Chapter 16: Proptosis

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The term proptosis as generally used is synonymous with exophthalmos. The term signifies that one or both eyes are displaced in a direction anterior to their normal position within the orbit. In addition to being proptotic the eye may be displaced up, down, medially, or laterally. The eye is rarely displaced in one of these directions without proptosis. Whether an eye is proptotic or not can usually be determined by a careful visual inspection of the relative position of the two eyes. An exophthalmometer can be used to obtain an objective measurement of a proptotic eye. An exophthalmometer is also valuable for determining bilateral proptosis and for following the clinical course of proptosis.

Plan of Evaluation

There are several subjective and objective features of proptosis that one needs to consider when examining a given patient.

Subjective Considerations

Age, sex, and race are important. Some tumors are found almost entirely in childhood. Examples would be neuroblastoma, rhabdomyosarcoma, and medulloblastoma. Meningiomas are tumors of middle-aged white women. Nasopharyngeal carcinomas are more common in Chinese males.

One needs to inquire about the past history of a tumor that could have metastasized from a distant site or by direct extension. Skin, lung, breast, and central nervous system tumors are perhaps the most common. A history of hyperthyroidism may be helpful, since a patient may present in a euthyroid state and have thyroid orbitopathy. Trauma produces multiple complications that may lead to proptosis, eg, bleeding, edema, and infection. Pregnancy may be associated with a relatively rapid enlargement of both meningiomas and pituitary adenomas. Intermittent proptosis suggests orbital varices. Bilateral proptosis is usually secondary to hyperthyroidism, orbit pseudotumor, or cavernous sinus disease. The rapid onset of proptosis suggests hematoma, edema, or infection. Occasionally thyroid orbitopathy and orbital pseudotumor present with the rapid progression of proptosis. In children rhabdomyosarcoma is often rapid in course. In adults metastatic tumors may change rapidly. If proptosis responds to steroids one would suspect orbital pseudotumor. Occasionally meningiomas and hemangiomas are somewhat responsive.

A family history of von Recklinghausen's disease or retinoblastoma of the eye may provide a clue to the etiology of the proptosis.

Objective Considerations

A clinical impression of the consistency of the "mass" may be obtained by retropulsing the eye into orbit. If the orbit is very soft it suggests a benign tumor or encephalocele. If the eye is very difficult to retropulse it is suggestive of a malignant lesion. Thyroid orbitopathy falls in between.
Lid signs are important from two aspects. Ecchymosis of the lids suggests dyscrasia or neuroblastoma. A mass involving the lid structures suggests a tumor in the anterior part of the orbit, such as a dermoid cyst, a lacrimal gland tumor, or a hemangioma. Orbital varices may be seen in the lower fornix when the lid is retracted.

Vascular signs are of importance because they suggest either collagen vascular disease, Graves' disease, cavernous sinus disease, or vascular tumors. The signs to look for are edema of the lids, edema of the conjunctiva, and dilated vessels of the conjunctiva. Pulsating proptosis would suggest a carotid cavernous sinus fistula or an encephalocele, primarily. An orbital bruit may be heard in the presence of a vascular tumor or a fistula.

A decrease in vision and the presence of diplopia are indicative of an infiltrating lesion. If a tumor is present it is more likely malignant. In the absence of visual loss and the presence of diplopia one would think of a benign orbital lesion.

Associated neurologic and systemic symptoms and signs need to be considered. Intracranial neurologic signs may suggest a tumor of the intracranial cavity or cavernous sinus disease. Ear, nose, and throat symptoms may suggest the possibility of disease in the sinus such as a mucocele or tumor. Pharyngeal signs may suggest a nasopharyngeal tumor. Systemic signs may help one detect dyscrasia, histiocytosis, sarcoid, neuroblastoma, or tumors of the breast and lung that have involved the orbit.

All of the cases of proptosis in the following discussion are considered as unilateral. Less commonly, bilateral proptosis is seen. It may be congenital as in craniofacial dysostosis. Osteopathies may cause bilateral proptosis. Examples are fibrous dysplasia, osteitis deformans, infantile cortical hyperostosis, leontiasis ossea, osteopetrosis, and acromegaly. The most common cause would be hyperthyroidism. Occasionally nonspecific pseudotumor or specific pseudotumor (eg, Wegener's granulomatosis) may cause bilateral changes as well as cavernous sinus disease. Rarely, neoplasia such as lymphoma or histiocytoma is a cause. Systemic amyloidosis has rarely been associated with changes in both orbits.

**Radiographic Studies**

**Soft Tissue.** Soft tissue studies may be helpful in the differential consideration of proptosis. Abnormal collections of calcium (often seen better with the CT scan than with conventional x-rays) would suggest old infection or trauma, or the possibility of a tumor (dermoid, retinoblastoma, meningioma). An air-fluid in a sinus would indicate trauma or infection in most cases. Occasionally an adjacent orbital tumor will exhibit this finding. Soft tissue studies may indicate a mass in the orbit with distinct edge. This would suggest a tumor rather than pseudotumor. Free gas in the orbit suggests mucocele, wall fracture or, rarely, infection by gas-producing organisms.

**Bone Studies.** Dense, thick bone may be diagnostic of a dysplasia of bone causing the proptosis (fibrous dysplasia, Paget's disease, osteopetrosis, or Hurler's disease) or a tumor adjacent to bone (meningioma or hemangioma). Bone destruction almost always indicates a tumor (probably malignant) or infection of bone. Rarely, pseudotumor may do this. A mucocele may displace bone. Enlargement of normal orbital dimensions of foramina may indicate specific diagnoses: enlarged optic canal (optic glioma, craniopharyngioma,
nasopharyngeal tumor, pituitary adenoma), enlarged superior orbital fissure (neurofibromatosis, meningioma), or enlarged lacrimal fossa (lacrimal gland tumor).

**Vessel Studies.** Arteriovenous malformations, aneurysms, carotid cavernous fistulas, and vascular tumors (meningioma, angiofibroma, or hemangioma) can be detected by carotid angiography. Orbital venography is essential to establish the diagnosis of orbital varices or cavernous sinus thrombosis.

**Specific Examples of Proptosis**

**Sudden Onset of Proptosis within a Few Hours of Trauma**

**Subjective Complaints**

The proptosis comes on a few hours after relatively severe trauma, caused by a blunt object, and remains stable. Blurred vision and diplopia occur in association with the proptosis. The orbit feels very uncomfortable as if it were going to explode. The eyelids are quite black and blue.

**Objective Findings**

Examination indicates vision is moderately reduced but it is difficult to determine because of the swollen lids. The pupil on the side of the injury is mildly dilated and does not respond as well to light as the pupil on the other side. The eye moves but is somewhat limited in all directions. There is no specific restriction of one direction, which would suggest a cranial nerve palsy. Visual fields are full to a confrontation finger-counting method. The ophthalmoscopic examination of the eye does not reveal any obvious damage. Examination of the orbit and lids reveals the lids to be markedly swollen and ecchymotic. Retropulsion of the eye is very difficult to accomplish. The orbit seems very tense and it is painful to push on the eye.

**Assessment**

The history of sudden onset of proptosis following blunt trauma is suggestive of hemorrhage or edema in the orbit. The markedly swollen ecchymotic lids would be further evidence for this. The fact that the orbit is very tense and painful when one attempts to push the eye back in the orbit also would further suggest this possibility. The fact that the pupil is slightly dilated and does not respond as well as the other pupil to light indicates a concussion effect on the iris. The fact that the eye moves poorly in all directions and does not have loss of movement in one plane favors a mechanical limitation of movement within the orbit, eg, edema or blood rather than a cranial nerve palsy. The differential between hemorrhage into the orbit and edema of the orbit would be primarily based on the fact that edema develops over a number of hours and hemorrhage is essentially immediate. Edema does not produce as tense an orbit as hemorrhage does. Additionally, pain is more severe with an orbital hemorrhage.
Plan

The workup would include skull and orbit x-rays and a blood count. The presence of fractures of the orbital walls would further complicate the problem of management. For example, a blowout fracture of the floor or medial wall of the orbit might entail surgery to correct that problem. However, the absence of fracture would allow the physician to concentrate on the question of whether to decompress the orbit and remove the hematoma, or alternatively, to observe the patient. Because most patients are very uncomfortable it is difficult to get the patient to cooperate well in order to obtain the best possible vision. The concern about severe ocular damage is largely alleviated if the eye examination is normal. If there is further question, tomograms of the optic nerve canal may be done to exclude the possibility of fracture in that area compromising the optic nerve. In general both hemorrhage and edema into the orbit may be managed by observation. In the event that optic nerve function is gradually decreasing in the face of edema and dilated veins of the optic nerve head, one may elect to decompress the orbit. There are four accepted approaches to decompression of the orbit. The more benign procedures would involve removal of the lateral wall, orbital floor, or ethmoid air cells. The transfrontal removal of the orbital roof is done by some but involves the possible complications of the neurosurgical procedure. It would be rare that adequate decompression could not be obtained by a combination of the benign approaches.

Sudden Onset of Proptosis 3 Days after Trauma

Subjective Complaints

Injury occurs 3 day before the sudden onset of proptosis. The proptosis develops over a period of 1 day. Part of the trauma may have been caused by a sharp instrument that perforated the lids. Intense pain is associated with the onset of proptosis. Pain had not been present in the first 2 days post-trauma. Vision was fairly suddenly reduced. The eye is painful to move or even to light touch. The patient feels generally ill as if he might have the flu.

Objective Findings

Vision is significantly reduced. The pupil is the same size as the other eye but reacts poorly to direct light. The other pupil, of the normal eye, reacts very briskly to direct light. This defines a defect in afferent transmission of the optic nerve on the proptotic side. Ocular mobility is almost zero. The patient will not attempt to move the eye in one direction or another. He complains of severe pain when asked to do this. It is difficult for the patient to count fingers of the visual field. The optic nerve is swollen with dilated veins. This indicates external pressure on the optic nerve. Examination of the orbit reveals the lids are very swollen and tender. They are also red. The conjunctiva is swollen. Even the slightest attempt to push the eye back into the orbit is painful and the patient does everything possible to resist this maneuver. The patient is not febrile in spite of his complaints of a flu-like feeling. Careful examination of the swollen lids reveals what appears to be a small puncture wound in the upper lid in the central area.
Assessment

The delay in development of sudden proptosis associated with severe pain, loss of vision, and other signs suggestive of infection points toward an orbital cellulitis. The swelling of the conjunctiva, while it may be found in hemorrhage or edema of the orbit, makes one particularly concerned about the possibility of infection. The marked swelling of the lid with redness in the absence of ecchymosis also would support this suggestion. The extreme tenderness of the eye and orbit are further indications of infection.

Plan

Orbit and optic canal x-ray views are obtained to be sure there is no fracture complicating the problem. In children fractures of the ethmoid sinus may be associated with orbital cellulitis. This is particularly true if this is associated with chronic infection in the sinus. The x-rays help exclude the possibility of a foreign body within the orbit. Most metallic foreign bodies are tolerated well within the orbit but organic material almost routinely causes infection. A CAT scan may help define a nonmetallic foreign body. A blood count is done and reveals an elevated white count of 10,000-15,000 WBC with a shift to the left. This further confirms the suspected cellulitis. The area of perforation may be probed to obtain purulent material. The abscess may be drained through the primary site of the penetrating injury. Further orbital surgery may not be necessary. Occasionally cellulitis or abscess of both lids will appear to make the eye proptotic. If the lids are swollen but the conjunctiva is not and the eye is not displaced relative to the other eye, the diagnosis would be periorbital cellulitis. This has a much better prognosis in regard to vision and eye movement than does an orbital cellulitis. This connotes that the orbital septum, which separates the lid structures from the orbit, has not been perforated and so the orbital space is not infected.

"Sudden" Development of Proptosis over 3 Days in a Child, Unassociated with Trauma

Subjective Complaints

Usually it is a child under 5 years of age who presents with the sudden development of proptosis. The proptosis will have developed over a period of 3 days and the maximum amount of development is usually over a 1-day period. The child will have been perfectly well until 3 weeks before the proptosis. At that point the relatively sudden onset of pain develops about the left side of the face with fever after a mild cold. These symptoms may have resolved when the proptosis begins. The lids will be quite swollen and red, and the eye is very tender to touch.

Objective Findings

Examination confirms that the eyelids are swollen and red. The conjunctiva is swollen. The patient resists all efforts to touch or manipulate the eye and will not move the eye voluntarily. The patient seems to be in a considerable amount of pain and is found to be running an elevated temperature. Vision is blurred but cannot be tested well. The field of vision is poor. The pupil reacts poorly to direct light and the optic disc is swollen.
Assessment

The history suggests ethmoid sinusitis followed within a short time by proptosis. Pain and the associated fever suggest orbititis. The history is quite sudden for a tumor of the orbit.

Plan

Sinus x-rays show the ethmoid sinuses are opaque. The white count is elevated with a left shift. The plan at this point would be to decompress the ethmoid sinus directly and drain the sinus into the nose. Systemic antibiotics are begun, using ampicillin or other agents effective against *Haemophilus influenzae* plus penicillin until sensitivities are obtained. If the orbit seems quite soft at the end of the procedure the periorbita needs not be opened. If the space inside the orbital periosteum seems quite tight, the periosteum (periorbita) over the ethmoid plate can be opened in an "H"-type incision to allow decompression of the orbital contents.

"Sudden" Development of Proptosis over 2 Weeks in a Child, Unassociated with Trauma

Subjective Complains

The history is that the progression of proptosis has been relatively rapid but not of the 1- to 2-day variety that was seen with the orbital cellulitis. This proptosis will have developed over a period of 2 weeks. The child is usually about 4 years old and has been in perfect health until the onset of proptosis. There has been no fever or any other constitutional symptoms until the proptosis started. Since that time the child has been fussy and has not eaten well. The lids have not been particularly red or edematous but there has been ecchymosis of one upper lid starting about the time the proptosis was first noticed.

Objective Findings

Examination indicates that vision is quite good, the pupil reacts normally to light, but ocular motility is markedly restricted. It does not suggest a cranial nerve palsy but suggests something in and about the muscles that is restricting the movement of the eye. Visual field examination is grossly normal. Examination of the fundus reveals only mild edema of the optic disc. The eye is not particularly tender although the child does not want the eye touched. The attempts to push the eye back into the orbit reveal that the orbit is essentially filled with a solid lesion. The globe is proptotic but the lids are only mildly swollen and not particularly red. The conjunctiva is only minimally swollen. Ecchymosis is noted in the upper lid. The presence of ecchymosis suggests the possibility of a tumor such as a neuroblastoma. Rhabdomyosarcoma would also be a possibility. The child is afebrile and the white count is normal. No other signs of disease are found but the child seems fussy.

Assessment

The development of proptosis over a 2-week period in a child, 4 years of age, without trauma, suggests the possibility of a rapidly growing mass in the orbit. The two significant diagnoses would include rhabdomyosarcoma and neuroblastoma.
Plan

Workup includes x-rays of the orbit, adjacent sinuses, and the optic canal. These examinations are normal. Further examination indicates that the blood count and physical examination are within normal limits. Computerized tomography reveals a large fairly localized mass in the posterior medial orbit without any intracranial component. At this point a careful search is made of the abdomen and chest by radiology to determine if there are masses along the anterior paraspinal area suggesting a neuroblastoma. A suspicious posterior mediastinal mass may be noted. An intravenous pyelogram (IVP) would be done to evaluate the condition of the perirenal tissues. If this is normal, a neuroendocrine workup is done to determine if there is an increase in adrenaline and non-adrenaline substances being excreted in the urine. The findings of elevated values would suggest a neuroblastoma. A bone marrow should be done, to look for abnormal cells suggesting neuroblastoma. Medical and surgical oncology should then be consulted in regard to chemotherapy and radiation therapy of the marrow and the mediastinal tumor. The orbit probably will not require surgery but would be treated by chemotherapy and radiation. If the other studies are normal, an orbital exploration would be carried out to determine whether a rhabdomyosarcoma was present. If that were present, orbital exenteration would be followed by chemotherapy and radiation therapy.

"Sudden" Development of Proptosis in an Adult, Unassociated with Trauma

Subjective Complaints

In this case there is a sudden development of proptosis without any history of previous malignant disease. The signs and symptoms are very similar to those noted for the metastatic breast tumor (see preceding section).

Objective Findings

Visual acuity is reduced slightly. The pupil reacts in a relatively normal fashion. Ocular motility is restricted in a similar fashion to that seen in orbital metastasis. Visual fields are relatively full. Examination of the eye does not reveal any abnormalities. Cranial nerves do not reveal dysfunction. The orbit is quite firm to retropulsion but is not particularly painful and there is only mild edema of the lids and conjunctiva with some dilated vessels of the conjunctiva. The general physical examination is normal.

Assessment

This symptom complex suggests the possibility of a pseudotumor of the orbit. As can be seen it resembles quite closely the metastatic tumor problem described above. On occasion these problems are also associated with other granulomatous disease such as Wegener's midline granuloma. The chance of a primary malignancy of the orbit is possible but somewhat unlikely without a clear history for a primary.
Plan

X-rays of the orbit, paranasal sinuses, and optic canal do not reveal any abnormality with the exception that there is some erosion of bone along the ethmoid plate. Computerized tomography will reveal a very ill-defined mass filling most of the posterior part of the orbit. There is no intracranial component. At this point the decision is made to explore the orbit. Fibrous "tumor" is found that fills most of the orbital cavity. It surrounds muscles and nerves and could not be removed in its entirety without a risk of destroying visual function. Therefore, it is biopsied. It is found to be made up of fibrous tissue with chronic inflammatory cells, lymphocytes, and plasmacytes. Surgery is then terminated. A permanent section confirmed probable pseudotumor of the orbit. The appropriate treatment is with steroids.

Recurrent Sudden Proptosis in a Young Adult Male

Subjective Complaints

The patient will have had multiple recurrences since childhood of sudden proptosis in which there has been pain and some visual difficulty. Each episode lasts for several days to a couple of weeks. Usually the eye is quite red and lids and conjunctiva are swollen. Usually the problem has resolved without treatment. In between the episodes the orbit and eye are perfectly normal.

Objective Findings

The orbit is moderately stiff to retropulsion but the conjunctiva and lids are swollen. The vessels over the conjunctiva are quite dilated. The eye does not move well but vision, pupils, and fields are normal. There are no cranial nerve findings. Two or three dilated vessels are noted in the inferior fornix.

Assessment

The fact that the patient has had multiple episodes of sudden proptosis lasting for 2 days to 2 weeks and that resolve, leaving the eye and orbit normal, is almost pathognomonic for orbital varices.

Plan

The plan in addition to the routine x-rays of the orbit, paranasal sinuses, and optic canal is to do orbital venography. It may be difficult to fill the varices during the time there is proptosis. Between episodes orbital venography and angiography may reveal large varices in the orbit. The usual treatment is observation with the possibility of added steroids. The varices can be removed if they are anterior to the orbital septum or if they are in the anterior part of the orbit and compromise only one or two large channels.
Gradual Development of Proptosis in an Adult, Associated with Prominent Diffuse Vascular Signs

Subjective Complaints

There is gradual development of proptosis associated with very little pain or disturbance in vision. The prominent feature aside from the proptosis has been swelling of the lids, conjunctiva, and dilated vessels over the conjunctiva. Otherwise there is no complaint.

Objective Findings

Findings indicate exactly what the patient says. Vision is essentially normal with normal pupil reaction. Visual fields are full. Ocular motility is only minimally limited, if at all. The fundus of the eye and the rest of the ocular structures are normal. Cranial nerves are normal. Examination of the orbit reveals mild proptosis with edema of the lids and conjunctiva and dilated vessels of the conjunctiva. The eye can be retropulsed into the orbit quite easily and painlessly.

Assessment

This chain of events suggests the possibility of an inflammatory or vascular lesion of the orbit. The primary concern would be one of an orbital myopathy related to hyperthyroidism or collagen vascular disease.

Plan

Workup includes the usual x-rays of the orbit, sinuses, and optic canal, which are normal. Blood workup further is done to evaluate the possibility of vasculitis and thyroid disease. It is found that the vasculitis workup, including RA, ANA, CBC, sedimentation rate, blood sugar, VDRL, and sickle cell disease are all normal. In addition the thyroid function studies, routine T3 and T4, are normal. At this point the patient is re-questioned and it is found that 2 years before his illness the patient had an episode of nervousness, increased sweating, increased appetite, and weight loss of 20 pounds. This was self-limited over 6 months and the patient then felt well again. The patient didn't think this important to mention in the original history. A T3 or T4 suppression test can be done to determine whether the thyroid is responding normally to its usual control. It is found that the radioactive iodine uptake of the thyroid does not suppress appropriately when T3 is administered. It is therefore felt that this is an orbital myopathy related to a previous episode of hyperthyroidism. To be certain that there is no other vascular disease being overlooked, angiography and computerized tomography may be considered. The CT scan often reveals thickened extraocular muscles. Orbital ultrasound may also be considered but usually would be normal in the diffuse infiltration that is associated with the orbital complications of hyperthyroidism.

There are three avenues of treatment that are available. One would be to try systemic steroids or steroids injected into the periocular tissues. These may be helpful in reducing the inflammation of the orbit in about 50% of cases. In addition to this, selective radiation to the posterior orbit may be tried; again this is helpful in a number of cases. In the event that the proptosis is massive or increasing and the other methods of treatment have not been helpful,
orbital decompression may be considered. If thyroid-related orbitopathy was not found but instead collagen vascular disease was found the orbit would be treated along with the general disease.

**Gradual Development of Proptosis in a Middle-aged Patient, Associated with Tortuous Conjunctival Vessels**

*Subjective Complaints*

Onset of symptoms is very similar to that just described for Graves' disease. On careful questioning, however, no past history suggestive of Graves' disease can be determined. In addition there has been no trauma. The patient does mention that there are some very "odd" dilated vessels over the conjunctiva.

*Objective Findings*

It is found that the examination is very similar to that seen with the Graves' disease patient. The lids are edematous. The conjunctiva is filled with edema and it is slightly reddened. The different feature is that in Graves' disease the conjunctiva is diffusely reddened, with perhaps a bit more redness over the medial and lateral rectus muscles. In this case there may be six or eight very dark tortuous vessels that radiate directly away from the cornea. These are arteriolized veins.

*Assessment*

This characteristic appearance of the dilated tortuous veins of the conjunctiva suggests the possibility of a fistula, with arterial blood being shunted into the venous channels, distending them. The most common site for the fistula would be in the cavernous sinus.

*Plan*

The patient has the usual orbital x-rays which are normal. Ultrasound of the orbit is normal. Computerized tomography does not reveal any abnormalities of the orbit. Selective carotid angiography is done which reveals a carotid cavernous fistula with a huge draining vein, the superior ophthalmic vein. In a fairly young patient in whom there is relatively high flow the possibility of vascular surgery may be considered. If the patient is old or if the flow is quite slow the situation probably would be kept under observation, since a number of these will resolve spontaneously.

**Gradual Development of Proptosis over Several Weeks in a Young Patient, Associated with Minimal Conjunctival Redness**

*Subjective Complaints*

The patient, in addition to the above symptoms, described the fact that he has had some edema of the lids and conjunctiva. There has been no pain or disturbance in vision. There's been no double vision.
**Objective Findings**

Vision, pupils, and fields are normal. The eye moves quite fully. Examination of the eye does not reveal any abnormalities. Cranial nerves are intact but examination of the orbit reveals that there is moderate stiffness to the orbit. The lids are swollen and the conjunctiva is swollen and red. The eye can be retropulsed, but with some difficulty. Careful digital examination of the tissues between the eye and the orbital rim reveals that there is fullness over the upper lid. Further examination then reveals a slight fullness to inspection with perhaps minimal ptosis of that lid.

**Assessment**

The symptoms and signs suggest that there may be a benign vascular tumor in the orbit.

**Plan**

Includes orbit, sinus, and optic canal x-rays. These are normal. Computerized tomography and orbital ultrasound reveal a mass localized in the superior orbit. It has only moderate tissue density and A-ultrasound. The CT does not reveal any cranial component and sinus x-rays do not reveal any opacity. Careful examination of the nasopharynx is carried out and determines there is no tumor involving that area. A vascular orbital tumor is seen on carotid angiography, but it does not reveal any tumor of the sinuses, nose, nasopharynx, or the intracranial compartment. Surgical exploration determines that the tumor is a hemangioma of the orbit. It is excised. Differential diagnosis would include angiofibroma and meningioma. Both of these would be expected to produce bony changes.

**Gradual Development of Painless Proptosis in a Child with a White Eye**

**Subjective Complaints**

The patient will have been asymptomatic until a few months before. Then proptosis will have gradually begun to develop without redness or swelling of the lids. The proptosis will be mild in extent. There is no pain or visual complaints. The family history reveals neurofibromatosis to be present and the child may have several brown birth marks.

**Objective Findings**

Mild proptosis will be present. The eye is white and there is no edema of the conjunctiva. The lids are not swollen. The eye retropulses as easily as the other eye. There is no pain. Vision, pupils, and visual fields are normal. Ocular structures are normal. Cranial nerves are likewise normal.

**Assessment**

The constellation of normal symptoms and signs suggests that this is a bony abnormality of the wall of the orbit, an encephalocele. The key objective finding here in
addition to the normal neurologic function and absence of vascular and mass signs would be the fact that the eye pulsates one to two times a second.

**Plan**

Start out with the routine orbit, sinus, and optic canal views. These indicate that the greater wing of sphenoid is largely missing. Computerized tomography is done to determine if there is a mass in the orbit and this is normal or may reveal brain tissue in the orbit. This problem will be observed.

An optic glioma of the orbit and/or chiasm may present in a very similar fashion. The differences would be the following: The proptosis is usually moderate to marked; there is a slow, progressive loss of visual acuity and field of vision; ocular mobility may become quite restricted; the eye is difficult to retropulse; the eye does not pulsate; and x-ray studies show a large optic canal usually, possibly a J-shaped sella and a fusiform mass of the optic nerve and/or chiasm (CT or pneumoencephalogram).

**Gradual Development of Proptosis in a Young Adult with a White Eye**

**Subjective Complaints**

There is slow development of painless proptosis over several months. The eye is white. There is no double vision or visual loss. The patient is perfectly well.

**Objective Findings**

The proptosis may be moderate but it is difficult to retropulse the eye into the orbit. There are no vascular signs such as lid edema or dilated vessels over the conjunctiva. No mass is palpated in the anterior part of the orbit. Vision, pupils and fields are normal. Ocular structures are normal. Ocular motility is normal and there are no cranial nerve signs.

**Assessment**

This combination of symptoms in which there are no associated disease, no neurologic signs, no vascular signs, and no palpable mass suggests the possibility of a benign nonvascular tumor of the orbit. It is different from the vascular tumors of the orbit in which there are vascular signs of the lids and conjunctiva. This eye is white without lid edema. This suggests a benign tumor involving nerve or fibrous tissue.

**Plan**

The x-rays of the orbit, canal, and sinuses are normal. Orbital ultrasound and computerized tomography reveal a mass isolated to the medial part of the orbit. It is quite circumscribed and round. The rest of the examination is normal. Surgical exploration is carried out and a neurinoma of the medial orbit is present. It is excised in its entirety. The other possibilities are fibroma or dermoid or epidermoid tumor. Dermoid tumors are more likely to occur in children and usually in the superior lateral orbit. Presentation of all of these
would be very similar. Simple excision is usually sufficient to take care of the problem. Another nonvascular tumor would be a lacrimal gland tumor. This would present in a similar fashion except for the following differences: There may be double vision with the eye displaced downward and medially; a mass is almost always palpable in the area of the lacrimal gland; the globe will commonly have striae over the superior-medial aspect; orbital x-rays will usually show an enlarged lacrimal fossa or bony erosion, and the tumor may extend from the lacrimal fossa to fill part or all of the orbit.

Gradual Development of Proptosis in a Middle-aged Adult, Associated with Facial Asymmetry

Subjective Complaints

The patient is perfectly well but has noticed a slight, slowly developing proptosis over about a year. This has been painless. Vision has been normal and there has been no diplopia. The eye has been white. She has otherwise been perfectly well.

Objective Findings

The examination is totally normal with the exception of mild proptosis. The orbit is quite firm to retropulsion and there is no tenderness or pain. Vision, pupils, motility, and visual fields are normal. The ocular fundus is normal.

Assessment

This is very similar to the benign tumor of the orbit discussed in the preceding section in relation to the young adult, with the exception that the development of the proptosis has been somewhat slower. This suggests the possibility of a problem involving the paranasal sinus or orbital bone. Examples would be fibrous dysplasia and mucocele.

Plan

Orbital, sinus, and optic canal x-rays reveal very thickened bone over the sphenoid wing and roof of the orbit. This is thought to be fibrous dysplasia. If other areas of involvement are not seen, it may be decided to follow this problem. Complications could involve narrowing of foramina. The optic nerve canal could be compromised. The other problem is the cosmetic aspect. If the bone were normal but the sinuses were cloudy and there were some suggestion of bony displacement of sinus walls into the orbit, a mucocele would have been considered. Some of these have essentially no history of prior infection. In this case, surgical exploration of the sinuses and orbital wall adjacent to the sinus would reveal the diagnosis, and correct this problem.
Gradual Development of Proptosis in a Middle-aged Patient, Associated with Slow Progressive Optic Atrophy

Subjective Complaints

The patient will have been perfectly well except for the slow development of painless proptosis, the loss of vision, and the occurrence of double vision. There is no other history that is contributory.

Objective Findings

Vision is moderately reduced and there is an afferent defect of the pupil. Visual fields show a rather dense central scotoma with some contraction of the peripheral field. Ocular motility indicates that there is mild ptosis on that side and a moderate degree of paresis of cranial nerve III. The eye does not move up, in or down well. Ocular structures are normal with the exception of mild atrophy of the optic nerve head. Orbital examination reveals that the orbit is relatively firm to retropulsion. The proptosis is mild, 2 mm. There are no vascular signs. Further examination may reveal that the patient is anosmic on the side of the proptosis. There also may be mild hypesthesia of the forehead and cornea.

Assessment

The combination of neurologic signs in the absence of vascular signs suggest the possibility of a slowly growing tumor that is invading the apex of the orbit.

Plan

The x-rays of the orbit, sinuses, and optic canal reveal that there is erosion around the superior orbital fissure and the roof of the optic canal on the side of the proptosis. Intracranial x-rays show hyperostosis. The latter finding might only be confirmed on tomography of the optic canal area. Computerized tomography reveals a mass in the orbital apex but also involving the tuberculum sellae area intracranially. Cerebral angiography demonstrates further that there is a vascular mass in the area of the tuberculum sellae intracranially. The suggestion is now made that this is probably a meningioma, and surgical exploration is in order. Less frequently craniopharyngiomas or pituitary adenomas may present with extension from the intracranial cavity into the apex orbit. If the patient is a middle-aged Oriental male, a nasopharyngeal carcinoma would be a significant consideration. In a child medulloblastoma would be one of the significant considerations. Occasionally an aneurysm of the ophthalmic-carotid junction will present in this way.