Mantle cell lymphoma of orbit in a middle aged female-a case report

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ABSTRACT

A 31 years female presented with painful swelling, proptosis of right eyeball and headache of 9 months duration. There were absense of perception of light, pupillary reaction and extraocular movements. Exposure keratopathy with proptosis was observed. She was provisionally diagnosed a case of orbital cellulitis. The computerized tomography revealed lymphoma. Her right orbit was exenterated. The histopathologic examination revealed non-Hodgkin's lymphoma of diffuse small type. Immunohistochemistry demonstrated malignant cells of B cell origin and it was classified as mantle cell lymphoma. Orbital lymphoid tumors typically affect older persons, but occasionally, it can involve middle aged adults also. Mucosa associated lymphoid tumors are the most common affecting eye and orbit, but mantle cell lymphoma is extremely rare.

Keywords: non-Hodgkin's lymphoma, mantle cell lymphoma, diffuse small B cell lymphoma, orbit.

Orbital lymphomas are infrequent manifestation among ocular neoplasm. Lymphoma as such is a systemic disease and various types of lymphoma can involve it.[1] Non-Hodgkin's lymphomas (NHL) of orbit have been reported in the literatures which is less than 1.0% of all NHL. Although NHL of orbit is mostly indolent in nature, uncommonly it can transform into aggressive form to metastasize directly to affect eye-adnexa manifesting from swelling, compression, diplopia, ptosis, exophthalmus, sometimes enophthalmus[2] to loss of vision. Early diagnosis and proper management can save the vision and debility of the patients. Orbital lymphoid tumors typically affect older persons, but rarely, it can involve younger adults also. The latest classification of WHO divides non-Hodgkin lymphoma into B or T cell type and almost all orbital lymphomas consist of B cell tumors.[3] Mucosa associated lymphoid tumor (MALT) has been reported as the commonest tumor affecting eye and orbit, but mantle cell lymphoma is extremely rare in a middle aged person and thus it is being reported.

CASE REPORT

A 31 years female patient presented with complaints of painful swelling, proptosis of right eyeball and headache of 9 months duration. The complaints aggravated for the last 2 months followed by loss of vision. On examination, there was no perception of light, absent pupillary reaction, no extraocular movement and exposure keratopathy owing to marked degree of proptosis. The left eye was found to be normal. There was swelling and tenderness over frontal sinus. Mild lymphadenopathy of the right axilla and anemia were detected. She was provisionally diagnosed a case of orbital cellulitis, and treated with steroids and antibiotics. The symptoms relieved to some extent. The total leucocyte count was 12,500 with lymphocytosis. The computerized tomography (CT) revealed lymphoma, having a diffuse enhancing soft tissue mass within muscle cone of the right orbit replacing retro orbital fat and pushing the eyeball laterally and forward causing exophthalmos with evidence of no bony erosion. The left orbit appeared intact (Fig. 1). After intradepartmental discussion, her right orbit was exenterated under general anaesthesia and tissues were sent for histopathology examination. The tumor mass was glandular in consistency. The hematoxylin and eosin stained histopathologic examination revealed predominantly small cells, with intermediate lymphocytes in diffuse pattern having irregular nucleus and inconspicuous nucleoli. Fibrous tissue proliferation and orbital fatty tissue infiltration by neoplastic cells were seen. It was

Fig. 1. The CT scan showing diffuse enhancing soft tissue mass in the right orbit replacing retro orbital fat.

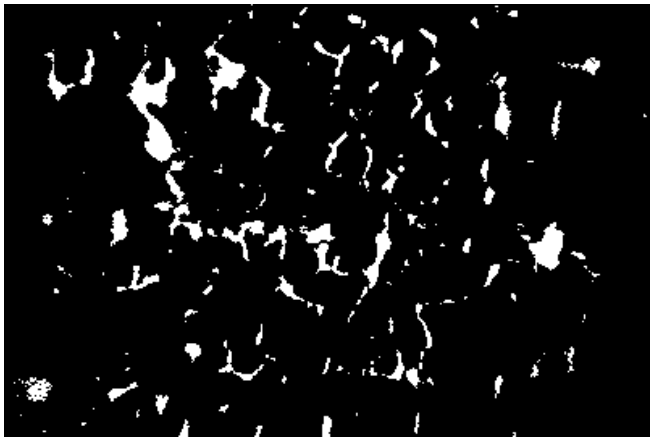

Fig. 2. Photomicrograph showing small lymphocytes in diffuse pattern (H & E x 40).

concluded with NHL of diffuse small type (Fig. 2). Immunohistochemistry demonstrated malignant clonal cells of B cell origin (CD19, CD20, CD5 positive) coexpressing surface IgM and IgD and chromosomal translocation of t (11;14). Finally it was classified as mantle cell lymphoma. CT scan of the chest and abdomen did not reveal any evidence of lymphomatous involvement. The bone marrow biopsy showed reactive lymphoid cells. The patient with preliminary advice was referred for radiotherapy for any residual involvements. She is doing well without any remarkable complaints for the last 2½ years.

DISCUSSION

The incidence of primary ocular lymphoma is although rare, it affects various parts of eye. The present case was a middle aged female. Lymphomas usually affect older age group persons above 60 years as it was reported in a 63 years female[1]. However, sometimes it has been reported in below 60 years of age. Lutz *et al* reported it in a case of 47 years male.[4] Inflammatory lesions are the most common to involve the orbit. Pseudotumour and lymphomas are next in incidence. Ptosis may result due to excessive weight of edema fluid on upper lid which creates difficulty in diagnosis as was seen in the present case with painful swelling and tender frontal sinus. In the early stage the present case was provisionally differentiated as orbital cellulitis owing to its presentation,[5] and initially prednisolone and antibiotics were advised in the case as also done in a 61 years female with complaints of headache and eyeball pain.[6] Lymphoma most commonly involves conjunctiva followed by eyelids and orbit.[7] Mantle cell lymphoma was reported involving conjunctiva in a 78 years male.[8] This lymphoma demonstrates diffuse growth pattern on histology. The microscopy of the present case showed the same predominantly of small lymphocytes with few intermediate cells. This was demonstrated histologically in 95.0% cases in a study.[9] Mantle cell lymphoma is an aggressive tumor, but MALT has been implicated as the most common tumor which involves eye and adnexa.[7] Chemotherapy and radiotherapy are vital for survival depending upon spread of disease. The role of radiotherapy in localized lesions is satisfactory and regresses the orbital signs.[4,5,9] However, in some cases, recurrence has been reported.[6] The extremely rare type of mantle cell lymphoma can affect a middle aged patient, but early presentation and investigations for diagnosis is mandatory to direct the managements.

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