Malignant Lymphoma of the Orbit

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Left eyelid swelling and drooping, which had been worsening slowly over the last 5 to 6 months, prompted a 77-year-old man to seek medical evaluation. There was no pain and no discharge from the eye. The patient stated that his vision was not adversely affected, and he denied diplopia. There was no history of ocular injury or surgery.

Palpation of the eyelid and periorbital area identified a nontender, rubbery tumor (A). An MRI scan of the orbits revealed an ill-defined, irregularly shaped, homogeneously enhancing mass that involved the left upper eyelid and supero-anterior aspect of the orbit (B). The mass was extraconal. A biopsy demonstrated a mildly vascular, friable solid tumor. Pathologic analysis of material from the lesion revealed a nodular infiltrate with small lymphocytes that showed cleaved nuclei. The stained lymphocytes revealed leukocyte common antigen, B-cell antigen, and lambda light chain. T-cell antigen and kappa light chain were not found. The diagnosis was malignant lymphoma, follicular, small-cleaved lymphocyte, grade 1 (C).

Clinically, malignant lymphomas are the most common lymphoproliferative disorders that affect the orbit. They occur in older persons and usually do not arise in children. Generally, these tumors progress slowly and cause painless proptosis, when they originate from behind the globe, or ptosis, when they originate from the upper orbit or lacrimal gland. Sometimes, only the conjunctiva is involved and exhibits a fleshy, salmon-colored mass or subconjunctival swelling. Roughly 75% of malignant lymphomas are unilateral and 25% are bilateral; 60% present as localized lesions and 15% of these subsequently disseminate.¹

An orbital biopsy needs to be obtained for histologic assessment and proper staging of the malignant lymphoma. Radiotherapy is the standard treatment of localized disease, such as for that seen in this patient. Usually, radiation is given to the involved orbit in divided doses over 3 or 4 weeks while the eye is shielded. Systemic chemotherapy is used when the tumors are part of a disseminated process. Localized conjunctival tumors have been treated with cryotherapy. The prognosis is excellent for patients whose disease is identified early and treated appropriately. Six-month follow-up visits to an ophthalmologist and an oncologist are needed to monitor for local or systemic recurrence or side effects from radiation therapy.

REFERENCE:

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