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Original Research Paper

Pathology

ORBITAL TUMORS: A FIVE YEAR STUDY OF CLINICOPATHOLOGICAL SPECTRUM IN A TERTIARY CARE HOSPITAL

Dr. Sai Sridevi Krishna*

Assistant Professor, K.A.P.Viswanatham Government Medical College, Trichy. *Corresponding Author

ABSTRACT 140 cases of orbital tumors and 4 cases of pseudotumors were diagnosed among the 3409 biopsies examined during 2014-2019 five year period. The cases were analysed based on the patient age, sex, clinical presentation and tumor histology. The most common benign tumors were dermoid, lipoma, cavernous hemangioma and pleomorphic adenoma with a bimodal age presentation. Most of the tumors were benign. Non Hodgkin's lymphoma was the most common malignant tumor. Pseudotumors were common in the older age group. Role of immunohistochemistry was also analysed.

KEYWORDS : Orbital tumors, Chloroma, Solitary fibrous tumor

INTRODUCTION

Orbital cavity diseases require a multidisciplinary approach for its treatment. Due to its specific anatomic location and complex structure, they need a closer attention. The treatment includes meticulous insight into clinical presentation and evaluation radiologically. The World Health Organisation classification of orbital tumors include benign and malignant soft tissue tumors, hematologic and lymphoid tumors, pseudotumors of orbit and other primary tumors of orbit. Secondary and metastatic tumors form a separate entity in this classification[1]. The most common site of metastases include uvea followed by orbit[2].

Pseudotumors are inflammatory pathology and are histopathologically classified into three types: sclerosing, granulomatous and lymphoid[3]. They mimic lymphoma. Orbital tumors usually occurs unilaterally[4]. The most common presentation is proptosis.

The aim of this study is to present the clinical and pathological spectrum of orbital tumors and pseudotumors diagnosed by biopsy during 2014-2019. Out of 3409 biopsies received, 140 cases of orbital tumors and 4 cases of pseudotumors were diagnosed.

MATERIAL AND METHODS

140 cases of orbital tumors and 4 cases of pseudotumors diagnosed during the five years study period of 2014 -2019 were analysed for patient sex, age, histopathological features, clinical presentation and immunohistochemistry. Retrospective analysis was done using formalin fixed paraffin blocks. Immunohistochemistry was done using DAKO kit. Tables and figures were used to present the result.

RESULTS

Our study showed 140 orbital tumors and 4 cases of pseudotumors, out of which the following types were identified- Skin and soft tissue tumors (n=56), Vascular tumors(n=23), Neurogenic tumors (n=14), Lymphoid tumors (n=16), Lacrimal gland/sac tumors (n=24) and secondary extensions (n=11) (Table 1).

Table 1: Types of orbital tumors and pseudotumors diagnosed.

SKIN AND SOFT TISSUE	VASCULAR TUMORS	LACRIMAL GLAND/SAC TUMORS
Dermoid - 28	Capillary hemangioma -6	Pleomorphic adenoma - 14
Lipoma - 15	Cavernous hemangioma-12	Myoepithelioma - 1
Benign fibrous histiocytoma -2	Composite hemangioma -2	Mucoepidermoid carcinoma - 2
Solitary fibrous tumor-1	Lymphangioma-2	Adenoid cystic carcinoma -3
Nodular fasciitis-1	Hemangioendothelioma- 1	Adenocarcinoma - 1
Angiolymphoid hyperplasia with eosinophilia-1	**NEUROGENIC TUMORS**	Mucocele - 3
Inflammatory pseudotumor-4	Neurofibroma - 10	**SECONDARY EXTENSIONS**
Malignant fibrous histiocytoma-1	Schwannoma -3	Retinoblastoma - 4
Chloroma-1	Meningioma -1	Malignant melanoma -3
Embryonal Rhabdomyosarcoma-1	**LYMPHOID TUMORS**	Basal cell carcinoma -2
Small round cell tumor -1	Non Hodgkin's lymphoma - 16	Squamous cell carcinoma -2

The various specimens received included debulking(1.5%), anterior orbitotomy(7%), lateral orbitotomy(43%), medial orbitotomy(33%), incisional biopsy (18%), wedge biopsy (3.5%) and exenteration(4%). The clinical presentations included proptosis, swelling of eyes, ptosis, watering of eyes, ulcerative growth, pain and diplopia, of which the most common presentation was proptosis (33%). The male to female ratio was 1: 1.25 with a slight female predominance. All the tumors were unilateral. There was a bimodal age presentation with peaks in second and fifth decades. The most common location was lateral(36%) and the other locations were as follows – superior(33%), medial (9%), inferior (18%) and central /intraconal (4%).

The size of the lesions varied from 0.2 to 6.5cm, majority being 2 to 4cm in size. 72% of the tumors were benign. Neurogenic tumors were more common in females whereas rest of the tumors were common in males. The most common lacrimal gland tumor was pleomorphic adenoma. The vascular lesions included capillary hemangioma, cavernous hemangioma, composite hemangioma, lymphangioma and hemangioendothelioma. All the neurogenic tumors were benign and included neurofibroma, schwannoma and meningioma. A five year old boy presented with chloroma. One rare case of solitary fibrous tumor was seen. Metastases and secondary extensions from retinoblastoma, malignant melanoma, basal cell carcinoma and squamous cell carcinoma were seen. These data are consistent with literature reports.

DISCUSSION

Vascular tumors

Vascular tumors of orbit were classified based on their natural history by Mulliken and Glowacki[5] and based on

hemodynamic flow by the Orbital Society[6]. Capillary hemangiomas demonstrate small capillaries surrounded by benign endothelial cells. Cavernous hemangioma show large dilated vascular channels lined by flattened endothelial cells.

Neurogenic tumors

Out of all orbital tumors, neurofibroma form 0.5% to 2.4%. Peripheral nerve tumors required local surgical excision whereas advanced malignant cases were treated by exenteration and radiotherapy[7].

Lacrimal gland/sac tumors

Lacrimal gland tumors account for 10% of orbital tumors[8]. They occur in the superotemporal orbit. In our study, most common benign tumor was pleomorphic adenoma as reported in literature.

NonHodgkin's lymphoma

Extranodal non Hodgkin's lymphoma is seen in head and neck for 60% of cases[9]. Primary orbital disease occurs rarely [10,11,12]. In our study, immunohistochemistry was done and subtyping was done. Majority were of B cell lineage.

Inflammatory pseudotumor

The following points help to diagnose pseudotumors in cases of exophthalmos: 1) may be bilateral; 2) a more acute onset; 3) later age at onset than the primary neoplasm; 4) edema and pain of eyelids and conjunctiva in 50% of cases; and 5) regression with steroid treatment [13].

Chloroma

Leukemia in children formed less than 2% of orbital tumors[14]. Orbital involvement following or preceeding acute myeloid leukemia may occur unilaterally or bilaterally. Granulocytic sarcoma is the most commonly used terminology for chloroma because of the colour produced on exposure to myeloperoxidase (Figure 1).

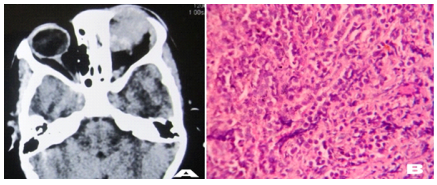

Figure 1: A) CT scan image showing unilateral orbital mass. B) Histopathology showing myeloid blasts. 100x H&E.

Solitary fibrous tumor

Though the awareness of this tumor is heightened, orbit still remains to be a rare location for this tumor. Microscopically, this tumor was composed of fascicles of spindle cells admixed with fibrocollagenous stroma and thick collagen bundles. Pericytomatous arrangement of tumor cells were seen around vascular channels. Immunohistochemistry study showed that the tumor cells were positive for CD34 and vimentin but negative for cytokeratin and S100 (Figure 2). They usually have a benign course and metastasis is rare. Local recurrences have been reported. Enbloc surgical removal provides best prognosis[15].

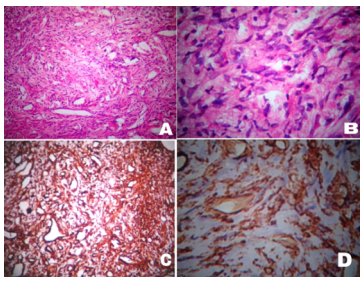

Figure 2: A) Histopathology image showing solitary fibrous tumor 40x H&E.
B) Spindle cells in a fibrocollagenous stroma. 100x H&E.
C) Vimentin positivity in tumor cells.
D) CD 34 positivity in tumor cells.

CONCLUSION

Orbital lesions still pose a diagnostic and therapeutic challenge even today. Immunohistochemistry along with histopathology pave way for a accurate diagnosis. Appropriate treatment enables complete cure and preserves vision. It is of utmost importance to diagnose the orbital tumors from pseudotumors. It is concluded that these tumors have bimodal age presentation and proptosis is the most common presentation. Non Hodgkin's lymphoma is the most common malignant tumor in our study.

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