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Diagnostic Pitfalls: A Case Series of 5 Patients with Lacrimal Gland Mass

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Abstract: Background: The orbit is infamous to harbor various mass lesions, out of which lacrimal gland tumors contribute to about 10% of all tumors. The peak incidence is in the middle-aged adults who present with upper eyelid mass. These tumors have a high morbidity and mortality rate due to their misdiagnosis. The diagnosis is still a dilemma on clinical examination and even its very difficult to get a specific diagnosis after imaging. Thus, biopsy can be an important tool to help in diagnosis and their suitable treatment. Objective: To provide specific, accurate and definitive diagnosis by correlating clinico-radiological findings with pathological examination to optimize management. Methods: We present a case series of 5 patients who presented with lacrimal gland mass with or without proptosis, defective vision, diplopia or restricted motility. These were evaluated radiologically which provide differential diagnosis that leads to a diagnostic challenge and their further management. So to reach a definitive diagnosis they were then correlated with histopathology with or without immunohistochemical stains by taking biopsy from the mass. Result and Conclusion: In our case series, we concluded that to make a confirmative diagnosis of the lacrimal gland tumors, the clinical and radiological findings should be correlated with the histopathological examination with or without immunohistochemical stains by biopsy and thus helps in rationalizing their further treatment.

Keywords: Orbit Tumors, Lacrimal Gland Mass, Proptosis

1. Introduction

A wide variety of space occupying lesions may be encountered in orbit. Out of which, lacrimal gland lesions constitute 10% of all lesions [1, 2]. They are further divided into benign and malignant lesions. Benign lesions present typically with painless mass of long duration in supero-temporal orbit which may or may not be associated with proptosis, defective vision, diplopia or restricted motility and can be differentiated from a malignant lesion by a rapid onset of symptoms, proptosis and evidence of adjacent bone destruction seen radiologically. Histologically, the lacrimal gland is composed of acini with an inner layer of cuboidal to columnar cells and outer layer of myoepithelial cells. These lesions pose diagnostic difficulties. Sometimes, these clinical and radiological diagnosis do not co-relate with their pathological and immunohistochemical diagnosis, which thus causes diagnostic dilemmas. We reviewed a case series of five patients by analyzing the clinical features, radiological and histopathological findings for definitive diagnosis of lesion for proper management and estimating prognosis.

2. Case Report

Case 1:

A 55years/male patient presented with proptosis and defective vision in left eye for 1 year. On examination there was a palpable mass in superolateral orbit with restricted motility, RAPD and optic atrophy. (Figure 1). VA: OD 6/24, OS: PL.

On the basis of MRI report diagnosis made was Lymphoma / Soft tissue sarcoma which showed well defined oval lesion in lateral extraconal space of left eye abutting and infiltrating lacrimal gland, lateral rectus medially and superior rectus superiorly and extending intraconally with optic nerve infiltration. It was extending up to orbital apex, showing restricted diffusion with suspicious infiltration of lateral orbital

wall and minimal intracranial extension noted (Figure 2).

Histopathology reported poorly circumscribed tumor tissue arising from lacrimal gland. The tumor tissue was formed by basaloid epithelial and myoepithelial cells in tubular and predominant cribriform pattern with intraluminal myxoid substance. Areas of neurotropic arrangement, replicated basement membrane material seen. Peripheral margins were infiltrated by tumor nests. Figure 3

Adenoid Cystic Carcinoma/Polymorphous Low Grade Adenocarcinoma

Case 2:

A 39 years old female presented with swelling in left eye since 1-year, occasional pain and redness, no defective vision /diplopia. On clinical examination it was a firm, non-tender mass noted in superotemporal orbit causing inferonasal dystopia and restricted motility (Figure 4). VA: OD 6/12, OS: 6/24.

The MRI report showed a mass in the lateral aspect of left orbit, intermediate signal on T1 and hypo intensity on T2, no diffusion restriction. It involved lacrimal gland and lateral rectus muscle and tendon, superiorly it was in contact with superior rectus muscle. No obvious bony erosion, no extension along superior and inferior orbital fissure (Figure 5) -- Lacrimal Gland Neoplasm

Histopathology reports structures of preserved lacrimal acini with diffuse infiltration of polymorphic lymphocytes, plasma cells, histiocytes, interspersed karyorrhexis and hyalinization. Interspersed areas of endothelial proliferation with entangled RBCs, hemorrhage and hemosiderin deposits. Fungal stains negative (Figure 6).

IHC: CD3: SCATTERED POSITIVE.

CD20: DIFFUSE POSITIVE.

CD31: Not done.

Inflammatory Proliferative Lesions of Lacrimal Gland with Low Grade. Hemangioendothelial Proliferation.

Case 3:

A 45 years/ Female, presented with painless swelling in left eye for 6 months without any defective vision/ diplopia. On examination there was a palpable non tender swelling, mildly restricting the motility without any proptosis (Figure 7). VA: OU: 6/6.

MRI: showed well defined multilobulated, multicystic lesion with septations and few solid components in extraconal compartment. The lesion also seen in inferior aspect of left lacrimal gland, medially extending upto lateral rectus (Figure 8) -- Lymphangioma

Histopathology: Poorly circumscribed tumor tissue arising from lacrimal gland. The tumor tissue formed by basaloid epithelial and myoepithelial cells in tubular and predominant cribriform pattern with intraluminal myxoid substances, areas of perineural invasion and collagenous spherulosis like arrangement seen (Figure 9). *Adenoid Cystic Carcinoma – Cribriform Type and Perineural Invasion.*

Case 4:

A 52years/ Male, presented with protrusion of right eye for 6 months and defective vision for 1 month. On clinical examination of proptosis there was restricted motility with no palpable mass seen, posterior segment showed choroidal folds in right eye (Figure 10). VA: OU:6/6.

MRI report presents well defined sharply marginated extraconal mass in superolateral aspect involving lacrimal gland causing mass effect with displacement of lateral rectus. Lesion was isointense to hyperintense in T1 and T2 with significant diffusion restriction. Scalloping of lateral bony wall seen (Figure 11) – Lymphoma.

Histopathology: Structure of fibrocollagenous tissue with diffuse lymphocytic and predominant plasma cell and eosinophilic infiltration. Extensive stromal sclerosis and associated karyorrhexis and perivascular hyalinization seen. Figure 12.

IHC: CD45: Diffusely positive.

CD20: Diffuse and scattered positive.

CD3: diffuse and scattered positive.

Idiopathic Inflammatory Orbital Disease.

Case 5:

There was a 41 years/Male who presented with protrusion of left eye for 3 months with mild pain.

There was no defect in vision or diplopia. On examination there was an axial proptosis with mild restricted motility but no palpable mass (Figure 13). VA: OU:6/6.

MRI depicts well defined heterogenous soft tissue lesion which was isointense in T1 and hyperintense in T2, antero supero-lateral aspect of post septal compartment with extraconal and intraconal components involving lacrimal gland with superior rectus infiltration. No optic nerve invasion (Figure 14) -- Pseudotumour.

Histopathology: Structure of fibro-collagenous tissue with sheets of monomorphic atypical lymphoid cells with mitosis, necrosis and lymphoplasmacytic differentiation. Hyalinization seen. Figure 15

IHC: LCA: strong and diffuse positive.

CD20: Diffusely positive.

CD3: Scattered positive.

Non- Hodgkins Lymphoma – B Cell Type, Medium to Large Size.

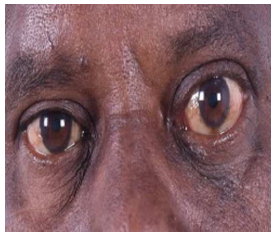

Figure 1. *Proptosis and palpable mass in left eye.*

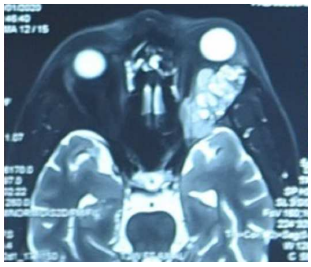

Figure 2. *MRI: lymphoma/Soft tissue sarcoma.*

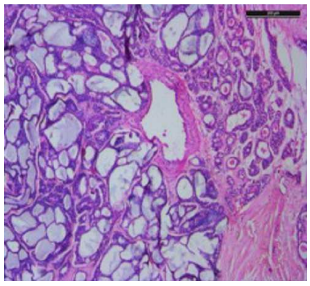

Figure 3. *HPE: Adenoid cystic carcinoma/polymorphous low grade adenocarcinoma.*

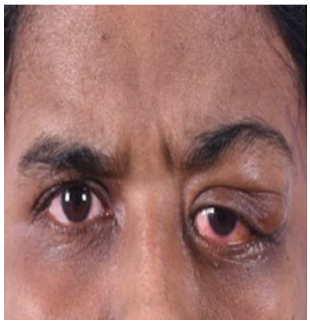

Figure 4. *Left eye dystopia with palpable mass.*

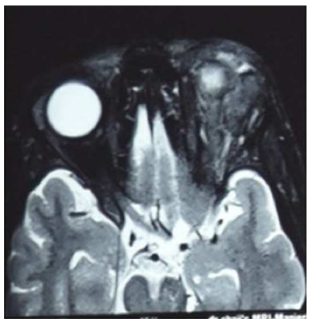

Figure 5. *MRI: Lacrimal gland neoplasm.*

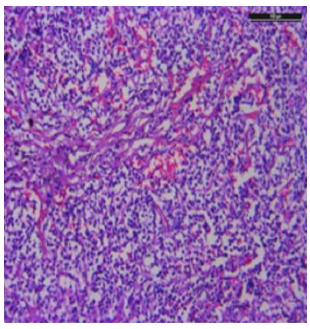

Figure 6. *HPE: Low grade hemangioendothelial proliferation.*

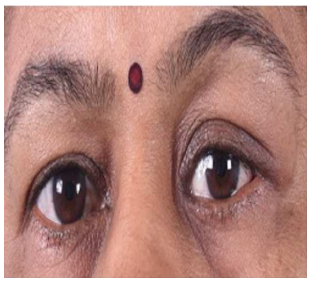

Figure 7. *Left eye palpable mass with no proptosis.*

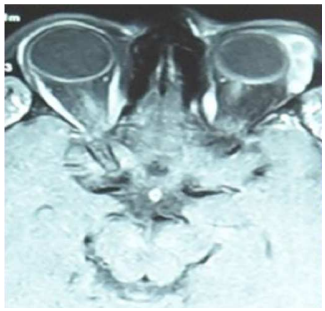

Figure 8. *MRI: Lymphangioma.*

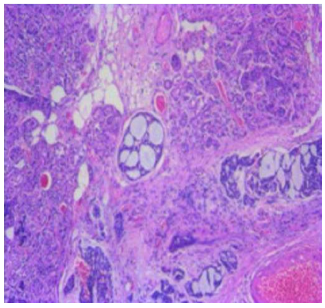

Figure 9. *HPE: Adenoid cystic carcinoma – cribriform type with perineural invasion.*

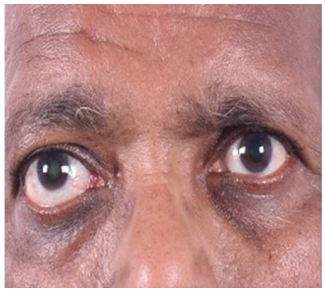

Figure 10. *Right eye proptosis with no palpable mass.*

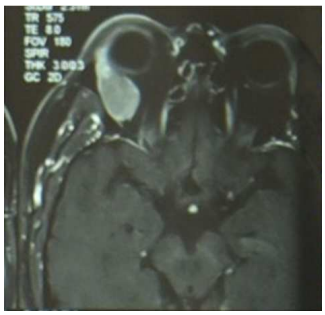

Figure 11. *MRI: Lymphoma.*

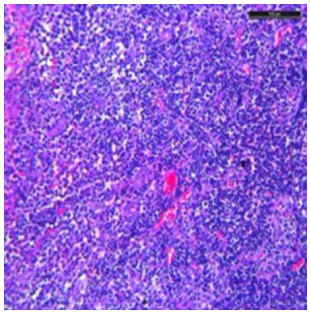

Figure 12. *HPE: Idiopathic inflammatory orbital disease.*

Figure 13. *Left eye proptosis with no papable mass.*

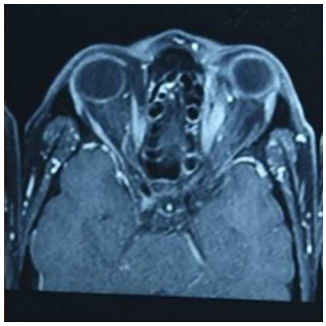

Figure 14. *MRI: Pseudotumour.*

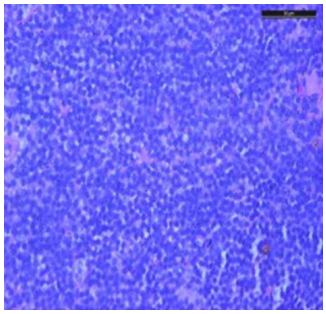

Figure 15. *HPE: Non-Hodgkin Lymphoma – B cell type.*

3. Discussion

Tumors of lacrimal gland are a rare condition in clinical practice, constituting about 10% of all orbital tumors [1]. They are further divided into benign and malignant lesions [2]. These typically present with supero- temporal mass lesion with or without proptosis, pain, restricted motility and palpable mass. Their diagnosis is of utmost importance for their management and estimating prognosis. They are diagnosed clinically and radiologically and then confirmed histopathologically by taking biopsy as also depicted in some studies [3, 4].

In our case series, in case 1, patient was diagnosed with lymphoma / Soft tissue sarcoma, based on clinical and radiological findings but was confirmed to be Adenoid cystic carcinoma / Polymorphous low-grade Adenocarcinoma (Figure 1), on histopathology. The preferred treatment for adenoid cystic carcinoma is complete surgical resection followed by radiotherapy to reduce relapses whereas in lymphoma complete excision is difficult, so role of surgery is limited to biopsy and chemotherapy followed by radiotherapy is the main stay of treatment [5-7]. Jerry A. Shields et al in a clinicopathologic case report described a case of adenoid cystic carcinoma of lacrimal gland in a 9 year old that simulated a dermoid cyst clinically and radiographically [8].

Case 2 was diagnosed as Lacrimal gland neoplasm. To get accurate diagnosis, biopsy was done, which revealed inflammatory proliferative lesions of lacrimal gland with low grade hemangioendothelial proliferation. In a case report by Neeta Misra et al, diagnosis of malignancy of maxillary antrum was made but the diagnosis of hemangioendothelioma orbit was confirmed by histopathology [9]. Hemangioendothelioma of lacrimal gland, is an uncommon benign low grade vascular tumor usually occurring in young adults, requiring complete excision of the mass. Whereas, lacrimal gland neoplasm is treated by complete excision followed by chemotherapy and/or radiotherapy, based on the type of neoplasm [10]. Thus, these diagnostic dilemmas change the treatment protocols.

In our case series, case 3 was diagnosed as lymphangioma. Incisional biopsy revealed Adenoid cystic carcinoma of Cribriform type and perineural invasion. Pattern of tumor cells help in prognosticating the disease, with better prognosis and survival rate in cribriform type. In a case report, Nigel G. Rawlings et al had diagnosed an orbital tumor as hemangioma, based on MRI but histopathological examination was disclosed as orbital schwannoma [11].

Case 4 was diagnosed with lymphoma clinically and radiologically but histologically as idiopathic orbital inflammatory disease. A comparative observational study by M N Islam et al in 2013 demonstrated the role of computed tomographic scan to detect clinically suspected orbital mass in 47 patients which were compared and correlated with Fine needle aspiration cytology or histopathology [12]. In another study by Sarah Linea von Holstein et al in 2013 described various epithelial tumors of lacrimal gland and also various tumor specific histological characteristics which determines the subsequent treatment regimen and important clues regarding prognosis [13]. Treatment modalities available for lymphoma are surgery, radiotherapy, chemotherapy and immunotherapy and prognosis depends on the histology, grading and staging whereas systemic steroid therapy is the mainstay of treatment for idiopathic orbital inflammation [14, 15].

Case 5, Diagnosis of Pseudotumor was made based on clinical and radiological examination. Systemic oral steroids were given as the treatment modality but since there was no resolution, incisional biopsy was planned to reconsider the diagnosis. Based on incisional biopsy, diagnosis of Non-Hodgkin lymphoma – B cell type was made and radiation therapy given. A case report by Strianese Diego et al in 2013 had the presumptive diagnosis of Pleomorphic adenoma based on clinical and radiological features and histological features were consistent with low grade B -cell non-Hodgkin lymphoma [7].

4. Conclusion

Orbital space occupying lesions can present in a similar fashion but their accurate and specific diagnosis is a challenge for proper treatment and their prognosis. These lesions are diagnosed based on clinical, radiological and histopathological findings but sometimes does not co relate and thus create diagnostic dilemma. In our case series, we concluded that along with clinical and radiological findings, histopathological examination with or without immunohistochemical stains help in correct diagnosis and thus rationalize their further treatment.

Abbreviations

VA- Visual Acuity, OD – Right Eye, OS – Left Eye, OU – Both Eyes, PL – Perception of Light, RAPD – Relative Afferent Pupillary Defect, IHC – Immunohistochemistry.

Conflicts of Interest

All the authors do not have any possible conflicts of interest.

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