Henderson

ORBITAL TUMORS

Second Edition

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Second Edition

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(*Above*) A very early illustration of an orbital tumor in a 56-year-old man. The tumor was removed with the surgical tool lying on the table in front of the patient.
(*Below*) The enucleated pathologic specimen. The patient survived the operation.
(*From Fabricius von Hilden W* [*Fabricii Hildani G*]: Wund-Artzney, gantzes Werck, und aller Bücher, so viel deren vorhanden. [Auss dem Lateinischen in das Teutsche Sprach übersetzt durch Friderich Greiffen.] Frankfurt am Main, Johann Beyers, 1652, pp 3, 13.)

Affectionately dedicated to
Nadine, Sally, and Holly

Preface

Eight years have passed since the manuscript was finalized for the first printing of *Orbital Tumors*. In this interval, several major advances have occurred in this field. Foremost in overall importance has been the burgeoning application of computed tomography to orbital diagnostics. In this short interval, the technology of computed tomography has passed from its infancy to a sophisticated family of fourth generation scanners. This has provided a window to the orbital deep by which soft tissue masses are now visualized; a wonderment beyond imagination at the time I started clinical practice.

Also noteworthy have been the changing concepts in the terminology, diagnosis, and treatment of some of the individual tumors. Malignant lymphomas, for one, have undergone such changes in their histopathologic classification that new names have made passé such old and familiar terms as *reticulum cell sarcoma* and *lymphosarcoma*. In addition, the use of immunochemistry in the entire spectrum of lymphoproliferative diseases may, in the future, provide a supplementary means for differentiating the malignant from the nonmalignant tumors as well as the individual lymphomas from each other. The scourge of orbital rhabdomyosarcoma has been definitely mitigated by the use of adjuvant chemotherapy and radiotherapy, and the treatment protocols of cooperative treatment centers have made possible a controlled judgment of prognosis. In addition, newer orbital lesions, such as Wegener's granulomatosis, have been sufficiently documented to warrant inclusion in the classification of orbital tumors. Electron microscopy has become a most useful supplement to light microscopy in determining the tissue genesis of some tumors of equivocal origin. All of this and more has made parts of the first edition woefully out-of-date.

Finally, in the eight-year interval, there has been a priceless opportunity to extend further the follow-up observation of the original consecutive series of 465 patients and to enlarge the scope of our study to a total of 764 consecutive cases. This has permitted an observation of some patients for 27 years or longer and has revealed new information concerning the long-term (30 to 40 years) or natural history of some tumors. Such data have revealed the minatory behavior of many benign recurrent mixed tumors and orbital meningiomas, and the lethal course of orbital hemangiopericytomas; features heretofore suspected but not well documented. Our concept of the clinical course of these as well as other tumors has been revised accordingly by the lengthened period of study.

All this has made a major revision of *Orbital Tumors* desirable, but such an endeavor could not be undertaken without consideration of the economic realities of such a publication. Simply stated, the spiraling costs of printing must be weighed against the still narrow readership appeal of the subject relative to the more populated subspecialty groups. Our publisher, Brian C. Decker, believed such a project feasible and W.B. Saunders, publisher of the first edition, graciously released the various copyright restrictions.

In keeping with budgetary guidelines, all copy of the first edition was carefully revised and re-edited so as to make room for all new subject material without increasing the original size of the text. Also, in the interests of conserving space, we have followed the custom of the former edition in listing only key references at the conclusion of each chapter rather than duplicating in-depth bibliographies that can be found elsewhere in the literature. We believe these alterations will still provide an attractive updating of the text for the benefit of the faithful readers of the first edition and provide a stimulating overview of the subject to a newer generation of ophthalmologists.

In preparing this revision I have again relied heavily for advice and counsel on my close collaborater, Dr. George Farrow, and his associates in the Department of Surgical Pathology of the Mayo Clinic. Dr. Peter Banks and Dr. Edward Soule, also of this department, generously provided data important to the updating of the chapters on malignant lymphoma and rhabdomyosarcoma. Other standbys from the staff of the Mayo Clinic who helped in the preparation of this as well as the first edition were Dr. Omer Burgert, Jr. (Pediatrics), Dr. Martin Van Herik (Radiophysics), Dr. Colin Holman (Radiology), and Dr. Kenneth Devine (Surgical Oncology of the Head and Neck). Dr. Thomas McDonald (Otorhinolaryngology) shared with me his viewpoint on some of the tumors secondarily invading the orbit from sources in the nose, nasopharynx, and paranasal sinuses. Other sources of information and new figures are carefully acknowledged in the text. Mr. John Prickman of the Department of Publications, Mayo Clinic, and Mary Donchez of New York shared in editing and assembling the revised manuscript. Dolores Sexton willingly typed the many addenda so necessary to a revised manuscript. Most important, during the two-year period of preparation, was the steadfast backing of the publisher, Brian C. Decker.

Now back, once again, to the cushioned chaise lounge for rest and recuperation, but ever curious about the progress of orbital tumor patients.

John Warren Henderson, M.D.

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SECTION I

Applied Anatomy of the Orbit

Diagnosis of Orbital Tumors

1

Applied Anatomy of the Orbit

Although the orbit is defined by some in terms of its bony components and contours, others regard it as a cavity or space in which are housed those tissues and organs contributing to the function of the eye. The former is properly termed the *bony orbit* and the latter, the *orbital cavity*. In medical parlance the word *orbit* frequently includes both the space and its surrounding bone, that is, the *bony socket*. Throughout this text the word orbit will be used with its broadest denotation, and the reader may judge from the context whether the reference is to orbital bone or socket cavity, or both. The eye, of course, is the principal resident of this compartment, the bones and adnexal structures being carefully adapted for its protection and support. Nature has designed this orbit so nicely that the eye, particularly anteriorly, fits as smoothly as a hand in a glove and almost as snugly as a cork in a bottle.

It follows, then, that when something goes amiss with this contact arrangement – such as the edema of inflammation, the swelling of an expanding tumor, or the dilatation of vascular channels – direct pressure is exerted on the eye. The result is forward expulsion of the eye from the protective environs of its orbit.

GENERAL ORBITAL TOPOGRAPHY

Among the various descriptions of the orbit, Whitnall's (1932) is one of the most graphic. He compared the orbit to a pear, the anterior orbital aperture corresponding to the base of the pear and the optic canal, to its stalk. The volume of this orbital spaced is only 29 ml. The *measurements* of the principal meridians of the orbital cavity are noted by all texts of anatomy and, as these values are so nearly alike, it is convenient to consider its size (in adults) by taking a standard of 40 mm for each meridian: that is, for height, width, and depth.For practical purposes the anterior aperture of the orbit in diameter is about the size of a half-dollar; in children and infants these values are correspondingly less.

Just as the body of the pear has a greater circumference than its base, so too the roomiest portion of the orbit is about 15 mm inside the orbital rim. This space would be quite adequate for the exploring finger were it not for the protective overhang of the circumference of the orbital rim. Access to the orbital cavity through this narrow space is easier on the nasal and inferior sides of the orbit. The width of these avenues of approach is about a finger's breadth. To facilitate palpation of orbital structures through these narrow apertures, some physicians use the little finger, but the proprioceptive sense of this digit is not nearly as acute as that of the index finger.

The *anterior portion* of the orbit, wherein are lodged the eye and lacrimal gland, is roughly quadrangular. Posterior to these structures there seems less need for support, and the bony floor merges into the other three walls of the compartment, which now becomes triangular. Although the volume of the orbit is reduced, adnexal tissues are not as crowded in this area as they are more anteriorly. The transfrontal and lateral surgical approaches, by circumventing the eyeball and lacrimal gland, have proved more useful in reaching this retrobular space than the older route of anterior orbitotomy.

At the *apex*, where the three remaining walls merge, are located the posterior outlets of the orbit: the *optic foramen* and *superior orbital fissure*. The former, with its prolongation posteriorly making the optic canal, is frequently unroofed by the neurosurgeon either during his dissection for meningioma or during the excision of a glioma extending along the optic nerve. This remote area is approximately 5 cm from the nearest skin surface. The ophthalmic surgeon also must be well acquainted with this apical area in order to facilitate removal of "intra-orbital" meningiomas, which are encountered rarely in the course of lateral orbitotomy, and resection of long sections of the optic nerve in cases such as intraocular retinoblastoma and optic nerve glioma. However, neither neurosurgeon nor ophthalmologist is anxious to disturb structures adjacent to the superior orbital fissure, for through it pass at least eight nerves of assorted diameters and several blood vessels. This is a veritable "no man's land."

The student or the young surgeon rummaging around the rear of the orbit for the first time usually has some misconceptions as to the exact location of these orifices. Figure 1 shows their position as viewed from the front, but the photographer or medical illustrator often positions the skull at an angle to reveal best all the major bony structures and foramina of the orbit (Fig. 2). In such illustrations, the optic foramen appears to be located close to the intersection of lines drawn through the vertical transverse diameters, and the illusion is given that the orbit is cone-shaped in a direct anteroposterior axis, the apex of the cavity being nearly equidistant from the four sides. The observer may not realize, however, that such views do not represent a true front view of the orbit. Instead, the line of visual regard is actually parallel to the axis of the optic canal and, when such a line is projected forward, it passes through the anterotemporal portion of the orbit, not the true anteroposterior meridian of the skull. Actually, the position of the optic foramen is more medial and superior to that represented in such photographs. If one aligns the orbital cavities of a skull with the observer's eye level, an imaginary line running through the sagittal plane of the observer's nose and the nasal bone of the cadaver, the optic foramina will be directly posterior along the medial walls of the orbit (Figs. 1 and 2). If the observer's head is kept steady and not moved laterally, the right optic foramen of the skull will almost be concealed from the view of the observer's left eye by a slight lateral bulge of the medial wall as it passes along the ethmoid sinus; the situation is similar when the observer tries to locate the position of the left optic foramen with the right eye. This knowledge of the true versus the false location of the apex of the orbit is pertinent to those surgical situations wherein the orbit is approached from the anterior or lateral aspect; the optic foramen is further from the surgeon and, surprisingly, more inaccessible than

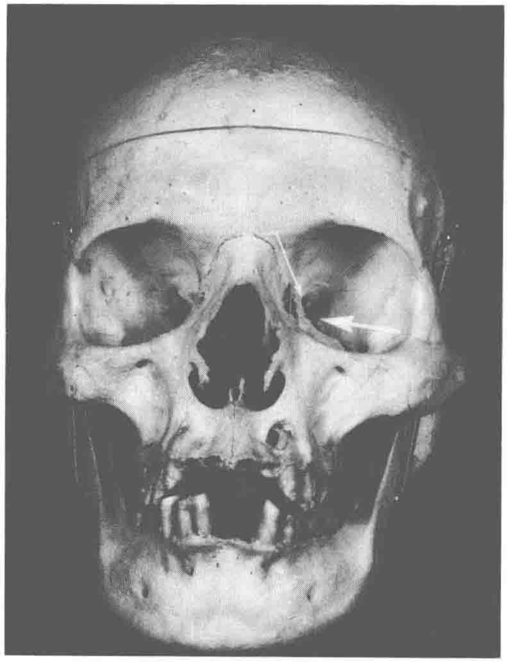

Figure 1. True anterior view of orbits. Camera has been positioned along midline axis of skull. Right optic foramen is concealed by slight lateral bulge of medial wall of orbit as it passes along ethmoid sinus. Left optic foramen is only slightly visible as a small dimple (*small arrow*). *Large arrow* superior orbital fissure.

Figure 2. Anterolateral view of orbit. Camera has been shifted 35° along an arc away from midline. Here true size of optic foramen is clearly visible, but opening appears centered along rear wall of orbit. (*Small arrow*, optic canal; *large arrow*, superior orbital fissure.)

expected. Although the visual axis of the eye is on a saggital plane, the antero-posterior axis of the orbit is offset laterally.

ORBITAL WALLS

The makeup and configuration of the bony walls and rim of the orbit and the application of this knowledge to the problem of orbital tumors are chiefly the concern of the roentgenologist and the surgeon. The superior, nasal, and inferior walls are of greater interest to the former, for it is through study of these barriers that roentgenographic evidence of invasion of the orbit by tumors in adjacent cavities is usually found, and primary tumors may erode these walls in their effort to escape the orbit. (Further comment on the nature of these changes will be found in Chapter 2.) From a surgeon's standpoint the orbit may be entered from any direction, but those anatomic features relating to the superior and lateral rims and walls are more pertinent to the modern surgical approaches.

Superior Wall. The superior wall, or roof, is rather thin throughout its extent, and the periorbita easily peels away from its undersurface. On the intracranial side the dura can be lifted almost as easily. Because the roof is perforated neither by major nerves nor by blood vessels, it is easy to nibble away this bony partition with rongeurs without serious consequences, a fact that is utilized in the transfrontal approach to the orbit. Except when eroded by mucoceles or epidermoid cysts, the roof is an effective barrier that prevents accidental contact with dura underneath the frontal lobes during anterior and lateral orbitotomy.

Lateral Wall. Because the lateral orbital surgical approach is popular, the lateral wall concerns the ophthalmic surgeon more than other bony partitions. In practice, it has some similarities to the roof: it is almost as devoid of foramina, and its anterior portion can be broached without serious hemorrhage; the zygomaticotemporal vessels and nerves piercing the bone just posterior to the midportion of the lateral rim usually do not pose a problem; and the periorbita is reasonably easy to separate from the bone, even in the area of the orbital tubercle where several fascial bands and tendons insert. However, at this point any similarity to the roof of the orbit ceases.

Any exterior approach to the lateral side of the orbit must take into account the tough and thick *lateral rim*, which is but the forward extension of the lateral wall (Fig. 2). This rim is the strongest portion of the orbit, and even the suture line between the zygoma and the frontal bone is not easily jimmied except by the heaviest chisels, saws, and drills. Even so, malignant tumors of the lacrimal gland seem to erode this tough bone as easily as they exert pressure on the eye. That portion of the zygomatic bone constituting the lateral rim and the anterior one-third of the lateral wall protects the posterior half of the eye; the anterior half of the eye thus has an unencumbered vista laterally (Fig. 3). Palpation of retrobulbar tumors, therefore, should be easier from the lateral rather than from the nasal side of the eyeball, but the lacrimal gland, which fills the anterior portion of the superotemporal quadrant, largely nullifies this anatomic advantage. The lateral wall, which is thick along its front surface, becomes thinner as it passes posteriorly to its junction with the forward and lateral extension of the greater wing of the sphenoid bone.

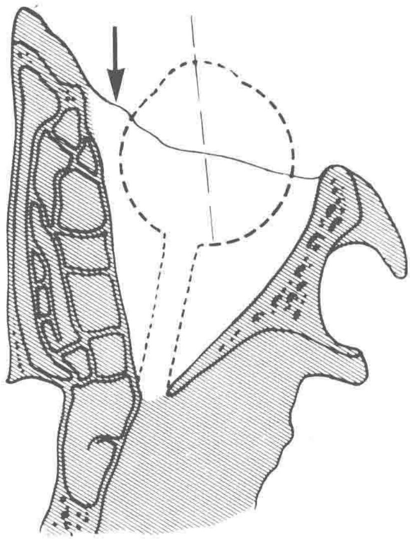

Figure 3. Transverse section of right orbit with diagrammatic representation of eye in relation to nasal and lateral bony walls. Anterior half of eye on lateral surface is not protected by bone. (*Arrow* indicates approximate line of superior bony rim of orbit superimposed on drawing.)

This portion of the sphenoid bone, comprising the posterior two-thirds of the lateral wall, is also wedge-shaped but in a direction inverse to the wedge of the adjacent zygomatic bone. This configuration of the sphenoid makes it difficult to dissect or remove bone posterior to the suture line; biting forceps or drills encounter thicker and thicker bone with but little additional exposure of the orbital contents. Another hindrance to extensive removal of bone beyond this point is the possibility of encountering orbital branches of the middle meningeal artery and vein that penetrate the sphenoid bone posterior to the zygomatic-sphenoid suture; bleeding from this source in the depth of the orbit can unexpectedly complicate manipulations and can be difficult to stem. The zygomatic-sphenoid suture is an important landmark because along this line, bone invariably fractures when one is creating the flap common to all modifications of the Krönlein operation (Fig. 4.) Once the bone flap has been turned, the

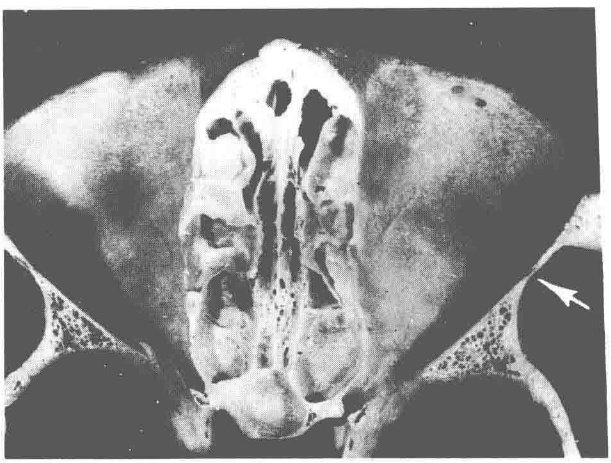

Figure 4. Mid-horizontal section through orbits (superior halves): note triangular shapes of bony orbits, broad expanse of orbital roofs, parallel position of medial walls, and thin portion of lateral walls (*arrow*) corresponding to suture line between left sphenoid and zygomatic bones. Dehiscence is present in roof of right orbit. (From Berke RN: A modified Krönlein operation. Trans Am Ophthalmol Soc *51*:193–227, 1953. By permission of the American Ophthalmological Society.)