

What location of a periocular lymphoma is associated with the lowest risk of systemic lymphomas?

- A. lacrimal gland
- B. eyelid skin
- C. conjunctiva
- D. orbit

Correct Answer

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### Explanation & Notes

Discussion

BCSC Excerpt

References

Notes



59% of respondents answered correctly.  
A. 9% B. 22% C. 59% D. 8%

#### Section 7: Orbit, Eyelids, and Lacrimal System Chapter 5: Orbital Neoplasms and Malformations

Neural Tumors

Mesenchymal Tumors

**Lymphoproliferative Disorders**

Lacrimal Gland Tumors

Secondary Orbital Conditions

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#### Lymphoid Hyperplasia and Lymphoma

Lymphoproliferative lesions of the ocular adnexa constitute a heterogeneous group of neoplasms that are defined by clinical, histologic, immunologic, molecular, and genetic characteristics. Lymphoproliferative neoplasms account for more than 20% of all orbital tumors.

The vast majority of orbital lymphoproliferative lesions are non-Hodgkin lymphoma (NHL). In the United States, the incidence of NHL in all anatomic sites has been increasing, and it is one of the most common malignancies affecting the orbit. Workers with long-term exposure to bioactive solvents and reagents are at increased risk for NHL, as are older adults and individuals with chronic autoimmune diseases.

#### Identification and classification of lymphoproliferative disorders

Non-Hodgkin lymphoma encompasses a heterogeneous group of malignancies and includes many subtypes. The Revised European-American Lymphoma Classification applies immunophenotypic and genetic features to identify distinct clinicopathologic NHL entities, including extranodal sites such as the orbit. The World Health Organization's classification elaborates on this approach. Orbital extranodal disease appears to represent a biological continuum and behaves unpredictably. By molecular genetic studies, approximately 90% of orbital lymphoproliferative disease is monoclonal, and 10% is polyclonal; however, both types of lesions may involve prior, concurrent, or future systemic spread. Approximately 20%–30% of periocular lymphoproliferative lesions have a history of previous or concomitant systemic disease, and an additional 30% develop it over 5 years. The risk of systemic disease remains elevated for decades after the original lesion is diagnosed, regardless of the initial lesion's location in the orbit or its clonality.

The risk of having or developing systemic NHL is lowest for conjunctival lesions, greater for orbital lesions, and highest for lesions arising in the eyelid. Lymphoid lesions developing in the fossa of the lacrimal gland may carry a greater risk of systemic disease than those occurring elsewhere in the orbit. Bilateral periocular involvement markedly increases the risk of systemic disease, but such involvement is not definitive evidence of systemic disease.