CRANIO-ORBITAL LESIONS
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There is a wide variety of intracranial lesions that involve the orbit. Survey of these lesions as a group emphasizes the anatomical complexity of the retro-orbital region, to which the author would like to refer as the sphenosellar recess, illustrated in Fig. 1. This concave area houses a number of important structures, namely portions of six cranial nerves, the cavernous sinus, the internal carotid artery and branches, hypothalamic pathways and frontotemporal cerebrum. In some respects, one may compare this neurovascular crossroad with the cerebellopontine angle. The considerations for surgical approach to the sphenosellar recess are certainly no less weighty than those in regard to its subtentorial counterpart.

Fig. 1. The term “sphenosellar recess” refers to the anteromedial aspect of the middle cranial fossa. A section of the internal carotid artery is shown lateral to the optic nerve. The extra-ocular nerves are embraced by the cavernous sinus.
It is the purpose of this presentation to review indications for surgical intervention in cases of cranio-orbital lesions and to exchange information on surgical management in such cases. Details of a series of type cases will be shown rather than a statistical survey of all categories. Excluded from the subject matter will be aberrant pituitary tumors, vascular lesions and infectious disorders. Apology for delineation may be offered by a quotation from the writings of Harvey Cushing: "the time is ripe for special studies of special tumors in special localities, particularly if the surgical treatment of these difficult lesions is to be perfected."

**CLINICAL FEATURES**

The patient with a cranio-orbital lesion usually presents one or more of the following signs: (1) exophthalmos, (2) diplopia, (3) ambylopia, and (4) orbital pain. The presence of a lesion can be established in most cases by clinical observation and roentgenograms of the skull. Special contrast studies, such as arteriogram or air injection, may be necessary for diagnosis. Arteriography is not necessarily a commitment to surgery. However, as will be stressed later, if surgery is planned, carotid arteriography should be performed preoperatively even though it may not be necessary for localization of the lesion itself.

Clinical onset of these lesions may be insidious in patients showing decrease of visual acuity and defect in visual fields. The main pitfall to be avoided is assumption of "multiple sclerosis" as the diagnosis, based on absence of pain, normal roentgenograms of the skull and normal spinal fluid findings. A contrast study may be extremely revealing. Mention of this problem of differential diagnosis perhaps seems superfluous but catastrophic errors of omission are being made currently even in this day of refined neuro-ophthalmological considerations.

**SELECTION OF CASES**

Once the presence of a sphenosellar lesion is established, the indications for surgery deserve careful consideration. Factors of cosmetic state, unilateral visual defects, or radiological abnormalities should not convince the surgeon that something has to be done at once. The primary factors that provide indication for surgical intervention are: (1) uncontrollable pain, (2) progressive threat to vision in both eyes, and (3) increased intracranial pressure.

Elsberg et al.² pointed out that the radiological changes seen in cases of metastatic lesions may resemble the findings often associated with meningioma. However, if the lesion can be identified as metastatic malignancy, the course of action is considerably different from that with benign or relatively benign lesions. With metastatic tumors, the exophthalmos and pain often are not related to the size of the tumor mass but are the result of infiltration in the cavernous sinus (Fig. 2).

Surgical decompression of the orbit in cases of malignant invasion offers
very little in relief of either exophthalmos or pain. The operative choice is control of exophthalmos by tarsorrhaphy and a procedure for relief of pain, such as trigeminal neurectomy or prefrontal lobotomy. Fig. 3 illustrates a case of long-term nasal malignancy producing left-sided exophthalmos and trigeminal pain. This patient was relieved satisfactorily by tarsorrhaphy and trigeminal neurectomy during the 2-year period he survived. The exception to this form of management occurs in cases of radiosensitive malignancies, such as metastatic lymphoma. In some of these cases, surgical decompression of the orbit may be deemed indicated prior to roentgen-ray therapy.
SURGICAL MANAGEMENT

The pre-operative work-up of patients with cranio-orbital lesions selected for direct surgical approach should include the following:

1. **Endocrine Evaluation.** The proximity of these lesions to hypophysial-hypothalamic structures produces a slowly progressive defect in hormonal and electrolytic relationships. Operative interference in the same locality may plunge the patient from borderline maintenance to profound deficiencies with fatal terminus. Pre-operative studies of levels of ketosteroids and electrolytes are essential, and preparation with ACTH followed by electrolyte supportive therapy, may make the difference in survival of the patient.

2. **Carotid Arteriography.** As mentioned above, pre-operative carotid arteriography is highly important and serves as a guide to the neurosurgeon as a map does to the navigator. It is possible to enter the calvarium and remove the lesion without an arterial guide, just as it is possible to navigate by means of dead reckoning. But knowledge of the vascular pattern can influence the operative result significantly. Regional cranial nerves may already have been impaired, so that the technical goal of the neurosurgeon is removal of the lesion with preservation of vascular pathways. It is interference with the carotid artery or its branches that produces the major neurological defects secondary to surgery in this area. The direction of displacement of the internal carotid artery on the arteriogram indicates whether the surgeon’s approach is better in front of or below the carotid bifurcation, namely frontal or temporal—or possibly combined. Radiographic demonstration of a displaced anterior cerebral artery offers valuable information relative to the need for bifrontal approach, with section of the longitudinal sinus and falk. Pre-operative study of the angiograms charts the course of entry and once the lesion is encountered, the surgeon benefits from knowledge of arterial positions in his differential dissection of the troublesome intrinsic vessels of the lesion from the major blood supply of the cerebrum.

CASE PRESENTATIONS

Details of a series of cases are presented as follows:

**Meningiomas.** Statistics relative to incidence and location of meningiomas have been well reviewed by Cushing and Eisenhardt. Seriousness of surgical attack on meningiomas in the sphenosellar region of the skull has been emphasized by Olivecrona et al. and by Grant. These lesions are slow-growing and best handled conservatively unless they present one or more of the operative indications listed above.

**Case 3 (E.D.).** Fig. 4A illustrates a slowly progressive right-sided ophthalmoplegia in a 54-year-old woman with no pain and fortunately enough ptosis to avoid double vision. Roentgenograms of the skull showed osseous reaction about the sphenoid ridge and clinoid process.

Carotid arteriography (Fig. 4B) gave evidence of a relatively small lesion, presumably meningioma, elevating the internal carotid bifurcation and first part of the middle cerebral artery.
The patient has been followed over a 3-year period without change in her status, and surgery is not considered indicated under these circumstances.

Case 4 (G.G.). A 37-year-old woman with progressive defects in visual acuity and fields had been treated elsewhere for "multiple sclerosis." She had demonstrated the misleading triad: absence of pain, negative roentgenograms of the skull, and normal findings on lumbar puncture.

A carotid arteriogram showed a large lesion of the right sphenosellar region extending to the midline. Extirpation of the lesion was performed. The chief problem was dissection of the lesion from the carotid artery and its bifurcation. A small edge of capsule embracing the carotid artery was left in place.

Her immediate course was satisfactory. However, on the 3rd postoperative day, the patient had sudden onset of hemiplegia and coma and then died, presumably from thrombosis of the carotid artery. Postmortem examination was refused by the patient's family.

Case 5 (E.S.). This illustrates a more happy outcome. A 36-year-old woman presented a major visual defect in the right eye and threat of loss of vision in the other. This type of bilateral visual problem provides indication for surgery. Roentgenograms of the skull showed bony reaction about the right anterior clinoid process. Carotid arteriogram indicated lateral displacement of the anterior cerebral artery.

Craniotomy was performed and a meningioma was removed from the region of the right anterior clinoid process. She has residual useful vision in the left eye, 4 years after surgery.

Case 6 (B.S.). This further outlines indications for surgical intervention. A 60-year-old man was admitted in 1954 with exophthalmos and ophthalmoplegia on the
left side. Roentgenograms of the skull were not diagnostic. An arteriogram showed displacement of vessels, compatible with meningioma.

He was followed conservatively because the moderate exophthalmos and unilateral visual problem were not considered indications for surgery. However, in 1956, pain developed in the left periorbital region, not controlled with medication. He was unable to work.

Left transfrontal craniotomy and orbital decompression were performed with resection of a meningioma along the sphenoid ridge. Postoperatively the patient has been relieved of his pain and has returned to full-time work—an interval of 1 year to date. Fig. 5A shows the pre-operative state, and Fig. 5B, his appearance 6 months after surgery.

**Neurofibroma.** An unusual case of neurofibroma is presented.

*Case 7 (D.H.)*. A 2-year-old child had a tumor involving the cranium and orbit, causing not only exophthalmos, ophthalmoplegia and blindness, but also intractable pain. Fig. 6 shows the serious deformity of the orbital structures and pigmented hairy nevus over the temporal region.

The lesion had been biopsied elsewhere, with pathological verification as benign neurofibroma. Unfortunately, tight application of dressing produced pressure necrosis of skin over the forehead and a skin graft was necessary—leaving the scar shown in Fig. 6. Roentgenograms of the skull showed distortion of the bony orbit and sphenoidal ridge.
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Fig. 6. Case 7 (D.H.). (A) Two-year-old child with severe proptosis and solid masses of tumor extending into periorbital tissues. Scar on forehead is a residual of pressure necrosis from a tight dressing, following biopsy elsewhere. (B) Lateral view indicates size of masses extending into upper lid and shows the hairy nevus over the temporozygomatic region.

Because of constant pain, transfrontal craniotomy was performed with orbital decompression including resection of the sphenoidal ridge. The tumor masses were closely related to structures of the ocular nerve in the superior orbital fissure, fanning out forward to the orbit and posteriorly to the extradural cranial region. Fig. 7 demonstrates the type of elongated masses that were resected.

Postoperatively, there has been continuation of some of the periorbital swelling, presumably from interference with venous and lymphatic drainage, but the general status is considerably improved since the child has been relieved of the former pain.

Glioma of Optic Nerve. Most gliomas of the optic apparatus have progressed to bilateral extension before diagnosis or surgical therapy may be accomplished, as indicated by Ingraham and Matson.9 There is the occasional case of unilateral optic-nerve involvement which may have a large intracranial component, without apparent extension to the opposite optic nerve.

Case 8 (R.H.). A 3-year-old boy was admitted with exophthalmos, ophthalmo-plegia and blindness of the right eye. Roentgenograms indicated an enlargement of the right optic foramen, and the possibility of a glioma was entertained. On the morning of the day that surgery had been planned, there developed abruptly signs of increased intracranial pressure. An emergency ventriculogram was performed, demonstrating a large right frontal mass, inferior to the ventricle.

Transfrontal craniotomy and orbital decompression were performed, and a large dumb-bell type of encapsulated mass was found arising from the right optic nerve at the optic foramen. The intracranial and intraorbital portions were completely resected and the right optic nerve was sectioned behind the orbit and just in front of the optic chiasm. The gross appearance of the lesion at the time of surgery
was considered compatible with neuroma of the optic nerve; however, pathological examination established the diagnosis of glioma.

In the 3-year period since surgery, the patient has shown no defect of visual acuity or field in the left eye.

**Congenital Lesions.** Various forms of congenital lesions present cranio-orbital problems. Surgical exploration may be necessary to establish the exact differential diagnosis as related to dermoid, epidermoid, teratoma, chordoma, mucocele and other lesions. It is of interest to note that the cystic dermoids and epidermoids tend to produce rather marked deformity of the skull and exophthalmos without the impairment of optic or ocular nerve seen with the solid lesions, such as chordoma and teratoma.

**Case 9 (D.K.).** A 43-year-old woman presented a 4-year history of right-sided ophthalmoplegia, amblyopia and exophthalmos. Roentgenograms of the skull taken 4 years previously showed sizable calcification in the sphenosellar recess. There had been recent development of orbital pain and generalized headache.

Roentgenograms of the skull showed an increase in the size of the calcified lesion in the right sphenosellar recess. Angiograms indicated that the internal carotid bifurcation was elevated above the lesion and that surgical approach, if made, should be along the temporal route. There seemed to be no opportunity for cure, but the patient and her family were anxious for any available interval of survival. She proved
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Fig. 8. Case 9 (D.K.). Calcification in right sphenosellar recess. Tumor was removed at operation. Pathological diagnosis: chordoma.

to be in extreme endocrinological deficit and, following preparation with hormones and electrolytes, craniotomy was performed. We were able to remove the gross lesion, the location of which is marked by the calcification in the right sphenosellar recess shown in Fig. 8.

It is of interest to note that the surgery was begun in the right temporal fossa and extended across the sella turcica to the medial aspect of the left temporal fossa. The patient followed a satisfactory course for about 1 year and then recurrence of the lesion developed with secondary complications and she died at her home in another city. The value of palliative surgery in cases of progressive lesions is always debatable but the interval of remission in this case seemed worth while.

DISCUSSION

The cases illustrated are representative of cranio-orbital problems that often confront the ophthalmologist and then the neurosurgeon, as outlined by Davis and Martin. Those patients, such as Case 4, who present decrease in visual acuity and field may require repeated examinations before the presence of a space-occupying lesion can be established. The combination of exophthalmos and a palpable mass may indicate to the ophthalmologist the need for biopsy or resection by an anterior approach, described by Henderson. These problems merit careful evaluation as to, first of all, the need for an intervention and, secondly, the proper approach if operation is indicated.
Once a neoplasm has invaded the sphenosellar recess, either primarily or secondarily, there is an imbalance established in a closely confined area containing important neurological and vascular structures. From then on, the situation does not remain static. Treatment is ordinarily in the nature of a compromise. The primary challenge is not only the matter of diagnosis, but also the design of therapy that will afford the most satisfactory solution to the particular problem that faces the patient.

The factors that ordinarily require surgical intervention have been outlined. There are exceptions to any such criteria. Coordinated observations by the ophthalmologist and the neurosurgeon can guide the patient’s course, even as it may change from one phase to another, as demonstrated in Case 6. The various factors that indicate a degenerative disorder in one case, or the acceptance of monocular vision in the presence of an obvious lesion, or the need for radical surgical approach in a different type of case, have all stimulated the progressive development of that important subspecialty, known as neuro-ophthalmology.

SUMMARY

Attention is directed to the anatomical relationship of the sphenosellar recess and to the clinical features of lesions in that region. The indications for surgery and certain guiding factors are discussed. Details of selected cases are presented.

REFERENCES