
Abstract:

Patient with a history of Marginal Zone Lymphoma complains of dry eyes. Upon physical examination he appears to have an unusual orbital fat prolapse. Radiological imaging and biopsy display Lacrimal and Conjunctival Lymphoma.

I. Case History

- Patient demographics
  - 85 year old African American Male
- Chief complaint
  - Patient reports intermittent blurring of vision in current spectacles. In addition, he complains about dryness and foreign body sensation, where the right eye is worse than left eye.
- Ocular, medical history
  - PMHx:
    - Lymphoma, Non-Hodgkin’s
    - Spinal Stenosis of Lumbar Region
    - Osteoarthritis
    - Mild Cognitive Impairment, so stated
    - Constipation
    - Esophageal Reflux
    - Osteoporosis
    - Shingles
    - Anemia
    - Elevated PSA
    - Erectile Dysfunction
    - Hypertrophy of Prostate
  - POHx:
    - Normal Tension Glaucoma
    - Age related Macular Degeneration, Category 2
    - MGD
    - Cataract OU
    - H/O PRK c Lasik Enhancement
    - Refractive Error
- Medications
  - ASA 325 mg
  - Miconazole Topical
  - Finasteride 5mg
  - Artificial Tears
  - Gabapentin 100mg
  - Latanoprost
  - Multiple multivitamins

II. Pertinent findings

- Clinical
  - Pink fleshy elevated lesions of the superior palpebral conjunctiva OU, right worse than left.
- Physical
  - Enlarged upper lid resulting in a ptosis, right worse than left.
- Laboratory studies
  - Elevated: PSA, ESR
  - Low: HGB, HCT, MCHC, MPV, ALT (sgpt)
- Radiology studies
PET/CT images show metabolic evidence of extensive lymphadenopathy, which extends from the left oropharyngeal region, neck, axillary, down to inguinal and femoral nodes, compatible with recurrent lymphoma.

- Enlargement of both conjunctival tissue and lacrimal glands, right greater than left. They demonstrate fairly homogenous signal intensity and mild enhancement as well as subtle restricted diffusion. The findings are more compatible with lymphomatous infiltration, particularly in light of the clinical history. Other etiologies such as Sjogren’s disease or sarcoidosis are felt to be relatively unlikely. There is dilatation of both superior ophthalmic veins which is likely a result of the enlargement of the lacrimal gland. The cavernous sinuses demonstrate normal enhancement.

III. Differential diagnosis

- Primary/leading

- Others
  - Sarcoidosis
  - Tuberculosis
  - Sjogren’s Disease

IV. Diagnosis and discussion

- Elaborate on the condition
  - Marginal Zone Lymphoma is a rare B-cell lymphoma, which accounts for 5% of NHL.
  - There are 3 distinct subtypes: nodal, extranodal and splenic.
  - Extranodal MZL of mucosa associated lymphoid tissue (MALT) usually results in the skin or mucosal surfaces, which include the GI tract, lungs and upper respiratory tract.
    - It’s less commonly found in the thyroid, salivary, lacrimal, conjunctival and breast glands.
  - The condition may be associated with chronic infection or autoimmune diseases causing the conversion of normal lymphoid cells to MALT lymphoma.
  - MALT’s name arises from the resemblance to the marginal zone that surrounds the germinal center of normal lymph nodes.

- Expound on unique features
  - MZL presents with peripheral lymphadenopathy
    - The head and neck are most commonly involved.
    - Primary extranodal MZL of the ocular adnexa is rare and is characterized by local disease.
    - Orbital lymphoma patients will have systemic lymphomas in 75% of cases.
    - MALT lymphomas have been associated with systemic H.pylori, B.burgdorferi, and C.psittaci.
  - Ocular adnexal lymphomas occur in approximately 1-2% of NHL patients and are the most common malignant tumors of the orbit.
    - The majority are B-cell non-Hodgkin lymphomas 98%.
    - Clinically, ocular involvement is slowly progressive and can affect the eyelid, orbit, lacrimal gland, or conjunctiva.
    - Lacrimal gland lymphomas only represent 7-26% of ocular adnexal lymphomas.
    - Conjunctival lymphomas make up 25% of all adnexal lymphomas.

V. Treatment, management

- Treatment and response to treatment
  - Patient elected to be monitored every 3-4 months, as this condition has been indolent since diagnosis in 2012.

- Refer to research where appropriate
  - For localized ocular involvement where Chlamydia has been associated, high dose doxycycline has shown regression of condition.
  - External beam radiotherapy is the treatment of choice for primary ocular MZL and follicular lymphoma
    - Complications secondary to radiation treatment include damage to the lacrimal and meibomian glands with possible resulting dry eye and corneal damage, cataract formation, and radiation retinopathy.
  - Chemotherapy is indicated in secondary OAL.
Bibliography, literature review encouraged


VI. Conclusion

Clinical pearls, take away points if indicated:
- Primary systemic Non-Hodgkin’s Lymphoma rarely presents with ocular involvement, but it may occur anytime during the disease course. NHL should be suspected in a patient who presents with lid fullness, ptosis, proptosis, or diplopia.