CLINICAL VIGNETTE

Orbital Lymphoma

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A 37-year-old Caucasian male presented to his ophthalmologist with a pea sized lesion in the medial canthus of his right eye. He had noticed this lesion initially one month prior. Ophthalmologist noted a right eye ptosis on exam without any visual impairment. Patient also did not have any fevers, weight or appetite changes, pain or fatigue. An MRI revealed a lobular unilateral right-sided extracoronal mass involving the anterior, medial, and inferior right orbit measuring 1.4x 2.5x 1.3 cm. There was no involvement of adjacent extraocular muscles. He underwent excision that removed 90% of the mass.

Pathology of the right eye mass was consistent with a high grade B cell lymphoma favoring a diffuse large B cell lymphoma. The characteristics of the tumor were most consistent with a Germinal center, B cell like lymphoma measuring 1.2 x 1.0 x 0.7 cm with a proliferative index of 85%. Tumor was positive for BCL6, CD10, CD20. Tumor was negative for MUM-1, and negative for MYC gene rearrangement.

Discussion of Orbital Lymphomas

Orbital lymphomas constitute only 1% of all non-Hodgkin's lymphoma.[1,2] Lymphomas are the most common primary orbital tumor in adults 60 years of age and older.[2] The majority of orbital lymphomas are extranodal marginal-zone B cell lymphomas of mucosa-associated lymphoid tissue (MALT) type lymphomas, but mantle cell lymphoma and diffuse large B cell lymphomas also occur to a lesser frequency.[3] The most common signs and symptoms are periorbital tumor mass, exophthalmos, ocular pain, eye motility and visual restrictions.[1]

There are no standard guidelines for treatment of orbital MALT lymphomas, but there is a high rate of local relapse with surgery alone due to difficulty of a complete resection. Radiation therapy has been reported to be highly effective in MALT lymphoma of the orbit.[4,5] Radiation alone can also have high rates of local and distant failure with recurrence rates of 25%-47% in some studies.[6] Radiation can also cause late complications including dry eyes, keratitis, and cataract formation.

Combination chemotherapy is usually not administered for MALT lymphoma due to effectiveness of radiation therapy. Most often chemotherapy is used post operatively, post radiation, in advanced stages or with diffuse large B cell lymphomas.

Discussion of Diffuse Large B Cell Lymphoma (DLBCL)

The oncogenes, MYC, BCL2 and BCL6 are the most common chromosomal translocations in DLBCL. BCL2 translocations are seen in one third of DLBCL mostly in the more favorable form Germinal Center B cell molecular subtype. By itself the BCL2 translocation, does not appear to portend a less favorable survival and expression of BCL2 is not correlated with the t (14:18). BCL6 translocation is found in up to one third of DLBCL.[7,8] The MYC oncogene translocation are present in 5-15% of cases of DLBCL and appear to confer a worse prognosis following treatment with anthracycline based combination chemotherapy such as RCHOP.[9]

High risk double or triple "Hit" lymphomas are characterized by a MYC translocation combined with a BCL6 and /or BCL2 translocation. These subtypes of DLBCL are more difficult to treat and have worse prognosis. The patient did not have a MYC translocation classifying his lymphoma as a Germinal center.

He underwent further imaging for completion of staging with PET scan that did not show any evidence of distant metastasis rendering him a stage IE status. Due to subtype of lymphoma (Germinal center B cell like), his age and patient preference he was offered sequential chemotherapy for 3 cycles with R-CHOP followed by radiation therapy to the inner canthus of right eye, versus chemotherapy alone for six cycles. He wanted to avoid complications of radiation and chose chemotherapy alone. He tolerated his treatments and had no evidence of residual tumor after his third cycle of treatment.

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