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Orbital Tumors and Tumor like Lesions: A Hospital Based Study

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ABSTRACT

Background: Orbital tumors have rare incidence, still they play a significant role in terms of morbidity and mortality. Orbital tumors may be primary, secondary or metastatic. These consist of benign and malignant lesions with extreme variations in pediatric and adult groups. These lesions can have acute or chronic onset, slow to rapid progression with or without bony destructions leading to vision loss, deformity and sometimes death.

Methods: This retrospective cross-sectional study was carried out in the Department of Ophthalmic Pathology and Laboratory Medicine in Biratnagar Eye Hospital. Fifty-one patients who underwent histopathological evaluation for their orbital lesions from June 2018 to December 2019 were included in the study.

Results: Orbital tumor and tumor like lesions comprised 27 cases (52.94%) in adults and 24 (47.06%) in paediatrics. Histopathologically, the most common pediatric benign tumor was dermoid cyst and malignant was secondary to orbital extension of retinoblastoma. Also, the most common adult benign orbital tumor was cavernous hemangioma and malignant was non Hodgkins lymphoma. The clinico-pathological accuracy for diagnosis was 68.63%. Association between age groups and nature of orbital lesions and between clinical and histopathological diagnosis was found to be statistically significant ($p<0.05$).

Conclusions: Orbital tumors and tumor like lesions are uncommonly encountered. These masses showed significant variation in incidence in children versus adults. Combined efforts by different specialties help in early and prompt management of the orbital tumors.

Keywords: Adult; neoplasms; Nepal; orbit; pediatrics.

INTRODUCTION

Orbital tumors and tumor like lesions are rarely encountered and comprise 3.5-4% of all ophthalmic pathologies.[1-2] According to the American Cancer Society, the incidence of orbital tumors is less than 1 per 100,000 population.[3] Incidence of tumors depend on source and geography of material studied.[4-6]

Various benign and malignant neoplasms, non-neoplastic and inflammatory conditions develop within the orbit. Orbital tumors can be primary, secondary or metastatic based on their site of origin. These masses can cause visual impairment, deformity, eye movement deficit, cosmetic problems and be life threatening as well. Orbital tumor and tumor like lesions in children reveal a varied histologic pattern than in adults.

We found it interesting to evaluate the histopathologically proven orbital lesions in the first newly established ophthalmic pathology department in Nepal. The main aim was to determine the histopathological spectrum of orbital tumors in children and adults and to evaluate the correlation between clinical findings and histopathological diagnosis.

METHODS

This hospital based retrospective cross-sectional study was carried out in the Department of Ophthalmic Pathology and Laboratory Medicine in Biratnagar Eye Hospital from June 2018 to December 2019. This study was conducted after the approval from Institutional Review Committee of Biratnagar Eye Hospital.

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Whole sampling method was used and all surgically resected orbital lesion specimens, both biopsy and orbitotomy, sent for histopathological evaluation were included in this study. Secondary tumors with orbital extension on histopathological evaluation were likewise included. Orbital cases that were dealt clinically like capillary hemangioma, thyroid orbitopathy and eyelid or intraocular tumors with no orbital extension were excluded from the study. These orbital specimens were sent in containers with 10% buffered normal saline along with a requisition form containing the demographic details, clinical history and diagnosis with radiological reports to Department of Ophthalmic Pathology and Laboratory Medicine. No repeat biopsy was done in any case.

Gross findings were noted emphasizing on the shape, size, consistency, color and margins. Representative sections of the specimens were processed, embedded and stained using conventional haematoxylin and eosin for histopathological evaluation.

The patient details including the age, gender and clinical diagnoses were obtained from the submitted form filled by the surgeons. All files, computer records for histopathological diagnosis were reviewed. Orbital lesions were grouped into non- neoplastic lesion, benign tumor and malignant tumor. Further, non-neoplastic and benign tumor were grouped together as benign tumor and tumor like lesions.

Data were entered in Microsoft Excel and statistical analysis was done using SPSS 20.0. Categorical variables were presented as frequencies and percentage. Chi-square test was used to find the association. Differences were regarded as statistically significant when p value was less than 0.05.

RESULTS

Among the 51 cases, adults comprised of 27 cases (52.94%) and children 24 cases(47.06%). Out of the 27 adult orbital masses, 20 were benign and 7 were malignant. Similarly, the pediatric group had 15 benign and 9 malignant orbital masses. Maximum cases were noted in children ten years and younger. Association between age groups and nature of lesions was found to be statistically significant (p<0.0025) (Table 1).

Considering the benign orbital conditions in children, cystic lesions were the most common followed by inflammatory lesions. Dermoid cyst was the most encountered cystic condition whereas orbital cysticercosis was the most common inflammatory lesion. All of the malignant cases in the pediatric group were orbital extension of retinoblastoma (Table 2).

Considering the benign orbital cases in adults, the maximum cases were of inflammatory lesions followed by vascular and cystic conditions. Cavernous hemangioma was the most common benign entity. The lymphoid tumors were the most common malignancy in adults. All lacrimal gland tumors were of epithelial in origin (Table 3).

The clinical findings were correlated with histopathological diagnosis and had accuracy rate of 68.63% of the total cases. The association between clinical and histopathological diagnosis was found to be statistically significant (p<0.0057). The orbital tumors and tumor like lesions that were histologically different from clinical evaluation are listed below (Table 4) .

Table 1. Distribution of Orbital tumor and tumor like lesions.

Categories		Benign	Malignant	Total (%)	p-value*
Population grouping	Paediatric(<=18years)	15	9	24 (47.06)	0.373
	Adult (>18 years)	20	7	27 (52.94)	
	Total (%)	35 (68.6)	16(31.4)	51 (100)	
Gender	Male	20	10	30 (58.82)	0.718
	Female	15	6	21 (41.18)	
	Total (%)	35(68.6)	16(31.4)	51(100)	
Age groups in years	0-10	6	9		0.0025
	11-18	9	0		
	19-30	7	0		
	31-40	8	2		
	41-50	0	1		
	51-60	5	2		
	61-70	0	2		

* Chi square test with statistically significant at p<0.05

Table 2. Orbital tumor and tumor like lesions in children.

Groups	Category	Histopathological Diagnosis		Sub-total	Total (Percentage)
Primary (62.5%)	Benign (62.5%)	Cystic	Dermoid cyst	5	8(33.3)
			Microphthalmia with cyst	3	
		Inflammatory	Parasitic granuloma (Cysticercosis)	2	3(12.5)
			Tubercular granuloma (Tuberculosis)	1	
		Vasculogenic	Lymphangioma	1	1(4.2)
		Myogenic	Leiomyoma	1	1(4.2)
		Neural	Schwannoma	1	1(4.2)
		Lacrimal	Dacryoadenitis	1	1(4.2)
Secondary (37.5%)	Malignant (37.5%)	Orbital extension	Retinoblastoma	9	9 (37.5)
	Total				24(100.0)

Table 3. Orbital tumor and tumor like lesions in adults.

Groups	Category	Histopathological diagnosis		Sub-total	Total (Percentage)
Primary (92.6%)	Benign (74.0%)	Inflammatory	Chronic inflammation	3	6 (22.2)
			Parasitic granuloma (Cysticercosis)	1	
			Granulomatous inflammatory reaction	1	
			Sclerosing orbital inflammation	1	
		Vascular	Cavernous hemangioma	5	5 (18.5)
		Cystic	Dermoid cyst	3	5 (18.5)
			Simple Epithelial cyst	1	
			Microphthalmia with cyst	1	
		Neural	Neurofibroma	1	2 (7.4)
			Schwannoma	1	
		Lacrimal	Pleomorphic adenoma	2	2 (7.4)
	Malignant (26.0%)	Lymphoid	Non Hodgkins Lymphoma	4	4 (14.8)
		Lacrimal	Adenoid cystic carcinoma	1	1 (3.7)
Secondary(7.4%)		Orbital extension	Lower lid squamous cell carcinoma	1	2 (7.4)
			Uveal melanoma	1	
	Total				27 (100.0)

Table 4. Orbital tumor and tumor like lesions with no clinic-pathological concordance.

Clinical Diagnosis	**Histopathological Diagnosis**
Ruptured Anterior Staphyloma	Retinoblastoma with orbital extension
Calcified Vitreous Hemorrhage	Retinoblastoma with orbital extension
Phthisical eye	Retinoblastoma with orbital extension
Pleomorphic Adenoma	Dacryoadenitis
Pleomorphic Adenoma	Cavernous Hemangioma
Dermoid cyst	Simple Epithelial cyst
Benign epithelial cyst	Parasitic granuloma (Cysticercosis)
Benign epithelial cyst	Tubercular granuloma (Tuberculosis)

Table 4. Orbital tumor and tumor like lesions with no clinic-pathological concordance.

Clinical Diagnosis	Histopathological Diagnosis
Lymphoma	Chronic Inflammation
Lymphoma	Granulomatous inflammatory reaction
Lymphoma	Schwannoma
Lymphoma	Sclerosing orbital inflammation
Retention cyst	Leiomyoma
Optic nerve meningioma	Chronic Inflammation
Hemangiopericytoma	Schwannoma
Hemangioma	Neurofibroma

DISCUSSION

This study revealed adults being more affected (52.94%) by the orbital tumors and tumor like lesions compared to children (47.06%) with predominance of male population (58.82%). Toopalli et al and Priya et al have concluded similar findings.[7,8]

This study concluded 68.6% benign and 31.4% malignant orbital cases. Similar findings with greater proportion of benign orbital lesions were reported by Toopalli et al,[7] Priya et al,[8] G.Bonavolonta et al [9] and He et al.[10] In our study, benign cases exceeded malignant ones because may be the malignant cases were referred directly to higher cancer centers.

The present study revealed 78.4% primary orbital tumors and tumor like lesions which was consistent with study by Shields et al (82%)[11] but was significantly higher than reported by Parashkevova et al (56%)[1] and Sen DK (59.9%)[12] and lower compared to findings by Priya et al (89%)[8] and G. Bonovolanta et al (88%).[9] These studies had vasculogenic, cystic, inflammatory and lymphoproliferative tumors as the main contributors which corresponded well to our findings. We encountered 21.6% secondary tumors with maximum cases of orbital extension of retinoblastoma. Similarly Sen DK reported 35.8% secondary tumors in which the highest contribution was made by orbital retinoblastoma (42%).[12] A higher percentage of secondary tumors were reported by I. Gunalp et al (48.9%), which included 34.9% of eyelid lesions.[13] Shields et al had 11% secondary tumors with orbital extension of uveal melanoma (29%).[11] We had only one such case. No metastatic tumors were found in this study. Johansen et al, G.Bonavolonta et al and Shields et al had breast carcinoma as the most common metastatic tumor. [6,9,11]

The pediatric population comprised 47.06% cases of total orbital tumors and tumor like lesions. The maximum cases belonged to the category of malignant secondary orbital extension of retinoblastoma (37.5%) followed by benign cystic lesions (33.3%) and inflammatory lesions (12.5%) of the pediatric orbital cases. Contrary to this study, several literature series revealed benign conditions as the most encountered pediatric orbital case. H. M Alkatan et al,[14] Abdallah et al[15] and Kodsi et al,[5] concluded maximum of cystic cases (23-43%) followed by vascular tumors (14-17%) in their respective series. Study by Shields concluded cystic cases (52%), inflammatory (16.4%), vasculogenic (6.8%) and adipose containing lesions (6.8%) in their series.[4] Study by Johansen et al reported optic nerve lesions as the most common (32.2%) followed by cystic lesions (13.8%).[6] In this series, the most common benign orbital lesion in children was dermoid cyst (20.8%). The finding was consistent but comparatively higher in occurrence than reported by I.Gunalp et al (8%),[13] Modi et al (13.3%)[2] and Kodsi et al (16.7%)[5] while relatively lower than H.M.Alkatan et al (27.1%)[14] and Abdallah et al (26.1%).[15] Johansen et al reported optic nerve glioma (26.3%) as the most common benign tumor in children.[6] Kodsi et al emphasized that cystic lesions have increased in incidence in children because of wider availability of general anaesthesia with its safe removal.[5] Shields et al revealed that those studies that have included dermoid cyst located at the antero-lateral orbital rim have increased cases compared to those studies that haven't stating that it may not be a true orbital lesion.[4] Several studies reported capillary hemangioma as the second most common benign tumor (10-12%),[15-16] but we had no any case of it as no surgical intervention was done. Wait and watch or intralesional steroids were used for these cases. The orbital retinoblastoma (37.5%) was the only malignancy encountered in children in this study. Similar findings with malignant orbital retinoblastoma as the most common orbital lesion were reported by Johnson et al (32%),[16] I.Gunalp et al (32.7%)[13] and Modi

et al (61.3%).[2] Our findings were discordant with other literature findings. Study by Templeton in Africa reported Burkitt's lymphoma as the most encountered pediatric orbital neoplasm causing proptosis.[17] Shields et al,[4] Kodsi et al,[5] Johansen et al,[6] Toopalli et al,[7] H.M.Alkatan et al[14] and Abdallah et al[15] reported rhabdomyosarcoma as the most common malignancy (4-10%) in childhood. Though rhabdomyosarcoma happens to be the most prevalent extraocular orbital malignancy in children,[18] we did not receive any such case due to lower incidence of these tumors in this country or direct referral to higher cancer centers.

Orbital retinoblastoma occupying 37.5% cases highlights the fact that it is still a big problem in our part of the world. Microscopic orbital retinoblastoma included those cases of intraocular retinoblastoma that presented with full thickness scleral infiltration, episcleral extension or invasion of the transected cut end of the optic nerve on histopathologic examination. Retinoblastoma though an intraocular tumor, presented to us with secondary orbital extension due to late approach, lack of health awareness, poverty and refusal to enucleation due to cultural beliefs. All our cases were present in children less than 9 years of age. H.M.Alkatan et al specified that orbital retinoblastoma have decreased in their country as a result of advancing techniques in its management.[14] Kodsi et al concluded that though the frequency of malignant primary orbital tumor in children remained same in 60 years, the overall incidence of malignancy decreased due to reduction in number of secondary and metastatic neoplasms and that was because of the changes in management of orbital metastatic lesions and earlier recognition with treatment of intraocular retinoblastoma.[5]

The adult population comprised 52.94% cases of total orbital lesions. The maximum cases belonged to the inflammatory conditions (22.2%), followed by cystic and vascular lesions (each18.5%) and lymphoid tumors (14.8%) of the adult orbital cases. The higher incidence of inflammatory orbital lesions attribute to granulomatous and infectious lesions.[19] Shu Fen Ho et al concluded majority of lymphoproliferative lesions (34.6%) and inflammatory conditions (10.3%) in their study.[20] Alsalamah et al also had maximum cases of lymphoproliferative conditions (26.4%) and vascular lesions (21.8%).[21] Johansen et al reported secondary tumor (23.24%) as the most common lesion with maximum cases of orbital extension of choroidal melanoma in adults.[6] In this study, cavernous hemangioma (18.5%) was the most common benign orbital mass in adults. Similar findings with different occurrence rates were reported by I.Gunalp et al (4.6%)[13] and Alsalamah et al (18.18%)[21] respectively in their studies. Shu fen Ho et al concluded benign lymphoid hyperplasia (16.9%) as the most common benign entity of adult orbital cases.[20] Non Hodgkins lymphoma (14.8%) was the most common malignancy in adults in this study. Shu fen Ho et al (16.2%) [20] and Alsalamah et al (21.8%) [21] reported similar findings in their series. Shields et al (10%)[11] and Demirci et al (24%)[22] reported malignant lymphoma as the most common malignancy in older population.

The incidence of malignant lymphoma is increasing in the world. Study by Ohtsuka et al revealed increased incidence of malignant lymphoma in Japan due to racial or environmental differences and use of Polymerase chain reaction to diagnose lymphoproliferative disorders.[23] Demirci et al concluded lymphoma, which may be primary or secondary to systemic disease, as the most prevalent orbital neoplasm in older adults and that systemic evaluation must be done in these cases.[22]

The proportion of lacrimal gland lesions (7.8%) in this study was high compared to study by I. Gunalp et al (2.8%)[13] but is significantly lower than studies reported by G.Bonavolonta et al (10%),[9] Tanushree et al (10%),[24] Toopalli et al (22%)[7] and Priya et al (22%).[8] All lacrimal gland tumors were epithelial in origin in the present study. Both cases of pleomorphic adenoma were in age range 25-35 years. In this study, adenoid cystic carcinoma was the only malignant lacrimal gland tumor encountered in a female in her mid-30s. Priya et al had 77.7% epithelial lesions and 55.5% malignant lacrimal gland tumors. Adenoid cystic carcinoma was seen in the third decade.[8] I. Gunalp et al reported epithelial tumors four times more common than non-epithelial tumors.[13] Tanushree et al had 80% benign and 20% malignant cases and all were epithelial in origin. Mucoepidermoid carcinoma was the only malignant case in their study.[24] Toopalli et al reported 75% benign and 25% malignant cases. They had maximum cases of pleomorphic adenoma while all malignant cases were adenoid cystic carcinoma.[7]

In our study, clinico-pathological accuracy was found to be 68.63%. Sim et al reported 21.7% patients had clinical and pathological concordance and 62.7% had clinical, radiological and pathological concordance.[25] Clinical pathological correlation was present in 72.3% cases in a study by Shrestha GB et al.[26]

CONCLUSIONS

The present study emphasized on the details of orbital tumors in our area, represented by a tertiary eye care center in Biratnagar, Eastern Nepal. In this study, children and adults showed extreme variation in the occurrence of the tumors. So, knowledge about the incidence of

orbital tumors in a particular area could help to consider it in differentials in further practice. Our eye hospital with first established ophthalmic pathology department in Nepal provided a privilege to ophthalmologists and pathologists to work under the same roof that made a positive impact in early and accurate management of the tumors.

CONFLICT OF INTEREST

None

REFERENCES

1. Parashkevova B, Balabanov C, Stateva D. Orbital tumors: clinical cases presentation. J Int Med Assoc Bulgaria. 2007 Jan 1;13(1):47-50.[Download PDF]

2. Modi PJ, Shah NA, Bhalodia JN, Gonsai RN. Orbital tumors in children: a descriptive study at tertiary care centre. Natl J Med Res. 2013;3;362–6.[Download PDF]

3. Markowski J, Jagosz-Kandziora E, Likus W, Pająk J, Mrukwa-Kominek E, Paluch J, et al. Primary orbital tumors: a review of 122 cases during a 23-year period: a histo-clinical study in material from the ENT Department of the Medical University of Silesia. Med Sci Monit. 2014;20:988-94.[Article]

4. Shields JA, Bakewell B, Augsburger JJ, Donoso LA, Bernardino V. Space-occupying orbital masses in children. A review of 250 consecutive biopsies. Ophthalmology. 1986;93(3);379–84.[Article]

5. Kodsi SR, Shetlar DJ, Campbell RJ, Garrity JA, Bartley GB. A review of 340 orbital tumors in children during a 60-year period. Am J Ophthalmol. 1994;117(2);177–82. [Article]

6. Johansen S, Heegaard S, Bøgeskov L, Prause JU. Orbital space- occupying lesions in Denmark 1974–1997. Acta Ophthalmol Scand. 2000;78(5);547–52.[Article]

7. Toopalli K, Koonamala RD, Pandharpurkar M. Orbital tumors and pseudotumors-A clinicopathological study. Scholars Journal of Applied Medical Sciences. 2018;6(9):3490-95.

8. Priya MT, Kishore N. Study of orbital tumors in a tertiary care eye hospital. Indian Journal of Pathology and Oncology. 2020;7(3):458-70.[Article]

9. Bonavolontà G, Strianese D, Grassi P, Comune C, Tranfa F, Uccello G, et al. An analysis of 2,480 space-occupying lesions of the orbit from 1976 to 2011. Ophthalmic Plast Reconstr Surg. 2013;29(2):79-86.[Article]

10. He Y, Song G, Ding Y. Histopathologic classification of 3476 orbital diseases. Zhonghua Yan Ke Za Zhi. 2002;38(7):396-8.[Article]

11. Shields JA, Shields CL, Scartozzi R. Survey of 1264 patients with orbital tumors and simulating lesions: The 2002 Montgomery Lecture, part 1. Ophthalmology. 2004;111(5):997-1008.[Article]

12. Sen DK. Aetiological pattern of orbital tumours in India and their clinical presentations: A 20-year retrospective study. Orbit. 1990;9(4):299–302.[Article]

13. Gunalp I, Gunduz K. Biopsy-proven orbital lesions in Turkey. A survey of 1092 cases over 30 years. Orbit. 1994;13(2):67-79.[Article]

14. Alkatan HM, Al Marek F, Elkhamary S. Demographics of Pediatric Orbital Lesions: A Tertiary Eye Center Experience in Saudi Arabia. J Epidemiol Glob Health. 2019;9(1):3-10.[Article]

15. Abdallah AM, Abdellatif MA, Elhwary AM, Hassan AA, Kamel AG, Saman ISE, et al. Paediatric orbital tumors in Upper Egypt: A 3-year retrospective analysis at a university hospital. Journal of Clinical ophthalmology. 2019;3(1):108-20.[Article]

16. Johnson TE, Senft SH, Nasr AM, Bergqvist G, Cavender JC. Pediatric orbital tumors in Saudi Arabia. Orbit 1990;9(4):205–15.[Article]

17. Templeton AC. Orbital tumours in African children. Br J Ophthalmol. 1971;55(4):254-61.[Article]

18. Chung EM, Smirniotopoulos JG, Specht CS, Schroeder JW, Cube R. From the archives of the AFIP: Pediatric orbit tumors and tumorlike lesions: nonosseous lesions of the extraocular orbit. Radiographics. 2007;27(6):1777-99.[Article]

19. Kim UR, Khazaei H, Stewart WB, Shah AD. Spectrum of orbital disease in South India: an aravind study of 6328 consecutive patients. Ophthalmic Plast Reconstr Surg. 2010;26(5):315-22. [Article]

20. Ho SF, Othman R. Adult orbital tumors: a Southeast-Asian experience. Asian J Ophthalmol. 2017;15(4):202-12.[Article]

21. Alsalamah AK, Maktabi AM, Alkatan HM. Adult Orbital Lesions in Saudi Arabia: A Multi-centered Demographic Study with Clinicopathological Correlation. J Epidemiol

Glob Health. 2020;10(4):359-66.[Article]

22. Demirci H, Shields CL, Shields JA, Honavar SG, Mercado GJ, Tovilla JC. Orbital tumors in the older adult population. Ophthalmology. 2002;109(2):243-8. [Article]

23. Ohtsuka K, Hashimoto M, Suzuki Y. A review of 244 orbital tumors in Japanese patients during a 21-year period: origins and locations. Jpn J Ophthalmol. 2005;49(1):49-55.[Article]

24. Tanushree V, Gowda HTV, Balakrishnan U, Kulkarni A. Clinical and Histopathological Study of Orbital Tumors. Int J Curr Res. 2015;7(3):13954–8.

25. Sim R, Young SM, Agrawal R, Sundar G. Clinical, Radiological and Histological Correlation in diagnosis of orbital tumors. Cogent Medicine. 2019;6:1607128. [Article]

26. Shrestha GB, Karmacharya PC, Shrestha JK, Shrestha GS. Profile of Pathology in Patients with Orbital Diseases. J Ophthalmol & Vis Sci. 2017; 2(1): 1014.[Download PDF]