

Orbital Tumors – Retrospective Study of 24 Years

Faiz M. Halepota, Khalid Iqbal Talpur, Mahesh Kumar Luhano, Late Sher Muhammad Shaikh, Abdul Rehman Siyal

Pak J Ophthalmol 2014, Vol. 30 No. 1

See end of article for authors affiliations

Correspondence to:
Faiz M. Halepota
Hayat Medical Centre
Satellite Town
Mirpurkhas, Sindh 6900
mklohana@yahoo.com

Purpose: To evaluate incidence of uncommon tumors with unique clinical presentation and rising incidence of Lymphomas.

Material and Methods: This study was conducted at Chandka Medical College Larkana, Peoples Medical College, Nawabshah; Liaquat University of Medical and Health Sciences, Eye Hospital, Hyderabad; and two private hospitals of Sindh province namely Hajiani Hospital, Pathan Colony Hyderabad; and, Hayat Medical Center, Satellite Town Mirpurkhas. A total of 42 cases and conditions simulating tumors were identified and in all cases diagnosis was confirmed by histology.

Results: A rising incidence of Lymphoma, 06 cases (2.52%), followed by tumors of eyeball and lids (squamous cell CA, Retinoblastoma) 06 cases each (2.52%) were noted. Fibroangio sarcoma 03 (1.26%) lacrimal gland tumor 03 (1.26%), socket tumors 03 (1.26%) Nerve sheath tumor 03 (1.26%) (schwannoma and Neurofibroma) porocarcinoma and metastatic tumor one case each were identified (0.42%). Miscellaneous non malignant lesions were 08 (3.36%) Lymphoid Hyperplasia of lacrimal gland 02 case (0.84%).

Conclusion: Our study shows variety of tumors, few uncommon which affect orbit, with lately rising incidence of Lymphomas, revealed by employing modern investigation techniques. Ever increasing incidence of orbital lymphoma need to be confirmed by further studies in future.

Orbital tumors may be primary, secondary, metastatic extension from adjacent tissues – sinuses, lids, Eye ball or manifestation of leukaemia. Tumors may be benign or malignant; in children 90% are benign (Cystic) and 10% malignant. Studies by various authors have given variable incidence of different tumors, depending on age, race, region and study period.

Lymphoid tumors, inflammatory lesions (Pseudo-tumor mucocoele) vascular and cystic lesion (dermoid and epidermoid are most common¹. There are conditions, which mimic orbital tumors such as thyroid ophthalmopathy usual presentation is axial proptosis, decreased vision, restriction of E.O.M, pain, inflammation and cosmetic disfigurement. The initial clinical evaluation of patient with orbital mass (lesion

is frequently inclusive to arrive at corrective diagnosis clinical examination, blood tests, X-Ray P.N.S and Rhinological examination, B-Scan ultrasound, CT Scan and MRI are done. Biopsy remains gold standard; ultrasonography is a good diagnostic tool especially in tumors of solid, cystic variety and thyroid orbitopathy².

Primary orbital tumors if not treated on time and adequately cause morbidity and mortality by local extension and systemic metastasis.

MATERIAL AND METHODS

This is a retrospective study includes all cases confirmed by histology. We excluded cases of thyroid ophthalmopathy, congenital bony anomalies, orbital varicose veins, pseudo tumors who responded to

retrobulbar and systemic steroids. The cases where histopathological slides showed multi cellular and anaplastic cell appearance and diagnosis was speculative were also excluded.

We sought opinion of paediatrician, radiologist and clinical pathologist where necessary biopsy was done in all cases. Blood tests, CT Scan, MRI were done in selected cases to assess site, size, spread to surrounding structures and plan surgical approach.

CT Scan and MRI helped clinically and morphologically to differentiate orbital infections from benign and malignant tumor of epithelial and connective tissue origin. Tumor within muscle cone or advanced tumors invading sinuses on anterior cranial fossa were referred to ENT and Neuro Surgeon respectively.

RESULTS

Age of study cases varied from 6 months to 80 years and gender wise male were 23 and female 19 (Table 1). The most common tumors were Lymphoma, squamous cell carcinoma and retinoblastoma comprising 6 of each (Table 2). While other tumors included Fibro Sarcoma, Lacrimal Gland Tumor, Socket Tumor, tuberculous granuloma, neurofibromatosis (3 each), optic nerve meningioma, Dermoid Cyst, Lymphoid Hyperplasia of Lacrimal Gland (2 each) and Schwannoma, Metastatic Tumor, Porocarcinoma (1 each) (Table 2).

In squamous cell carcinoma 04 cases were extension from limbus (Fig. 6) and 2 cases were direct spread from lid (Fig 7).

Regarding Fibrosarcoma, first case of two year old female child had grown to larger dimensions just within three months time. It shows rapid and aggressive growth in children¹.

In a second case of 24 year old male, it had grown slowly and was well differentiated. In a third case 55 year old woman, it had grown slowly over a period of 5 years and was painless (Fig 3). On attempted exentration, it was massive growth which bled profusely and had eroded bony walls of maxillary sinus, lateral wall of nose and roof of orbital fossa. Patient died three weeks post-operatively due to concurrent infection.

As regards lacrimal gland tumor adenoid cyst carcinoma occurred in a 40 year old female. This tumor had local infiltrative and metastatic potential. It responds to radio therapy but is not radio curable¹.

Rest three tumors were one case epidermoid carcinoma of lacrimal gland origin and two cases of lymphoid hyperplasia.

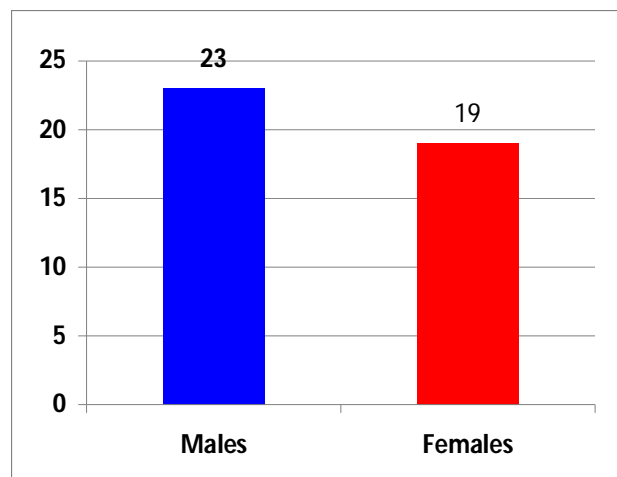


Table 1: The gender breakup of cases

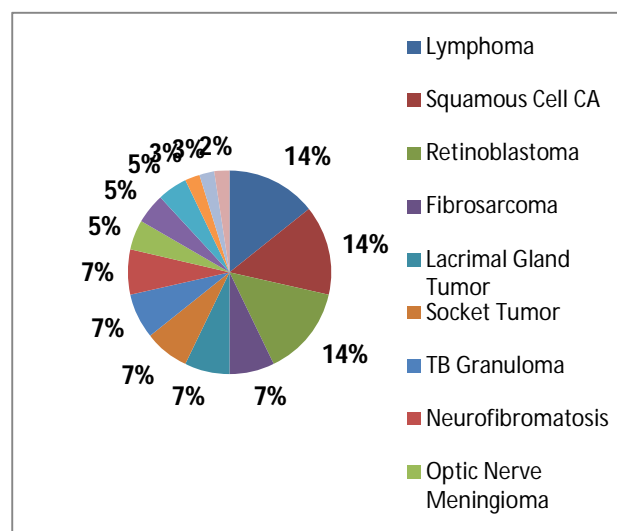


Table 2: The percentage of different types of tumors



Fig. 1: Fibrosarcoma

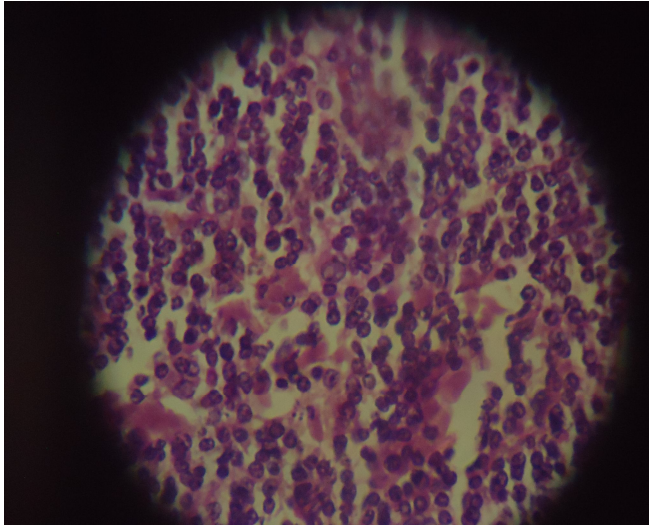


Fig. 2: Lymphoma H.E Staining

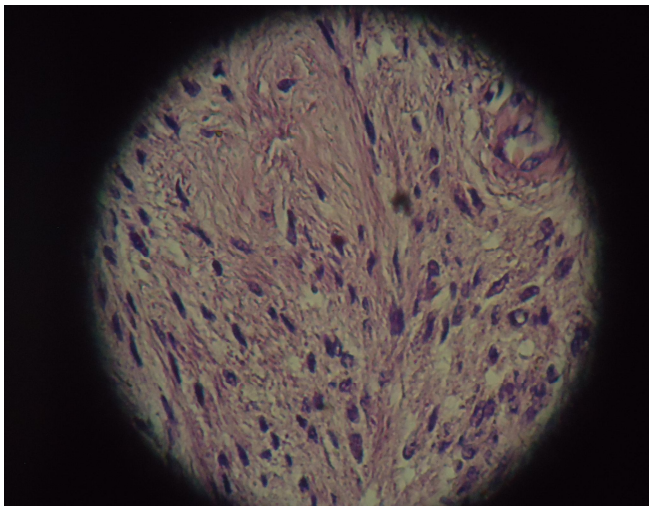


Fig. 3: Schwannoma H.E Staining



Fig. 4: Meningioma Optic Nerve

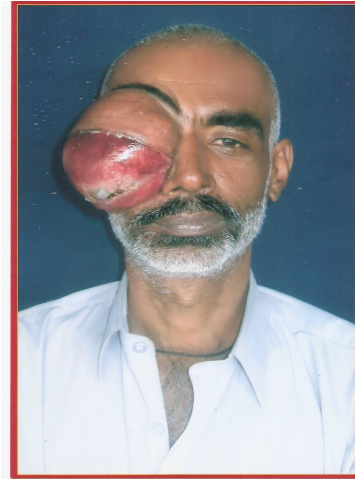


Fig. 5: Schwannoma



Fig. 6: Squamous Cell Carcinoma

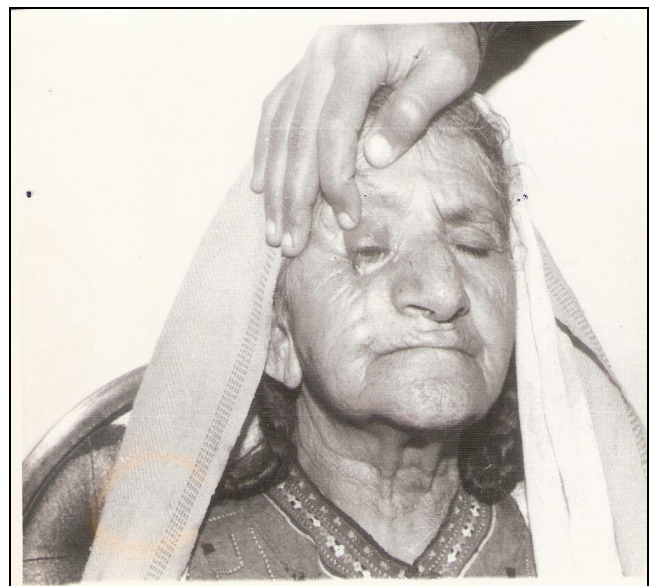


Fig. 7: Squamous Cell Carcinoma Lid.

Two cases of optic nerve meningioma were recorded. In both patients enucleation was done: one 24 year old male had progressive proptosis, pain, lid oedema, chemosis (Figure 4), for such growths similar description has been described with age less than 10 years in literature¹¹.

Regarding nerve sheath tumors, one schwannoma (Neurolimoma) and two Neurofibroma were recorded. Schwannoma has been reported in two studies^{5,8} but without mention of histological pattern and duration of onset.

Our case (Fig 3) with 5 year duration is unique: bulky mass and easily enucleated out without blood loss contrary to our expectations. Microscopically both Antoni A-Spindle cells in cords and whorls plus Antoni B-stellate cells with mucoid stroma, coexisted in some cross sectional view¹.

A nine year old boy presented with painless proptosis of right eye of recent onset. After clinical examination retrobulbar leukaemia deposits were suspected. Peripheral blood film revealed blast cells. Patient was referred to hematologist and was lost to follow up.

In 50 year old man with proptosis of few years duration exentration was done. A diagnosis of poro carcinoma was made by Histopathologist after examination of orbital contents.

Incidentally secondary tumors with extension from adjacent sinuses, nose brain were not encountered in our collection of 24 years duration.

DISCUSSION

Incidence of Lymphoma was higher as reported in recent studies^{3,4} followed by squamous cell carcinoma and Retinoblastoma (Table 2). The two latter tumors are listed equal to Lymphoma but are not primary tumors of orbit.

Out of lymphomas, 04 cases were non Hodgkin's (two large and two small) and 02 were Hodgkin's type. Our incidence was 13% but 7 – 20% has been reported in literature and same reported by Jawaid in his study⁵. However in Hodgkin's type search for extra orbital involvement was not attempted. One case of orbital Burkitt's tumor (B-cell) arising from ethmoid sinus was reported in a 5 year old boy⁶. In another report⁷ primary non Hodgkin's Lymphoma which involved left orbit and upper lid in a 4 year old girl.

Editorial by Awan⁸ quoted 24 year study (1962 – 1986) about 750 orbital tumors although there was no

mention of Burkitt's tumor. He further stated that in early nineties 200 – 300 cases were reported annually and that such tumors were not recognized or misrecognized by the pathologists. Both Hodgkin's and Non Hodgkin's Lymphoma have been reported in immuno deficiency syndrome^{9,10}.

Out of 228 ophthalmic Lymphoma adenexal and ocular reported¹ during 1980 – 2005 more than 50% were located in orbit with rapid rising incidence. Complete remission or significant reduction of lymphoma lesions following antibiotic therapy for Chlamydia psittaca infection – suggest its role in aetiology of lymphoma¹¹. Some authors also noted higher incidence of non Hodgkin's Lymphoma in Asians than Europeans and blacks.

Relationship between lymphoma and Chlamydia psittaci with regard to aetiology and response to antibodies was previously reported by Ferri A et al in two separate papers with different team of coauthors^{12,13}.

Galieni et al.¹⁴ reported fifteen patients with localized orbital lymphoma and low grade mucosa associated lymphoid tumor (MALT) which were treated with chemotherapy, radiotherapy and surgical excision with local relapse in three but disease spread was never recorded.

Hodgkin's and Non Hodgkin's classification depends on variable histological cellular pattern and morphology picture¹³⁻¹⁵.

Non Hodgkin's Lymphomas are classified into B and T cells, B cells are much more common and consist of large B cells, small cells and marginal cells. Burkitt's tumor is composed of large B Cells. Biopsy is sent to molecular diagnostic laboratory and following methods applied.

- Immuno histochemistry with surface cell markers for B and T cells¹⁵.
- Florometry

Genetic study (Chromosomal dependant) for definitive diagnosis and subtyping of lymphoma^{16,17}.

In our opinion, modern diagnostic techniques mentioned were non existent/non-available to pathologists of late 20th century, could explain lack of proper histological diagnosis typing and subtyping.

CONCLUSION

The study revealed rising incidence and prevalence of orbital lymphoma. Its incidence was even higher than that of reported. Lymphoma tumor is localized, stationary, and occasionally assuming large size.

Modern laboratory techniques have revealed higher incidence as reported in recent international study. Rising incidence and occurrence of this tumor need to be confirmed by further studies in future.

Author's Affiliation

Prof. Faiz M. Halepota
Department of Ophthalmology
Muhammad Medical College,
Mirpurkhas, Sindh – Pakistan.

Prof. Khalid Iqbal Talpur
Chairman, Department of Ophthalmology
Liaquat University Eye Hospital Hyderabad,
Sindh – Pakistan

Dr. Mahesh Kumar Luhano
Clinical Pathologist, Department of ophthalmology
Liaquat University Eye Hospital Hyderabad,
Sindh – Pakistan

Late Sher Muhammad Shaikh
Department of Pathology
Chandka Medical College, Larkana
Sindh – Pakistan

Dr. Abdul Rehman Siyal
Department of Pathology
Muhammad Medical College, Mirpurkhas
Sindh – Pakistan.

ACKNOWLEDGEMENTS

Authors extend thanks to Dr. Aijaz Ali Khooharo, Associate Professor and Incharge Director, Advanced Studies, Sindh Agriculture University Tandojam for his input in interpretation of results.

REFERENCES

1. Greer ocular pathology. 4. Blackwell Science; 1989. 241-57.
2. **Khalil M.** Diagnostic role of ultrasonography in orbital disorders. *Ophthalmology Update* 2010; 8: 17-21.

3. **Moslehi R, Devesa SS, Schairer C, Fraumeni JF Jr.** Rapidly increasing incidence of ocular Non-Hodgkin Lymphoma. *J Natl Cancer Inst.* 2006; 98: 936-9.
4. **SJO LD, Ralfkiaer E, Prause JU, et al.** Increase incidence of ophthalmic lymphoma in Denmark from 1980 to 2005. *Invest Ophthalmol Vis Sci.* 2008; 49: 3283-8.
5. **Jawaid MA.** Management of orbital tumors. *Pak J Ophthalmol.* 2005; 21: 44-8.
6. **Muhammad Z, Khan D.** Orbital Burkitt's Lymphoma, arising from Ethmoid sinus. *Pak J Ophthalmol.* 1991; 17: 87-9.
7. **Talpur KI.** Orbital – Blephro lymphoma. *Pak J Ophthalmol.* 2001; 17: 134-6.
8. **Awan K.** (Editorial), Burkitt's Lymphoma in Pakistan. *Pak J Ophthalmol.* 1991; 7: 85-90.
9. **Park KL, Gonis KM.** Hodgkin Lymphoma of orbit associated with acquired immunodeficiency syndrome. *Am J Ophthal.* 1993; 116: 111-2.
10. **Antle CM, White VA, Horsman DE, Rootman J.** Large cell orbital lymphoma in patient with acquired immunodeficiency syndrome. *Ophthalmology.* 1990; 97: 1494-8.
11. American Academy of Ophthalmology 2006. *Ophthalmic pathology; Intraocular tumors section 4.* AAO, San Francisco. 209-10.
12. **Ferreri AJ, Guidobomi M, Ponzoni M De Conciliis C, Dell'Oro S, Fleischhauer K, Caggiari L, Lettini AA, Dal Cin E, Ieri R, Freschi M, Villa E, Boiocchi M, Dolcetti R.** Evidence for association between Chlamydia psittaci and ocular adenexal lymphomas. *J Natl Cancer Inst.* 2004; 96: 586-94.
13. **Ferreri AJ Ponzoni M, Guidoboni M, De Conciliis C, Resti AG, Mazzi B, Lettini AA, Demeter J, Dell'Oro S, Doglioni C, Villa E, Boiocchi M, Dolcetti R.** Regression of ocular adenexal lymphoma after Chlamydia psittaci eradicating antibiotic therapy. *J Clin Oncol.* 2005; 23: 5067-73.
14. **Galièni P, Polito E, Leccisotti A, Marotta G, Lasi S, Bigazzi C, Bucalossi A, Frezza G, Lauria F.** Localized orbital lymphoma. *Haematologica.* 1997; 82: 436-9.
15. **Freedman AS, Nadler LM.** Immunologic markers in non-Hodgkin's lymphoma. *Hematol Oncol Clin North Am.* 1991; 5: 871-89.
16. **Clack C Glaser SL, Dorfman RF, Bracci PM, Eberle E, Holly EA.** Expert View of Non Hodgkin Lymphomas in population based cancer registry reliability of diagnosis and subtype classifications cancer epidermoid. *Biomarkers Prev.* 2004; 13: 138-43.
17. **Harris, N. Jaffe ES, Stein H, Banks PM, Chan JK, Cleary ML, Delsol G, De Wolf-Peeters C, Falini B, Gatter KC, et al.** A revised Euro-American classification of Lymphoid neoplasms: a proposal from the International Lymphoma Study Group. *Blood.* 1994; 84: 1361-92.