

Unilateral Proptosis due to Sino-Nasal Pathology. Management of Thirty Cases

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A series of thirty cases of unilateral proptosis due to sino nasal pathology is presented and their management is discussed. The proptosis was due to encephalocele in 1, inflammatory processes in 13, neoplasms in 13, frontal sinus mucocoele in 2 and pseudotumour in one case. CT Scan in axial and coronal sections is a radiological study of first choice in evaluating unilateral proptosis. The treatment is according to the under lying cause. Majority of cases were treated surgically depending upon the stage of disease at initial presentation.

Key words: Unilateral proptosis, ENT causes

Proptosis is an abnormal anterior prominence of orbital contents. The term "Exophthalmos" is used interchangeably, although it is more specific to the organ involved.

The otolaryngologist and head and neck surgeon is frequently called upon to evaluate and treat the patients with unilateral proptosis and must work closely with the ophthalmologist, neurosurgeon and radiologist to ensure good prognosis.

Proptosis may occur from a variety of causes. Thyroid and primary orbital disease account for 90% of cases. In ENT practice, infections, inflammations and neoplasms are the predominant etiologies.

CT Scan is found most helpful in delineating the extent of the orbital involvement by disease process.

Surgical management depends upon etiology and extent of the disease.

Material and Methods

Thirty cases of non-thyroidal unilateral proptosis seen primarily in ENT Department Lahore General Hospital, Lahore during Jan 1996 to Dec 1999 were included in the study.

The underlying pathology was usually identified using a diagnostic approach outline in Table 1.

Table 1 Evaluation of unilateral proptosis

History and Physical Examination

Laboratory tests (CBS, ESR, Blood Sugar, Thyroid (function tests), Radiological studies.

Radiological studies

Standard Sinus Views

C.T.Scan

X-Ray Chest

Other studies (for special circumstances)

Angiography

Ultrasonography

Biopsy

Important aspects of history include duration and degree of globe displacement, change in vision, diplopia, epiphora, pain or fever. A history of allergies, sinus infections,

epistaxis, nasal discharge, airway obstruction, tearing and facial swelling suggested a sinonasal origin. The presence of thyroid disease, facial or orbital trauma or tumours elsewhere in the body ascertained.

In the routine ENT and Neck examination, focus was directed toward the nose, paranasal sinuses, nasopharynx and neck for cervical glands and enlarged thyroid gland.

Patients were reviewed by ophthalmologist for documentation of visual acuity and fields, extra ocular mobility, examination of fundi and retinal vessels and a measurement of intra ocular pressure. Proptosis was measured with "hertel exophthalmometer". Proptosis was established when the distance between corneal apex and lateral orbital margin was more than 21 mm or a difference of 2mm between the eye positions. Causes of pseudoproptosis such as macrophthalmia, ipsilateral lid retraction, contralateral ptosis and bony orbital abnormalities were excluded.

Laboratory investigations included complete blood count (CBC) with differential, an erythrocyte sedimentation rate (ESR) for evidence of inflammation or blood dyscrasia or immune status. Blood sugar random test was performed on all patients. Thyroid function tests including T3, T4 and TSH were performed when indicated.

Radiological studies included Water's view, Caldwell's view, the Law's Lateral view and skull base view were recorded in all cases. Chest X-Ray performed to screen for metastatic disease. Computerized tomography (CT) was performed in all cases to determine the bone erosion, extent of the disease and its relation to extraocular muscles optic nerve and brain.

Angiography was done when a vascular pathology was suspected by history of massive/ frequent epistaxis in young adolescent male.

Ultrasonography of orbit performed to classify masses as solid, cystic, angiomatous or infiltrative, Biopsy was taken when indicated.

Treatment was mainly surgical. The surgical procedures performed included external ethmoidectomy. Excision of angiofibroma with weber-furguson approach, total maxillectomy with and without orbital exenteration.

Prosthodontist was consulted when required.

Results

Out of 30 cases, 16 were male and 14 female. The age of the patients in this study ranged from 3 to 60 years, with average age 25 years. Duration of symptoms before admission in hospital was 1 month to 1 year in 16 cases, 1-2 years in 8 cases and beyond 2 years in 6 cases. Right eye was involved in 16 and left eye in 14 cases.

Unilateral proptosis was present in all the cases. Exophthalmometry confirmed the presence of proptosis of less than 1 cm in 26 cases and more than 1 cm in 4 cases. 3 cases were blind in the affected eye, 3 patients complained of diplopia.

Nasal obstruction was complained of by 26 cases, facial swelling in 13, headache and facial pain in 19 cases. Exopthalmos was presenting feature in 4 cases. Telorism was found in 7, unilateral nasal mass in 11, bilateral nasal mass in 7 and nasopharyngeal mass in 3 cases.

Two were febrile.

None of patients was diabetic or immunodeficient. The routine radiography demonstrated involvement of maxilloethmoid complex, bony erosion, periosteal mass or nasopharyngeal mass.

CT scan in axial and coronal planes with 5 mm cuts confirmed the extent of disease with involvement of orbit in all cases, intracranial extension in 3 cases, oral cavity/oropharynx in 2 cases and infratemporal fossa in 1 case.

Diagnosis was confirmed on histopathology. Inflammatory and neoplastic pathology accounted for 13 cases each, encephalocele in 1 case, mucocoele in 2 cases and pseudotumour in 1 case. Table 2 shows the diagnosis in 30 cases.

Surgery was the primary treatment modality in 19 cases, in combination with radiotherapy in 3 cases, in combination with radiotherapy and chemotherapy in 1 case. 4 patients with intracranial extension were treated with radiotherapy alone. 3 patients were treated medically.

Surgical procedures included external ethmoidectomy in 12 cases, total maxillectomy in 4 cases, with orbital exenteration in 1 case, osteoplastic flap operation in 1 case, lateral rhinotomy with medical maxillectomy in 1 case, Weber-Ferguson approach was employed twice. Nasal polypectomy was additionally performed in 8 cases.

Medical treatment consisting of steroid therapy for pseudotumour, antituberculous therapy for tuberculosis of maxillary sinus and high doses of broad spectrum intravenous antibiotics for acute ethmoiditis and early cavernous sinus thrombosis. Results of treatment are shown in table 2.

Patients with congenital, inflammatory pathology were cured except 1 patient of cavernous sinus thrombosis who died despite aggressive antibiotic therapy. Cases of mucocoele and pseudotumour were also cured. Amongst neoplastic cases, benign lesions (schwannoma and angiofibroma) were cured. Recurrence occurred in 1 case of angiofibroma which was dealt with surgery. All patients with inoperable malignant disease or disease extending intracranially died. 1 case of rhabdomyosarcoma is alive with disease. Both cases of Ewing sarcoma were lost to follow-up. Four patients with malignant sino-nasal disease were disease free on regular follow-up.

Table 2 Diagnosis, treatment and results in 30 cases of unilateral proptosis.

n	Diagnosis	Treatment	Result
1	Encephalocele	Excision	Cured
13	Inflammatory		
	Infective		
	Acute	2 Medical	1 pt, with cavernous sinus thrombosis died.
	Chronic		
	Non-specific	3 External frontoethmoidectomy	Cured
	Specific (TB)	1 Antituberculous therapy	Cured
	Non Infective -allergic (Polyposis)	7 External frontoethmoidectomy with intranasal Polypectomy	Recurrence in 1 case dealt with simple polypectomy.
13	Tumours		
	Benign		
	Angiofibroma	2 Excision (Weber-Ferguson approach)	Recurrence in 1 case dealt with surgery
	Schwannoma	1 Lateral rhinotomy + Medical Maxillectomy	Cured
	Malignant		
	Squamous cell carcinoma	3 Total maxillectomy with / without orbital exenteration followed by radiotherapy. Radiotherapy alone in in-operable cases & also for SCC of nasopharynx. Fronto ethmoidectomy + Radiotherapy	4 patients disease free. 3 patients died (all with intracranial disease)
	Adenoid cystic carcinoma (Ethmoid)	2 Surgery + Radiotherapy + Chemotherapy Total maxillectomy	
	Transitional cell carcinoma	2	
	Rhabdomyosarcoma	1	Patient alive with disease.
	Ewing's sarcoma	2	Patient lost in follow up.
3	Others		
	Mucocoele	2 Osteoplastic flap operation (Frontal sinus) External fronto ethmoidectomy (ethmoid sinus)	Cured
	Pseudotumour	1 Debridement + Steroid	Improved

Discussion

Although 90% of the cases of unilateral proptosis have primary pathology in the orbit itself. Orbital involvement is not uncommon by a variety of pathologies arising in nose, paranasal sinuses and postnasal space. Patients with non-thyroidal unilateral proptosis mainly present to ENT department, few are referred from ophthalmologist.

Diagnostic approach as outlined in table 1 is found most helpful. Important aspects of the history include duration and degree of globe displacement, change in vision, diplopia, pain or fever. Infections, inflammatory diseases, pseudotumour and malignant neoplasms have rapidly developing symptoms and pain, whereas mucocoeles and benign or low grade neoplasms symptoms develop slowly.

Ophthalmologic evaluation is mandatory and includes documentation of visual acuity and fields, extra ocular motility, examination of the fundi and measurement of proptosis. Hertel's exophthalmometer determined proptosis accurately. The normal values range from 15-20 mm for the lateral projection of cornea. A difference of 2 mm or more between 2 corneas would indicate proptosis.

Routine radiology and ultrasonography are helpful in diagnosis, the later having the advantage of being non-invasive. Four general diagnostic patterns of mass lesions are identifiable with B scan ultrasonography: cystic, solid, angiomatous and infiltrative.

CT scan is the radiological study of choice in evaluating unilateral proptosis. Imaging in axial and coronal plan is the single most reliable investigation in confirming presence or absence of growth, delineating the extent of disease and in showing bony involvement. Carotid angiography is helpful in delineating the blood supply and preoperative embolization reduces blood loss in excision of angiofibroma.

Majority of cases demand surgical intervention. The extent of surgery is based on extent of disease initial presentation.

Infectious causes are treated with appropriate antimicrobial agents and surgical drainage, pseudotumour responds to steroid therapy.

Patients presenting with proptosis due to either infection or mucocoele of frontal or ethmoid sinus did quite well, but patients with proptosis secondary to malignant growth such as rhabdomyosarcoma and Ewing tumour did not generally do well even when early surgical intervention is done. Exenteration of the orbit is indicated in malignancies invading the eyeball.

To summarize, non-thyroidal unilateral proptosis is a

serious sign and must be treated in the early stage. For a suitable outcome, multidisciplinary approach comprising otolaryngologist, of ophthalmologist, a neurosurgeon, oncologist and radiotherapist is worth while.

References

1. Calcaterra TC and Trappe KT. Unilateral proptosis. *Otolaryngol Clin North Am* 1988; 21: 53-63.
2. Cheisman AD, Jainp D. Cysts, granulomas and tumours of the jaws, nose and sinuses. *Scott Browns Otolaryngology Head and Neck Surgery*. 6th ED. 1997; 1-40.
3. Gates GA. Current Therapy in Otolaryngology Head & Neck Surgery. 5th edition. Mosby, Baltimore, USA 1994; 391.
4. Healy GB, Chandler et al. The pathogenesis of orbital complications in acute sinusitis. *The laryngoscope* 1997; 107:441-46.
5. Hussain M, Hadi A, Qureshi R. Department of Neurosurgery, Sheikh Zayed Hospital Lahore. *Pak J Surg* 1996; 12:1.
6. Ikram M, Khan M A, Khan K. the midfacial degloving approach for excision of juvenile angiofibroma. *Pak J Otol Laryngol* 1997; 13:89-90.
7. Katreidev SA, Dortzback RK. Destructive cysts of the maxillary sinus affecting the orbit. *Arch Ophthalmol* 1988; 106:1398-1402.
8. Latt BS and manning SC. Blindness resulting from orbital complications of sinusitis. *J Otolaryngol Head Neck Surgery* 1991; 104:6.
9. Lund VJ. Extended applications of endoscopic sinus surgery--- the territorial imperative. *J Laryngol Otol* 1997; III: 313-15.
10. Lund VJ, Rulfe ME. Ophthalmic considerations in frontoethmoidal mucocoeles. *J Laryngol Otol* 1989; 103:667.
11. Mayerss E, et al. Management of inverted papilloma. *Laryngoscope* 1990; 100-481.
12. Nelson EG, Goldman ME, Hemmati M. Metastatic carcinoma of the ethmoid sinus. *J Otolaryngol Head and Neck Surgery* 1990; 103:120-23.
13. Osguthorpe JD. Sinus neoplasia. *Arch Otolaryngol Head and Neck Surgery* 1994; 120:19-25.
14. Stankiewicz JA, Newell DJ, Park AH. Complications of inflammatory diseases of the sinuses. *Otolaryngol Clin North America* 1993; 26:639-55.
15. Timon CI and Dwyer TPO. Ethmoidal Mucocoeles in children. *J Laryngol Otol* 1989; 103:284-86.
16. Waxman JE, Spectre JG, Sale SR, et al. Allergic aspergillus sinusitis concepts in diagnosis and treatment of a new clinical entity. *Laryngoscope* 1997; 261-66.
17. Zaidi SH. Unilateral proptosis in ENT practice. Department of ENT & Head and neck Surgery JPMC Karachi 1991; October 248-250.
18. Zulfiqar Ali and Anjum Khawar. Non Thyroidal unilateral proptosis. Department of ENT and Head & Neck Surgery PIMS Islamabad. *J Surg PIMS* 1993;5.