A Study of Tumours Giving Rise to Unilateral Proptosis

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Abstract Study of distribution, clinical features, radiological features, histopathological aspects, management and outcomes of tumours giving rise to unilateral proptosis. This is a prospective study including 40 patients with benign and malignant tumours of orbital, paraorbital and other sites giving rise to unilateral proptosis. Final diagnosis was made following detailed radiological and histopathological examination. Then, treatment modality was decided, which included surgery, radiotherapy, chemotherapy or required combination. Proptosis was measured by simple/plastic ruler exophthalmometry method. The maximum number of cases 8(20%) were in age group 31–40 years. The youngest patient was 4 year old and the oldest was 80 years. M:F ratio was 1.85:1. Malignant tumours were 32(80%) and benign tumours were 8(20%). 12 cases were having 3 mm proptosis (minimum) and one case having 18 mm (maximum). Only 4(10%) orbital tumours were responsible for unilateral proptosis. Paraorbital tumours 34(85%) was the major cause for unilateral proptosis and 2(5%) were from distant sites. Surgery was the mainstay of treatment 14(35%), followed by combined (surgery + radiotherapy) 5(12.5%), chemotherapy 4(10%). There was 4(10%) mortality, 16(40%) improved 3(7.5%) had same status and 17(42.5%) deteriorated. Thus nasal obstruction is the commonest ENT presenting complains in these patients. The major cause of unilateral proptosis was

found to be paraorbital tumours (34%). Thus, a case of proptosis should never be ignored but a thorough ENT and ophthalmic examination is necessary. As malignancy is one of the major cause of unilateral proptosis catching and treating them early will reduce mortality.

Keywords Unilateral proptosis · Paraorbital malignancy · Orbital tumours · Histopathology · Management

Introduction

Proptosis is defined as a forward movement of the globe in relation to the skull and generally measured by the protrusion of the apex of the cornea in front of the outer orbital margin, while the eye-look straight ahead [1]. Eye serves as a major cross road for the central nervous system, nose, nasopharynx, paranasal sinuses and all the structures related to support and functioning of the globe. The proximity and intimacy of these relations will lead to extension of tumours into orbit and ultimately to proptosis. Various ENT diseases may manifest as proptosis and numerous such disorders have been described. The signs and symptoms of such lesions may be non specific and do not help directly in reaching a diagnosis. Also many of the lesions cannot be examined directly since these are neither visible nor palpable; moreover, surgical exploration of orbit is an invasive procedure, which may lead to various ophthalmic complications. As stated above various pathological conditions may be responsible for proptosis but this study is conducted with the aim to determine only 'NEOPLASTIC' etiology for the same. An attempt has been made to analyze various investigative procedures used in the diagnosis of this condition so as to serve as a guideline for the management of this vast group of disorders.

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Materials and Method

The study was conducted for a period of 2 years; the study included those patients with unilateral proptosis due to benign or malignant tumour of orbit, paraorbital region and other region. Patient with unilateral proptosis due to other causes and bilateral proptosis were excluded from the study. All the patients were investigated radiologically with paranasal sinus X-rays, X-ray orbit and X-ray chest PA view.

B-scan of orbit, CT PNS and MRI was performed whenever required.

Direct FNAC from the mass was done whenever possible, USG guided FNAC was done whenever required and preoperative biopsy was done whenever required.

After final diagnosis treatment modality was decided, this included Surgery, Radiotherapy, Chemotherapy or Combination of these. Those patients who were not suitable for definitive treatment were treated symptomatically. Patients were followed up at the interval of 3 months, 6 months and 1 year.

Proptosis was measured by simple/plastic ruler exophthalmometry method [2]. In this method plastic ruler is kept at the outer orbital margin and protrusion of apex of cornea in relation to the outer orbital margin is measured. Normal readings are between 14 and 21 mm and reading greater than this indicates some degree of proptosis.

Results

The Maximum numbers of cases (8) were belonging to age group 31–40 years. The youngest patient was 4 years where as oldest patient was 80 years. The malignant tumours were 32 (80%) and benign tumours were 8(20%). These cases presented to us mainly with unilateral proptosis, nasal obstruction, nasal and facial swellings, etc. as shown in Table 1.

Table 1 Signs and symptoms

Eye	No.	ENT	No.
Proptosis (unilateral)	40	Nasal obstruction	25
Diminished vision	32	Nasal mass	22
Diplopia	30	Facial swelling	18
Diminished ocular motility	26	Nasal discharge	08
Epiphora	22	Headache	08
Chemosis	08	Epistaxis	07
Orbital mass	03	Oral lesion	05
Eye pain	03	Facial lesion	03
Ptosis	01	Trismus	02

Table 2 Proptosis (in mm)

Proptosis (mm)	No. of cases	Proptosis (mm)	No. of cases
3	12	11	–
4	09	12	–
5	05	13	–
6	03	14	02
7	01	15	–
8	04	16	01
9	02	17	–
10	–	18	01

Fig. 1 Pre and postoperative photo of recurrent angiofibroma

These cases underwent B-Scan (20 cases) and confirmed by CT Scan (36 cases) and MRI (four cases). The most of the lesions were mainly in right medial extraconal compartment.

The proptosis was measured in mm by ruler/simple exophthalmometry by which 12 cases were having 3 mm proptosis (minimum) and one case having 18 mm (maximum) as shown in Table 2.

Among these, in 19 cases origin of tumour was from maxillo-ethmoid complex, 4 from frontal sinus, four from nose, three from nasopharynx (Fig. 1 shows preoperative and postoperative photo of recurrent angiofibroma and histopathology of the same lesion is shown in Fig. 2), two from sinonasal site, and one each from Sphenoid, CNS, Thyroid (Fig. 3 shows a case of follicular CA of thyroid with secondaries in frontal sinus and Fig. 4 shows histopathology of follicular Ca of thyroid) and Parotid as shown in Table 3.

Total 18 types of histopathology were encountered and maximum 20 cases were of squamous cell carcinoma (SCC) (Fig. 5 shows a case of inverted papilloma which

Fig. 2 Histopathology of angiofibroma

Fig. 3 Follicular carcinoma with secondaries in frontal sinus

Fig. 4 Histopathology of follicular CA thyroid

Table 3 Origins of tumours invading orbit

Orbital	
Schwannoma	01
Retro orbital tumour (SCC)	02
Mixed tumour of lacrimal gland	01
Nose	
Rhabdomyosarcoma	01
Olfactory neuroblastoma	03
Nasopharynx	
Leukaemia	01
Juvenile angiofibroma	01
Carcinoma low grade	01
Sinonasal	
Giant cell lesion	01
S.C.C.	01
Meninges	
Meningioma	01
Paranasal sinuses	
Maxillo-ethmoid	
S.C.C.	16
Adenoid cystic carcinoma	01
Chondromyxoid fibroma	01
Non-hodgkins lymphoma	01
Frontal	
S.C.C.	01
Mucocele	01
Osteoma	02
Sphenoid	
Wegener's granuloma	01
Thyroid	
Follicular carcinoma	01
Parotid	
Rhabdomyosarcoma	01
Double Primary	
Adenocarcinoma (L) (parotid) with SCC (R)—retroorbital (included in retroorbital tumour)	01

later turned out to be SCC). The others being olfactory neuroblastoma (3), osteoma (2), rhabdomyosarcoma (2), schwannoma (1), adenoid cystic carcinoma (1), non-Hodgkin's lymphoma (1) (Fig. 6 shows a case of non-Hodgkin's Lymphoma in right maxilla), Wegener's granulomatosis (1), mucocele (1), giant cell lesion (1), leukemia (1), angiofibroma (1), meningioma (1), follicular carcinoma of thyroid (1), mixed tumour of lacrimal gland (1) adenocarcinoma of parotid (1) (Fig. 7 shows a case of retroorbital SCC with adenocarcinoma of left parotid) and low grade carcinoma (1), chondromyxoid fibroma (1) (Fig. 8 show a case of recurrent chondromyxoid fibroma).

Total 14 cases were subjected to Surgery (S) only, one case to Radiotherapy (R) only, 04 cases to Chemotherapy

Fig. 5 Inverted papilloma turned to be SCC

Fig. 7 A case of retroorbital SCC with adeno CA of left parotid

Fig. 6 Non Hodgkins' lymphoma of right maxilla

(C), five cases to Surgery and Radiotherapy (S + R), one case to Surgery + Radiotherapy + Chemotherapy, and one case to Surgery + Radionuclide iodine therapy and 14 cases received Symptomatic treatment.

While follow-up 12 cases developed complications like radiation induced mucositis and dermatitis (four cases), CSF rhinorrhoea (one case), etc. and all were managed symptomatically. Other complications included osteomyelitis, obesity, subconjunctival haemorrhage, sudden flared residual disease.

Coming to outcome of the treatment there were four cases (10%) improved, three cases (7.5%) had same status and 17 cases (42.5%) deteriorated as shown in Table 4.

Fig. 8 Recurrent chondromyxoidfibroma

Total four cases died on diagnosis, ten cases are alive with no evidence of disease, 18 cases are alive with evidence of disease and eight cases lost to follow-up after 6 months.

Discussion

The incidence of tumours giving rise to unilateral proptosis varies from place to place. It also varies in different age groups though some lesions occur in particular age groups, for example most of the malignancies are common in elderly patients.

Algernon B. Reese [3] studied records of 230 consecutive patients with unilateral proptosis. He found commonest

Table 4 Treatment outcomes

Sr. no.	Modality	Improved	Same	Detoriated	Died
01	S	12	–	02	00
02	R	–	01	–	–
03	C	02	01	01	00
04	S + R	02	–	02	01
05	S + R + C	–	–	01	–
06	S + Radio nuclide iodine	–	–	01	–
07	Symptomatic	–	01	10	03
Total		16	03	17	04

cause to be Endocrine Ophthalmopathy (37 cases), followed by Orbital Haemangioma (28 cases). He also found Malignant Lymphomas (22 cases), Pseudotumour (18 cases), Lacrimal gland epithelial tumours (17 cases), Meningioma (11 cases), Lymphangioma (ten cases), Glioma of optic nerve (eight cases), Metastatic malignant tumours (eight cases), Other tumours he reported are Peripheral nerve tumours, Dermoid cyst, Mucocoele, Rhabdomyosarcoma, Osteoma, Leukaemia, Fibrous-dysplasia, Sarcoid, Histiocytoma, Liposarcoma, etc.

According to A. Vincent Thamburaj [4] there are over 1,500 different tumours that can affect the orbit and the majority of these are benign. According to him, capillary hemangiomas are most common benign tumours in children that affect the orbit and rhabdomyosarcoma is most common malignant tumour in children that affects orbit. In adults cavernous hemangiomas are most common benign orbital tumour followed by meningioma. Metastases from lung or breast are most common malignant tumours affecting the orbit.

Esthesioneuroblastoma is an uncommon malignant neoplasm of the nasal vault, that in past was considered benign or low grade malignant. Figure 9 showing a Warms' eye view for proptosis in a case of esthesioneuroblastoma. Bradley Patrick et al. [5] described that large tumours should be considered for preoperative radiotherapy and post operative chemotherapy. Figure 10 shows the CT of the same patient with the mass. Clifford Chao et al. [6] studied the impact of treatment modality and found that with adjuvant radiation therapy, the surgical margin status did not influence local tumor control. Surgical resection plus adjuvant radiation therapy yielded the best treatment outcome. More effective chemotherapy agents with reproducible effectiveness are needed for patient with locally advanced esthesioneuroblastoma. It has sex predilection in males and occurs in all age groups with bimodal peak in second and sixth decades. Figure 11 showing the Postoperative photo following surgical resection. Figure 12 shows case of recurrent esthesioneuroblastoma.

Fig. 9 Eye view for proptosis in a case of esthesioneuroblastoma **Fig. 10** CT of the same patient of esthesioneuroblastoma

In 10 years retrospective study of malignant tumours of the maxillary sinus Dragoljub Popovic and Dusan Milisavljevic [7] found that out of total 20 patients all were of SCC. Eleven patients had lymphnode involvement at diagnosis. Symptoms initially included nasal obstruction, epistaxis etc. Three patients with orbital involvement presented with proptosis. Surgery with postoperative radiation remains the standard treatment for respectable carcinoma.

Galati [8] stated that early stage sinus malignancies are usually asymptomatic, making detection difficult. Larger

Fig. 11 Postoperative photo following surgical resection

Fig. 12 Another case of recurrent esthesioneuroblastoma

sinus tumour may extend into orbit, nasal cavity, or mouth. Resulting symptoms can mimic those caused by benign orbital, nasal or oral disease causing delay in diagnosis.

According to Kevin Katzenmeyer and Anna Pou [9], 75% of tumours are T3 or T4 stage at diagnosis. This leads to difficult decision about treatment. Use of preoperative or postoperative radiation is often used for positive margins. Chemotherapy is usually reserved for palliation of unresectable lesions, metastatic lesions or with recurrences.

Scott McCary et al. [10] studied preservation of the eye in the treatment of sino-nasal malignant neoplasms with orbital involvement. Out of 74 patients, 41 of whom

confirmed invasion of the orbit, with the use of preoperative radiotherapy and resection of involved periorbital with frozen section control, the eye can be spared in most instances without compromising oncologic safety.

A total of 79 cases of paranasal sinus tumours were reviewed by Johnson et al. [11]. Of these 47 (59%) demonstrated orbital involvement out of which 18 (38%) patients had proptosis and 16 (34%) had nasal obstruction as a presenting complaint. The maxillary sinus was the most common site of origin of paranasal sinus tumors (61%).

Zaidi [12] in a study of unilateral proptosis in ENT practices found 23.50% malignant tumours and 32.25% benign tumours responsible for unilateral proptosis. Rest of the causes for unilateral proptosis were inflammatory, congenital, traumatic, etc.

Vikas Sinha et al. [13] showed in a study of 50 consecutive cases of proptosis due to ENT causes showed a male female ratio of 7:3. Nasal obstruction and epistaxis are the commonest ENT complaint, while facial swelling and nasal mass are commonest findings in these cases. The commonest cause being Nasopharyngeal angiofibroma followed by Ca Nasopharynx and Ca Maxilla. CT scan with its high positivity rate is of primary importance in diagnostic work-up. USG is also good non invasive investigation.

Calcaterra et al. [14] stated that patients with unilateral proptosis warrant a systemic and multi-disciplinary work-up before surgical exploration or therapeutic regimen is undertaken. The otolaryngologist is concerned primarily with the diagnosis of disease process that arises adjacent to the orbital space in the nose, paranasal sinuses, nasopharynx and temporal fossae. Careful head and neck examination should be supplemented by recently refined investigations such as computed tomography, arteriography with subtraction, orbital venography, contrast orbitography, ultrasonography, etc.

The direction of exophthalmos may indicate the site of lesion. Tumours arising within the muscle cone like optic nerve glioma may give purely axial exophthalmos. Diseases of frontal or ethmoid sinuses may displace the eyeball downwards and/or laterally. Medial displacement of the eye often indicates tumours of the lacrimal gland or temporal fossa. Tumours of maxillary sinus may push the globe upward [14]. Vertical displacement can be assessed by placing a ruler across both lateral canthi and determining the relationship of the pupils to this straight line. Horizontal displacement is determined by comparing the distance from a mark on the center of the bridge of nose to each limbus [14].

In every case of unilateral proptosis thorough examination of the anterior surface of globe and eyelids should be done. Pulsations of the globe may indicate intra-cranial

pulsations being transmitted to the eye. Auscultation of the eye may reveal bruits indicative of arteriovenous malformations [14].

Palpation is very important in cases of unilateral exophthalmos. It yields important information particularly regarding anteriorly situated lesions. Inflammatory lesions and tumors tend to feel resistant, where as less resistance is noted with endocrine exophthalmos and hemangiomas. Fullness in infra-temporal fossa suggests lateral extension and with bony destruction there may be irregularities of orbital rim [14].

Clinically significant proptosis is any forward displacement of globe 2 mm greater than the other as measured by Hertel's/Krahn exophthalmometer [15].

The average volume of an adult orbit is 30 ml. An increase of orbital volume by only 5 ml will produce 4–5 mm of proptosis [16].

There are very few studies available about unilateral proptosis and these studies have included all the lesions giving rise to unilateral proptosis i.e. inclusive of congenital, traumatic, infective, hormonal lesions as well and in this study only the tumours are considered thus it is difficult to correlate. Therefore, tumour cases among these studies available are compared with the present study.

In the present study of 2 years a total 40 cases with unilateral proptosis were found and matches with the study of Deepak Rajenderkumar [1] who found 19 cases in 1 year but this is in contrast to Bernard Palmer and Denver [17] who found four cases in 1 year, Harimohan et al. [18] found 31 cases in 6 years, Somnath Saha et al. [19] found 31 cases in 4 years as shown in Table 5.

In present study most common age group was found to be 31–40 years (20%) which is in contrast with the study of Deepak Rajender Kumar [1] where in (50%) cases belonged to age group between 11 and 30 years. Somnath Saha et al. [18] found most common age group to be 40–49 years (25%).

Reese [3] studied records of 230 consecutive patients with unilateral proptosis and found commonest cause after endocrinal ophthalmopathy was orbital haemangioma. This

is contrasting to present study as no case of orbital haemangioma is reported and most common cause for unilateral proptosis was found to be Maxillo-ethmoidal tumours (47.5%).

In present study distribution of malignant and benign tumours nearly matches with the study of Hari Mohan et al. [18] who encountered with 77.41% malignant tumours and 22.59% benign tumours. In this study there are 80% malignant and 20% benign tumours, where as Deepak Rajender Kumar [1] reported 50% malignant tumours as cause for unilateral proptosis.

Lenworth Johnson et al. [11] reviewed a total number of 47 cases of para-nasal sinus tumours invading the orbit and found maxillary sinus was the most common site of origin of para-nasal sinus tumours (61%). This is comparable with present study as most of tumours were found to be originating from maxillo-ethmoidal region (47.5%). Deepak Rajender Kumar [1] found 50% cases of maxillo-ethmoidal complex and Somnath Saha et al. [19] found 25% cases of maxillo-ethmoidal region in a retrospective study.

Most common pathology in present study was found to be squamous cell carcinoma (50% cases). This is comparable with Harimohan et al. [18] who found (39%) cases of SCC but this is in contrast with the study of Somnath Saha et al. [19] who found (13%) cases of SCC and most common pathology he found was angiofibroma.

There were 57.5% right sided and 42.5% left sided proptosis which can be compared with study of Deepak Rajender Kumar [1] who found 50% right sided proptosis and 45% left sided and 1 case with bilateral proptosis. He also found maximum 8 mm of proptosis however in present study maximum 18 mm was recorded.

Conclusion

Thus, one of the major causes of unilateral proptosis is malignancy arising from the paraorbital region. Due to their location and non specific symptoms they are mainly detected at a later stage. Treating such lesion most probably lead to permanent facial disfigurement and still not provide much hope for disease control. Thus, even tough proptosis is an orbital symptom a thorough and complete ENT examination is necessary for detecting such tumours early, managing them at early stage and improve the morbidity and mortality of the patients suffering from such lesions.

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Table 5 Comparison between numbers of cases in each study

S. no.	Author	No.	Period in years	Prospective/retrospective
01	Bernard W. Palmer and Denver	04	01	Prospective
02	Hari Mohan et. Al	31	06	Prospective
03	Lenworth N. Johnson et. Al	47	–	Retrospective
04	Deepak Rajender Kumar	19	01	Prospective
05	Somnath Saha et. Al	31	04	Retrospective
06	Our Study	40	02	Prospective

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