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Original Research Article

Study of Orbital tumors in a tertiary care eye hospital

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ABSTRACT

Background: Tumors of the orbit are rare diseases in ophthalmic pathology-3.5 to 4%. They include primary and secondary tumors. Their diagnosis pose a great challenge for both clinicians and pathologists since they present with a variety of signs and symptoms and they are often difficult to diagnose at initial stages.
Aims and Objectives: To analyse the histomorphological features of orbital tumors and to compare the results of the present study with other studies in the literature.
Materials and Methods: This is a prospective and retrospective study done in Sarojini Devi Eye hospital, Hyderabad at the Department Of Pathology during December 2013 to September 2016. All surgical resected specimens of orbital tumors recieved at the Department of Pathology were included in the study. Variables examined included clinical details, radiological findings and histopathological findings.
Results: A Total of 54 cases were studied. Of these, 33 cases were males and 21 cases were females. Age group ranged from 5 years to 62 years. Incidence of the tumors showed bimodal age distribution. The most common tumor was Dermoid cyst.
Conclusion: Orbital tumors can occur at various age groups and because of their myriad presentation their diagnosis pose a great challenge. Although imaging studies graphically illustrate the tissue definition, pathological conditions can be assessed definitely only by obtaining tissue specimen surgically.

1. Introduction

The eye is not only an organ of vision but also an index of beauty in the human race.[1] The volume of the orbit is small and confined by bony walls on all sides except anteriorly. Within this space is a juxtaposition of numerous structures that subserve visual as well as extraorbital functions.[2]

The bony orbit is the smallest unit and can be defined as an enclosure bordered by bony structures and a space that contains all tissues and organs that contribute to the function of the eye. Tumors of the orbit arise primarily from soft tissues and bones.[3]

Tumors of the orbit are rare diseases in ophthalmic pathology-3.5 to 4%.[4,5] All anatomic structures of the orbit can give rise to neoplasm.[6] Majority originate between the bony orbital wall and the extraocular muscle cone.[7] Orbital tumors are classified into primary and secondary orbital tumors.[2]Primary orbital tumors include benign and malignant neoplasms. The direct extension from contiguous anatomical structures, lymphoproliferative disorders, and hematogenous metastasis result in secondary orbital invasion.[6] Also the proximity of the paranasal sinuses may lead to the secondary spread of sinus neoplasia and infections or inflammation into the confines of the orbit.[3]

Most common pediatric tumors are Dermoid cysts, Capillary hemangiomas and Rhabdomyosarcoma.[8,9] Most common adult tumors are Lymphoid tumors, Cavernous hemangiomas and Meningiomas.[5,6]The major presenting symptom is proptosis resulting from the mass effect. Changes in visual acuity or field of vision, diplopia, extraocular motility disturbances or pupillary abnormalities

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can result from invasion or compression of intraorbital contents secondary to solid tumor. Lid dysfunction and lagophthalmos or lacrimal gland dysfunction can result in exposure keratopathy,keratitis and thinning of cornea.[5]

Orbital tumors constitute a heterogenous array of lesions and as such pose numerous challenges in terms of diagnosis, imaging and management.[10]They call for a closer attention due to its specific anatomic structure and location. Orbital tumors generally require a multidisciplinary approach with cooperation of a number of medical specialities.[11]

2. Aims and Objectives

1. To analyse the histomorphological features of Orbital Tumors.
2. To compare the results of the present study with other studies in the literature.

3. Materials and Methods

The present study is a prospective and retrospective study done between December 2013 to September 2016. All surgically resected specimens of orbital tumors received by Department Of Pathology, Sarojini Devi eye hospital were included. Samples were obtained either through incisional or excisional biopsy. An informed consent was taken from all the study subjects. A total of 54 cases were taken up for the study.

3.1. Inclusion criteria

1. All ages.
2. Both sexes.
3. All benign and malignant tumors of orbit including primary and secondary tumors.
4. Lacrimal gland tumors.

3.2. Exclusion criteria

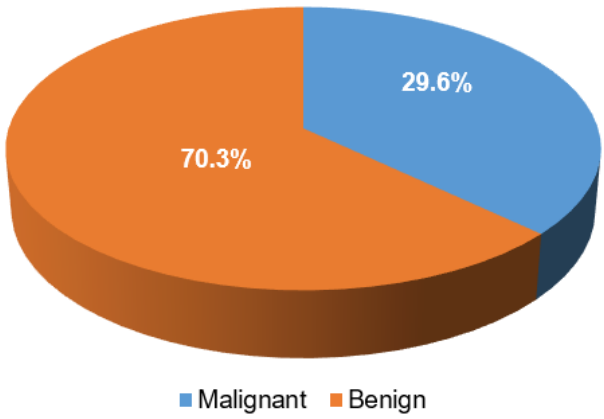
1. Intraocular tumors.
2. Infectious and inflammatory lesions of the orbit.
3. Eye lid tumors.

A detailed history taking followed by a detailed ocular examination was done. Routine blood investigations, Xray, MRI and CT scan were done wherever necessary. The lesions were approached as per their anatomical location, size, extent and suspected pathology. The surgically resected specimens were fixed in 10% formalin. Thorough gross examination of each mass for its size, shape and consistency was done.

4-5 sections of 1.2 mm thickness were taken from different areas of the specimen and processed in automatic tissue processor. Blocks were prepared with the help of leuckharts piece. The sections were stained by H&E in all the cases. Immunohistochemistry by different immune markers was done selectively. A histopathological diagnosis thus made was entered in the proforma.

4. Results

1. For the period Dec 2013 to Sep 2016: 54 patients with orbital tumors.
2. Sex ratio: M/F-1.5.
3. Age ratio: Adult / children – 1.5.
4. Majority of the patients presented with mass and proptosis.
5. 70.3% cases were benign and 29.6% cases were malignant.
6. Majority of childhood tumors were dermoid cysts.
7. 6 Out of 54 cases were secondaries.
8. 12 Out of 54 cases were lacrimal gland lesions.
9. 4 Out of 54 cases were meningiomas.



Graph 1: Percentages of benign and malignant tumors

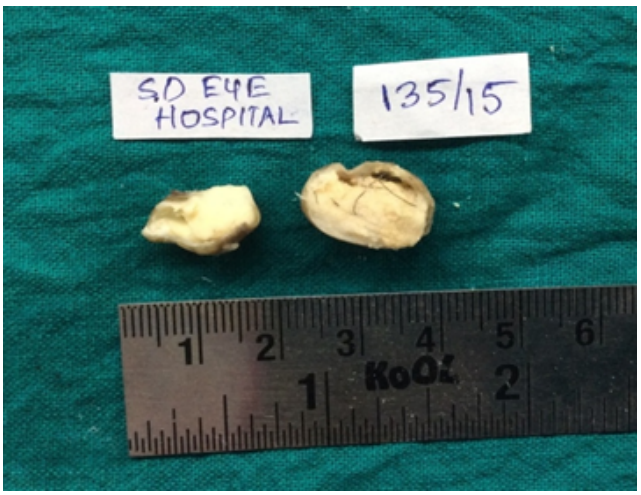


Fig. 1: Gross image of Dermoid cyst: Grey white cystic specimen measuring 1.5 x1 cm. Cut surface shows hair structures

Table 1: Classification of orbital tumors

Primary Orbital Tumors
Choristomas
Epidermoid cyst
Dermoid cyst
Teratoma
Ectopic lacrimal gland
Hamartomas
Phakomatoses
Hemangioma
1. Capillary hemangioma
2. Cavernous hemangioma
Arteriovenous communication
Telangiectasia
Lymphangioma
Mesenchymal Tumors
Vascular
1. Hemangiopericytoma
2. Glomus tumor
3. Hemangiosarcoma
4. Kaposi's sarcoma
Fatty
1. Lipoma
2. Liposarcoma
Fibrous
1. Reactive fibrous proliferations
• Nodular fasciitis
• Juvenile fibromatosis
2. Neoplastic fibrous proliferations
• Fibrous histiocytoma
• Fibroma and fibrosarcoma
• Solitary fibrous tumor
3. Giant cell angiofibroma
Muscle
1. Leiomyoma and leiomyosarcoma
2. Mesectodermal leiomyosarcoma
3. Rhabdomyoma
4. Rhabdomyosarcoma
Cartilage
1. Chondroma and chondrosarcoma
Bone
1. Aneurysmal bone cyst
2. Fibrous dysplasia
3. Giant cell tumor
4. Juvenile fibromatosis
5. Leontiasis ossea
Osteitis Fibrosa Cystica
Osteopetrosis
Paget's disease
Osteoma and osteogenic sarcoma
Neural Tumors
Amputation neuroma
Neurofibromas
Neurilemmoma
Juvenile pilocystic astrocytoma
Peripheral primitive neuroectodermal tumors (PNETs)
Miscellaneous Tumors
Meningioma
Nonchromaffin paraganglioma
Granular cell tumor
Alveolar soft-part sarcoma
Malignant melanoma
Endodermal sinus tumor
Epithelial Cysts and Neoplasms of Lacrimal Gland
Lacrimal ductal cysts
Benign mixed tumor
Malignant mixed tumor
Adenoid cystic carcinoma
Other types of carcinoma
Reticuloendothelial System, Lymphatic System, and Myeloid System
Langerhans' granulomatoses (histiocytosis X)
Eosinophilic granuloma
Hand–Schüller–Christian disease
Letterer–Siwe disease
Juvenile xanthogranuloma
Sinus histiocytosis
Inflammatory pseudotumor
Malignant lymphoma
Leukemia
Multiple myeloma
Monoclonal and polyclonal gammopathies
Secondary Orbital Tumors
Direct Extension
Metastatic

Table 2: Sex wise distribution of orbital tumors

S. No.	Sex	Total no. of tumors	No. of benign tumors	No. of malignant tumors
1.	Males	33(61%)	21	12
2.	Females	21 (39%)	17	4
Total		54 cases	38	16

Table 3: Age wise distribution of orbital tumors

S. No.	Age	Benign Tumors	Malignant Tumors
1.	1-10	11	1
2.	11-20	10	2
3.	21-30	3	3
4.	31-40	4	1
5.	41-50	7	4
6.	51-60	3	3
7.	61-70	0	2

Table 4: Percentage distribution of clinical features of orbital tumors

S. No.	Symptoms	Percentage
1.	Swelling	57.4%
2.	Axial Proptosis	35%
3.	Eccentric Proptosis	7.4%
4.	Others	12.9%

Table 5: No. of cases based on histopathological diagnosis

Benign	No. of cases	Malignant	No. of cases
Dermoid Cyst	18	Secondaries	06
Hemangioma	05	Lymphoma	05
Pleomorphic Adenoma of Lacrimal Gland	04	Adenoid Cystic Carcinoma of Lacrimal Gland	03
Schwannoma	02	Embryonal Rhabdomyosarcoma	02
Lacrimal Ductal Cyst	03		
Epidermoid Cyst	01		
Solitary Fibrous Tumor	01		
Meningioma	04		
Total	38	Total	16

Table 6: Percentage distribution of orbital tumors in childhood age group (first and second decades)

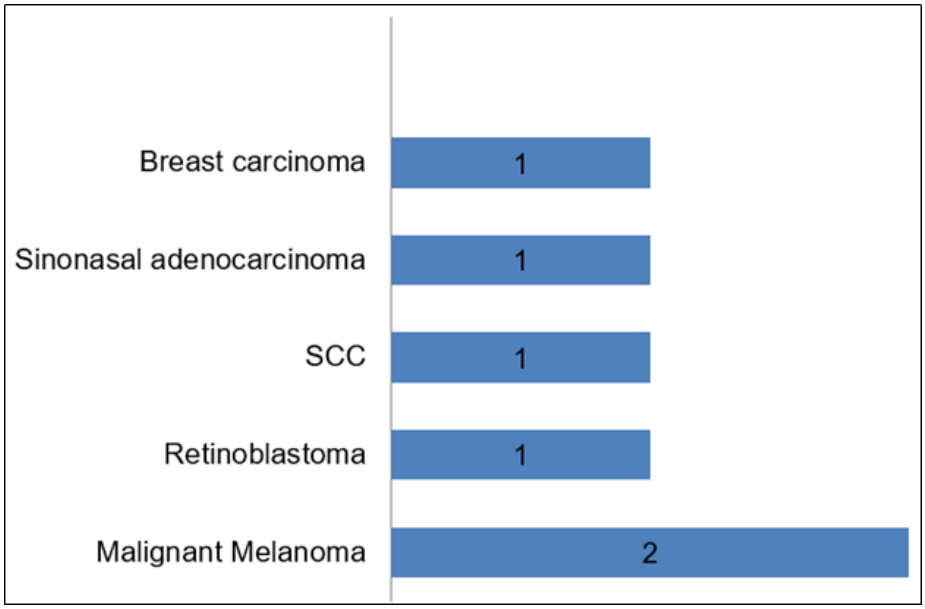
Benign	No. of cases	Malignant	No. of cases
Dermoid cyst	17	RMS	2
Capillary hemangioma	03	Retinoblastoma orbital extension	1
Meningioma	01		
Total	21	Total	3

Table 7: Case distribution of lacrimal gland lesions

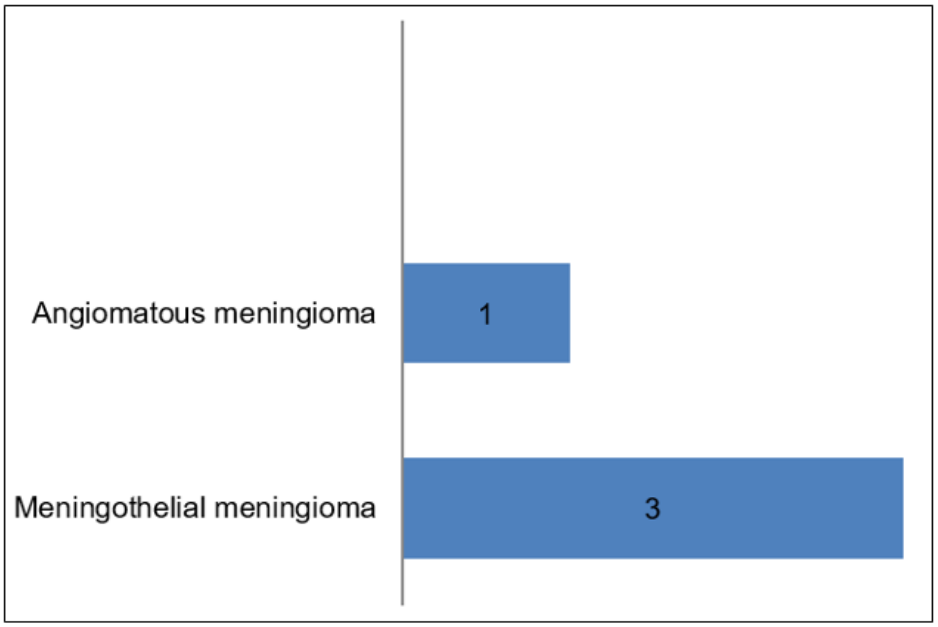
S. No.	Histopathological diagnosis	No. of cases
1.	Lacrimal ductal cyst	03
2.	Pleomorphic adenoma	04
3.	Adenoid cystic carcinoma	03
4.	DLBCL	02
	Total	12

Table 8: Distribution and typing of orbital lymphomas

S.No.	Site	No.of cases	IHC	Histopathological Diagnosis
1.	Lacrimal gland	2 cases	(CD 20,CD 79a)-positive ALK-1-negative	DLBCL
2.	Extraconal space	3 cases	2 cases-CD 20 positive And BCL2 positive in germinal center	Follicular lymphoma-Low grade
3.	-	-	1 Case-(CD 20,CD 79a)-positive, ALK-1-negative	DLBCL
4.	Total	5 cases		



Graph 2: Distribution of secondaries



Graph 3: Histopathological types of meningioma observed

Table 9: Comparative analysis of studies based on the no. of patients and the duration of period studied

Study	No. of patients studied	Year studied	Duration of study
Present study	54	2013 to 2016	3 years
Tanushree et al, 2015	48	2010 to 2014	4 years
Boriana et al, 2007	28	2001 to 2005	4 years
Radha et al, 2005	24	2003 to 2005	2 years
Jasna et al, 2004	24	1998 to 2003	5 years
Demirci et al, 2002	200	-	25 years

Table 10: Comparative analysis of Age and sex distribution of orbital tumors

Studies	Age (maximum no. of patients)	Sex (M)	Sex (F)
Present study	1^{st},2^{nd} and 5 th decades	61%	39%
Tanushree et al, 2015	2^{nd} and 3^{rd} decades	45.8%	54.1%
Boriana et al, 2007	6 th and 7^{th} decades	60%	40%
Radha et al, 2005	5^{th} decade	62.5%	37.5%
Jasna et al, 2004	1st and 7^{th}	60%	40%

Table 11: Comparative analysis of clinical presentation

S. No	Clinical symptoms	Present study	Tanushree et al, 2015	Radha et al, 2005	Demirci et al, 2002
1.	Swelling or mass	57.4%	27%	62.5%	26%
2.	Proptosis of eye	42.5% Axial -35% Eccentric -7.4%	63% Axial -32% Eccentric -11%	83.3%	18%
3.	Other –symptoms	Defective vision, pupillary abnormalities and optic atrophy-12.9%		Defective vision (33.3%) restricted mobility(25), Ptosis (8.3%)	Pain-15%

Table 12: Comparative analysis of percentage distribution of orbital tumors in various studies

Histopathological diagnosis	Present study	Tanushree et al, 2015	Boraina et al, 2007	Radha et al, 2005	Shields et al, 2002
Dermoid cysts	33.3%	33%	12.5%	4.2%	2%
Lacrimal gland tumors	16%	10	-	8.4%	9%
Secondaries	11%	4%	44%	4.2%	11%
Lymphomas	9.2%	4%	16%	33.3%	11%
Cavernous hemangiomas	9.2%	10%	4%	16.6%.	6%
Meningiomas	7.4%	6%	-	-	4%

Table 13: Percentage distribution of lacrimal gland tumors with epithelial and nonepithelial origin

S.No.	Study	Percentage of lacrimal gland tumors with epithelial origin	Percentage of lacrimal gland tumors with non epithelial origin
1.	Present study	77.7%	22.3%
2.	Tanushree et al, 2015	100%	-
3.	Radha et al, 2005	100%	-
4.	Jasna et al, 2004	100%	-

Table 14: Comparative analysis of orbital lymphomas in various studies

S. No.	Study	No.of cases	Histopathological diagnosis	Age group
1.	Present study	9.2%	3 cases –DLBCL 2 cases-Follicular lymphoma low grade	47-62 years
2.	Tanushree et al, 2015	4%	Non Hodgkin lymphoma	Older age group
3.	Boriana et al, 2007	3.5%	Non Hodgkin lymphoma	-
4.	Radha et al, 2005	20.8%	80% were MALTOMAS	54-72 years
5.	Jasna et al, 2004	16%	All typed as Diffuse B cell phenotype	71-80 years
6.	Bastola et al, 2013	37.5%	Non Hodgkin lymphoma	-

Table 15: Reporting of pattern of Follicular lymphoma

S. No.	Reporting of pattern	Proportion follicular
1.	Follicular	>75%
2.	Follicular and diffuse	25-75%
3.	Focally follicular	<25%
4.	Diffuse	0%

Table 16: Comparative analysis of secondaries in various studies

S. No.	Study	No. of cases of Direct extension	No. of cases of Metastatic deposit	Total no. of cases and percentage	Age group
1.	Present study	5	1	6 cases (11%)	5 cases-5th to 7th decades
2	Boriana et al, 2007	9	2	11 cases (44%)	-
3.	Tanushree et al, 2015	1	1	2 cases (4%)	-
4.	Radha et al, 2005	1	-	1 case (4.2%)	8th decade
5.	Jasna et al, 2004	3	-	3 cases (12%)	2 cases-8th decade 1 case-7th decade
6.	Shields et al,2004	-	-	142 cases (11%)	Older age group

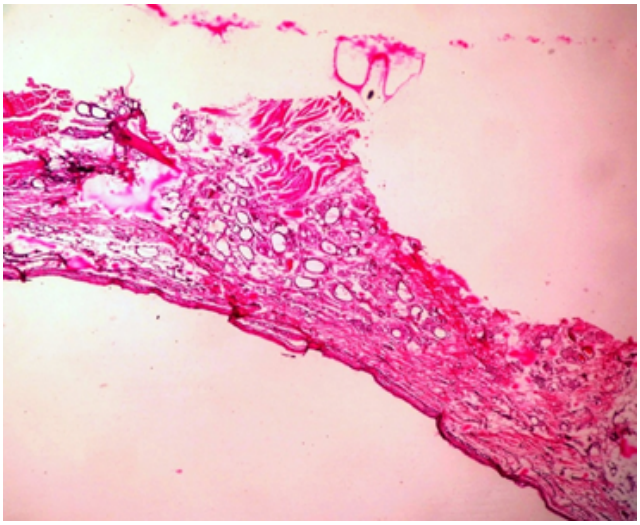


Fig. 2: H&E 10X Dermoid cyst lined by stratified squamous epithelium and subepiithelium shows dermal appendages

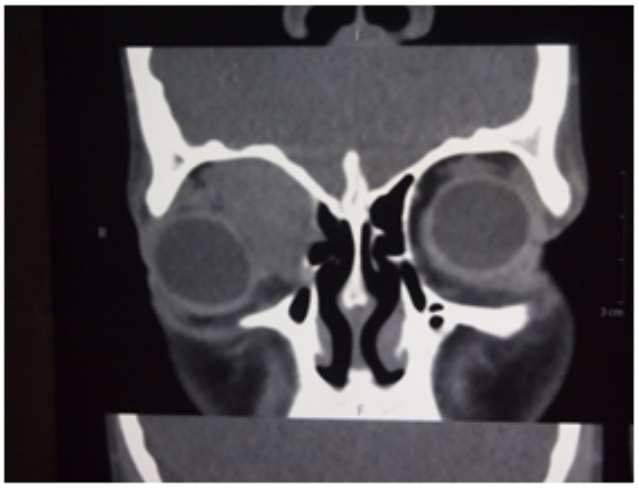


Fig. 3: Embryonal Rhabdomyosarcoma: 15 year old patient presented with right eye proptosis. CT scan image showing soft tissue mass in the superomedial orbital space

5. Discussion

Most common pediatric tumors are Dermoid cysts, Capillary hemangiomas and RMS.[8,9] Most common adult tumors are Lymphoid tumors, Cavernous hemangiomas and Meningiomas.[5,6] Most lymphomas are low-grade B-cell lymphoma, with extranodal marginal zone lymphoma (ENMZL) as the most common type.[12]

In the present study out of 54 cases, benign tumors (70.3%) were found to be common than malignant tumors (29.6%). In Tanushree et al 2015[13] out of 48 patients, benign tumors accounted for 90% and malignant 10% of the cases. In Radha et al 2005[14] out of 24 patients studied 70.8% accounted for benign tumors and 29.2% malignant tumors. Thus correlating with Tanushree et al, Radha et al and Jasna et al. But in Boriana et al 2007[15] malignant tumors were found to more common.

Among the benign tumors, dermoid cyst was found to be most common(33.3%). This correlated with Tanushree et al.[13] Other studies show varied percentages. 88% of the dermoid cysts were seen in 1^{st} and 2^{nd} decade. Dermoid cyst is the most common orbital cystic lesion in children. It accounts for over 40% of all orbital lesions of childhood and for 89% of all orbital cystic lesions of childhood that come to the biopsy or surgical removal.[16]

In the present study, maximum number of patients were in 1^{st}, 2^{nd} and 5^{th} decades. A total of 24 (44.4%) orbital tumors were seen in 1^{st} and 2^{nd} decades. Of

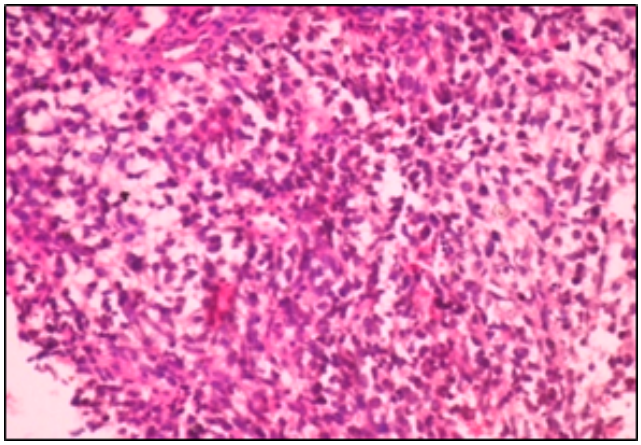


Fig. 4: H&E 10X Embryonal RMS: composed of primitive mesenchymal cells, small, polyhedral, oval and spindle cells with lightly amphophilic cytoplasm and cells with more cytoplasmic eosinophilia and elongated, "tadpole," strap," or "spider" cells and myxoid appearance of stroma

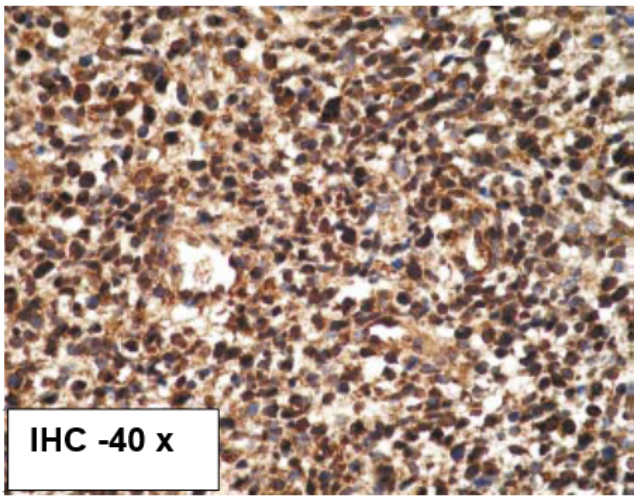


Fig. 5: IHC of Embryonal RMS showing myogenin positivity

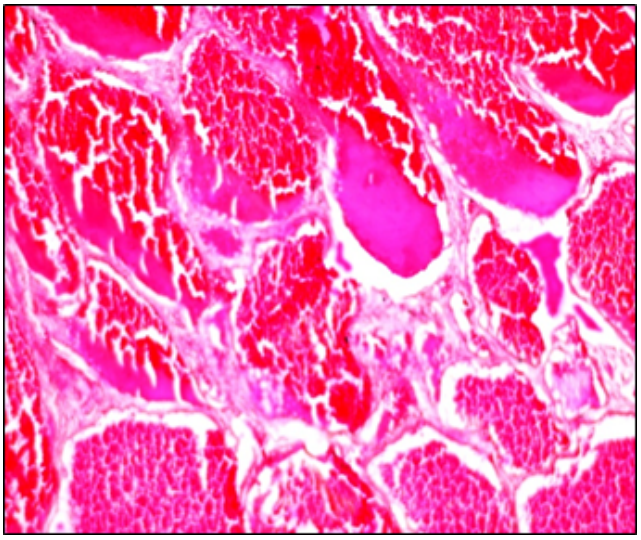


Fig. 6: H&E 40x Cavernous Hemangioma: vascular spaces lined by endothelium and filled with blood

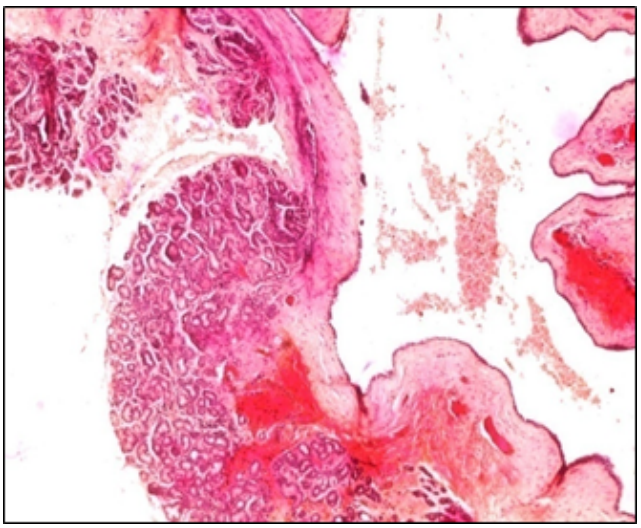


Fig. 7: H&E 10x –Lacrimal ductal cyst lined by cuboidal cells

these, 21 cases (87.5%) were benign and 3 cases (12.5%) were malignant. Thus benign tumors were the majority in the first and second decades. Of the benign tumors, dermoid cysts predominated. In the present study, among the malignant tumors in childhood age group, 2 cases of embryonal rhabdomyosarcoma were diagnosed based on the morphology and 1 case of secondary with direct extension from ocular retinoblastoma. Of the 2 cases of RMS, 1 case was seen in 13 year old male patient and other case was seen in 15 year old male child. Both cases presented with proptosis.

In Jasna et al 2004[17] and Saha somnath et al 2002[1] also, there were 2 cases of rhabdomyosarcoma. In Saha somnath et al 2002[1] pediatric tumors were rare (22.6%). Most of the orbital tumors of childhood are distinct from the tumors that occur in adults. The most common orbital malignancy in childhood is Rhabdomyosarcoma.[18] However, the majority of pediatric orbital tumors are benign and usually include developmental cysts, capillary hemangioma and hamartoma.[19]

Rhabdomyosarcoma is malignant soft tissue sarcoma that recapitulates the phenotypic and biological features of skeletal muscle.[20]RMS is the most common soft tissue sarcoma in children.[21,22] About 42% of patients with orbital rhabdomyosarcoma are aged 5–9 years; they present with proptosis or dysconjugate gaze.[23] Embryonal rhabdomyosarcomas constitute the most common subtype of rhabdomyosarcoma of the orbit by far. A special subtype of embryonal rhabdomyosarcoma is botryoid

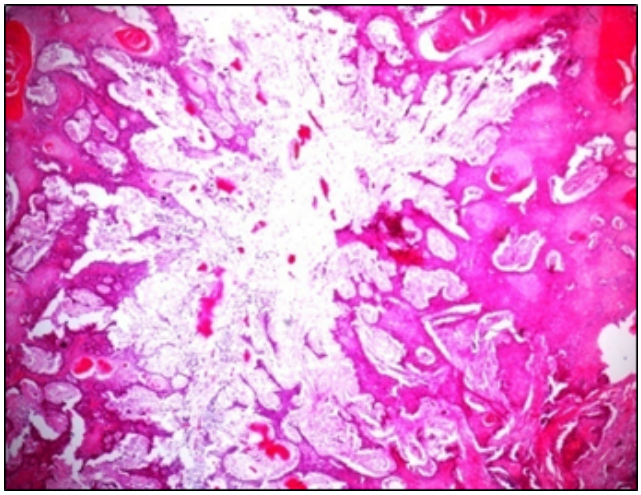


Fig. 8: H&E 10X Pleomorphic adenoma with epithelial component showing tubules and sheets and mesenchymal component showing chondromyxoid material

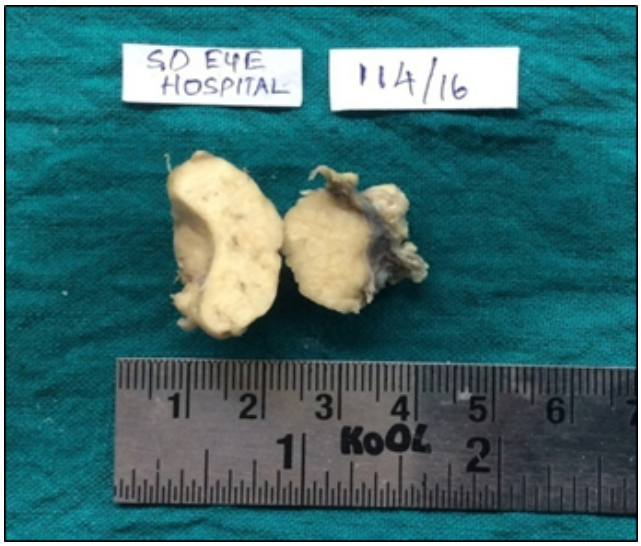


Fig. 9: Gross image of Adenoid Cystic Carcinoma: lacrimal gland mass measuring 2x1 cm. Cut surface showing grey white areas.

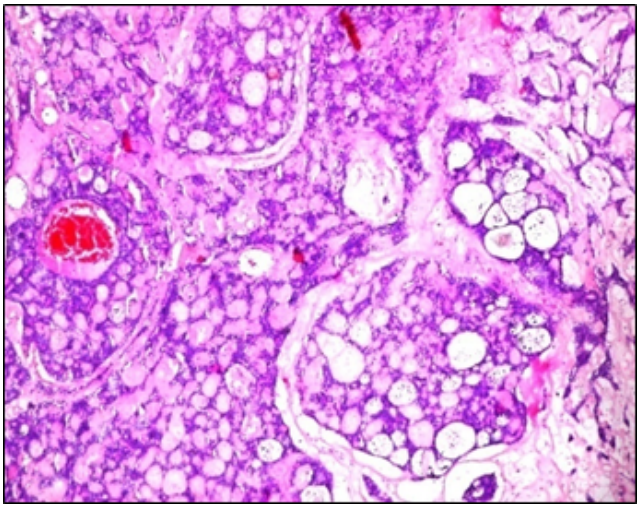


Fig. 10: H&E 10X Adenoid cystic carcinoma showing cribriform pattern with small hyperchromatic cells

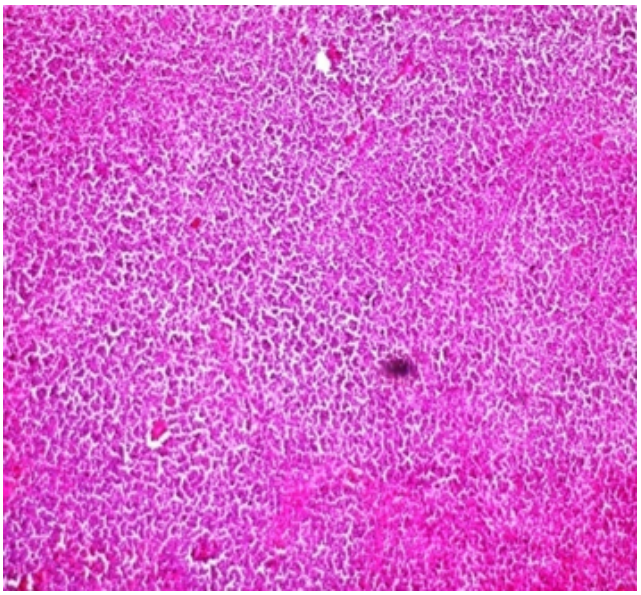


Fig. 11: H&E 10X DLBCL Diffuse sheets of monotonous cell population

rhabdomyosarcoma (involving the conjunctiva).[24] Overall 5-year survival rates have improved to more than 80% with the combined use of surgery, radiation therapy, and chemotherapy in patients with localized orbital disease.[23–25]

In this study, lacrimal gland lesions accounted for 12 cases (22.2%). Out of 12 cases, 3 cases were lacrimal ductal cyst, 4 cases pleomorphic adenoma, 3 cases adenoid cystic carcinoma and 2 cases non hodgkins lymphoma. In Tanushree et al[13] of 5 cases,4 cases were pleomorphic adenoma and 1 case of adenoid cystic carcinoma. In Radha et al[14] out of 2 cases, 1 case of pleomorphic adenoma and 1 case of adenoid cystic carcinoma. In Jasna et al 2004 out of 25 cases, 2 cases were pleomorphic adenoma. Thus all were of epithelial origin in Tanushree et al, Radha et al and Jasna et al. Lacrimal gland lesions represent 5–25% of orbital tumors, and the proportion in the literature that are epithelial in origin, range from 23% to 70% of biopsied cases.[26–29]

In the present study, among the lacrimal gland tumors, benign tumors were 44.4% and malignant 55.5%. Thus malignant tumors were slightly common than benign tumors. The age group ranged from 28 to 62 years. Thus in Radha et al[14] among the lacrimal gland tumors 50% benign and 50% were malignant.us the present study shows similar observation as seen in Radha et al study. In Tanushree et al[13] mong the lacrimal gland tumors, 80% were benign and

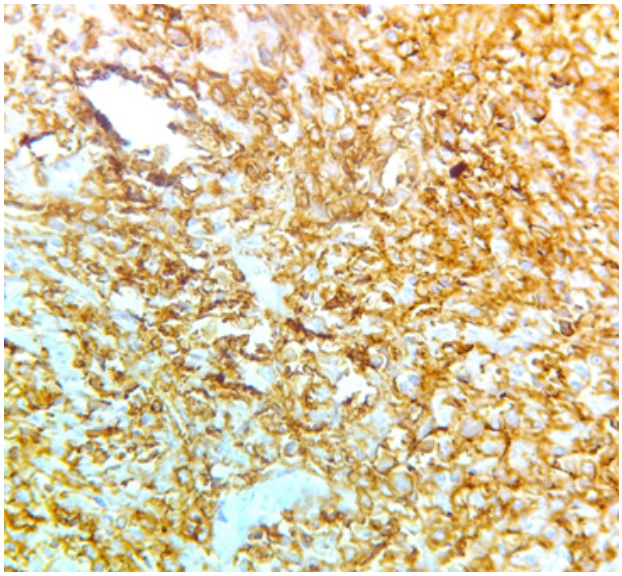


Fig. 12: IHC 40X DLBCL CD 20 Positive B cells

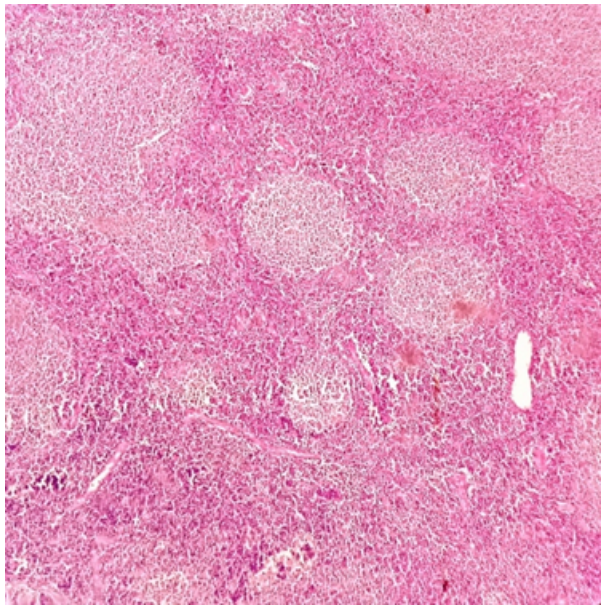


Fig. 13: H&E 10X Follicular Lymphoma: closely packed follicles and decreased interfollicular area

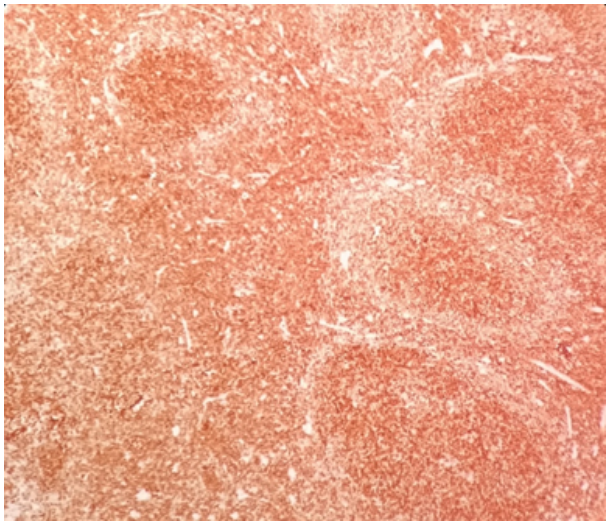


Fig. 14: IHC 10X Follicular Lymphoma: BCL2 Positive germinal center

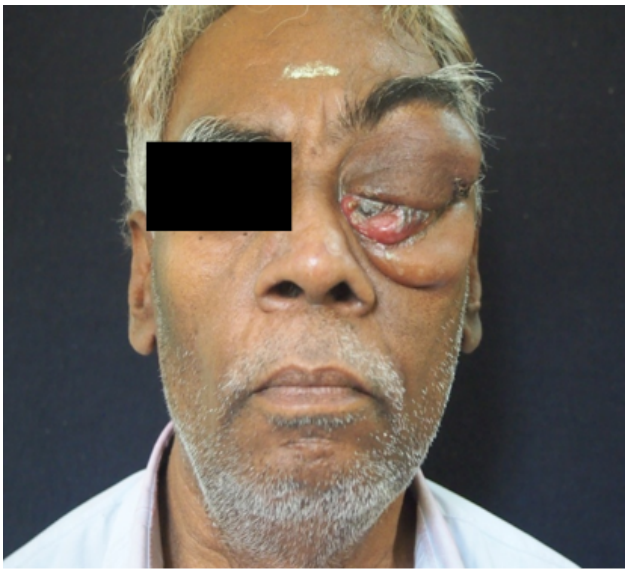


Fig. 15: Sinonasal Adenocarcinoma with orbital extension: 62 years male patient presenting with proptosis

20% were malignant. Thus, benign tumors were in much greater number in Tanushree et al. In Jasna et al, only benign tumors were seen.

Shields et al[17] in 2004 concluded that approximately 55% are benign and 45% are malignant. In Perez et al,[30] out of 18 cases, 66.7% were Adenoid cystic carcinoma, 27.8% were pleomorphic adenoma and 5.5% were carcinoma ex pleomorphic adenoma. This indicates a different distribution of benign and malignant epithelial lesions with a higher rate of malignancy.

In the present study, Pleomorphic adenoma was found to be most common benign epithelial tumor. Similar observation was found in Tanushree et al,[13] Radha et al,[14] Jasna et al.[18] Although pleomorphic adenomas are histologically benign, incomplete excision will lead to recurrences and even malignant transformation. Hence, when suspected, lateral orbitotomy is mandatory and entire tumor must be excised enbloc.[31]

In the present study,out of 9 cases of lacrimal gland tumors, there were 3 cases of adenoid cystic carcinoma. It was found to be most common malignant epithelial tumor. In Radha et al.[14]Whereas in Tanushree et

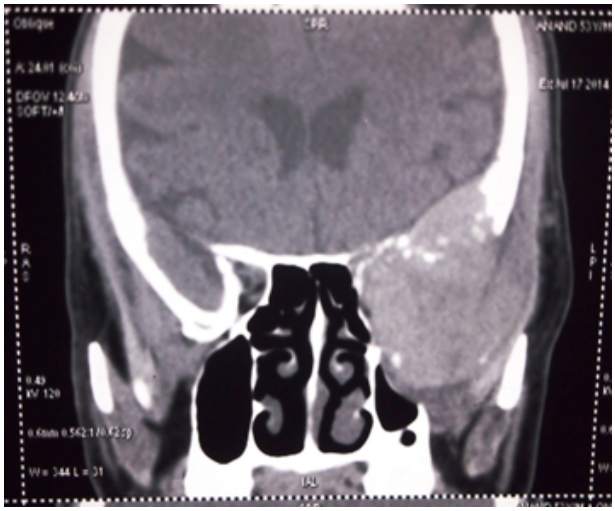


Fig. 16: CT scan image of the patient with Sinonasal Adenocarcinoma showing extension of the tumor to the orbital region

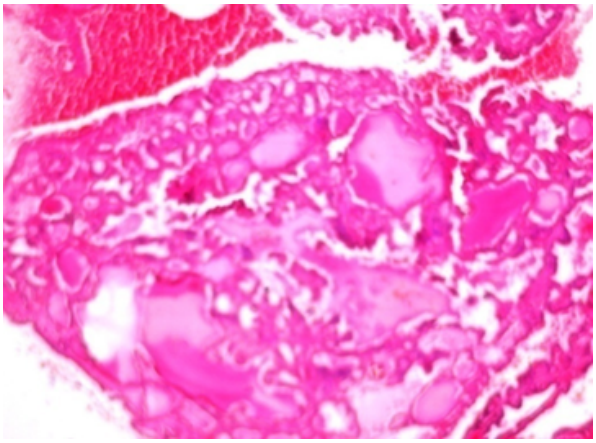


Fig. 17: H&E 40X of Sinonasal Adenocarcinoma showing glandular pattern lined by neoplastic tumor epithelial cells

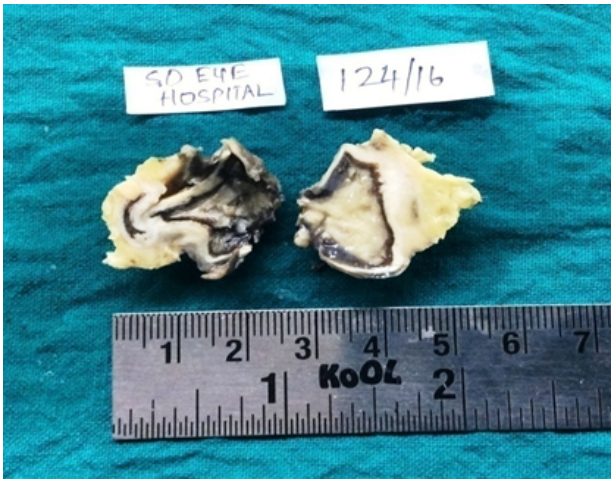


Fig. 18: Gross image of Orbital extension of Conjunctival Squamous Cell Carcinoma: Irregular mass measuring 2x2 cm. Cut surface shows grey yellow to grey black areas

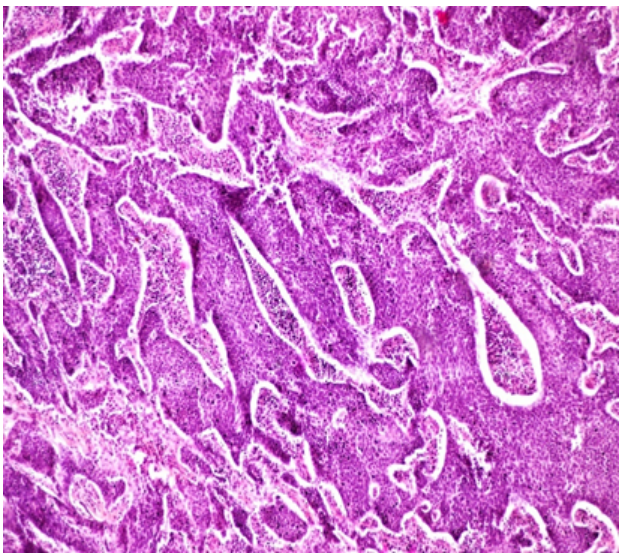


Fig. 19: H&E 40 X Conjunctival Squamous Cell Carcinoma: slands and nests of polygonal cell with abundant cytoplasm and pleomorphic nucleus

al,[13] mucoepidermoid was found to be common. In Jasna et al,[18] there were no malignant cases.

In the present study, all the three cases of adenoid cystic carcinoma were seen in 3rd decade (21-30). Thus it is seen in young adults.Whereas in Radha et al[14] it was seen in older age group (6th decade). Lacrimal gland ACC generally affects young to middle aged patients (range 6.5–79 years).[3]

The outcome for patients with ACC in the lacrimal gland is reportedly poorer than that with ACC in the salivary gland.[32] Thorough histologic examination of ACC in order to detect any solid or dedifferentiated component and evaluation of the margins as well as perineural invasion are important. Major cause of death is intracranial spread hastened by perineural invasion. Treatment is exenteration and post- operative radiotherapy.[14]

In the present study, out of 9 cases of lacrimal gland tumors, 2 cases (22%) were non epithelial. Of the non epithelial tumors, all were non hodgkins lymphomas. After running a panel of markers, all were confirmed as Diffuse Large Bcell Lymphoma. Thus DLBCL was found to be most common type of lymphoma among lacrimal gland tumors. In Sjo LD, et al[33] out of 15 primary lacrimal sac lymphomas, five cases (33%) were diffuse large B cell lymphomas.

The Ocular Adnexal Lymphoma (OAL) represents malignant lymphoid neoplasms, which can develop as primary or secondary tumor manifestations. Lymphoma

localized in the ocular region (eyelid, conjunctiva, lacrimal drainage system, orbit, and intraocular) is one of the most common orbital malignancies.[17,34] 78% of lymphomas involving the ocular adnexa are primary, whereas 22% are secondary.[12] OAL is primarily a disease of older adults (peak age 65 years) with a slight female preponderance.[33] The deep orbital tissue is considered as true extranodal/nonfunctional mucosa-associated tissue (ENMZL) whereas the lacrimal gland, the conjunctiva, and the lacrimal sac are considered as functional mucosa-associated tissues.[35] Mucosa associated lymphoid tissue lymphoma (MALT) is therefore not synonymous with ENMZL and is characterized by mucosal location and lymphoepithelial lesions. Lymphoma arising in the lacrimal sac was surprisingly predominantly DLBCL.[33]

FL are BCL-2 and CD10 positive in 85% of cases: BCL-2 is quite specific versus germinal centers. CD10 is quite specific versus other small B lymphomas. They also stain positive for BCL-6 and show expansion of follicular dendritic cell networks staining positive for CD21, CD23, and CD35. They are graded as low and high grade.

LOW GRADE -0 to 15 centroblasts per high power field.

HIGH GRADE –more than 15 centroblasts per high power field.[36]

In the present study, there were 6 cases (11%) of secondaries. Of these, 5 cases were of direct extension and 1 case of metastatic deposit. Of the 5 cases of direct extension, there was 1 case each of sinonasal adenocarcinoma extending from paranasal sinus, 1 case of squamous cell carcinoma extending from conjunctiva, 1 case of retinoblastoma extending from eye and 2 cases of choroidal malignant melanoma. Metastatic deposit was from breast carcinoma.

In the present study, out of 54 cases, one case of solitary fibrous tumor is seen on histopathology.IHC revealed CD 34 positivity. Hemangiopericytomas and Solitary fibrous tumors are uncommon neoplasms found in orbit. Local recurrences of SFT are possible and usually follow an incomplete initial excision.[37] In Furusato et al 2010,[36] Forty-one fibroblastic orbital tumors, originally diagnosed as hemangiopericytomas (n = 16), fibrous histiocytomas (n = 9), mixed tumors (hemangiopericytomas/fibrous histiocytoma) (n = 14), and giant cell angiofibromas of orbit (n = 2) between 1970 and 2009, were retrieved, the Ophthalmic Registry, at the Armed Forces Institute of Pathology. Slides and clinical records were reviewed, analyzed, and compared. IHC was performed for CD34, CD99, Bcl-2, Ki-67, and p53 and all cases were reclassified as solitary fibrous tumor (41/41).

In Furusato et al 2010,[36] overlapping features with soft tissue giant cell fibroblastoma were observed. IHC revealed positivity for CD34 in all cases (100%), p53 in 85%, CD99 in 67.5%, and Bcl-2 in 47.5%. Although Ki-67 labeling was seen in all cases, it ranged from less than 1% in 54.3% of cases to 5% to 10% in 20% of cases.

In the present study, 4 cases(7.4%) of Meningioma are seen and 2 cases(3.2%) of Schwannomas (confirmed on IHC–S100 positive). All the patients presented with axial proptosis and other findings such as diminution of vision, defective pupillary reaction and optic atrophy. Of these, 1 case was seen in 1^{st} decade and 4^{th} decade each and 2 cases seen in 5^{th} decade. All the cases presented as intraconal mass. Of these, 3 cases diagnosed as Meningothelial Meningioma, WHO grade 1 and 1 case as Angiomatous Meningioma WHO grade 1. In Tanushree et al 2015,[13] there were 3 cases (6%) of meningioma and 2 cases (4%) of schwannoma. All the patients had axial proptosis and other findings such as diminution of vision, defective pupillary reaction and optic atrophy. Thus correlating with Tanushree et al study. In Jasna et al 2004,[18] there were 4 cases(16%) of meningioma. In Saha Somnath et al 2002,[1] there was 1 case (3%) of schwannoma.

Meningiomas can arise from the optic nerve or extend into the orbit from adjacent structures.[38]The age at presentation of patients with optic nerve meningiomas is typically younger than for intracranial meningiomas.[39] A number of meningioma subtypes have been proposed through the years, and the current (2016) World Health Organization (WHO) classification includes 15 named entities.[40]

Intraorbital meningiomas were most frequently of the meningothelial or transitional subtypes and were WHO grade 1. Significant changes that affect tumor grade are further classified (1) meningiomas showing central nervous system invasion as grade II rather than grade III (2) meningiomas with 4 or more mitotic figures per 10 high-power fields (HPFs) as grade II (3) Clear Cell and Chordoid Meningiomas as grade II (4) Papillary and Rhabdoid Meningiomas as grade III, and (5) tumors with 20 or more mitotic figures per 10 HPFs as grade III.[41]

In Deepali Jain et al 2010,[42] a total of 51 intraorbital meningiomas were reviewed. Meningothelial type was most common (25 of 51 tumors; 49%). Most (47 of 51; 92%) of the tumors were WHO grade I. They concluded that Intraorbital meningiomas were most frequently of the meningothelial or transitional subtypes and were WHO grade I.

Schwannomas are benign peripheral nerve sheath tumors derived from Schwann cells. Nuclear atypia with hyperchromatic pleomorphic nuclei can be observed in Ancient Schwannoma.[43]Mitotic figures with counts of 6 mitoses per 10 HPF can be observed. True malignant change of Schwannoma is very rare.

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None.

7. Conflict of Interest

None.

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