Clinical Ophthalmic Oncology

Orbital Tumors

Catherine J. Hwang
Bhupendra C. K. Patel
Arun D. Singh
Editors

Third Edition

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Preface

Ophthalmic tumors are rare and diverse so their diagnosis can be quite complex. Treatment usually requires special expertise and equipment and in many instances is controversial. The field is advancing rapidly, because of accelerating progress in tumor biology, pharmacology, and instrumentation. Increasingly, the care of patients with an ocular or adnexal tumor is provided by a multidisciplinary team, consisting of ocular oncologists, general oncologists, radiotherapists, pathologists, psychologists, and other specialists.

For all these reasons, we felt there was a need for the new edition of the textbook providing a balanced view of current clinical practice. Although each section of *Clinical Ophthalmic Oncology* now represents a standalone volume, each chapter has a similar layout with boxes that highlight the key features, tables that provide comparison, and flow diagrams that outline therapeutic approaches. We gratefully acknowledge the contributions of Julian Perry, MD, for editing the previous two editions of the volume.

The enormous task of editing a multi-author, multi-volume textbook could not have been possible without support and guidance by the staff at Springer: Caitlin Prim, Melanie Zerah, ArulRonika Pathinathan, and Karthik Rajasekar. Michael D. Sova kept the pressure on to meet the production deadlines.

It is our sincere hope that our efforts will meet high expectation of the readers.

Cleveland, OH, USA — Catherine J. Hwang, MD
Salt Lake City, UT, USA — Bhupendra C. K. Patel, MD
Cleveland, OH, USA — Arun D. Singh, MD

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To my husband, JD, my life partner and who edited the first and second editions of this book. To my parents, Steve and Nancy, who inspired me to be the person and surgeon I am today.

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Examination Techniques

1

Catherine J. Hwang and Julian D. Perry

Introduction

Examination of a patient with orbital disease begins with a detailed history to discern the chronicity of symptoms, past medical history including systemic medical conditions or neoplasia, and past surgical history and review any corresponding imaging. Orbital examination techniques in the adult and child will help establish differential diagnoses and direct further studies.

History

The history aids in establishing a probable diagnosis and in guiding the initial workup and therapy. Important historical elements will be discussed in the following chapters of this section.

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Examination

External Examination

External examination with visual inspection is critical, assessing the position and symmetry of periocular structures, such as the brows, eyelids, canthi, surrounding soft tissues, and bony structures. Visual inspection should include observation for obvious globe deviation, including the direction. Grossly visible changes in the periocular skin and preauricular or submandibular lymph nodes and asymmetries are noted.

Pupils

All patients with suspected orbital disease should undergo a pupillary examination, to help aid in determining optic nerve function. The swinging flashlight test to determine the presence or absence of a relative afferent pupillary defect is helpful to ascertain possible compression of the optic nerve or disruption of the visual system between the optic nerve head and the apex of the orbit. Optic nerve function is further characterized by testing of visual acuity, color plates, and confrontational fields. The efferent pupillary pathway should be tested as well. Anisocoria should be recorded as worse in light (parasympa-

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thetic defect) or in dark (sympathetic defect), and pharmacologic testing can be performed.

Tumors of the lateral orbit may impair ciliary ganglion function to produce a parasympathetic defect, whereas cavernous sinus or superior orbital fissure tumors may result in sympathetic dysfunction.

Extraocular Motility

Extraocular motility (EOM) should be tested in every patient, documenting restriction in motility as well as diplopia. EOM restriction can be documented in either a percentage from 1 to 100, with 100 being normal, or on a scale ranging from −4 to +4, with 0 being normal. If a phoria or tropia is found, the cover–uncover test can be useful to help measure deviations with the aid of prisms.

Reduction of EOM can either be a restrictive process or a palsy. To differentiate restriction versus a palsy, patients may undergo forced duction testing. Classically, after a drop of topical anesthetic is placed, a cotton-tipped applicator soaked in 4% lidocaine solution is applied to the muscle away from the direction of gaze limitation for approximately 1 min. The anesthetized muscle is then grasped firmly with toothed forceps and rotated toward the direction of gaze limitation. Resistance indicates a restrictive disorder; free movement is more likely a palsy. If the patient is not amenable to such testing while awake, one can discern restrictive disease from paresis by looking for a "floating" saccade or, basically, the relative speed and comparison of the simultaneous saccades between the two eyes. Standing approximately 3–4 ft directly in front of the patient, the examiner should ask the patient to look at the examiner's nose and then quickly look at his or her finger on an outstretched arm in the four main positions: left, right, up, and down. For example, if the patient has an abduction deficit on the right from 6th nerve paresis, he or she will have a saccade that "floats" to the right, when compared to the fast adducting saccade on the left. If the abduction deficit is due to restriction, the right eye abducting saccade will be limited by a sudden stop.

Fields of single vision and double vision can be mapped using a penlight; Finoff transilluminator, aka muscle light; or a kinetic perimeter.

Eyelid Position and Function

Eyelid position is characterized by marginal reflex distances (MRD). The MRD1 represents the distance from the center of the upper eyelid margin to the corneal light reflex measured in millimeters. The MRD2 represents the distance from the center of the lower eyelid margin to the corneal light reflex. The action of the levator muscle (levator function) is measured as the extent of upper eyelid excursion from downgaze to upgaze with the brows fixated. If present, scleral show is measured from each limbus to the corresponding eyelid margin with the eye in primary position. Upper eyelid ptosis (Fig. 1.1) may imply either mechanical involvement of the levator muscle or palsy, whereas eyelid retraction (Fig. 1.2) suggests proptosis, such as thyroid eye disease or CNS disorder. The upper eyelid may be everted to inspect the palpebral lobe of the lacrimal gland (Fig. 1.3) or by having the patient look down and in and lifting up the upper eyelid. An s-shaped deformity characterized by ptosis and edema laterally is usually associated with

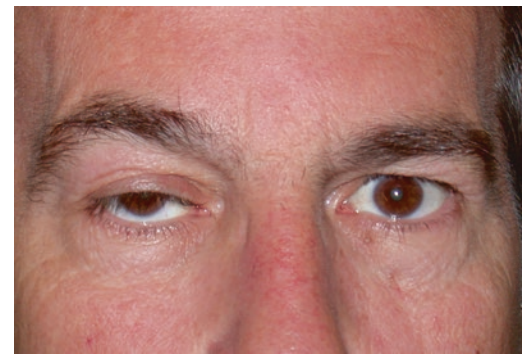

Fig. 1.1 Right upper eyelid with ptosis. Note the right brow is also elevated due to the patient's use of the frontalis muscle in an attempt to lift the ptotic right upper eyelid. The left upper eyelid is also pseudo-retracted and would likely descend to a more normal position with ptosis correction on the right

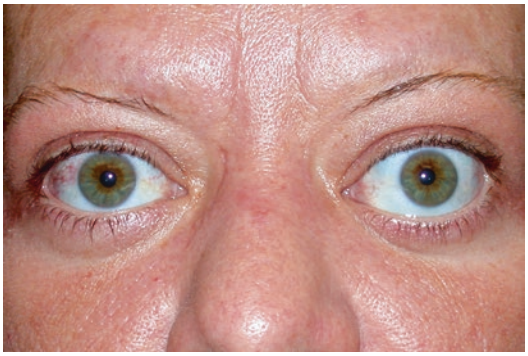

Fig. 1.2 Bilateral upper and lower eyelid retraction, left greater than right from thyroid eye disease

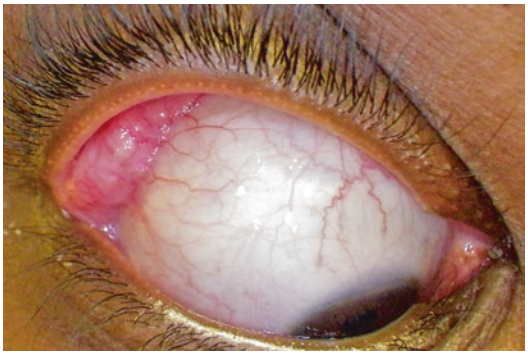

Fig. 1.3 Prominent palpebral lobe of lacrimal gland, visible beneath the upper eyelid

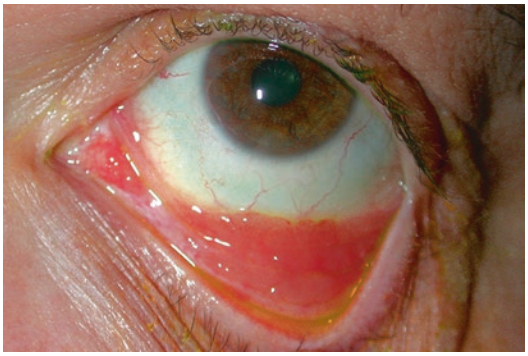

Fig. 1.4 Salmon-colored lymphoma in the inferior fornix

lacrimal gland enlargement. Lymphoma can result in a salmon-colored conjunctival mass that is visible upon inspection of the fornix (Fig. 1.4). Orbicularis strength, Bell's phenomenon, and lagophthalmos should also be evaluated as part of the cranial nerve exam detailed below.

Globe Position

Proptosis

By evaluating the patient in the submental view (chin-up position), the examiner can qualitatively look for globe protrusion or retrusion relative to the canthal angle and the nasion (Fig. 1.5). To quantify the degree, three common exophthalmometry tools exist: the Hertel, which is most commonly used (Fig. 1.6); the Naugle, which is useful for patients with abnormal lateral orbital rims (Fig. 1.7); and the Luedde, which is more feasible to use in children (Fig. 1.8).

The Hertel exophthalmometer quantifies the anterior protrusion of the eye by measuring the distance in millimeters from the anterior lateral orbital rim to the front surface of the cornea. The reading is taken with a base measurement of the separation of the positioning arms of the tool to help reference subsequent measurements on the same device. The Naugle exophthalmometer measures anterior globe position relative to the superior and inferior orbital rims. This method provides a more accurate assessment in those with lateral rim fractures, iatrogenic repositioning of the lateral rim, or orbital rim defects. The Luedde exophthalmometer measures globe protrusion unilaterally from the lateral orbital rim.

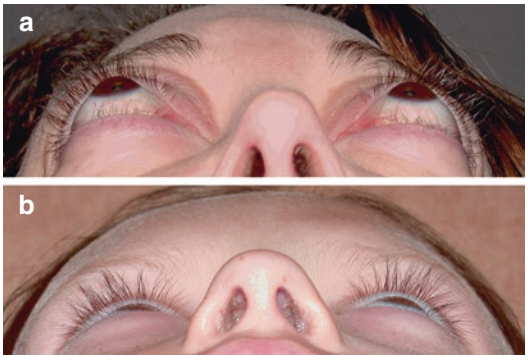

Fig. 1.5 Submental view of proptotic globes from Graves' disease (**a**). Child with left proptosis from orbital dermoid (**b**)

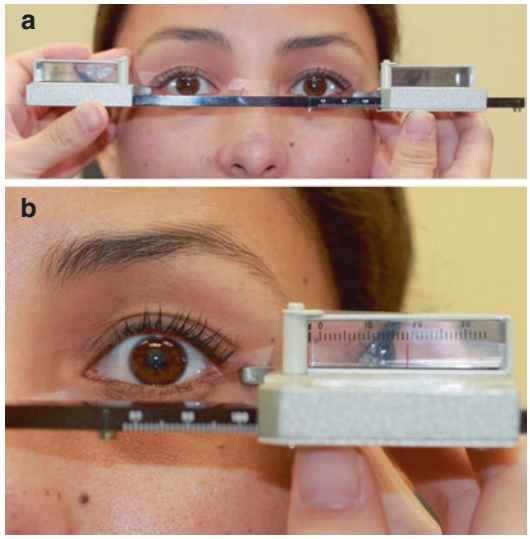

Fig. 1.6 Hertel exophthalmometer. While resting the Hertel instrument on both lateral rims, the base number is recorded on the ruler for consistency (**a**), and the amount of exophthalmos is measured by aligning the red bars then recording the number at which one sees the anterior surface of the cornea (**b**). The examiner and patient should be at eye level

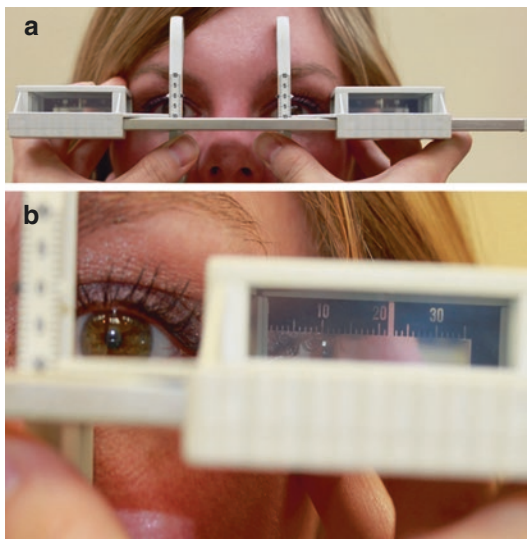

Fig. 1.7 Naugle exophthalmometer. In patients with lateral orbital rim defects, the Naugle can be used by resting the posts on the forehead and the maxillary prominence at the pupillary axis (**a**), aligning the red mark with the clear bar, and then recording the number at the anterior surface of the cornea (**b**)

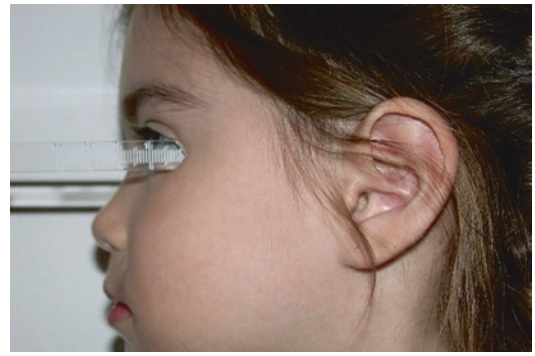

Fig. 1.8 In children, the clear Luedde ruler is placed at the lateral orbital rim, and the distance to the anterior corneal surface is measured

It consists of a clear bar with millimeter markers. The anterior corneal surface can be visualized through the bar to determine the millimeters of protrusion. This can be positioned on the lateral orbital rim without a device in front of the eyes and is easier to use in children who reflexively more away and close their eyes with the other tools.

Hyperglobus or Hypoglobus

Orbital or periorbital neoplasms often displace the globe. Nonneoplastic conditions such as thyroid eye disease, trauma, and silent sinus syndrome may cause similar examination findings, and further imaging studies, such as computed tomography (CT) or magnetic resonance imaging (MRI), may be indicated.

Horizontal and vertical globe displacements are measured in millimeters from the central pupil to vertical midline and horizontal canthal line, respectively. For vertical displacement, one can draw an imaginary line horizontally across a patient's pupillary axis and determine if the pupil of the other eye is higher or lower, which could suggest hyperglobus (Fig. 1.9) or hypoglobus (Fig. 1.10), respectively. Care must be taken to ensure the patient's head is in primary position, without any tilt, and that the line is parallel to the ground.