

71. Ocular Adnexal Lymphoma

Authors

Steffen Heegaard, Patricia Chevez-Barríos, Valerie A. White, Sarah E. Coupland, Paul T. Finger

Emerging Prognostic Factors for Clinical Care

- History of rheumatoid arthritis
- History of Sjögren syndrome
- History of connective tissue disease
- History of recurrent dry eye (sicca) syndrome
- History of IgG4 ocular adnexal disease
- Any evidence of previous or current hepatitis B, hepatitis C, or HIV infection
- Any evidence of *Helicobacter pylori* gastritis
- Any evidence of an infection caused by *Chlamydia psittaci*
- IGH-locus translocation or somatic mutation pattern (EMZL)¹
- Presence or absence of an A20 deletion²⁻⁴
- Presence of B symptoms^{5,6} (fever, night sweats, weight loss)
- Concordant /discordant bone marrow involvement (DLBCL)^{7,8}
- Centroblastic/immunoblastic (DLBCL)^{7,8}

Risk Assessment Models

The AJCC recently established guidelines that will be used to evaluate published statistical prediction models for the purpose of granting endorsement for clinical use.⁹ Although this is a monumental step toward the goal of precision medicine, this work was published only very recently. Therefore, the existing models that have been published or may be in clinical use have not yet been evaluated for this cancer site by the Precision Medicine Core of the AJCC. In the future, the statistical prediction models for this cancer site will be evaluated, and those that meet all AJCC criteria will be endorsed.

Recommendations for Clinical Trial Stratification

The authors have not provided any recommendations for clinical trial stratification at this time.

Bibliography

1. Coupland SE, Foss H-D, Anagnostopoulos I, Hummel M, Stein H. Immunoglobulin V \sim H Gene Expression among Extranodal Marginal Zone B-Cell Lymphomas of the Ocular Adnexa. *Investigative Ophthalmology and Visual Science*. 1999;40:555-562.

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2. Chanudet E, Ye H, Ferry J, et al. A20 deletion is associated with copy number gain at the TNFA/B/C locus and occurs preferentially in translocation-negative MALT lymphoma of the ocular adnexa and salivary glands. *The Journal of pathology*. 2009;217(3):420-430.
3. Hirsch B, Grünbaum M, Wagner F, et al. A novel A20 (TNFAIP3) antibody (Ber-A20) can be used to detect unmutated A20 by immunohistology. *Histopathology*. 2012;60(6B):E19-E27.
4. Bi Y, Zeng N, Chanudet E, et al. A20 inactivation in ocular adnexal MALT lymphoma. *Haematologica*. 2012;97(6):926-930.
5. Sjo LD, Heegaard S, Prause JU, Petersen BL, Pedersen S, Ralfkiaer E. BIOCHEMISTRY AND MOLECULAR BIOLOGY-Extranodal Marginal Zone Lymphoma in the Ocular Region: Clinical, Immunophenotypical, and Cytogenetical Characteristics-This is a very detailed and precise study on the extranodal marginal zone B-cell lymphomas (EMZL) arising primarily in the ocular adnexa including morphology, immunophenotyping, and cytogenetic analysis with correlation to clinical characteristics. *Investigative ophthalmology & visual science*. 2009;50(2):516.
6. Sjo LD. Ophthalmic lymphoma: epidemiology and pathogenesis. *Acta ophthalmologica*. 2009;87 Thesis 1(thesis1):1-20.
7. Munch-Petersen HD, Rasmussen PK, Coupland SE, et al. Ocular adnexal diffuse large B-cell lymphoma: a multicenter international study. *JAMA ophthalmology*. 2015;133(2):165-173.
8. Rasmussen PK, Ralfkiaer E, Prause JU, et al. Diffuse large B-cell lymphoma of the ocular adnexal region: a nation-based study. *Acta ophthalmologica*. 2013;91(2):163-169.
9. Kattan MW, Hess KR, Amin MB, et al. American Joint Committee on Cancer acceptance criteria for inclusion of risk models for individualized prognosis in the practice of precision medicine. *CA: a cancer journal for clinicians*. 2016.