motility. It is encapsulated. It must be completely excised because they may recur and undergo malignant degeneration.⁶ MRI and CT-scan is the choice of investigation but CT scan is the gold standard imaging technique in the orbital pathology. There is soft tissue mass located superonasally. In MRI there is T1 image isointense with respect to skeletal muscle, hypointense with respect to orbital fat. In T2 image the lesion is hyper intense to EOM and orbital fat. Also biopsy done in suspected cases, it may be incisional or excisional.⁷

Method:

A prospective interventional study was conducted in department of oculoplasty, National Institute of Ophthalmology & Hospital, Dhaka, Bangladesh; during the period of July, 2008 to June, 2010. A total of 60 patients with proptosis / lid growth irrespective of age and sex were selected for this study according to selection criteria. Informed written consent was taken from the patients prior to surgical techniques. All the patients were evaluated by complete history and meticulous preoperative assessment, which included best correct visual acuity, extra-ocular motility, measurement of proptosis, if any palpable orbital mass with their extension, surface, tenderness, consistency, papillary reactions, intraocular pressure measurement and fundus examination were noted. Method of medical and surgical treatment were recorded and postoperative details examination including visual acuity, amount of proptosis, cosmetic outcome had been done. Patients were followed after first day, first week, six weeks and six months of postoperative period. Pre & postoperative photographs and histopathological report was collected and assessed. Any postoperative complications such as ptosis, strabismus, enophthalmos, recurrence, visual loss, skin change, conjunctival change, optic neuropathy and

lagophthalmos were noted. The main objective of this study was to evaluate the outcome of management of different vascular lesions of orbit and adnexa attending at tertiary eye hospital. Collected data were compiled and analyzed statistically. Computer based statistical software SPSS (version 11.5) was used for analysis. P value < 0.05 was considered as significant and < 0.001 was considered as highly significant. Statistical analysis was done by t-test and Chi-square test.

Result:

Among the 60 patients, 32 (53.33%) patients were cavernous haemangioma, 16 (26.67%) patients were capillary haemangioma, 5 (8.33%) patients were orbital varices, 4 (6.67%) patients were lymphangioma and 3 (5%) were haemangiopericytoma. In this study, there were 39 (65%) cases with orbital vascular lesion, 12 (20%) adnexal vascular lesions and 9 (15%) cases combined orbit and adnexal vascular lesions. The patients were distributed into three age groups; such as 1-20 years, 21-50 years and >50 years. Thirty five (35%) patients were evaluated in 1-20 years age group. 56.67% patients were evaluated in 21-50 years age group and only 8.33% cases were evaluated in >50 years age group. Hundred percent (100%) capillary haemangioma cases were in 1-20 years age group and 90.63% cavernous haemangioma cases were 21-50 years age group. Sixty (60%) orbital varices were involved in 21-50 years age group and 20% were involved in 1-20 years and >50 years age groups respectively. Seventy five (75%) lymphangioma were in 1-20 years age group and 25% were in 21-50 years age group. 66.67% haemangiopericytoma were in >50 years age group. Among sex distribution of this study, it was observed that male patients were 27 (45%) cases and female were 33 (55%) in this study subjects. Male female distribution of patients was described in table 3. Found female predilection for cavernous and capillary haemangioma. The mean age, standard deviation, standard error, median age was described in table 4.

It was observed that right orbit and or adnexa were 34 (56.67%) cases and left orbit and or adnexa were 26 (43.33%). Right orbit and or adnexa were involved in 17 (53.13%) cavernous haemangioma cases, 9 (56.5%) capillary haemangioma cases, 3 (60%) orbital varices cases, 3 (75%) lymphangioma cases and 2 (66.67%) haemangiopericytoma cases. In this study, 23 (38.33%) cases were found skin change, 21

(35%) cases were found conjunctival change, 7 (11.67%) cases were found vision change, motility restricted were found in 4 (6.67%) patients, proptosis were found in 40 (76.67%) cases and retinal change were found in 4 (6.67%) cases of this study patients. The skin changes were found in 16 capillary haemangioma, 4 orbital varices and 3 haemangiopericytoma patients. Conjunctival change were found in 3 cavernous haemangioma, 10 capillary haemangioma, 4 lymphangioma, 2 orbital varices and 2 haemangiopericytoma cases. Visual effects change in 4 cavernous haemangioma and 3 capillary haemangioma cases. Fifty percent motility restricted was found in lymphangioma cases and 25% motility restricted were found in cavernous haemangioma and capillary haemangioma cases respectively. Proptosis were found in 32 cases of cavernous haemangioma, 4 cases of capillary haemangioma, 5 cases of orbital varices, 2 cases of lymphangioma and 3 cases of haemangiopericytoma. Retinal changes were found in only 4 cases of cavernous haemangioma. Among 60 patients, only 10 (16.67%) cases managed by medical treatment and bulk of the patients about 50 (83.33%) patients surgically managed. Ten (62.5%) patients of capillary haemangioma were managed by medically. In the 16 cases of capillary haemangioma, 10 cases were managed by observation, topical steroid and intralesional steroid. Among them 30% cases were static, 40% cases regress in size, 30% cases were resolved and only 10% cases were recurred. On observation, 66.67 percent patients were static lesion and 33.33 percent patients were regress in size. On topical low dose steroid therapy for 3 months, 33.33 percent were equally static, regress in size and resolution. Single dose of intralesional triamcinolone therapy causes 30% static condition, 40% regress in size and 30% resolution and 10% recurred. The surgical outcome of vascular lesion of the orbit and adnexa were evaluated in this study. Fifty percent patients were undergone surgery. We found skin change in 4% percent cases, conjunctival change in 4% percent cases, vision change in 2% cases, motility restricted in 6% cases, proptosis in 6% cases and retinal change in 2% cases after 6 weeks of surgery where as skin change in 26 cases, conjunctival change in 30% cases, vision change in 14% cases, motility restricted in 8% cases, proptosis in 92% percent cases and retinal change in 8% cases before surgery.

Table -I: Distribution of vascular lesion of the orbit and adnexa.

Type	Number	Percentage (%)
Cavernous Haemangioma	32	53.33
Capillary Haemangioma	16	26.67
Orbital varices	05	8.33
Lymphangioma	04	6.67
Haemangiopericytoma	03	5.00

Most of the (53.33%) vascular lesion of the orbit was cavernous haemangioma.

Table-II: Distribution of management of orbit and adnexal vascular lesion.

Type	Medical management	Surgical management
Cavernous Haemangioma	0	32 (100%)
Capillary Haemangioma	10 (62.5%)	6 (37.5%)
Orbital varices	0	5 (100%)
Lymphangioma	0	4 (100%)
Haemangiopericytoma	0	3 (100%)
Total = 60 cases	10 (16.67%)	50 (83.33%)

Table 2 shows - Most of the patients 50 (83.33%) surgically managed; only 10 (16.67%) cases managed by medical treatment. 62.5% (10) of capillary haemangioma patients were managed medically.

Table-III: Distribution of sex in the study subjects.

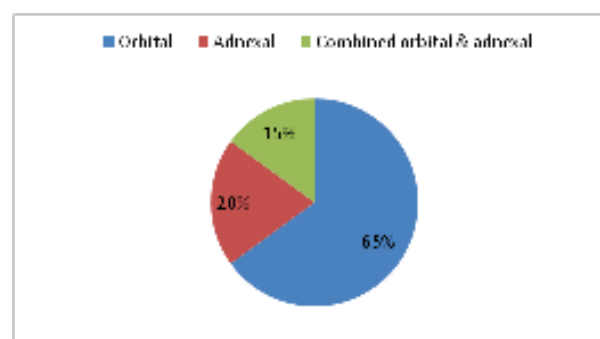
Type	Male	Female	χ^2 df P value
Cavernous Haemangioma	14 (46.67%)	18 (53.33%)	
Capillary Haemangioma	6 (37.5%)	10 (62.5%)	3.989 4 0.40751166
Orbital varices	3 (60%)	2 (40%)	
Lymphangioma	2 (50%)	2 (50%)	
Haemangiopericytoma	2 (66.67%)	1 (33.33%)	
Total = 60 cases	27 (45%)	33 (55%)	

Chi-square (χ^2) test
P value > 0.1
ns = not significant

Table 3 shows - Cavernous haemangioma female 53.33% (18) and capillary haemangioma female were 62.5% (10).

Table-IV: Distribution of Mean age, SD, SE, 95% CI and Median value of the patients.

Parameters	Cavernous haemangioma	Capillary Haemangioma	Orbital varices	Lymphangioma	Haemangiopericytoma
Mean age	41.53	5.74	33.60	13.50	50.67
SD	9.38	5.508	14.47	6.19	12.66
SE	1.66	1.377	6.47	3.10	7.31
95 % CI	36.15 to 42.91	2.809 to 8.679	15.64 to 51.56	3.65 to 23.35	19.21 to 82.12
Minimum	16 Y	0.4 Y	14 Y	08 Y	37 Y
Median	42 Y	04 Y	36 Y	12Y	53 Y
Maximum	59 Y	17 Y	51 Y	22 Y	62 Y



Graph 1: Distribution of the area of the vascular lesion.

Discussion:

The global distribution of vascular lesion of the orbit and or adnexa varies from Western hemisphere to Asian countries. The meticulous surgical outcome of orbital vascular lesion is excellent. The results of this study provide evidence in favor of a relatively common of cavernous haemangioma among orbital space occupying lesion 32 cases out of 60 cases which is 53.33%. In western countries, the rate of cavernous haemangioma is very high, among orbital vascular lesion in adult. In this study 90.63% cavernous haemangioma cases were 21-50 years age group; mean age was 41.53 years. Bilaniuk LT shown most of cavernous haemangioma occurs in female (60% - 70%) and mean age (43 - 48 years).⁸ In Orbital haemangioma, female predominance 70%, presentation at 4th to 5th decades of life and shows axial proptosis.³ Cavernous haemangioma was 9% of all orbital lesions. Cavernous haemangioma are the most common benign neoplasm of orbits in adults. Middle aged women are most commonly affected. Treatment consists of surgical excision if the lesion compromises visual function. These tumors rarely undergo spontaneously. In the largest survey reported involving 66 cases, women were more commonly affected than men.⁹ A painless, gradually progressive proptosis and visual disturbance were the main

clinical signs as Bilaniuk LT.⁸ The left orbit was affected in 123 cases and the right orbit in 86 cases. Ninety-five percent of them could be procedure of standard lateral orbitotomy in 36% of patients and anterior orbitotomy in 64% cases. Surgical complications were occurred in 24 (40%) patients. Among them, ptosis occurred in 2 (3.33%) cases, exotropia occurred in 4 (6.67%) cases; enophthalmos occurred in 1 (1.67%) case, recurrence occurred in 6 (10%) cases, optic neuropathy occurred in 1 (1.67%) case and stitch granuloma in 10 (16.67%) cases. There were no complications occurred in 30 (60%) cases. Our study was near to similar a study done by Wu ZY et al.¹⁰ They observed diagnosis and surgical management of 209 cases of orbital cavernous hemangioma and evaluated that 123 patients were females and 86 patients were males. Three patients had postoperative exotropia, and 1 enophthalmos. Two patient presented with recurrence few years after complete excision. All patients with adnexal lesion had satisfactory outcome. About 62% patients of capillary haemangioma were managed by medically. Vu BL et al. described the treatment options for capillary haemangioma was observation and topical, oral, or intralesional corticosteroid therapy. Interferon therapy, laser therapy and surgery typically reserved for potentially life-threatening complications.¹¹

Limitations:

The limitations of this study were relatively small sample size, single centre approached and lack of technical support. The rate of growth of the lesion and grade of proptosis were not estimated in this study. Carotid-cavernous fistula was not evaluated in this study. This study does not reflect the actual picture of vascular lesions of orbit and ocular adnexa.

Recommendation:

It can be recommended that the rate of cavernous haemangioma is higher among orbital vascular lesions in Bangladesh. Surgical outcome of the orbital and or adnexal vascular lesion is excellent if surgery is done meticulously. A long term study with a representative large sample will provide more specific results. Surgical management of orbit and adnexal vascular lesions still unsatisfactory in difficult cases. But proper evaluation and appropriate management, use of modern techniques and trained personnel are prerequisite to overcome difficulties.

Author's Contributions:

All the authors were contributed in various parts of

the publication from concept and design, acquisition of data, analysis & interpretation of data and drafting of the manuscript.

Declaration of Conflicts:

The authors declare that, there is no conflict of interest regarding the publication of this article.

Conclusion:

Analytical result of this study found that cavernous haemangioma is higher among vascular lesions of orbit and adnexa. Among 60 patients, only 10 cases managed by medical treatment.

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