Diseases of the Orbit

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Preface

Sufficient clinical and pathologic data have accumulated on diseases of the orbit so that this area has evolved as a distinct subspeciality within ophthalmology. The field has further developed due to technologic advances such as the diagnostic use of ultrasonography, computerized tomography, and electron microscopy. Greater precision in diagnosis has synergized with the emergence of new medical and surgical treatments. In preparing this volume we have drawn generously upon the knowledge of experts in the field of orbital disease to provide us with supplementary chapters. With the discovery of gaps in our knowledge that comes from preparing such a volume, we hope the spur for continuing improvements in the diagnosis and management of orbital disease will be sharpened.

We would particularly like to thank the following people for their assistance in the preparation of this volume: Ms. Carol Miller, Mrs. Pat Fallahy, and Mrs. Rita Dolan for their secretarial assistance; Mrs. Alice Roberts for cooperating in the collection of many fresh tissue samples that we have studied by electron microscopy; and Ms. Ida Nathan for her expert photomicrography, which contributes so much to the aesthetic and scientific quality of this book. We are particularly indebted to Dr. Algernon B. Reese and the Eye Cancer Foundation for financial support throughout this project.

Ira Snow Jones
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1979

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Patient Examination and Introduction to Orbital Disease

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A few remarks are in order regarding the organization and objectives of this section. We have attempted to write a comprehensive but not an encyclopedic review of neoplastic and nonneoplastic orbital diseases. Clinical and microscopic illustrations have been generously employed to facilitate familiarity with the material. We have also attempted to integrate some of the recent developments in pathology and the technologies of diagnosis, such as ultrasonography, computerized tomography, and electron microscopy, as they relate to orbital disease.

The first portion of this section deals with various ways of approaching orbital disease. Some material is covered from multiple standpoints such as diagnostic modalities, pathology, embryology, and neuro-ophthalmology. It is hoped that the reader will find this a suitable gridwork for handling the information that follows in the later chapters.

The organization of the later chapters relating to specific diseases was prompted by considerations of clinical logic and the relative clinical importance of various topics rather than by procrustean beds based on cell types and orbital locations. The subject of inflammation is covered extensively because the manifold types of inflammation are the most frequent cause of orbital disease and proptosis. Vascular disease, lymphomas, lacrimal gland tumors, peripheral nerve tumors, optic nerve glioma, rhabdomyosarcoma, and metastatic and secondary tumors are subjects of considerable clinical importance in terms of diagnosis and management.

We have not attempted to compress or telescope into this introductory chapter all that follows. At the end of this chapter, lists are provided of the various kinds of orbital disease and orbital findings (Tables 1-16 through 1-24); these are not meant to represent comprehensive enumerations but are only abbreviated ways of organizing rafts of data in a small space. The clinician will doubtlessly encounter diseases that do not fall into these categories, and he should not attempt to exculpate himself for failing to make a diagnosis because a list did not contain the correct diagnosis. This chapter also contains a brief review of the most common orbital diseases. It is helpful to have capsule outlines in one's mind of the most common orbital diseases and their clinical characteristics and settings.

BASIC ANATOMY AND TOPOGRAPHIC PATHOLOGY OF THE ORBIT

A full appreciation of the rich complexity of the three-dimensional relationships of the orbital contents is not required in order to understand most inflammatory and neoplastic diseases of the orbit. Figures 1-1 through 1-6 represent in simplified line diagrams the essentials of orbital anatomy. As a generalization, one can say that there is a definite tendency for inflammatory pseudotumors, tumors, and cysts of the orbit to occur in the upper quadrants.

ORBITAL BONES

Seven bones contribute to the orbital walls (frontal, sphenoid, maxillary, zygomatic, palatine, ethmoid and lacrimal) and create an enclosure with a volume of 26 to 30 cu ml (Figs 1-1 through 1-3). The orbital aditus is quadrangular, and the orbital contour is pear-shaped, the stem being the optic canal. The narrowest portion of the orbit is the apex, which is found medially; from this deep point the slant and progressive widening of the orbit are directed laterally. Thus rapidly developing orbital tumors situated in the retrobulbar space, eg, rhabdomyosarcoma, have a tendency to push the globe forward and down and out, in the path of

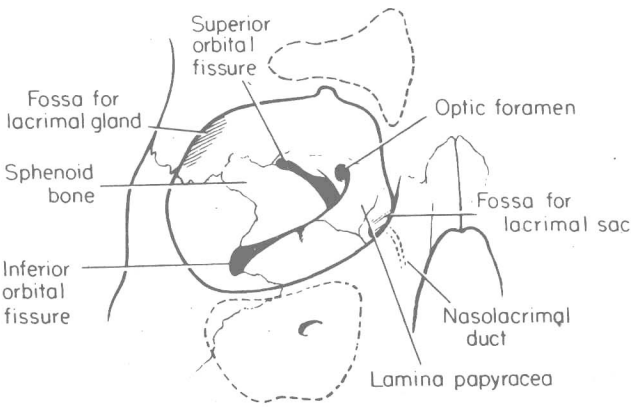
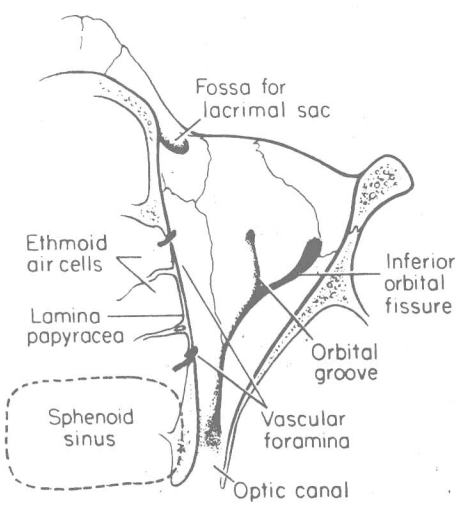


Fig 1-1. View of right orbit from front. Major posterior openings are the optic foramen which transmits optic nerve, ophthalmic artery, and sympathetic fibers; the superior orbital fissure which transmits central retinal and superior ophthalmic veins, cranial nerves III, IV, V-1 and VI, some sympathetic fibers, and anastomotic artery from the middle meningeal to the ophthalmic or lacrimal artery; and the inferior orbital fissure which transmits inferior ophthalmic vein. Sphenoid bone contributes to lateral and posterior orbital walls and provides optic strut. Dotted lines delimit the frontal sinus (above) and maxillary sinus (below).

least resistance dictated by the lateralward conformation of the orbit. The thinnest bone is the lamina papyracea, which separates the medially situated ethmoidal air cells from the orbit. It can be easily breached by inflammatory and neoplastic processes that originate in the region of the ethmoidal air cells as well as by rough dissection during surgery. Small foramens in the medial orbital wall allow communication between the vasculature of the orbit and ethmoidal sinus; similar communications are not found between the orbit and the frontal, maxillary, and sphenoidal sinuses.

Fig 1-2. View of floor of right orbit from above. Ethmoidal air cells are separated from orbit by thin lamina papyracea. Anterior and posterior ethmoidal arteries (branches of ophthalmic artery) pass through foramens in the medial wall. Sphenoidal sinus is situated posterior to ethmoidal air cells, and pathologic processes in sphenoidal sinus usually involve posterior ethmoidal cells as well.



Dermoids are found at sutural sites where surface ectoderm is presumably pinched off during embryonic development of the orbital bones. Although the superotemporal orbital rim is the usual site for dermoids, they can also be found deep in the orbit at the apex, along the superior orbital fissure, and, occasionally, intradiploically in the sphenoid bone.

Major defects in the sphenoid bone complex occur in neurofibromatosis and result in pulsating exophthalmos, since the arterially induced cerebrospinal fluid pulsation is transmitted directly to the orbital soft tissues. Anteromedially situated encephaloceles may cause bony defects, although not uncommonly a portion of brain tissue and/or meninges may become sequestered within the orbit without an apparent bony defect. It may also be difficult to identify preoperatively an encephalocele that extends through a preexistent normal aperture, such as the superior orbital fissure. Posterior, deep orbital encephaloceles can cause pulsating exophthalmos on the same basis as occurs in neurofibromatosis.

Benign tumors that grow slowly in juxtaposition to the orbital bones create fossae. Benign tumors of the lacrimal gland inconstantly produce a fossa superotemporally, whereas dermoids create regularly contoured, rather more punched-out defects in the same region. Elsewhere in the orbit, usually superiorly, fossae are due to slow-growing tumors such as neurofibromas, neurilemomas, leiomyomas, and cavernous hemangiomas.

In older individuals, irregular bone destruction in the region of the lacrimal gland should suggest a malignant tumor; cancers originating in the ethmoidal or maxillary sinuses produce bone dissolu-

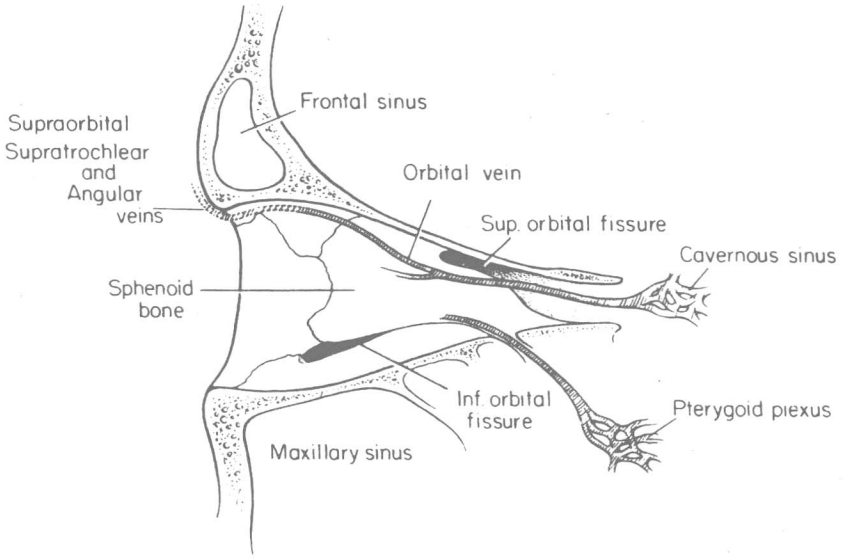


Fig 1-3. View of lateral aspect of right orbit from nasal side. Most of the orbital venous blood travels through the superior orbital fissure to the cavernous sinus. Anastomoses are shown between superior orbital (ophthalmic) vein and supraorbital, supratrochlear, nasofrontal, and angular veins of forehead and medial face and nose. Inferior orbital (ophthalmic) vein passes through the inferior orbital fissure to the pterygoid plexus.

tion of the medial and inferior orbital plates, respectively. Meningiomas, especially the en plaque type originating along the sphenoid, produce reactive hyperostosis. Specialized x-ray techniques, such as hypocycloidal tomography and computerized tomography, may be required to demonstrate subtle degrees of orbital bone destruction. Fibrous dysplasia has a tendency to involve the roof of the orbit and the sphenoid bone and may cause visual acuity problems if the sphenoid lesion encroaches on the optic canal. In fibrous dysplasia, x-ray changes may suggest a meningioma, but patients with fibrous dysplasia are younger than those with sphenoidal meningiomas. Ossifying fibromas can involve the orbital and sinus walls and are more aggressive, recurrent lesions than those of fibrous dysplasia. Osteogenic sarcoma usually begins in the bones of the sinuses or in the bones at the base of the skull rather than in the orbital bones. Patients with retinoblastoma who have received radiotherapy are liable to develop osteogenic sarcoma.

Tumors may begin in the orbital bones and erode into the orbit. Two tumors have a predilection for the maxilla, from which they invade the orbit: Burkitt's lymphoma and the rare retinal anlage tumor (benign pigmented neuroectodermal tumor of infancy). Among histiocytic neoplasms, eosinophilic granuloma is commonly situated at the superotemporal orbital rim and can resemble a dermoid; however, x-ray studies may reveal more irregular bone destruction in eosinophilic granuloma than that seen in dermoids. Hand-Schüller-Christian disease produces punched-out skull lesions, and if these tumors involve the base of the skull and orbital bones, diabetes insipidus and exophthalmos can result. Although most carcinomas metastasize to the soft orbital tissues, some malignancies may metastasize to the orbital bones: neuroblastoma has a tendency to metastasize to the zygomatic bone. Conversely, although prostate, urinary bladder, kidney, and thyroid carcinomas all have a propensity to settle in bone, they rarely metastasize to the orbit. Granulocytic sarcoma may present as an orbital osseous or periosteal mass before blood or bone marrow signs of leukemia are present; these signs almost always develop within several months of the orbital tumor. Lymphomas usually do not produce orbital bone destruction; exceptions include lethal midline granuloma, now considered in some cases to be an atypical, central facial reticulosis, and other lymphomas originating in the sinuses or craniofacial bones. Rare instances

of multiple myeloma may involve the orbital bones. Wegener's granulomatosis may also destroy the orbital bones by spreading from adjacent sinuses.

Concentric enlargement of the orbit can be seen in lesions presenting congenitally, such as teratoma, encephalocele, rhabdomyosarcoma, and in neurofibromatosis, but can also be seen in any long-standing slowly growing tumor. Practically any tumor, from cavernous hemangioma to lymphoma, can cause this change. Unusual orbital bone configurations and shallow orbits can be seen in the various craniostenoses and dysostoses. Optic atrophy in many of these diseases is not so much due to narrowing of the optic canal as to the associated hydrocephalus.

Blunt trauma to the orbit can cause an orbital floor fracture with entrapment of the inferior rectus muscle resulting in an inability to look up, a mechanical or restrictive problem and not a neurogenic one. More massive trauma to the central face can cause trimalar fractures involving the orbital rim, LeFort I and II fractures of the facial bones, and basal skull fractures.

SINUSES

Four sinuses (Figs 1-1 through 1-3) surround the orbit: superiorly, the frontal sinus; posteromedially, the sphenoidal sinus; medially, the ethmoidal air cells; and inferiorly, the maxillary antrum. Inflammations are the most common diseases of these structures. Since the ethmoidal air cells are the first to develop in childhood, acute ethmoiditis predominates as the chief childhood sinus disease. In children, ethmoiditis, if unchecked, gains ready access to the orbit through the thin lamina papyracea; therefore, it is one of the most common causes of proptosis and of orbital cellulitis in children.

The frontal and ethmoidal sinuses are the most common sites for the development of mucoceles. Interference with drainage causes slow expansion of the sinus in all directions. Frontal sinus mucoceles can cause enlargement of the forehead and downward displacement of the eye, whereas ethmoidal mucoceles can cause lateral displacement of the eye. Mucoceles create little difficulty with visual acuity, but they may interfere with extraocular motility in extremes of gaze. Maxillary and sphenoidal mucoceles are quite rare. When sphenoidal mucoceles do develop, they encroach on the posterior ethmoidal air cells, so that a precise origin is impossible to determine; these are best described as sphenoethmoidal mucoceles. Sphenoidal mucoceles are the most difficult to diagnose; they can cause headaches that simulate migraine and can even cause cranial nerve palsies.

Osteomas are another important sinus lesion. Osteomas of the frontal and ethmoidal sinuses are most likely to encroach on the orbit; maxillary sinus osteomas are also common.

Carcinomas that originate in the maxillary and ethmoidal sinuses usually invade the orbit; most of these are squamous cell carcinomas. The ethmoidal sinus is also a source of adenocarcinoma. Both the frontal and sphenoidal sinuses appear to be relatively immune to the development of carcinoma.

Fungal infections that invade the orbit usually originate in a sinus. Mucormycosis (phycomycosis) and aspergillosis are the two most frequent examples of this type of disease. The clinical course of mucormycosis is rapid and usually fatal; aspergillosis causes a more insidious exophthalmos but is also quite dangerous and often fatal. Wegener's granulomatosis and lethal midline granuloma are other "inflammatory" conditions that can invade the orbit from surrounding sinuses and cause bone destruction. The former may additionally cause orbital disease via vasculitis of the orbital vessels.

ORBITAL OPENINGS

The optic canal, superior orbital fissure, inferior orbital fissure and groove, and ethmoidal foramens are the major, normally occurring interruptions in the orbital walls. The optic canal is 5 to 10 mm long and is created by the two roots of the lesser wing of the sphenoid. It transmits the optic nerve, the ophthalmic artery, and sympathetic nerves. The optic canal can be enlarged by optic nerve gliomas and meningiomas. In x-ray studies of the optic nerve glioma, the outline of the canal is smooth and regular; in x-ray studies of meningiomas, there may be sclerosis of the margins and cloudlike calcifications. Intracanalicular meningiomas pose a real diagnostic challenge; any "optic neuritis" that is progressive over months or years should suggest intracanalicular meningioma. Optic canal enlargement by gliomas can be the result of intracanalicular spread of an orbital or an intracranial chiasmal tumor; a third possibility is that a benign arachnoidal hyperplasia extending beyond the tumoral glial tissue has enlarged the foramen. Rarely, a fungal infection, such as an aspergilloma, or a bacterial infection, such as a syphilitic gumma or a tuberculoma, can settle in the optic canal and mimic a neoplasm. Enlargement of the canal can also occur in a host of conditions, eg, sarcoidosis, neurofibroma, adhesive arachnoiditis, arachnoidal cysts, and chronic hydrocephalus. Fibrous dysplasia and ossifying fibromas of the

sphenoid bone can involve the canal and compromise its dimensions.

The superior orbital fissure outlines the medial extent of the greater wing of the sphenoid and is separated from the optic canal by a thin strip of the sphenoid bone, the optic strut (Fig 1-2). This fissure transmits the third cranial nerve (oculomotor), fourth nerve (trochlear), first division of the trigeminal nerve (ophthalmic), sixth nerve (abducens), and some sympathetic fibers to their respective orbital destinations. Most of the venous drainage from the orbit and the globe is conducted through the superior orbital fissure to the cavernous sinuses; additionally, an arterial inosculation between the middle meningeal and lacrimal arteries is transmitted through this fissure. When idiopathic inflammation preferentially involves the superior orbital fissure, the Tolosa-Hunt syndrome (painful ophthalmoplegia) results: the nerves to the extraocular muscles are injured by the inflammation as they pass through the fissure. The pain in this syndrome results from the inflammatory involvement of the first division of the trigeminal nerve; the frequently observed suffusion of the lids and orbit can be ascribed to interference with venous drainage through the inflamed fissure. Tuberculous and syphilitic periostitis involving the superior orbital fissure are curiosities today. Neoplasms may produce a superior orbital fissure syndrome by involving one cranial nerve after another, but this neoplastic form of the syndrome lacks the explosive, abrupt onset characteristic of the inflammatory Tolosa-Hunt syndrome. Both types of syndrome, however, may respond to steroids. Since the superior orbital fissure is intimately related to the optic foramen, being separated only by the optic strut, a posteriorly situated apical inflammatory or neoplastic lesion may compromise both the optic nerve and vision as well as the structures passing through the fissure, giving rise to an orbital apex syndrome. Enlargement of the superior orbital fissure can be seen with carotid-cavernous sinus fistula, intracavernous carotid aneurysm (with erosion of the anterior clinoid processes), in A-V malformations, and with orbital varices and hemangiomas.

There are usually two foramens in the medial orbital wall that transmit the anterior and posterior ethmoidal arteries. These openings can allow infections and tumors to spread from the ethmoidal sinus into the orbit. Similar foramens do not exist between the other sinuses and the orbit.

The inferior orbital fissure transmits venous drainage from the inferior orbit to the pterygoid plexus and ascending neural branches from the sphenopalatine ganglion. The infraorbital groove conducts the infraorbital nerve and artery. Traumatic orbital floor fractures commonly injure the infraorbital nerve precipitating hypesthesia of the infraorbital skin.

PERIORBITA

The periorbita is the periosteal membrane covering the bones of the orbital walls (Figs 1-4 and 1-5). Posteriorly it is continuous with the dura of the optic nerve where the dura is fused to the optic canal; at the superior orbital fissure, it also blends with the intracranial dura. Anteriorly it blends into the anterior orbital septum which partitions the lids from the anterior orbital tissues. The periorbita is a tough membrane that restrains periosteal hematomas and can temporarily provide resistance to the spread of infections and tumors from the sinuses and bones (osteomyelitis) into the orbit; however, it is eventually dissolved by these processes. In Caffey's disease (infantile cortical hyperostosis) the periosteum becomes idiopathically inflamed and produces puffy cheeks as well as proptosis and increased intraorbital pressure of such magnitude that a transient glaucoma may develop. The disease is benign and self-limited. Granulocytic sarcoma has a predilection for the periosteum and bones of the orbit. The periorbita may be the only separation between the orbital contents and the mucoceles and dermoids. The potential space between the periorbita and bone provides a convenient plane of dissection around lacrimal gland tumors and for removing the orbital contents during exenteration.

OPTIC NERVE AND MENINGES

The optic nerve (Figs 1-4 through 1-6) is divided into four sections: an intracranial, prechiasmal portion of variable length, averaging approximately 10 mm; an intracanalicular portion approximately 5 to 6 mm in length; an intraorbital stretch approximately 25 mm in length; and an intraocular portion, where it enters the globe through the sclera posteriorly, approximately 1.5 mm in length. Where the optic nerve enters the orbit at the optic foramen, approximately 2 inches behind the supraorbital margin, its dura is fused to the foraminal bone and blends with the periosteum (periorbita). In the orbit the nerve is not taut but has some slackness and describes an S shape which enables it to adjust to the movements of the globe. The fibrous annulus of Zinn, formed by conver-

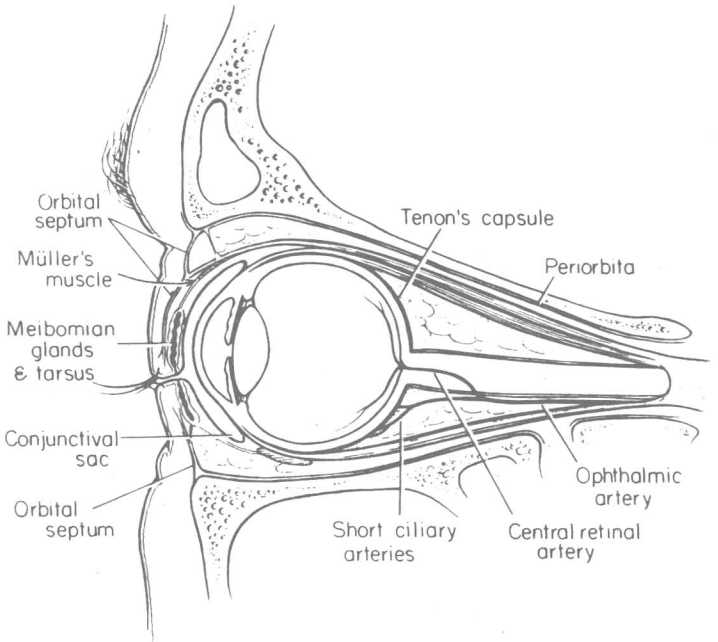


Fig 1-4. Sagittal section through lids, globe, and orbit. Lids are separated from the orbit by the orbital septum, a continuation of the periorbita (orbital periosteum). Müller's muscle is composed of smooth muscle and is interposed between the levator palpebral muscle and the tarsus. Meibomian glands are embedded in dense fibrous plaques (tarsus) in the lids. Tenon's capsule is an outer fibrous tunic that loosely adheres to sclera and blends posteriorly with dura of the optic nerve and anteriorly with muscular fascia. Ophthalmic artery enters the orbit from the optic foramen and divides into the central retinal artery and posterior ciliary arteries among many others.

gence of the origins of the rectus muscles, is also fused to the dura of the optic nerve at the optic canal. Consequently, when the optic nerve is inflamed, as in multiple sclerosis, tugging on optic nerve during movements of the globe causes pain.

Other inflammations, such as tuberculosis and luetic infections of the optic nerve, are rarely encountered today. Mucormycosis and aspergillosis may involve the optic nerve by direct extension. Uncontrolled orbital cellulitis may damage the optic nerve by impeding its blood supply or by direct infection. Inflammatory pseudotumor situated at the orbital apex or perioptically is one of the most common orbital inflammatory causes of visual loss, sharing with Graves' disease similar vasogenic mechanisms of damage to the optic nerve.

The two most common tumors of the optic nerve are optic nerve gliomas in children and meningiomas in adults. Gliomas grow within the dura and expand the nerve in a sausage-shaped fashion, whereas meningiomas break through the dura and grow luxuriantly in the orbital tissues. Both may cause forward proptosis, papilledema, retinal striae, and optic atrophy, depending on their relationship to the globe and to the point of entrance of the central retinal artery and vein. Opticociliary shunt vessels on the optic nerve head are seen in intraorbital meningiomas.

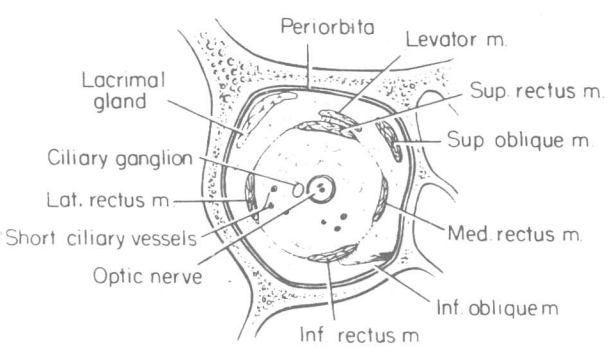


Fig 1-5. Coronal section behind globe. The four rectus muscles are connected by an intermuscular membrane that separates the central (intraconal) space from the middle orbital space. Ciliary ganglion is located lateral to the optic nerve in the posterior orbit.

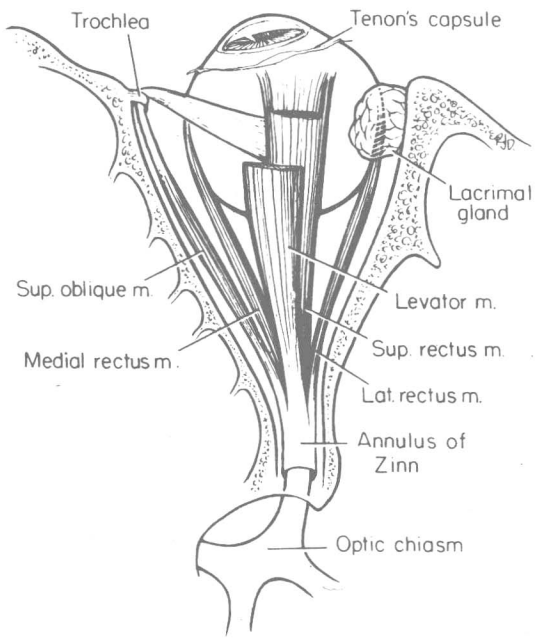


Fig 1-6. View of orbital contents from above. Rectus muscles originate from a fibrous ring at the optic foramen, annulus of Zinn. Superior oblique muscle has a separate origin medially in the posterior orbit, and its anterior tendinous portion passes through a cartilaginous pulley (trochlea) in the superonasal aspect of the anterior orbit. Tenon's capsule fuses with sclera anteriorly near the limbus.

EXTRAOCULAR (STRIATED) MUSCLES

The globe is moved by four rectus muscles and one inferior and one superior oblique muscle (Figs 1-4 through 1-6). The levator of the upper lid is located above the superior rectus muscle. Rare abiotrophies and muscular dystrophies can affect these structures. Myasthenia gravis is an important cause of dysfunction, and ptosis (levator dysfunction) is an early sign. Dermatomyositis may occasionally create clinically discernible extraocular motility difficulties. Trichinosis may settle in the extraocular muscles.

The two most common inflammatory diseases that affect the extraocular muscles are Graves' disease and idiopathic inflammatory pseudotumor. One of the earliest signs of Graves' disease is injection and conjunctival edema over the insertions of the horizontal rectus muscles. Inflammatory pseudotumor is an unpredictable disease that can attack any site in the orbit or any combination of orbital tissues. If the extraocular muscles are predominantly affected, the term "orbital myositis" can be applied to the disease. Exceptionally, the orbital muscles can become inflamed following viral infections, suggesting a pseudotumor. Ultrasonography has been particularly helpful in distinguishing the swollen muscles of Graves' disease, which tends to be bilateral, from those in pseudotumor, which is unilateral. Multifocality of the inflammatory disease, eg, extraocular muscles plus a separate orbital inflammatory mass, suggests pseudotumor.

Embryonal rhabdomyosarcoma is a primitive, malignant tumor of striated muscle that paradoxically does not usually develop in preformed orbital striated muscle but rather in embryonic mesenchymal tissue. Alveolar rhabdomyosarcoma and the rarer, better differentiated pleomorphic rhabdomyosarcoma of older individuals do develop in a muscle. Rhabdomyoma, a benign tumor, has yet to be shown unequivocally to involve the extraocular muscles.

The four rectus muscles are joined together by an intermuscular fascial membrane. The conical central area of the orbit thus created is sometimes referred to as the inner or intraconal space. This method of subdividing the orbital volume is carried further by referring to the area between muscles and the periorbita as the middle space and to the potential area between the periorbita and the bone as the peripheral space. The classification of orbital tumors on the basis of their statistical likelihood of being encountered in one of these spaces does not seem to be very practical to us. The information obtained from this schema is either trivial (eg, optic nerve gliomas must always be located in the inner space) or not very helpful (eg, hemangiomas, pseudotumors or neurofibromas can be found in either the inner or middle space).

ADIPOSE TISSUE

The orbital contents are invested by the ubiquitous presence of adipose tissue (Fig 1-5), which is subdivided by fine fibrous septums into lobules. The retrobulbar fat pad provides a resilient cushion or bed that supports the globe and yields to the actions of the extraocular muscles. As the fascial layers of the anterior orbita and lids slacken with age, the orbital fat sometimes prolapses subconjunctivally or protrudes through the weakened orbital septum (Fig 1-4) into the lids.

Inflammatory pseudotumor invariably involves the orbital fat to some degree. At times, the disease is localized primarily in this tissue. The fat cells

degenerate, releasing their lipid content which further augments the inflammatory process. The inflammation is progressively subdued through fibrosis, and a sclerosing lipogranuloma eventuates. Ruptured dermoids release their irritating contents into the orbital fat and also incite a lipogranulomatous response. Trauma to the orbit can cause fat necrosis and is another cause of orbital lipogranuloma.

In orbital cellulitis, the unresistant orbital fat is quickly overrun by the infection; if the infection is not arrested, the fat liquefies and is the site of an orbital abscess which may require surgical drainage for cure. All kinds of chronic granulomatous disease, either infectious, such as fungal infections, or noninfectious, such as Wegener's granulomatosis, involve the orbital fat. The only systemic disease wherein the orbital fat is selectively injured among the other orbital contents is relapsing febrile, nodular, nonsuppurative panniculitis (Weber-Christian disease, an inflammatory disease of the subcutaneous and abdominal fat); however, this is an extremely infrequent type of orbital disease.

Since the orbital fat is the filler substance of most of the retrobulbar space, metastatic tumors and infections expand at its expense. Thus, metastatic carcinoma, lymphomas, and rare parasitic conditions, such as hydatid cyst (*Echinococcus granulosus*) and the cysts of cysticercosis, are found in the retrobulbar fat.

Primary tumors of the orbital fat are among the rarest of orbital neoplasms. Prolapse of the orbital fat must be distinguished from lipomas. Liposarcoma of the orbit is extremely rare and originates from primitive mesenchymal cells related to the orbital fascia rather than from a lipoma or preexistent adipose tissue.

TENON'S CAPSULE, CONNECTIVE TISSUE, AND FASCIA

Lying upon the scleral tunic of the eye is Tenon's capsule, a fibrous membrane separated from the sclera by a potential space, the episcleral space (Figs 1-4 and 1-6). Inflammatory pseudotumor may inexplicably be confined to Tenon's capsule (tenonitis) and reach such a florid state as to produce proptosis. B-scan ultrasonography can very nicely identify this type of periocular inflammation. Posterior geographic scleritis and intense choroiditis may cause secondary inflammations of Tenon's capsule. The extraocular spread of tumors such as melanoma and retinoblastoma encounters an ineffectual barrier in Tenon's capsule; in melanoma, however, a small episcleral nodule may be rendered sufficiently circumscribed so that the tumor can be completely excised during enucleation, making exenteration unnecessary.

Tenon's capsule is related by various reflections to the aponeuroses and check ligaments of the extraocular muscles and to the anterior conjunctiva and globe, to which it is fused near the limbus. This network of interconnections facilitates the extension of tenonitis to the orbital tissues. It should be remembered that the rectus muscles are connected to one another by an intermuscular fascial membrane. The trabeculae of the orbital fat are also part of this extensive fascial connective tissue system of the globe and orbit. In Graves' disease and early pseudotumor, the trabeculae of the orbital fat thicken, giving the fat a tough, gritty texture.

Nodular fasciitis is a reactive pseudosarcomatous proliferation of the fascial connective tissues of the globe and orbit. It is usually a rapidly developing nodule situated epibulbarly in the vicinity of the anterior aponeuroses of the extraocular muscles; on occasion, it occurs in the retrobulbar fascial planes and in the lids. Although it evinces disturbing histologic features, the condition is benign.

Fibrous histiocytomas (fibrous xanthomas) represent a group of tumors that combine features of the fibroblast and histiocyte. These tumors most likely originate from fibroblast-related orbital cells that can adopt some histocytic traits such as lipidization. They are usually benign but create problems because of their infiltrative growth potential. Some classifications of soft tissue neoplasms place these tumors in a reactive category related to nodular fasciitis. Fibrous histiocytomas have a predilection for the orbit; this fact has been little appreciated in the past because they have often been misdiagnosed as other tumors, frequently as neurogenic tumors. In the orbit they usually behave as true neoplasms and some have behaved malignantly. They are the most common primary fibrous tumors of the orbit; primary fibromas and fibrosarcomas of the orbit are virtually unknown. This latter fact is surprising given the high reactivity and universal distribution of fibroblasts. In the orbit, most fibroblastic neoplasms take the form of fibrous histiocytomas.

NERVES

The sensory innervation of the orbit and globe is supplied by the first division (ophthalmic) of the trigeminal nerve. The sympathetic nervous supply travels with the ophthalmic artery through the optic canal; a lesser contribution reaches the orbit through the superior orbital fissure. The parasympathetic contributions to the ciliary body and sphincter mus-

cle of the iris travel with the third (oculomotor) cranial nerve to synapse in the ciliary ganglion. The nerves to the extraocular muscles all pass through the superior orbital fissure: the third cranial nerve (oculomotor) to the levator, superior, medial, and inferior rectus muscles; the fourth cranial nerve (trochlear) to the superior oblique muscle; and the sixth cranial nerve (abducens) to the lateral rectus. A precise description of the intraorbital course and ramifications of these nerves is discussed elsewhere; it is sufficient for our purposes here to note that the lacrimal and frontalis nerves (branches of V-1), as well as the trochlear nerve, enter the fissure outside of the annulus of Zinn, while the nasociliary (another branch of V-1), the superior and inferior ramuses of the oculomotor and the abducens nerves pass between the two heads of origin of the lateral rectus within the annulus of Zinn.

There is debate as to whether the progressive external ophthalmoplegias represent disease of muscle or of the nerves that supply them or a combination of both. Functional abnormalities of nerves include the various misdirection-regeneration syndromes (eg, following aneurysms) or abnormal neural functions in familial dysautonomia (Riley-Day syndrome) with its attendant poor tearing and hypersensitive pupillary responses to parasympathomimetic drugs.

Tumors of the orbital nerves comprise neurofibromas, which are usually unencapsulated, and schwannomas (neurilemomas), which are encapsulated by the perineurium of the nerve of origin. Granular cell tumors, formerly referred to as "granular cell myoblastomas" but now called "schwannomas," represent an unusual degenerative condition of a small group of benign schwannomas and are rarely seen in the orbit. Plexiform neurofibromas are pathognomonic of neurofibromatosis; involvement of the skin creates a diffuse hypertrophy referred to as "elephantiasis neuromatosa." Traumatic neuromas giving rise to "phantom" sensory illusions are sometimes seen after enucleation; however, it is remarkable how infrequently they occur. In the multiple mucosal neuroma syndrome, spontaneous nerve tumors resembling traumatic neuromas are seen subconjunctivally.

Any branch or radicle of the orbital nerves can randomly give rise to a peripheral nerve tumor. Peripheral nerve tumors may even originate where the ciliary nerves penetrate the sclera. Malignant Schwann cell tumors are exceptionally rare in the orbit. The upper nasal quadrant in the region of the supraorbital or supratrochlear nerves is a site of predilection for malignant Schwann cell tumors; patients with neurofibromatosis are at risk to suffer a malignant degeneration of one of their neurofibromas (schwannomas are relatively immune to this phenomenon), but this most exceptionally supervenes in orbital lesions.

CILIARY GANGLION

This structure (Fig 1-5) is located on the temporal aspect of the optic nerve toward the apex of the orbit and measures 2 mm in length and 1 mm in width. It is the synaptic waystation for parasympathetic motor fibers to the ciliary body and iris sphincter muscles that originate in the Edinger-Westphal nucleus of the oculomotor nerve. Sympathetic and trigeminal sensory fibers also traverse the ganglion but do not synapse there, joining with the parasympathetic axons to form 8 to 20 short posterior ciliary nerves that enter the back of the globe. The sympathetic fibers are destined for the dilator muscle of the iris and also the intraocular vasculature.

The ciliary ganglion may become inflamed (ganglionitis) during herpes zoster infections as well as after a number of childhood and adult viral infections. The resultant depletion of ganglion cells causes a secondary tonic pupil, which is most often idiopathic (Adie's pupil). In Adie's myotonic pupil, there may be an associated postural hypotension and absent tendon reflexes; the pupil displays denervation sensitivity to dilute solutions of Mecholyl. Temporal arteritis may also interrupt the blood supply to the ciliary ganglion and produce a tonic pupil.

Paraganglioma (chemodectoma) is a benign, organoid tumor composed of cells containing bioamine granules; it presumably originates from cells of the ciliary ganglion or in its vicinity. Alveolar soft part sarcoma may well be the malignant counterpart of paraganglioma. It has been described in the orbit on the temporal side and must be distinguished from alveolar rhabdomyosarcoma since they both occur in children; however, the latter has a tendency to localize in the inferior orbit. The development of a primitive neuroblastoma as a primary orbital tumor (if this ever occurs) might be from the ciliary ganglion anlage.

ORBITAL VESSELS

The ophthalmic artery originates intracranially at an acute angle from the carotid artery, once the carotid has left the cavernous sinus and has traversed the dura. Within the optic canal the ophthalmic artery lies inferolateral to the optic nerve. After entering the orbit (Fig 1-4), it continues to

lie inferolateral to the nerve for a short distance but soon crosses over the nerve and runs medially. The first important but very small intraorbital branch is the central retinal artery; a host of other orbital ramifications quickly develop, eg, two important ciliary arteries, six to eight short posterior ciliary arteries, the lacrimal artery, the ethmoidal arteries, numerous muscular arteries, and the supratrochlear artery. An important potential collateral source of orbital blood supply is provided by the anastomosis between the orbital branch of the middle meningeal artery (which passes through the superior orbital fissure) and the lacrimal or other branches of the ophthalmic artery. This anastomosis may be utilized if obstruction to the proximal ophthalmic artery occurs. The central retinal artery enters the optic nerve approximately 12 mm behind the globe.

The ophthalmic artery may undergo medial calcification of the Mönckeberg type in older individuals. Aneurysms can develop at the point of origin of the ophthalmic artery from the carotid, in the optic canal, in the orbit, or along the intraneural portion of the central retinal artery; visual loss or intraorbital hemorrhage can occur, and arteriography is required to make the diagnosis.

Vasculitis of the orbital arteries can be seen in older individuals with temporal (cranial or giant cell) arteritis and in somewhat younger patients with Wegener's granulomatosis or polyarteritis nodosa. Some types of orbital pseudotumor may display a vasculitic component in the absence of a systemic disease, particularly in the young and in those with allergic rhinitis. Rampant orbital cellulitis can lead to destruction of the orbital arteries and necrosis of the globe. Mucormycosis has a predilection for vascular invasion and thereby causes extensive infarction of the orbit and ocular tissues.

Tumors containing arterial elements are usually arteriovenous malformations. The Wyburn-Mason syndrome, in which arteriovenous (racemose) malformations extend from the midbrain, through the optic canal to the orbit, optic nerve, and retina, typifies this type of lesion. Arteriovenous malformations may develop primarily in the orbit and may also extend into the orbit through the superior and inferior orbital fissures.

The orbital venous drainage (Fig 1-3) is accomplished mostly via the superior orbital (ophthalmic) vein which egresses through the superior orbital fissure to the cavernous sinus; a lesser role is played by the inferior orbital (ophthalmic) vein which sends a tributary to the superior ophthalmic vein and which exits through the inferior orbital fissure to the pterygoid plexus. The angular vein, via the nasofrontal vein, and supraorbital vein join at the upper nasal aspect of the orbit to form the superior ophthalmic vein, which travels posterolaterally into the orbit, penetrates the muscle cone in the midorbit, receives the venous drainage from the globe, and leaves the orbit near the annulus of Zinn via the superior orbital fissure. Because of the valve-free communication of the superior ophthalmic vein with veins of the face, nose, and forehead, infections in these sites that go unchecked can spread posteriorly to the cavernous sinus to cause a septic thrombosis. Likewise, the inferior ophthalmic vein, formed by a confluence of veins in the orbital floor and medial wall, may serve as a conduit for infections of the lower lid, sinuses, and lacrimal sac.

There are no valves in the orbital veins. Thus, in a carotid cavernous sinus fistula, whether the result of trauma, atherosclerosis, or a vascular malformation, blood regurgitates untrammeled into the orbit and causes suffusion of the orbital veins and devitalization of the orbital and ocular tissues attendant on blood stasis. Orbital varices may cause intermittent exophthalmos that may often be elicited by postural head changes; these postural changes lead to filling of the enlarged orbital veins. The orbital varices may be either primary or secondary, the latter resulting from the shunting of blood from an intracranial vascular malformation through the valveless veins that bridge the superior orbital fissure. Venous angiomas, which are true neoplasms containing vessels with muscle in their walls, may also cause intermittent exophthalmos. Thrombophlebitis of the orbital veins is a rare idiopathic condition wherein the inflammation attacks the veins rather than the arteries. It causes signs and symptoms of blood stasis. It should be remembered that pseudotumor of the superior orbital fissure (Tolosa-Hunt syndrome) may compromise the veins in this region and create orbital and lid venostasis. In the Klippel-Trenaunay-Weber syndrome, an orbital venous angioma may coexist with similar lesions of the limbs and associated acral hemihypertrophy; vascular and nonvascular visceral tumors may also occur.

Various orbital vascular tumors contain elements of the orbital microvasculature instead of recapitulating the more elaborate structure of arteries and veins. These tumors are referred to as monomorphous hemangiomas because they are composed predominantly of one cell type normally present in blood vessels: endothelial cells, pericytes, or smooth muscle cells. These tumors are to be contrasted with more complex polymorphous tumors, such as arteriovenous malformations, composed of many cell types. In childhood the most common monomorphous tumor is the capillary hemangioma (benign hemangioendothelioma)