

Sino-orbital Lymphoma: Case Report and Review of Literature

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Abstract

The orbit is involved in 10% of all lymphomas. Paranasal sinuses and nose are other sites for extranodal lymphoma that are involved in upto 2.6 to 6.7% of lymphomas involving head and neck region and are second most common presentation of extranodal lymphoma. In nearly 22.5% of patients, sinonasal lymphoma and orbital lymphoma may coexist. The present report describes such patient with first presentation to an ophthalmologist with an attempt to review the present literature of coexistent orbital and paranasal sinuses lymphoma.

Keywords: Proptosis, paranasal sinuses, orbit, lymphoma.

INTRODUCTION

The non-Hodgkin's lymphomas (NHL) are a group of hematologic malignancies which encompass any lymphoma other than Hodgkin's lymphoma. About one-third of NHL arise from sites other than lymph nodes, spleen, or the bone marrow, and even from sites which normally contain no native lymphoid tissue.^{1,2} The orbit is involved in 10% of all lymphomas, 5 to 8% of all extranodal lymphomas and 1.3 to 2% of all primary orbital extranodal tumors.³⁻⁵ Such lesions usually present as gradually increasing painless mass in the orbit or subconjunctival space. Paranasal sinuses and nose are other sites for extranodal lymphoma that are involved in upto 2.6 to 6.7% of lymphomas involving head and neck region⁶ and are second most common presentation of extranodal lymphomas⁷ in Asian population. In nearly 22.5% of patients, sinonasal lymphoma and orbital lymphoma may coexist⁸ and such patients need collaborative management effort by ophthalmologists, otorhinolaryngologists and oncologists. Extranodal orbital non-Hodgkin's lymphoma may present bilaterally along with involvement of paranasal sinuses. The present report describes such patient with first presentation to an Ophthalmologist. A review of literature of coexistent orbital and paranasal sinuses lymphoma is also reported.

CASE REPORT

A 68 years old female patient of Indian origin presented with history of nasal blockade and bilateral painless forwards protrusion of eyes for last 8 months. There was no history of visual blurring, or diplopia. There was no history of systemic complaints. Ocular examination revealed bilateral proptosis along with subconjunctival reddish mass temporally (Fig. 1) bilaterally. A CT scan was requested that revealed bilateral symmetrical homogenous moderately enhancing orbital mass along with homogenous diffuse sinus mass involving bilateral frontal sinuses, ethmoid sinuses and sphenoid sinuses (Fig. 2) with minimal haze in maxillary sinuses. A fine needle aspiration cytology of orbital mass was done that revealed features of reactive lymphoid hyperplasia. As the clinical suspicion of lymphoma was high, an incisional biopsy of orbital mass was carried out that revealed diffuse sheets of atypical lymphoid cells (Fig. 3) with nodular appearance. The cells were positive for CD 20 immunostain and negative for CD 3. This was consistent with the diagnosis of low grade B cell non-Hodgkin's lymphoma. The patient was planned to undergo chemotherapy using CHOP regimen (cyclophosphamide 750 mg/m² IV, vincristine 1.4 mg/m² IV, prednisone



Fig. 1: Clinical photograph of the patient showing bilateral proptosis, eyelid swelling and conjunctival mass temporally

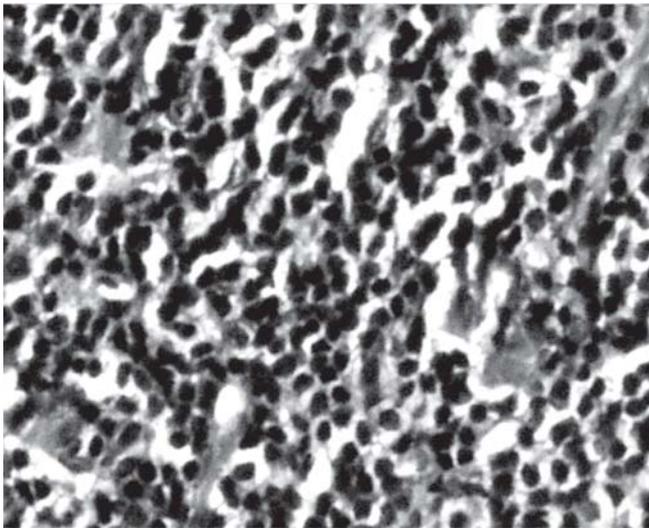


Fig. 2: CT scan orbits showing bilateral diffuse orbital mass with involvement of bilateral frontal, ethmoid and sphenoid sinuses

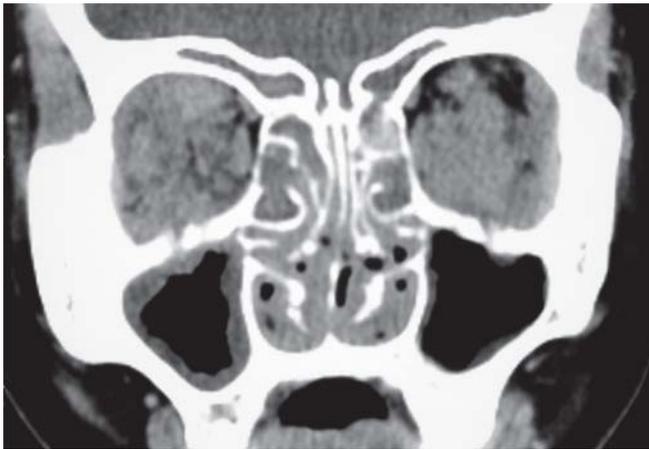


Fig. 3: Photomicrograph of incisional biopsy of mass showing features of non-Hodgkin's lymphoma; stained with hematoxylin eosin viewed under 10x

100 mg/m² orally, and doxorubicin 50 mg/m² IV) after hematological investigations.

COMMENT

At least 25% of all non-Hodgkin's lymphoma arises in tissues other than lymph nodes.⁹ These primary extranodal lymphomas can arise in the ocular adnexa and paranasal sinuses. Primary lymphoma of ocular adnexa refers to non-Hodgkin's lymphoma arising in orbit or subconjunctival location without any systemic manifestation of disease. Orbital lymphoma is commoner than ocular lymphoma¹⁰ and needs to be managed differently as ocular lymphoma is frequently associated with systemic lymphoma. Most of the orbital lymphomas are low grade lymphomas¹⁰ while the rest are intermediate or high grade lymphomas. Orbital lymphoma generally presents in sixth or seventh decade.¹¹ Any part of orbit may be involved and upto 25% of patients present with bilateral involvement.¹² Clinical presentations of orbital lymphoid neoplasms are most commonly characterized by slow, painless onset of swelling of the eyelid, palpable mass, and proptosis. Other common symptoms may include diplopia, diminished visual acuity, pain, ptosis, and restricted eye movements. Histologically, orbital lymphomas are commonly derived from B cell lymphocytes.¹⁰ The most common sites of disseminated disease for patients with orbital lymphoma are the lymph nodes (34%), skin (19%), bone marrow (11%), and spleen (10%). The risk of disseminated disease is related to histology; it is found in approximately 20% of marginal zone lymphoma and in 50% of diffuse large cell lymphomas at the time of initial diagnosis. Lymphoma of the conjunctiva or deep orbit has the lowest risk of dissemination compared with the lacrimal gland or eyelid. Bilateral clinical signs and local extension of the disease to the subcutaneous tissues around the eye or temporalis fossa are significant risk factors for future dissemination.¹³ According to REAL classification; the commonest sub-type of lymphoma originating in the orbit is the extranodal marginal zone lymphoma of B-cell origin. Sinonasal lymphomas are the commonest nonepithelial malignancies of the nasal cavity and paranasal sinuses. In Asian population, over 90% of these sinonasal lymphomas are T cell lymphomas.¹⁴ Sinonasal lymphomas are less commonly seen in females as compared to males. Sinonasal lymphomas usually spread from their site of origin in nasal cavity and invade adjacent structures including orbits. Most T cell lymphomas are intermediate grade or high grade lymphomas.¹⁵ The usual symptoms of sinonasal lymphoma are similar to any sinonasal mass and clinical diagnosis is usually not possible without histopathological evaluation. Sinonasal lymphoma may involve orbit

secondarily or may coexist with orbital lymphoma. In the Ophthalmic literature, sinonasal lymphomas with orbital involvement have been largely reported as sporadic cases.¹⁶⁻²¹ Cuadra-Garcia et al published a series of 58 sinonasal lymphomas, of which 16 involved the orbit of which 13 were diffuse large B-cell lymphomas.⁸ Pleg et al reported a series of 11 patients of sinonasal lymphoma with orbital involvement.²² The average patient age in their series was 68.3 years with male predominance. The authors reported multiple sinus involvement in 64% and destruction of orbital walls in 82% of their patients with subsequent spread of lymphoma. Baldini and coworkers reported a series of 19 patients with orbital and adnexal lymphoma.²³ In their series, only 1 patient had involvement of paranasal sinus. Cheung et al reported 113 patients with sinonasal lymphomas.²⁴ These authors reported orbital involvement in 39% of patients; majority of these patients had intermediate or high grade lymphoma. As the treatment used by various authors is different others in the above series, no definite recommendation can be given regarding the management of sino-orbital lymphomas. Radiotherapy has been said to be associated with excellent response;^{25,26} no controlled studies have however been conducted in such patients that compare radiotherapy with chemotherapy or any other treatment.

In the present case, the orbital mass was bilateral, symmetric and associated with bilaterally symmetrical sinus involvement without any bone destruction. To the best of our knowledge, such presentation or extranodal non-Hodgkin's lymphoma has not been described before. Our patient also had low grade B-cell lymphoma which again is less common variety in sino-orbital lymphomas. We emphasize the fact that in patients with diffuse sinus mass and bilateral orbital involvement, non-Hodgkin's lymphoma should be kept as a possible etiology and managed as early as possible.

REFERENCES

- Zucca E, Roggero E, Bertoni F, Cavalli F. Primary extranodal non-Hodgkin's lymphomas. Part 1: Gastrointestinal, cutaneous and genitourinary lymphomas. *Ann Oncol* 1997;8:727-37.
- Zucca E, Roggero E, Bertoni F, Conconi A, Cavalli F. Primary extranodal non-Hodgkin's lymphomas. Part 2: Head and neck, central nervous system and other less common sites. *Ann Oncol* 1999;10:1023-33.
- Witherspoon AC, Diss TC, Pan L, et al. Primary low-grade B-cell lymphoma of the conjunctiva: A mucosa-associated lymphoid tissue type lymphoma. *Histopathology* 1996;28:129-34.
- Shirai K, Okada Y, Saika S, Ohnishi Y. A case of mucosa-associated lymphoid tissue (MALT) lymphoma of presumed lacrimal gland origin. *Ganka Rinsho Iho (Jpn Rev Clin Ophthalmol)* 1996;90:604-6.
- Coupland SE, Krause L, Delecuse HJ, et al. Lymphoproliferative lesions of the ocular adnexa: Analysis of 112 cases. *Ophthalmology* 1998;105:1430-41.
- Ho F, Choy D, Loke SL, et al. Polymorphic reticulosis and conventional lymphomas of the nose and upper aerodigestive tract. *Hum Pathol* 1990;21:1041-50.
- Ng CS, Chan J, Lo S, et al. Immunophenotypic analysis of NHLs in Chinese. A study of 75 cases in Hong Kong. *Pathology* 1986;18:419-25.
- Cuadra-Garcia I, Proulx G, Wu CL, Wang CC, Pilch B Z, Harris NL, Ferry JA. Sinonasal Lymphoma: A Clinicopathologic Analysis of 58 Cases from the Massachusetts General Hospital. *Am J Surg Pathol*. Nov 1999;23(11):1356-69.
- E Zucca, E Roggero, F Bertoni, A Conconi, F Cavalli. Primary extranodal non-Hodgkin's lymphomas. Part 1: Gastrointestinal, cutaneous and genitourinary lymphomas. *Annals Oncol* 1997;8:727-37.
- E Zucca, E Roggero, F Bertoni, A Conconi, F Cavalli. Primary extranodal non-Hodgkin's lymphomas. Part 2: Head and neck, central nervous system and other less common sites. *Annals Oncol* 1999;10:1023-33.
- Darsaut TE, Lanzino G, Lopes B, Newman S. An introductory overview of orbital tumors. *Neurosurg Focus*. 2001;10:1-9.
- Smith MC, Donaldson SS. Radiotherapy is successful treatment for orbital lymphoma. *Int J Radiation Oncol Biol Phys*. 1993;26:59-66.
- Gedik S, Gur S, Maral T, Akova YA, Demirhan B. Bilateral synchronous primary orbital lymphoma. *Advances in Therapy* 2006;23(3):433-38.
- Kamath MP, Kamath G, Bhojwani K, Pai M, Shameem A, Agarwal S. Sinonasal lymphoma a case report. *ENT Journal* 2006;85(5):325-27.
- Cleary KR, Batsakis JG. Sinonasal lymphomas. *Ann Otol Rhinol Laryngol* 1994;108:411-19.
- Balayre S, Gicquel JJ, Mercie M, et al. Orbital extension of sinus plasmacytoma secondarily transforming into multiple myeloma: A case study. *J Fr Ophtalmol* 2004;27:67-71.
- Curi AL, Ferreira TC, Saddy JC, et al. Maxillary sinus non-Hodgkin's lymphoma with orbital and intraocular involvement in the acquired immunodeficiency syndrome. *Br J Ophthalmol* 2001;85:501-02.
- Derbel M, Ben Zina Z, Sellami D, et al. Exophthalmus and blindness disclosing an ethmoidal-maxillary malignant non-Hodgkin's T-cell lymphoma. Apropos of a case. *J Fr Ophtalmol* 1999;22:566-70.
- Desai UR, Peyman GA, Blinder KJ, et al. Orbital extension of sinus lymphoma in AIDS patient. *Jap J Ophthalmol* 1992;36:205-14.
- De Smet MD, Rootman J. Orbital manifestations of plasmacytic lymphoproliferations. *Ophthalmology* 1987;94:955-1003.
- Font RL, Laucirica R, Patrinely JR. Immunoblastic B-cell malignant lymphoma involving the orbit and maxillary sinus in a patient with acquired immune deficiency syndrome. *Ophthalmology* 1993;100:966-70.

22. Pleg A, Heran MKS, White VA, Chang WY, Rootman J. Malignant Lymphoproliferative disorders extending into the orbit from the paranasal sinuses. *Orbit* 2009;28:80-87.
23. Baldini L, Blini M, Guffanti A, Fossati V, Colombi M, La Targia ML, Bertoni F, Alietti A, Neri A, Bertoni G. Treatment and prognosis in a series of primary extranodal lymphomas of the ocular adnexa. *Annals of Oncology* 1998;9:779-81.
24. Michael MC Cheung, John KC Chan, WH Lau, William Foo, Paddy TM Chan, CS NG, Roger KC Ngan. Primary Non-Hodgkin's Lymphoma of the Nose and Nasopharynx: Clinical Features, Tumor Immunophenotype, and Treatment Outcome in 113 Patients. *J Clin Oncol* 1998;16:70-77.
25. Ping Z, Andrea K, Barbara S, Sigui L, Ling H, Peter MM. Radiation therapy for orbital lymphoma. *Int J Radiation Oncology Biol Phys* 2005;63(3):866-71.
26. Pelloski CE, Wilder RB, Ha CS, Hess MA, Cabanillas FF. Clinical stage IEA-IIIEA orbital lymphomas: Outcomes in the era of modern staging and treatment. *Radiother Oncol* 2001;59:145-52.