

## Spectrum of Asymmetrical Proptosis

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### ABSTRACT:

**Objectives:** To discuss different aspects of unilateral proptosis in our institutions.

### Material and Methods:

**Study Design:** Observational study.

**Place and Duration:** Department of Neurosurgery/Head and Neck, PGMI, Govt Lady Reading Hospital, Peshawar. Pakistan from May 2004 – April 2008. All patients presented to outpatient clinic of Neurosurgery and ENT departments along with referred cases to these both departments were included in this study. These patients were analyzed in detail. Apart from local ENT examination and neurological examination, ophthalmological and systemic examination was also carried out. Relevant investigations were carried out to see the effects on anatomy as well as on physiology of globe. Patients with bilateral proptosis, relative proptosis due to crowzen's syndrome, Apart's syndrome and post traumatic proptosis were excluded. All these patients were managed surgically either individually or as team case by combined approach in the department. The data was analyzed using SPSS.

**Results:** Thirty eight (38) patients with asymmetrical proptosis in this study were analyzed with regard to their sex, age, site of proptosis, origin and extent, surgical approach adapted, per operative and histological diagnosis. There were 22 male and 16 female patients with male and female ratio of 1.4 and 1. The age range was from 8-65 years with median age of 36.5 years. X-ray skull, PNS and CT brain including orbits and fronto nasal sinuses were done in all cases and MRI in 18 cases. Right sided proptosis was seen in 28 and left side in 10 cases. There were 11 cases of pediatric age group and 27 young and adult groups. A variety of different disorders were observed responsible for unilateral proptosis. These disorders were Orbitonasal encephalocele, Angiofibroma, Metastatic deposits, Hydatid cyst, Aneurismal bone cyst (ABC), Lymphoma, Meningioma, Fibrous dysplasia, post traumatic growing skull fracture and Plexiform neurofibroma. Surgical procedures adopted during surgery were lateral, medial orbitotomy, lateral rhinotomy, skull base sub frontal craniotomy and extradural transcranial orbitotomy.

**Conclusion:** Variety of causes is responsible for unilateral proptosis which present in clinical practice to both ENT and Neuro surgeons. Team work helps a lot in making proper protocol.

**Key words:** Asymmetrical proptosis, lateral orbitotomy, skull base tumor, orbital tumor.

### INTRODUCTION

Patients with non endocrine mediated protrusion of eyeball are labeled as proptosis. The protrusion of globe is based on increase in volume within the fixed bony orbital confines. Orbit is widest at its anterior aspect and conical shape at posterior aspect, thus orbital contents are displaced easily anteriorly in proptosis.

Variety of causes is responsible for these structural anomalies of globe which is diagnosed and graded after clinical and imaging examinations. Based on type and degree of proptosis due different pathological causes, asymmetrical proptosis is managed by a variety treatment options.

Neglected cases of proptosis can compromise visual functions and integrity of the eye leading to

significant morbidity and mortality. Thus a standard management protocol should be followed for better outcome of these patients.

## MATERIAL AND METHODS

A total number of 38 cases of asymmetrical proptosis who presented to neurosurgery and ENT departments of lady reading hospital Peshawar were included in this study. The duration of study was from May 2005 to April 2009. Patients with bilateral proptosis, proptosis in different syndromes like crowzen's and apart were excluded from this study. All the patients in this study underwent detailed local, neurological, ophthalmological and systemic examination. Radiological examination in the form of x-ray, CT scan and MRI were done for proper preoperative diagnosis. This also helped us in evaluation of the extent of spread of these lesions. Proper documentation was made about these patients. Surgical approach was discussed a day before. These lesions were approached by different surgical lateral and medial orbitotomy and transcranial sub frontal approaches and lateral rhinotomy approaches. Patients and their relatives were informed regarding the objective of operation. All these patients were operated under general anesthesia, preoperative findings were documented during surgery and tissue was sent for histopathological studies. Post operatively adjunct therapy and oncologist help was taken, where needed.

## RESULTS

**Table 1:** Sex incidence (n = 38).

Sex	No of Patients	% age	Ratio
Male	22	57.9%	1.4
Female	16	42.1%	1

### Age Incidence

Age of the patients ranged from 8 to 65 years, with the median age of 36.5 years. Of these 11 patients were of pediatric age group and 27 adults.

**Table 2:** Side of proptosis (n = 38).

Side	No of Patients	% age
Right	28	73.7%
Left	10	26.3%

**Table 3:** Clinical features

Clinical Feature	No of Patients	% age
Headache	32	88.9%
Vomiting	22	61.11%
Nasal obstruction	04	11.11%
Proptosis	38	100%

**Table 4:** Imaging done.

Image Name	No of Patients	% age
X-ray (skull / PNS)	38	100%
CT (Brain with orbit and frontonasal sinuses)	38	100%
MRI (Brain with orbit and frontonasal sinuses)	18	47.4%

**Table 5:** Causes of unilateral proptosis.

Cause	No of Patients	% age
Orbitonasal encephalocele	03	07.89%
Angiofibroma	03	07.89%
Metastatic deposits	05	13.16%
Hydatid cyst	02	05.26%
Aneurismal bone cyst (ABC)	01	02.63%
Lymphoma	03	07.89%
Meningioma	05	13.16%
Fibrous dysplasia	04	10.53%
Post traumatic growing skull fracture	01	02.63%
Plexiform neurofibroma	01	02.63%
Hemangioma	03	07.89%
Fungal infection	05	13.16%
Dermoid	02	05.26%
Rhabdomyosarcoma	01	02.63%

**Table 6:** *Surgical procedure performed.*

Procedure Name	No of Patients	% age
Lateral, orbitotomy,	13	36.11%
Medial orbitotomy,	06	16.66%
Skull base sub frontal craniotomy	11	30.55%
Extradural transcranial orbitotomy	08	22.22%

**DISCUSSION**

Eye is considered as an index of beauty in human civilization and thus its natural architecture can sometimes be distorted by orbital and paraorbital tumors of the structures around orbit which also affect and change the natural look of orbital contents.<sup>14</sup> The causes of proptosis are numerous. It may be due to neoplastic, vascular, traumatic, metabolic or inflammatory in aetiology. The neoplastic causes can be further subdivided into two groups : Primary orbital tumors and paraorbital tumor or PNS tumor involving the orbit. Majority of primary orbital tumors originate between the bony orbital wall and extraocular muscle cone and 90% of primary orbital tumors present with proptosis.<sup>15</sup> Apart from lesion of orbit itself, sino-nasal and intracranial lesions also manifest through orbital cavity. Rarely intracranial, sometimes paraorbital and often primary orbital tumor causes proptosis. The causes of proptosis can be therefore orbital cellulitis, orbital pseudotumors, cystic lesion in the orbit and tumor of the orbit. Intracranial tumor or skull base tumors from the paranasal sinuses extending or eroding orbital wall, like angiofibroma, meningioma, capillary haemangioma, fungal infection, metastatic deposits, post traumatic haematoma and arterio-venous fistula also manifest as proptosis.<sup>1,2,3,10</sup>

Out of 38 patients managed surgically there were 22 male and 16 female with male female ratio of 1.4:1. There ages ranged from 8 – 65 years with mean age of 36.5 years. *Abdul – Haq* et al has reported the male to female ratio of 1.5 : 1 with average age of 27 years from sindh province. Their patients presented mainly with nasal symptoms in the form of obstruction and epistaxis but the common symptom in our study was proptosis and headache with nasal obstruction in 4 cases only. Nasal obstruction and epistaxis was also observed by other studies as a result of paraorbital lesions.<sup>9</sup> The reason is probably that their study was

purely of ENT cases without neurosurgical participation, while our study was in collaboration with ENT colleagues that is why neurosurgical cases were picked more. Margalit and his colleagues studied 41 patients who undergone surgery for orbital tumors. The main presenting complaints in their patients were also proptosis and headache as we had in our study.<sup>4</sup>

*Somnath* et al have reported 31 case series of proptosis during 4 years time with maximum operative patients in the age group 40 – 49 years. They studied their cases with different aspects. They reported 17 cases due to paraorbital tumor (55%) and one case due to primary orbital tumors. we noted 13 cases out of 38 patients as primary orbital while in the rest 25 cases extension from paraorbital area.

Majority of our cases were having intra cranial extension like fungal infection, skull base carcinoma, ABC of frontal sinus and naso – orbital areas. We observed 11 (30.55%) paediatric patients with orbital masses. Children presented with proptosis secondary to Orbitonasal encephalic lymphoma, dermoid and rhabdomyosarcoma of rectus. Orbital Hydatid was observed as rare cause of secondary Hydatid cyst. Two children had this problem. Both the children belonged to villages near the afghan border. They had other multiple Hydatid cysts in Liver and Peritoneal cavity. S. saha has documented 22.6% in his cases of proptosis in paediatric age group.

Capillary haemangiomas are the tumors of the childhood causing proptosis,<sup>10-12</sup> we had three patients with haemangioma as cause of proptosis and these were in the paediatric age group.

Among young and adult, skull base tumor and fungal infection made large group of these cases. We observed 5 patients with intracranial fungal infection with extension in to orbit and malignant skull base tumor invading orbital cavity in 4 cases. *Majekodunmi* Studied 75 Nigerians and observed that there were 12% (5 cases) with primary lesions and 83% (34) with secondary lesions as cause of proptosis. He had 29% of patients with malignant cause and 52% cases with cellulitis as cause of unilateral proptosis.<sup>5</sup> Plexiform angiofibroma also causes facio – orbital deformity and proptosis. We had one patient with proptosis due to plexiform angiofibroma.<sup>8</sup>

Radical excision was made possible for all benign lesions and biopsy was taken for some malignant tumors. Lateral orbitotomy was performed in 13 cases, transcranial approach for 11 cases, medial orbitotomy approach in 6 cases and transcranial orbital extradural approach in 8 cases. Nasopharyngeal angiofibroma

extending toward orbit and cranial cavity were excised through combined transcranial sub frontal with rhinotomy approach and skull base sub frontal craniotomy approach was adopted in 5 cases. Transcranial<sup>5</sup> and sub frontal approaches were also adopted in other studies for orbital lesions.<sup>12,13</sup>

There was no immediate mortality in our series except one patient died on 3rd postoperative day due to severe per rectal bleeding probably for perforated duodenal ulcer. He was on steroids for long period and surgical stress was further provocative factor for his seriousness.

## CONCLUSION

As orbit is the cross road of many compartments, so disease of the orbit presenting with proptosis should be managed by joint approach. Unilateral proptosis needs in time diagnosis and a proper protocol management. The common causes may be benign in majority of cases in younger age group but highly malignant cases like skull base carcinoma, metastatic deposits and primary malignant tumor of the orbit made a good number of cases. Combined ENT and Neurosurgical work help further in diagnosis and making a good surgical plain during management.

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