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

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

Correct Answer: C. conjunctiva

Explanation & Notes

It is important to understand the risk of systemic lymphoma in a patient with periorcular lymphoma. More than half of periorcular lymphomas involve prior, concurrent, or future systemic spread (20%-30% have previous or concomitant systemic disease, 30% develop it over 5 years). This risk is somewhat predicted by the site of periorcular origin: The risk is lowest for conjunctival lesions, greater for orbital lesions and highest for eyelid lesions. Lesions in the lacrimal fossa may carry a greater risk than those occurring elsewhere in the orbit. Bilateral periorcular involvement greatly increases the risk of systemic disease, but it is not a definitive sign of systemic disease. The risk of systemic disease is increased for decades after the original lesion is diagnosed, regardless of the initial lesion's location.

The prediction of biologic behavior of periorbital lymphoproliferative lesions is limited, despite clinical, histopathologic, and immunohistochemical studies. Pathologically, lymphoproliferative lesions with lymphoid follicles with germinal centers, a polymorphous mixture of plasma cells, and mature lymphocytes favor a diagnosis of reactive lymphoid hyperplasia. More monotonous, small, round lymphocytes are seen in low-grade malignant lymphomas, whereas high-grade lymphomas demonstrate more malignant nuclei and frequent mitoses. Immunohistochemistry demonstrating mono- or polyclonality is helpful on tissue sections or by flow cytometry.