Ophthalmol Ina 2023;Supplement Ed.: 157-164

ORIGINAL ARTICLE

REVEALING ORBITAL TUMORS: 2-YEAR EVALUATION IN CLINICAL PROFILE, HISTOPATHOLOGICAL FEATURES AND SURGICAL TECHNIQUE

Silmi Lisani Rahmani[1], Agus Supartoto[1], Purjanto Tepo Utomo[1], Banu Aji Dibyasakti[1], Datu Respatika[1], Irene Titin Darajati[1]

[1]Department of Ophthalmology, Faculty of Medicine, Public Health and Nursing Gadjah Mada University, Sardjito General Hospital, Yogyakarta, Indonesia

Email: silmilisani@gmail.com

ABSTRACT

Introduction: *Incidence of orbital tumors is relatively low, but the delay in diagnosis, even if benign, can lead to vision loss and deformity.*

Purposes: *This study aims to review clinical and demographic information of orbital tumors in Sardjito General Hospital, Yogyakarta.*

Method: *We retrospectively reviewed clinical data of 138 patients who diagnosed with orbital tumors during July 2021 - March 2023. Data extracted included clinical characteristics, operative procedures, and histopathologic characteristics of orbital tumor.*

Result: *There were 138 orbital tumors managed at hospital during 2-year period. Among 138 patients, 82 patients (59%) were female and 56 patients (40%) were male. Twenty-one patients (15%) were in childhood age and 117 patients (84.8%) were in adulthood age. The common clinical manifestations were proptosis (83.3%) and decreased vision (10.4%). The main procedure used was anterior orbitotomy (54.34%). Eighty-eight patients (63.7%) have primary tumors and 51.4% of the orbital tumors were malignant. The most cases in children were benign tumors (61.9%) meanwhile in adults were malignant (54.7%). The main orbital tumor in childhood age was a Non-Hodgkin Lymphoma (NHL) (19%) and cyst (14.3%). On the other hand, in adult patient, the most common orbital tumor was meningioma (18%) and NHL (15.7%).*

Conclusion: *Orbital tumors are challenging group of tumors with proptosis become the most frequent clinical characteristic that found. The orbital tumors in childhood and adulthood differ significantly in their incidence and tumor type with the percentage of malignant tumors increasing with age.*

Keyword: *orbital tumor, orbital neoplasm, orbitotomy.*

INTRODUCTION

The orbit is a compartment with a fixed cavity and complex structure comprised of bone, fat, extraocular muscles, vascular, nerve, connective tissue, and glandular. Numerous benign and malignant tumors may cause proptosis, and decreased vision, and affect extraocular muscle function by compressing the adjacent structure.(1) Orbital tumors are relatively rare compared to other ocular disorders, occurring at an estimated incidence rate of 2.02 per million person-years in the USA, with a diverse clinical presentation from minor cosmetic conditions to visual loss and plenty of cases are malignant and life-threatening.(2,3)

According to cellular origin, the orbital tumor can be divided into primary, secondary, and metastatic tumors. Primary tumors are lesions that originate from the orbit and most often are managed surgically; secondary include lesions that extend to involve the orbit from neighboring structures, most often malignant, and similar to metastatic tumors often managed with multiple modalities.

However, the frequencies and types of orbital tumors are different depending on geographic area, ethnicity, age, and sex.(4) For example, the dermoid cyst is the most common orbital mass in children, accounting for 50% of all orbital tumors in the pediatric population, this tumor is rarely found in the senior adult population. In addition, there is some evidence that senior adults may manifest an array of more malignant orbital tumors than middle-aged adults or children. (3,5)

The purpose of this study was to determine the incidence, demography, and clinical features of orbital tumors that were diagnosed histopathologically in Sardjito General Hospital.

METHODS

A retrospective chart review was conducted on patients diagnosed with orbital tumors during two years in the Division of Reconstructive, Oncology, and

Oculoplastics, Department of Ophthalmology at Sardjito General Hospital. A designed data collection sheet was used and completed during a detailed review of the medical records including the demographic information (age and sex), initial presentation, clinical examination findings, surgical approach (enucleation, exenteration, orbitotomy anterior, lateral, posterior), tumors characteristics (histopathology, primary, secondary and metastasis).

All patients with a diagnosis of orbital tumors that were determined using preoperative computed tomography (CT) and underwent surgery and histological examination in Sardjito General Hospital from 2021 to 2023 were included. Lesions confined to the eyelid, conjunctiva, and intraocular lesions were excluded. However, lesions originating from the orbit and extending to involve the eyelid and conjunctiva were included in this study. We also included intraocular lesions with orbital involvement and metastatic lesions to the orbit.

Using a modification of orbital tumors published by Shield's study, the diagnosis was categorized under one of the major groups of lesions (cystic, vasculogenic, rhabdomyosarcoma, lymphoproliferative, histiocytic, fibrocytic, osseous/fibro-osseous/cartilaginous, peripheral nerve tumor, optic nerve tumors, infantile/embryonic, secondary, and metastatic).

Microsoft excel is used for the process of data entry and data coding. Clinical characteristics, socio-demographic, tumor origin, type of operative procedure, and

Ophthalmol Ina 2023;Supplement Ed.: 157-164

histopathological results were analyzed using descriptive statistics (mean, percentage, and frequency) and presented in table.

RESULTS

We identified 138 patients with orbital tumors and histopathologically verified tissue diagnosis who were seen and managed during the period encompassed by this study (2 years). The total subjects included in this study were 56 males (40.58%) and 82 females (59.42%). The age ranged from 1 to 84 years, with the mean age at presentation of 44.2 years. The number of pediatric patients was 21 (age 0-19 years old) and adults (>19 years old) 117 patients based on *World Health Organization* (WHO) criteria.

Proptosis was the most common encountered presenting symptom in 135 (83%) patients, followed by decreased vision in 17 (10.49%) patients, chemosis (2.47%), lagophthalmos (2.47%), and pain (1.23%). The frequency of presenting complaints in patients with orbital tumors is depicted in figure 1.

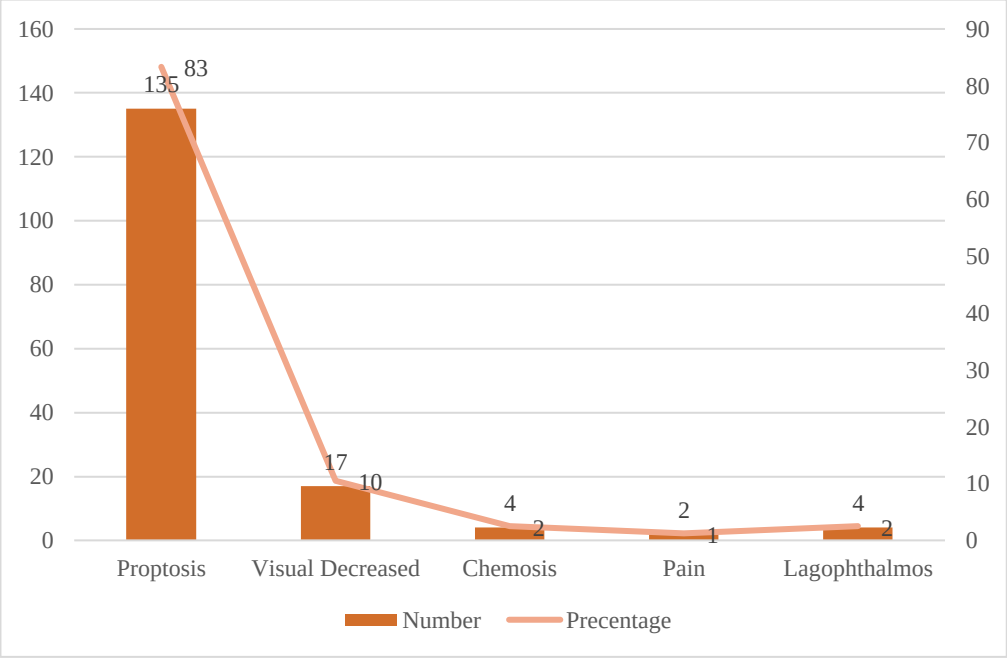

Figure 1. Frequency of the presenting complaints of 138 consecutive patients with orbital tumors

The most common orbital tumors among the 138 patients were primary tumors (63.77%) followed by secondary tumors (34.05%) and metastatic tumors (2.17%). The surgical approach that was performed either for tumor removal or excisional biopsy including anterior

orbitotomy 75 (54.34%), medial 3 (2.17%), lateral 21 (15.21%), posterior 7 (5.07%), exenteration 32 (23.18%) (Table 1).

Table 1. Demographic characteristics of patient with orbital tumor

Specificity	Patients number (%)
Sex	
Male	56 (40.58%)
Female	82 (59.42%)
Age Range	
Children (0-19 yo)	21 (15.2%
Adolescent (>19 yo)	117 (84.78%)
Malignancy	
Benign	66 (47.8%)
Malignant	71 (51.4%)
Origination	
Primary	88 (63.77%)
Secondary	47 (34.05%)
Metastatic	3 (2.17%)
Surgical Approach	
Anterior	75 (54.34%)
Medial	3 (2.17%)
Lateral	21 (15.21%)
Posterior	7 (5.07%)
Exenteration	32 (23.18%)
Complication	
Lagophthalmos	1 (0.7%)
Decrease in Vision	5 (3.3%)
Strabismus	2 (1.3%)

The distribution of tumors according to their histology was as follows: most cases in children were benign tumors (61.9%) followed by malignant tumors (61.9%) meanwhile in adults were malignant (54.7%) followed by benign (45.2%) (Figure 2). Among children, Non-Hodgkin Lymphoma (NHL) ranks first at 19% followed by fibrous dysplasia and epidermoid cyst at 14.3%. Hamartoma ranks third among tumors in children at 9.5%. Among adults, the distribution was as follows: meningioma (17.9%), NHL (15.4%), and SCC (13.7%) are among the highest percentage followed by adenoid cystic carcinoma (6%), epidermoid cyst (5.1%) and others. The histopathologic classification of the orbital lesions is shown in table 2.

Ophthalmol Ina 2023;Supplement Ed.: 157-164

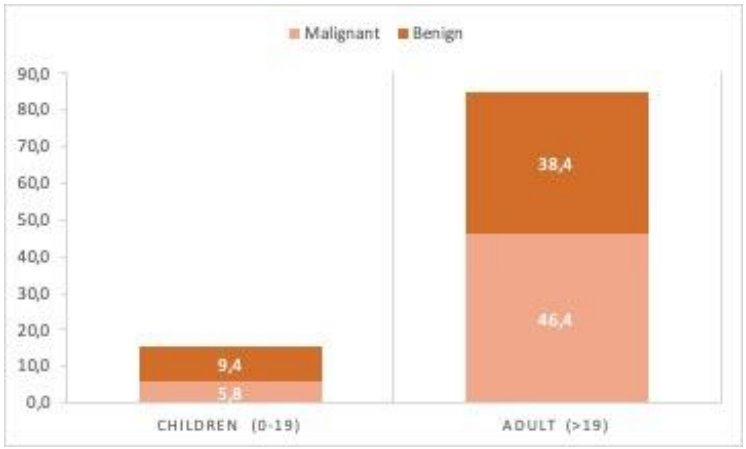

Figure 2. Distribution by age and malignancy

Table 2. Distribution of Histopathology of Orbital Tumor

	Children n (%)		Adult n (%)	
Malignancy				
Malignant	8 (38.09%)		64 (54.7%)	
Benign	13 (61.9%)		53 (45.2%)	
Tumor Type				
	Neurogenic Tumor - 4 (19%)		**Tumor Limfatik – 20 (17%)**	
	Meningioma	1	NHL	18
	Neurofibroma	1	Lymphoid hyperplasia	2
	Neuroblastoma	1		
	Schwannoma	1		
			Tumor Neurogenik – 26 (22.2%)	
	Lymphatic Tumor – 4 (19%)		Meningioma	21
	NHL	4	Neurofibroma	3
			Schwannoma	2
	Vascular Tumor – 1 (4.7%)			
	Hemangioma Cavernosa	1	**Tumor Epitelial – 24 (20.5%)**	
			SCC	16
	Mesenchymal Tumor– 1 (4.7%)		BCC	5
	Rhabdomyosarcoma	1	Melanoma Maligna	1
			Inverted papilloma	2
	Lacrimal Gland – 1 (4.7%)			
	Adenoid Cystic Carcinoma	1	***Developmental* – 1 (0.8%)**	
			Kista	1
	Epithelial Tumor – 1 (4.7%)			
	Epitheloid Sarcoma	1	**Tumor Vaskular – 6 (4.27%)**	
			Hemangioma Cavernosa	5
	Osseus, Fibro-osseus, Cartilaginous – 3 (14%)		**Tumor Glg Lacrimal – 19 (16.2%)**	
	Fibrous dysplasia	3	Pleomorfik Adenoma	4
			Adenoid Cystic Carcinoma	7
	Developmental – 6 (28.5%)		Adenocarcinoma	6
	Cyst	4	Mucinous carcinoma	1
	Hamartoma	2	Mucoepidermoid carcinoma	1
			Tumor Mesenkimal – 3 (2.56%)	
			Lipoma	1

	Chondrosarcoma	2
	Pseudotumor – 2 (1.7%)	
	Pseudotumor	2
	Developmental – 12 (10%)	
	Cyst	12
	Osseus, fibro-osseus, cartilaginous – 6 (5.12%)	
	Fibrous dysplasia	4
	Giant Cell Tumor	1

SCC: squamous cell carcinoma, NHL: Non-Hodgkin Lymphoma, BCC: Basal Cell Carcinom

DISCUSSION

The main function of the orbit is to give support and protection and protection to the eyeball and its structures. The volume of the orbital cavity of an adult is approximately 30cc. The border of the orbit measures about 40mm horizontally and 35mm vertically. Due to the fact it is a compartment with a fixed cavity that cannot be widened, SOL in the region may compress the adjacent structure. (1)

Clinical presentation of primary orbital malignancies often reflects the site of the lesion due to the effects on nearby tissues and structures. The most frequent presenting symptoms of space-occupying orbital mass in adults in progressive proptosis. In children, more rapid or sudden-onset proptosis can be seen and is highly suggestive of malignancy. The direction of the globe displacement can help localize the tumor site, such as inferonasal displacement from the lacrimal fossa lesion, and superior displacement from a lesion originating from the maxillary sinus. In contrast, more extensive lesions involving the extraconal and/or intraconal space such as RMS can present with a generalized proptosis.(6)

Other clinical manifestations result from the compressive effect of tumors on nearby structures due to the rigid, confined space of the orbit. As such, orbital tumors can also be classified based on their location within the orbit as intraconal, extraconal, and intracanalicular lesions. Intraconal tumors can exert a mass effect on the globe, optic nerve, and extraocular muscles. These patients tend to present with vision loss, axial proptosis, and diplopia secondary to decreased ocular motility. Extraconal lesions can present similarly; however, vision loss is usually seen later in the course. Intracanalicular tumors, such as optic nerve meningiomas or gliomas, often display minimal or delayed proptosis but can cause early vision loss with associated optic nerve head atrophy, swelling, and optociliary shunt vessels on funduscopic examination. (6)

Ophthalmol Ina 2023;Supplement Ed.: 157-164

In this study, proptosis was the most common encountered presenting symptom in 135 (83%) patients. This is consistent with the previous study by Bagheri et alwith proptosis and lump sensation being the most common clinical presentation in 42.4% and 31.7% of patients, respectively.1 Study by Demirci et al also reports palpable mass followed by proptosis (18%) as the main clinical feature.5 Decreased vision (10.49%) was the second most common clinical manifestation in our study. Visual impairment is a result of the optic canal invasion, optic nerve compression, periorbital tissue infiltration, or orbital apex invasion by the tumor. (7)

Because of the varied locations of primary orbital neoplasms, a number of surgical approaches are required to approach and remove them. The goal of these approaches is to provide adequate access without undue risk to important structures (optic nerve and its blood supply, sensory and motor nerves, extraocular muscles). Generally speaking, these approaches can be subdivided according to the orbital wall entered: lateral, medial, superior, or inferior orbitotomies. When surgeons select surgical approaches, in addition to the location of the tumor, other factors, such as the size of the lesion, goal of the surgery (biopsy,debulking, or gross-total excision), and the characteristics of the tumor, must be considered.(8)

The main procedure used was anterior orbitotomy (54.34%) followed by lateral orbitotomy (15.21%). Exenteration and enucleation is indicated for primary malignant orbital tumors or secondary tumor invasion to orbit from eyelid and intraocular tumors which may be life-threatening and those which does not respond to other conservative modes of treatment like chemotherapy or radiation. (9)

In this study, the most common orbital tumors among the 138 patients were primary tumors (63.77%) followed by secondary tumors (34.05%) and metastatic tumors (2.17%). This result is in line with a previous study by Bagheri et al that showed primary lesions were 11.1 times more common than secondary and metastatic lesions. The age-specific incidence of a primary malignant orbital tumor is approximately a 2/million population until the sixth decade of life when it increases to a 4/million population in those older than age 60 years and further increases to 10/million population in those older than age 80 years.(5) Secondary tumors were dominated by the result of invasion from the periorbital spaces such as paranasal sinuses.

The final histopathological diagnosis from a total of 138 patients was malignant (51.4%) and benign (47.8%). We categorized predominant benign and malignant lesions of the orbit according to age at presentation. Benign tumors (61.9%) were more prevalent in the children age group meanwhile malignant lesions (54.7%) were the most common tumor in the adult group. This result was consistent with a study that involved 375 patients by Bagheri et al that showed benign lesions were more prevalent in lower age groups and the frequency of malignant

lesions increased with age. (1) A ten-year epidemiological study in Japan also found that patients with malignant orbital tumor were significantly older than those with benign tumors.(2)

Major differences were found when comparing the histopathological type of orbital tumors in adults and those in children. Despite the fact that the relative incidence of tumors in children was mostly benign, Non-Hodgkin Lymphoma (NHL) become the greatest number of tumors in children. This result was followed by fibrous dysplasia and epidermoid cyst at 14.3%. NHL accounts for 5% of childhood cancers in children ages 0 to 14 and 7% of cancers in teens ages 15 to 19.(10) Furthermore, regardless of malignant tumors being the most common result found in adults, meningioma was more prevalent histopathological type in this study. This result was accompanied by NHL (15.4%), and SCC (13.7%) in the second and third rank. Meningiomas are the most common primary intracranial tumors that account for approximately 19% of primary intracranial tumors. However, orbital meningiomas account for only approximately 4-8% of total orbital lesions. Primary ectopic orbital meningiomas are very rare with a few case reports. Optic nerve meningioma (4%) was the most frequently encountered tumor in the central intraconal space.(11)

CONCLUSION

Orbital tumors are a challenging group of tumors with proptosis becoming the most frequent clinical characteristic that found. The orbital tumors in childhood and adulthood differ significantly in their incidence and tumor type with the percentage of malignant tumors increasing with age.

REFERENCES

1. Bagheri A, Ashtar-nakhaie P, Aletaha M, Kheiri B. Original Article A Survey on Orbital Space-Occupying Lesions during a Twelve-Year Period from a Referral Center in Iran. 2023;202–11.
2. Shimizu N, Oshitari T, Yotsukura J, Yokouchi H, Baba T, Yamamoto S. Ten-year epidemiological study of ocular and orbital tumors in Chiba University Hospital. BMC Ophthalmol. 2021;21(1):1–8.
3. Mombaerts I, Ramberg I, Coupland SE, Heegaard S. Diagnosis of orbital mass lesions: clinical, radiological, and pathological recommendations. Surv Ophthalmol. 2019;64(6):741–56.
4. Goto H, Yamakawa N, Komatsu H, Asakage M, Tsubota K, Ueda S ichiro, et al. Clinico-epidemiological analysis of 1000 cases of orbital tumors. Jpn J Ophthalmol. 2021;65(5):704–23.
5. 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.
6. Laplant J, Cockerham K. Primary Malignant Orbital Tumors. J Neurol Surgery, Part B Skull Base. 2021;82(1):81–90.
7. Wierzbowska J, Zegadło A, Patyk M, Rękas M. Spheno-Orbital Meningioma and Vision Impairment—Case Report and Review of the Literature. J Clin Med. 2023;12(1).
8. Park HJ, Yang SH, Kim IS, Sung JH, Son BC, Lee SW. Surgical treatment of orbital tumors at a single institution. J Korean Neurosurg Soc. 2008;44(3):146–50.
9. Saiju R. An unwanted disfiguring eye surgery: Exenteration. IP Int J Ocul Oncol Oculoplasty. 2021;7(2):109–11.
10. Siegel RL, Miller KD, Wagle NS, Jemal A. Cancer statistics, 2023. CA Cancer J Clin. 2023;73(1):17–48.
11. 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. Ophthal Plast Reconstr Surg. 2013;29(2):79–86.