

Figure 5-18 Lymphoproliferative lesion. **A**, Ptosis of the right upper eyelid and fullness (arrow) in the superior orbit. **B**, Axial CT scan shows a homogeneous mass (arrow) with characteristic molding along the globe. **C**, Incisional biopsy through an anterior orbitotomy approach via the upper eyelid crease. **D**, H&E stain of this hypercellular lesion is consistent with B-cell lymphoma. (Parts A–C courtesy of Cat N. Burkat, MD; part D courtesy of Bobby S. Korn, MD, PhD.)

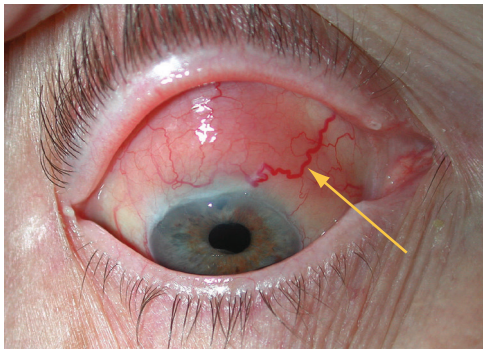


Figure 5-19 Lymphoproliferative lesion presenting as a salmon-patch subconjunctival lesion. Note the prominent feeder vessel (arrow) overlying the lesion. (Courtesy of Bobby S. Korn, MD, PhD.)

aspiration biopsy may establish all but the morphologic characteristics of the lesion. Conjunctival biopsy for follicular conjunctivitis can sometimes reveal a lymphoproliferative lesion.

Both reactive lymphoid hyperplasia and malignant lymphoma are hypercellular proliferations with sparse or absent stromal components. Light microscopy may reveal

a histologic continuum from reactive lymphoid hyperplasia to low-grade lymphoma to higher-grade malignancy; it may not by itself adequately characterize a given lesion. In such cases, immunopathology and molecular diagnostic studies aid in further categorization.

Malignant lymphomas are thought to represent clonal expansions of abnormal precursor cells. Immunologic identification of lymphocyte cell-surface markers can classify tumors as containing B cells or T cells. Specific monoclonal antibodies directed against surface light-chain immunoglobulins are used to determine whether the cells represent monoclonal (ie, malignant) proliferations.

Genetic analysis has shown that most lymphoproliferative lesions that appear to be immunologically polyclonal actually harbor small monoclonal proliferations of B lymphocytes. However, the finding of monoclonality, by either immunophenotype or molecular genetics, does not predict which tumors will ultimately result in systemic disease.

Management

Because there is considerable overlap among the various lymphoproliferative lesions in terms of clinical presentation and behavior, all patients with hypercellular lymphoid lesions (whether monoclonal or polyclonal) should be examined by an oncologist. Depending on the histologic type of the lesion, the examination may include a general physical examination, a complete blood count, a bone marrow biopsy, CT and/or MRI imaging, a positron emission tomography scan, and serum immunoprotein electrophoresis. The patient should be reexamined periodically because systemic lymphoma may develop many years after the occurrence of an isolated orbital lymphoid neoplasm.

For EMZL and FL, radiotherapy usually results in good outcomes, with 10-year survival rates of 92% and 71%, respectively. DLBCL and MCL have a poorer prognosis, with 10-year survival rates of 41% and 32%, respectively. Treatment of non-EMZL more often involves chemotherapy and immunomodulation in addition to radiotherapy. The optimal dose of radiation has not been established, with published amounts ranging from 20–40 Gy in fractionated doses. A surgical cure is usually not possible because of the infiltrative nature of lymphoid tumors. Alternative treatments include targeted therapies such as rituximab for CD20-positive lymphomas.

The management of low-grade lymphoid lesions that have already undergone systemic dissemination is somewhat controversial because indolent lymphomas are generally refractory to chemotherapy and are associated with long-term survival, even if untreated. Many oncologists take a watchful-waiting approach and treat only symptomatic disease.

Yen MT, Bilyk JR, Wladis EJ, Bradley EA, Mawn LA. Treatments for ocular adnexal lymphoma. A report by the American Academy of Ophthalmology. *Ophthalmology*. 2018;125(1):127–136.

Plasma Cell Tumors

Lesions composed predominantly of mature plasma cells may be plasmacytomas or localized plasma cell–rich masses. Multiple myeloma should be ruled out, particularly if there is bone destruction or any immaturity or mitotic activity of the plasmacytic elements. Some lesions are composed of lymphocytes and lymphoplasmacytoid cells and demonstrate the combined