OCULAR SIGNS OF MENINGIOMA*

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A meningioma is a connective tissue neoplasm which may involve the meninges any place in the central nervous system. It is the second most common brain tumor, only the glioma occurring more frequently. Hoessly and Olivecrona (1) report that of 5,250 verified brain tumors 1,004 were meningiomas. The tumor is of particular interest to ophthalmologists because of the frequency with which ocular symptoms first cause the patient to seek medical attention. Those meningiomas which occur at the base of the brain, particularly in the region of the sphenoidal ridge and the tuberculum sellae, initially present eye signs which, if recognized promptly and treated appropriately, may well permit survival of the patient, since the tumor is usually histologically benign.

Meningiomas are also known as dural endotheliomas, meningeal fibroblastomas, meningeal fibroendotheliomas, and similar terms suggesting fibroblastic or mesothelial origin. The tumor occurs in two forms: (1) a global type which grows into and compresses nervous tissue, and (2) a carpet-like or en-plaque type which spreads across the tissue. The tumors usually do not invade nervous tissue but only compress it, so that if they are removed early in their course there may be a resumption of normal neural function. They have an unusual property of spreading through bone and causing a marked hyperostosis, a process which is frequent with involvement of the outer one third of the sphenoidal ridge in the en-plaque type. In addition, the tumor tends to insinuate itself into the crevices, cracks, and foramina of the brain and cranial cavity and to become very widespread.

Numerous pathologic classifications have been proposed for the tumor. Cushing and Eisenhardt (2), whose authoritative monograph is monumental in this field, trace it back to a primitive meningiocyte from that part of the ectoderm giving rise to cell masses on the neural groove and neural crest.

Thus, these tumors are particularly prone to occur near the large

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venous lacunae of the skull about which arachnoid villi are clustered. Arachnoid villi form the pacchionian bodies which perform functions similar to those of the reticuloendothelial apparatus in other parts of the body. They have an active fibrocytic role in keeping the cerebrospinal pathway cleared of debris and an important role in the formation of new membranes associated with the reparative intracranial processes.

Globus (3) believes that cytologic studies of the meninges are unlikely to reveal the origin of the tumor, and he has directed attention to the phylogenetic and ontogenetic processes which take place in development of the meninges. He traced its origin to the mesenchyme which gives rise to the bone forming periosteum, the collagen-producing anlage of the dura, the anlage of the epithelial (endothelial or mesothelial) covering of the arachnoid, and the primordium of the pia. Thus these tumors may present a variety of cell types, and it is not uncommon to find several cytologic types in different portions of the same tumor. Bailey and Bucy (4), in 1931, classified the tumor into mesenchymatous, angioblastic, meningioendotheliomatous, psammomatous, osteoblastic, fibroblastic, melanoblastic and lipomatous types. Cushing and Eisenhardt presented nine types with variants within each group. Since then it has been recognized that the histologic origin plays a minimal role in prognosis.

Records of patients with histologically verified meningiomas seen at the University of Chicago Clinics are the basis of this report. Excluded from consideration were meningiomas involving the spinal cord and those tumors in which the diagnosis was not based on examination of the tissue. A total of 143 intracranial meningiomas are considered. The age distribution is shown in Table 1. In this series the tumor caused

TABLE 1. AGE DISTRIBUTION OF 143 CASES OF MENINGIOMA

Age	*Number of patients*
Under 20 years	9
20 to 29 years	7
30 to 39 years	28
40 to 49 years	39
50 to 59 years	43
Over 60 years	17

the majority of patients to seek care during the fifth decade. The youngest patient was five years old and the oldest was eighty years old.

Meningiomas occur predominantly in females, and in this series 91 females were seen compared to 52 males. Not infrequently the onset of

symptoms is initiated by a recent pregnancy, a topic recently discussed in detail by Weyand, MacCarty, and Wilson (5). The Negro is not involved as commonly as the white. There is frequent association between trauma to the skull and subsequent meningioma, both as to association with trauma and to location of the tumor.

TABLE 2. ANATOMICAL LOCATION OF 143 CASES OF MENINGIOMA

Parasagittal		46
Convexity		35
Frontal	20	
Temporal	6	
Parietal	2	
Occipital	3	
Parieto-occipital	4	
Sphenoidal ridge		15
Tuberculum sellae		14
Olfactory ridge		10
Cerebello-pontine angle		9
Middle and posterior fossa		9
Multiple meningioma		3
Optic nerve sheath		2

The location of the tumors in this present series is shown in Table 2. The distribution corresponds closely to that reported by Cushing and Eisenhardt (2) and Olivecrona (6). The tumors that arise from the cranial base slightly exceed in number those that underlie the cranial vault. It must be pointed out that the exact origin of the tumor may be difficult to localize after growth of the meningioma, and sometimes its probable origin is estimated only by the symptoms produced in its early period.

MENINGIOMAS OF THE TUBERCULUM SELLAE

Meningiomas of the tuberculum sellae arise from the meningeal covering of the anterior and posterior clinoid processes and the floor of the sella. Their classic presenting signs are bitemporal constriction of the visual fields and unilateral optic atrophy occurring in the absence of demonstrable roentgenographic disease. Because of the intimate association of the optic chiasm with the area, tumors here may cause signs very early in their course. However, the region is crowded with important structures and with moderate increase in size, carotid arteries and cranial nerves are implicated so that removal constitutes a formidable and occasionally insurmountable surgical task.

There have been several recent reviews of the ocular symptomology of meningiomas of the tuberculum sellae, notably those of Grant and Hedges (7; reporting 30 cases) and Uihlein and Weyand (8; reporting

53 cases). Females were involved predominately, and almost invariably loss of vision with an accompanying field defect was the outstanding symptom. The field defect was occasionally scotomatous, but usually peripheral, and was associated with an optic atrophy which was usually monocular but sometimes involved both eyes.

Although roentgenographic studies may indicate no abnormality in the majority of instances, erosion of the clinoids, enlargement of the sella turcica, or a calcified tumor may be demonstrated. It must be emphasized that roentgenographic studies demonstrating a normal skull when combined with optic atrophy by no means exclude the possibility of meningioma as a cause of the symptoms.

Fourteen patients were seen with meningioma involving the tuberculum sellae. Of this number 5 were male and 9 were female. The patients ranged in age from twenty-nine to fifty-eight years, with a median age of forty-two years. Their outstanding symptom was loss of vision involving one or both eyes, arising from primary optic atrophy. Twelve of the 14 patients were seen initially by an ophthalmologist because of ocular symptoms.

Analysis of the field defect at the time the patient was first seen was complicated by the frequency with which one eye was entirely blind before the patient sought care. A homonymous hemianopsia occurred in 3 patients and a bitemporal hemianopsia in 6 patients. In 5 patients vision was so poor in one or both eyes that it could not be determined if the field defect in the better eye represented a heteronymous or homonymous type of involvement. In only one patient was the scotomatous type of field defect found as described by Schlezinger *et al.* (9). This subsequently broke through to the periphery to be part of a superior temporal quadrantanopsia.

The extent to which visual defect had progressed in this group of patients when they were first seen is surprising. Eleven of the 14 patients had vision in one or the other eye of less than counting fingers at 2 feet. One patient was blind in both eyes, and others had vision reduced to finger counting in each eye. Patients had noted failing vision over a period of time ranging from six months to four years.

The optic atrophy seen in these patients, particularly when monocular, must be distinguished from that arising from vascular disease, glaucoma, and tabes. The patients were of an age group where their ocular symptoms might be attributed to glaucoma or a vascular accident involving the blood supply of the optic nerve or retina. The normal size of the blood vessels in the optic atrophy associated with meningioma serves to distinguish it from atrophy due to vascular occlusion.

The intraocular pressure is normal and the progress of the field defect is more rapid than that seen in glaucoma. Glaucoma provocative tests are, of course, negative in meningioma. There are no other signs suggesting central nervous system syphilis or other inflammatory causes of optic atrophy. Two patients, both with advanced visual field defects, sought medical attention because of retrobulbar pain, although each had been aware of the visual disturbance prior to the onset of this pain. Two other patients sought care because of divergence of a blind eye, although the decreased vision had not apparently alarmed them.

Two patients had amenorrhea of one year and two years' duration respectively, attributed to involvement of the pituitary gland. Other than this, the neurologic examination did not contribute to the diagnosis other than in regard to the ocular signs.

Roentgenograms were diagnostic or suggestive of the disease in 7 of the 14 patients. Findings included erosion or hyperostosis of the sella turcica, calcification in the tumor itself, displacement of the pineal gland, and decalcification of the posterior clinoids. In 5 patients roentgenograms of the skull and optic foramen revealed no abnormality, but the tumor was diagnosed by means of contrast radiography.

In 2 patients in whom roentgenograms of the skull and optic foramen indicated no abnormality, and angiography and pneumoencephalography were normal, a definite diagnosis was established only by craniotomy. The high index of suspicion necessary for early treatment of these patients is indicated in the following two case reports.

CASE REPORTS

CASE 1. A twenty-nine-year-old Negro female was first seen December 10, 1954, complaining of a decrease in vision in the left eye which had first been noted ten months earlier. Examination indicated vision O.D. 20/30, corrected to 20/20; O.S. counting fingers at three feet, unimproved with correction. A marked left optic atrophy was present, and there was questionable pallor of the right optic disc. Visual field examination indicated a nasal island of vision remaining on the left side. The right visual field was normal. Roentgenograms of the skull and optic foramina and left carotid angiography revealed no abnormality. Neurologic examination was normal and, except for the optic atrophy, no other disease could be found. On December 24, 1954, a left frontal craniotomy was performed by Dr. Joseph P. Evans. A meningioma of the tuberculum sellae was found which encompassed both optic nerves. The tumor was resected with a minimal postoperative reaction and the patient was discharged two weeks later. When last examined in March, 1955, vision in the left eye was counting fingers at three feet. The right eye was normal. Histologic examination indicated a meningioendotheliomatous meningioma.

CASE 2. A fifty-seven-year-old white male farmer was first seen June 8, 1954, complaining of blurred vision for the previous six years. Vision was O.D.

finger counting at one foot; O.S. 10/200, corrected to 20/30. Examination indicated temporal pallor of each disk, the right more marked than the left. Visual field examination demonstrated an irregular contraction of the visual field of the right eye and a central scotoma of the left eye. Roentgenograms of the skull demonstrated no abnormality. A carotid angiogram and pneumoencephalogram indicated no intracranial disease. On August 10, 1954, Dr. Ralph B. Cloward did a right frontal osteoplastic craniotomy and removed a meningioma the size of a tomato from the tuberculum sellae. A small berry-like aneurysm on the superior surface of the left carotid artery was an interesting additional pathologic finding. When seen six months after operation, vision was corrected to O.D. 20/40, O.S. 20/20. The visual field was full in each eye.

The importance of comprehensive study of a patient presenting signs of optic atrophy even in the presence of normal roentgenograms cannot be overstated. When the visual loss is progressive, diagnosis may require craniotomy even when other neurologic signs are absent and intracranial disease cannot be demonstrated with roentgenograms or contrast studies.

MENINGIOMAS OF THE SPHENOIDAL RIDGE

Kearns and Wagener (10) have recently reviewed the ocular signs in 106 patients with meningioma of the sphenoidal ridge, of whom 73 presented a disturbance of the eyes as the only or one of the chief complaints. The sphenoidal ridge demarcates the frontal from the middle fossa of the skull and extends laterally from the anterior clinoid process to the pterional region of the cranial vault. It is composed of portions of both the greater and lesser wings of the sphenoid bone and is in intimate association with the orbit, the optic foramen, and the superior orbital fissure.

At the time of operation it may be difficult for the neurosurgeon to distinguish between tumors originating at the inner one third of the sphenoidal ridge and those arising from the tuberculum sellae or even the olfactory groove. Careful anamnesis and analysis of the ocular symptoms occurring early in the course of the tumor frequently will indicate the probable area of origin.

Cushing and Eisenhardt (2) found meningiomas of the deep clinoidal one third of the sphenoidal ridge to be characterized by unilateral failure of vision and primary optic atrophy. In 8 of 13 cases the eye was blind on admission; in 9 patients, proptosis was present. In 6 patients contralateral papilledema was present and in only 3 was the contralateral eye normal in all respects. Oculomotor palsies were present in the majority of their patients.

Tumors of the middle sphenoidal ridge may reach a large size with-

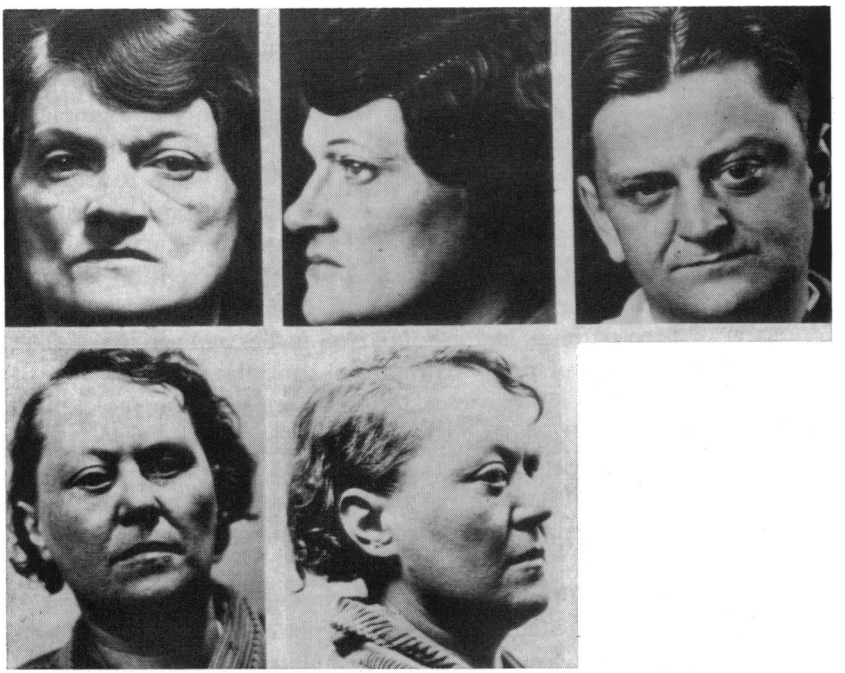

FIGURE 1. THE PROPTOSIS OF SPHENOIDAL RIDGE MENINGIOMA

out causing symptoms. Their chief ocular sign is bilateral papilledema, which is more marked on the side of the tumor.

Tumors of the outer one third of the sphenoidal ridge occur in both en-plaque and global form. The global type expands into the Sylvian fissure; it may remain asymptomatic until quite large, and causes an internal hydrocephalus with bilateral papilledema. This course was followed in 12 of the 13 patients who had a meningioma in this location, in Cushing and Eisenhardt's (2) series. The en-plaque tumor is prone to cause hyperostosis of the greater wing of the sphenoid with the development of a slowly increasing proptosis with reduction of vision and fullness of the temporal region. In 15 of 16 cases (Cushing and Eisenhardt) of en-plaque tumors there was exophthalmos; 10 patients had lost vision, but only 2 were blind.

Kearns and Wagener (10) found loss of vision, proptosis, diplopia, pain around one eye, and swelling of the lids to be the chief or one of the main complaints in 73 of 106 patients with sphenoidal ridge meningioma. The remaining 33 patients complained of headaches, convulsions, and mental changes. However, of these 33 patients without ocular

complaints, 15 had papilledema, 7 had optic atrophy, 3 had pupillary signs, and 1 had a visual field defect. Thus only 7 of the group of 106 patients were without eye findings.

In the present group, 15 patients were seen with meningiomas involving the sphenoidal ridge. Of this number, 11 were female and 4 were male. The patients ranged in age from thirty-one to fifty-seven years, with a median age of forty-one years. The outstanding symptom was decreased vision in the homolateral eye, which occurred in 10 patients. In 2 patients vision was reduced to light perception in the involved eye, and in the others central visual acuity varied from 20/200 to 20/20. Three patients had a homonymous hemianopic type of field defect. Of the group of 15, 10 had normal contralateral eyes.

Proptosis occurred in 8 patients. Of this number 2 were males, which is a circumstance not observed by Cushing and Eisenhardt (2), who found the en-plaque tumor of the pterional region exclusively in females. Proptosis was bilateral in 1 of the patients. All but one of these patients had roentgenographic evidence of an intracranial lesion involving the sphenoidal or orbital region.

Optic atrophy occurred in 5 patients, being bilateral in 2. Papilledema occurred in 6 patients; in 3 it was bilateral. In the remaining 3 patients papilledema occurred on the homolateral sides. Optic atrophy on the homolateral side preceded the development of papilledema in 2 of the 3 patients with bilateral swelling of the disks.

Three patients had ophthalmoplegia. It involved the third nerve in 2 of these patients, one of whom had internal ophthalmoplegia. The remaining patient had involvement of the sixth nerve. Of this group 1 had proptosis combined with optic atrophy with vision of 20/200 and 20/60, 1 had papilledema on the homolateral side, and 1 had vision of 20/200 and 20/20 with homolateral optic atrophy. None of them had involvement of the contralateral eye. These three patients with ophthalmoplegia most likely had the tumor originate at the clinoidal one third of the sphenoid ridge. The involvement of the motor nerves passing through the superior orbital fissure with resultant ophthalmoplegia surprisingly leads patients to seek care much earlier than when optic nerve involvement causes loss of central vision.

Pain in the eye was a prominent symptom in 3 patients, each of whom had proptosis and radiologic evidence of a sphenoidal ridge hyperostosis. In 2 of these patients a homolateral papilledema occurred.

General neurologic signs occurred in 7 patients, varying from headache to personality changes. Except for the ocular signs, neurologic examination did not contribute to the diagnosis.

Slowly progressive proptosis occurring in a woman of middle years is the characteristic symptom of sphenoidal ridge meningioma. The proptosis arises not only from hyperostosis of the orbital bones but, according to Knudtzon (11), may also be attributed to invasive growth into the orbit and congestive changes of the orbital tissue resulting from stasis of the cavernous sinus.

Roentgenographic examination of these patients will assist immeasurably in the diagnosis.

MENINGIOMAS EXTENDING ALONG OPTIC NERVE SHEATHS

The optic nerve within the orbit may be involved in a meningioma that originates within the cranium, at the optic foramen, or within the orbit itself. The chief criteria for orbital origin are that there be no cranial extension and that the proximal stump of the optic nerve adjacent to the optic foramen be normal and have a normal covering. Verhoeff (12), in 1932, stated that in each of 8 cases studied histologically the tumor had invaded the orbit from the cranium. He stated that on a priori grounds there was no reason why such a tumor should not occur but that it had never been demonstrated histologically. Coston (13) reported a case in 1936 which came close to fulfilling the criteria for intraorbital origin but was not definitely proven. Friedenwald (14) is credited with describing a case which fulfilled the requirements of orbital origin in 1937, and Craig and Cogela (15) described a single series of 17 cases of intraorbital meningioma, 9 arising within the orbit, 3 from the nerve within the optic foramen, and 5 apparently from other orbital structures.

The chief symptoms of optic nerve meningioma are decreased vision and proptosis which are slowly progressive. Either may occur without the other. Ocular movements are usually not disturbed. Roentgenographic examination usually indicates a normal optic foramen even though the tumor may extend through it. Intraocular involvement is uncommon but has been reported by Hudson (16), Coston (13), Dunn and Walsh (17), and Martin and Schofield (18). Craig and Cogela (15) do not mention this type of involvement although their series is by far the largest in the literature.

Two patients with meningioma extending along the optic nerve sheath were seen in this group. Proptosis was the outstanding symptom in the first and reduction of vision in the second. Because of the paucity of descriptions of this type of involvement the cases are presented in detail.

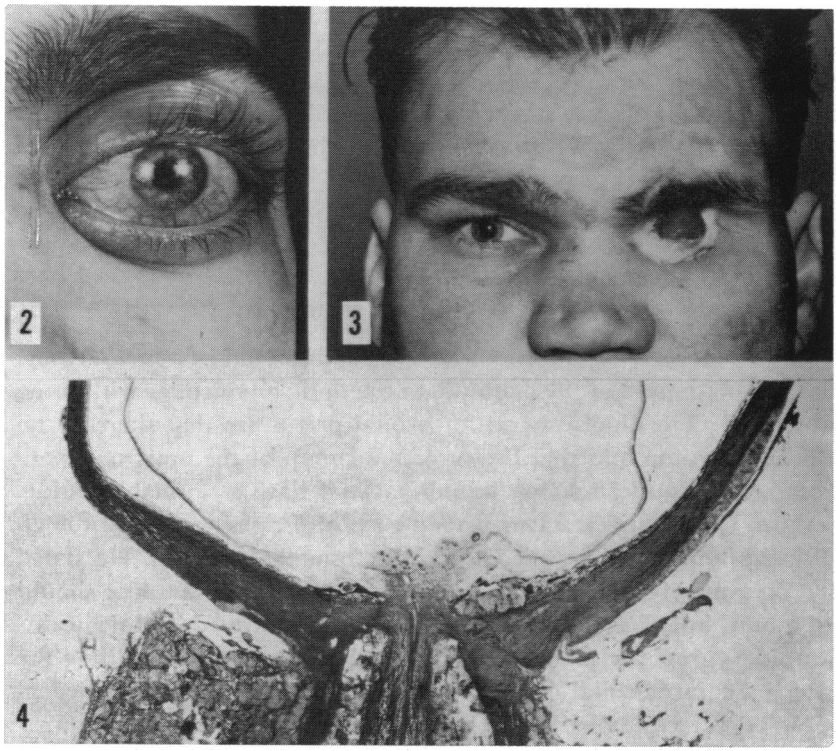

FIGURE 2. CASE 3 AT EIGHTEEN YEARS. SYMMETRICAL PROPTOSIS OF MENINGIOMA OF THE OPTIC NERVE

FIGURE 3. CASE 3 AT THIRTY-THREE YEARS. ORBITAL RECURRENCE OF MENINGIOMA

FIGURE 4. CASE 4. INVASION OF CHOROID AND OPTIC NERVE BY MENINGIOMA

CASE REPORTS

CASE 3. The patient, aged eighteen years, was first seen in October, 1935, complaining of proptosis of the left eye of two and one half years duration. Vision was O.D. 20/15, O.S. 20/40. The Hertel exophthalmometer reading was right 13 mm., and left 32 mm. Roentgenographic examination indicated the left orbit to be distinctly larger than the right with its posterior wall more dense than on the opposite side. The optic foramina were of equal size.

In December, 1935, the eye was enucleated because of a purulent inflammation of the cornea arising from exposure keratitis. Examination of the enucleated globe indicated a meningioma extending to the globe but not invading the optic nerve itself. The meninges of the proximal optic nerve contained tumor cells. The patient was not seen again until 1949, when he was

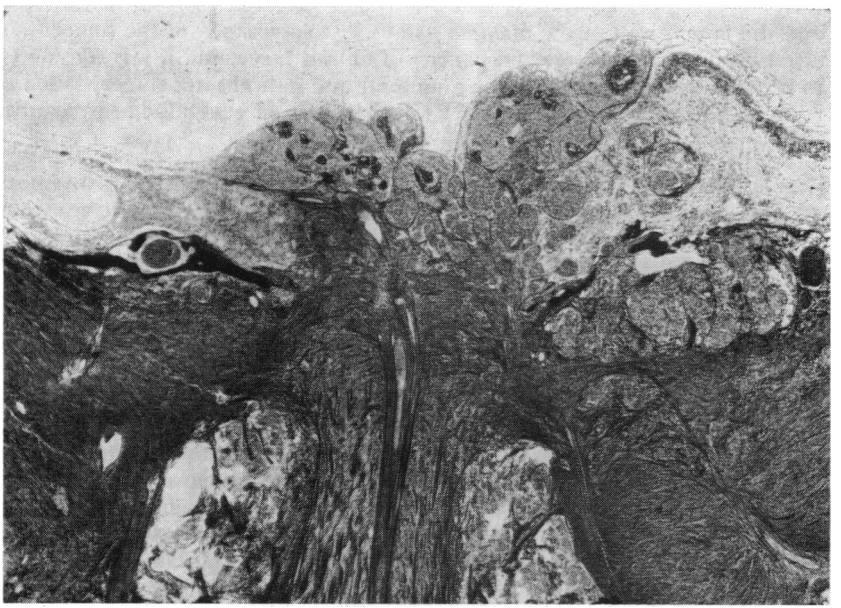

FIGURE 5. CASE 4. INVASION OF OPTIC DISC AND CHOROID BY MENINGIOMA

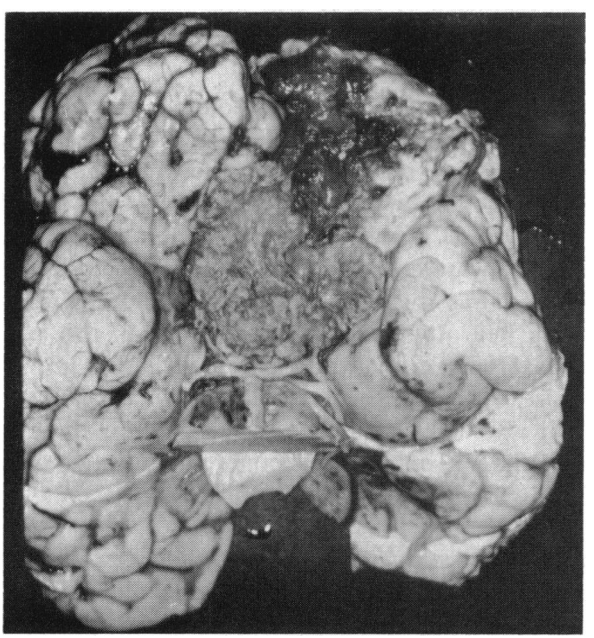

FIGURE 6. CASE 4. EXTENSION OF MENINGIOMA TO THE PONS FROM ORIGIN AT SPHENOIDAL RIDGE, COMBINED WITH ORBITAL INVOLVEMENT

thirty-three years old. At this time the tumor involved the roof of the orbit and the lateral wall of the maxillary sinus. Cytologic study of the tumor indicated many mitotic figures and paucity of fibrous tissue, and it was considered to be a malignant meningioma. Craniotomy and radical sinus surgery failed to remove the tumor completely and he died in 1952 of generalized intracranial involvement.

CASE 4. This patient was first seen in the Eye Outpatient Section, November 21, 1952, complaining of proptosis of the left eye during the past year. She stated that the eye had become suddenly blind twelve years earlier, and that during the past year it had become increasingly more prominent. Within the past three years, she had three cerebrovascular accidents which were attributed to hypertension. Examination indicated an orbital mass palpable in the lower fornix of the left orbit. A curd-like elevation of the nerve head 4 diopters in height was present in the left eye and measured approximately 2 diopters in diameter. Visual field examination indicated a constriction of the right temporal field. Roentgenographic examination of the left optic foramen indicated it to be larger than the right though within the normal limit of size. There were no neurologic findings other than ocular signs. A transfrontal craniotomy was performed and a meningioma of the sphenoidal ridge was found which extended posteriorly to the pons and anteriorly to the eye. The patient died sixteen days after the operation. Histologic examination of the globe removed at the time of postmortem examination showed invasion of the sclera and the choroid by tumor mass. The optic nerve and the meninges about it were infiltrated by tumor cells with extension into the eye at the disk. The retina was invaded about the disk by the neoplasm and showed cystic degeneration at the ora serrata.

The reports of patients with meningioma extending along the optic nerve sheaths together with the findings in these two patients indicate the futility of purely ocular surgery in their management. In the majority of cases it is extremely likely that the tumor has already extended intracranially even if it originated within the orbit. Since the optic foramen is not always enlarged there may be no indication of intracranial disease. Thus the removal of such tumors is probably in the province of the neurosurgeon who can unroof the orbit and inspect the optic foramen. However, the tumor is relatively slow growing and does not metastasize, so that in the event of discovery of the true nature of the neoplasm after the usual type of enucleation there has been no great harm to the patient, provided any remaining tumor is removed.

OLFACTORY GROOVE MENINGIOMAS

Olfactory groove meningiomas arise in that area which corresponds to the suture line separating the plane sphenoid and the cribriform plate. With olfactory groove tumors as well as suprasellar tumors, a disturbance of vision is likely to be the inaugural symptom in each

group from quite different causes. A suprasellar meningioma, while still quite small, serves to elevate the chiasm, thereby stretching the decussating fibers and causing bitemporal hemianopsia. Olfactory groove tumors attain a large size before vision is affected by downward pressure upon the nerves and chiasm from above. By that time papilledema and scotomas involve each eye equally. The olfactory groove meningioma has been described as the chief cause of the so-called Foster Kennedy syndrome, although it seems likely that meningiomas arising from the middle one third of the sphenoid ridge are more frequently at fault. From the location, the initial symptom is a unilateral anosmia. However, tumors may reach quite a large size before patients note an anomaly of their sense of smell.

Ten patients were seen with verified olfactory groove meningiomas. Their occurrence without production of symptoms of any type is indicated by finding the tumor in 3 of these patients on autopsy following death from quite unrelated disease. None of these three patients had complaints relating to their eyes or any symptoms or findings suggestive of an intracranial neoplasm.

Symptoms presented by the remaining patients related to personality changes in 3, headaches in 2, loss of smell in 2, and convulsive episodes in 3. Loss of vision was the chief complaint in only 1 patient although each of the patients presented ocular signs. The Foster Kennedy syndrome of optic atrophy on one side and contralateral papilledema was seen in only 1 patient, a fifty-five-year-old woman blind in one eye for the previous twenty-seven years. Two patients had bilateral papilledema with good central vision. Each of the remaining patients had bilateral optic atrophy with the defect far more severe on one side than the other. In all but one of the patients the eye was entirely blind. In the single patient without monocular blindness, vision was reduced to light perception.

The ocular signs observed in these patients are suggestive only of an intracranial tumor rather than being helpful as far as localizing signs are concerned. The associated neurologic symptoms, which are frequently severe, the commonly present roentgenographic findings, and the ocular signs should alert the ophthalmologist to the possibility of severe intracranial disease being present.

Tumors involving the remainder of the brain are not of specific ophthalmologic importance. The commonest types are the parasagittal meningiomas or those tumors which lie at either side of the parasagittal sinus and are adherent to the falx cerebri. If separated by normal cortex they fall into the classification of tumors of the cerebral convexity.

TABLE 3. OCULAR SIGNS IN 143 CASES OF MENINGIOMA

Location	*Number of patients*	*Diminished vision*	*Blindness*	*Papilledema*	*Optic atrophy*	*Exophthalmos*	*Field defects*	*Pupillary signs*	*Oculomotor lesions*	*Nystagmus*	*No ocular findings*
Olfactory groove	10	8	6	2	7	0	7	7	3	0	1
Optic nerve sheath	2	2	1	1	2	2	1	1	0	0	0
Multiple meningioma	3	1	0	3	1	1	1	2	1	0	0
Cerebellopontine	9	5	0	6	0	0	4	2	1	5	0
Tuberculum sellae	14	13	5	0	11	0	14	7	4	0	0
Sphenoidal ridge	15	11	1	6	4	9	10	2	4	0	1
Parasagittal	46	16	1	13	6	0	12	6	1	0	21
Convexity	35	15	1	23	4	1	20	5	6	0	7
Middle and post fossa	9	5	1	6	2	1	6	2	4	0	0
Total	143	76	16	60	37	14	75	34	24	5	30

These tumors, of course, give rise to ocular signs, but largely because of involvement of the optic tracts or ocular muscle nuclei and interference with ventricular drainage, with resultant papilledema. Symptoms and signs of these neoplasms are by no means specific, and they fall into the general class of brain tumors. The ocular changes observed in these patients are shown in Table 3. The ocular symptoms vary with the location and size of the tumor and are not characteristic of meningioma.

SUMMARY

A meningioma is a benign connective tissue neoplasm which may arise from the meninges any place in the central nervous system and which, if removed early in its course, may permit return of normal neural function. The records of 143 patients with histologically verified intracranial meningiomas were reviewed with special reference to the ocular signs.

Fourteen patients with meningioma of the tuberculum sellae were seen, 11 of whom had optic atrophy with vision in one or both eyes reduced to less than counting fingers at 2 feet. In 2 patients roentgenograms of the skull and contrast radiography did not indicate the abnormality, but the tumor was found by surgical exposure. It is urged that meningioma be considered a likely cause of the defect in middle-aged patients with progressive optic atrophy not arising from glaucoma, vascular disease, or syphilis.

Meningioma of the sphenoidal ridge was found in 15 patients. Symptoms varied with the portion of the ridge involved, but optic atrophy of the homolateral eye and proptosis were the outstanding signs. Roentgen-ray examination will frequently aid in the diagnosis, since meningiomas of this area, particularly those involving the pterional portion of the sphenoidal ridge, cause a characteristic hyperostosis.

Because of their tendency to insinuate themselves into crevices and foramina in the brain, meningiomas may extend into the orbit along the sheaths of the optic nerve. This causes proptosis, usually with a normal-sized optic foramen with no limitation of ocular rotation. Although meningiomas may have their origin from the meningeal sheaths of the optic nerve, this is so unlikely that it is urged that the neurosurgeon be invited to remove such tumors by means of a craniotomy, so that the full extent of the tumor can be assessed.

Tumors of the olfactory groove attain a large size before causing visual symptoms. Of 10 patients with meningiomas of this area, the majority had no ocular signs; the visual signs that did occur aided in localizing the tumor in only one instance.

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DISCUSSION

DR. WILLIAM L. BENEDICT. The signs in the ocular system caused by meningiomas may make it possible to differentiate the initial lesion from others in the same category. When considered in conjunction with other factors that characterize intracranial (including intraorbital) space-taking lesions, such as those revealed by roentgenography, neurologic deviations, history, exophthalmos,

disturbances of ocular motility, changes in visual fields and in visual acuity, and less distinctive systemic symptoms, these signs constitute the basis for assuming that, given certain findings, with exclusion of certain other findings, a space-taking lesion is present. The evidence narrows the probabilities to a small list of possibilities, of which meningioma is one. There are no signs that are pathognomonic of meningioma and the final diagnosis is made by histopathologic examination. Fortunately, the lesions in this category, consisting of tumors, cysts, and aneurysms, are mostly nonmalignant. Although they are not susceptible to irradiation and usually require surgical intervention, in most doubtful cases surgery may safely be deferred until definitive signs appear.

A peculiar feature of meningiomas is a tendency to expand along lines of least resistance and force their way into "all anatomical crannies and pockets." They may extend into the orbit through the superior orbital fissure or the optic foramen, along communicating blood vessels, and directly through the orbital walls. In the latter event hyperostosis occurs. By the time the tumor in the orbit produces proptosis the hyperostosis is quite clearly discernible in the roentgenogram. Exophthalmos with hyperostosis of the floor of the cranium is also produced by venous aneurysms and other benign space-taking lesions, and differential diagnosis cannot be made on such evidence. Once the tumor has invaded the orbit, resistance to its expansion is practically nil and extension may proceed in any direction. In any event, the increase in mass within the orbit causes proptosis. If the mass is mostly in the posterior portion of the orbit, the proptosis will likely be straight ahead; if it is in the anterior part of the orbit the globe will be displaced laterally or downward as well as forward and the tumor may be felt by palpation. While the weight of evidence is in favor of meningioma because of its statistical incidence among intracranial tumors, the possibility of lesions of another character must be considered. There are several reported cases of two or more tumors of different types, some simultaneous and others separated by a few years. Two or more tumors may be found in the orbit. One or both may originate outside or inside the orbit. Rarely will two or more tumors within the orbit be determined by clinical signs alone. Intraocular and extraocular orbital tumors of different pathological nature may, though rare, be present at the same time, for instance, a glioma and a meningioma of the same optic nerve (J. François & M. Rabaey, *Acta Ophth.* 30:203-21; 52f.).

Multiple primary malignant tumors are rare in children. Doctors Newell and Beaman have presented a large series of carefully annotated cases and a review of the literature. The relative incidence of signs of meningioma compares favorably with previous reports. The value of their contribution is therefore in its confirmation of opinions now held by ophthalmic surgeons.

In most cases of meningioma where the ocular signs are dominant, the oculist will be consulted early in the course of the disease. Exophthalmos in the absence of systemic or local disease is often considered to be an invitation to surgical exploration in hope of clearing the diagnosis by biopsy. This, I believe, is not good surgical judgment. In case of orbital meningioma, frontal exploration is futile. In many other types of tumors, the transcranial approach is far superior, even though the tumor be confined to the orbit.

The authors have pointed out the "characteristic complex" of meningiomas

originating in three situations where they are commonly found, namely, "ridge meningiomas" and "suprasellar meningiomas." This categorical classification is distinctly helpful in the management of any case.

DR. HAROLD HENRY JOY. This has been a most comprehensive analysis of the ocular signs of meningioma. I wish to stress the importance of early diagnosis of meningiomas of the tuberculum sellae. They are of great ophthalmologic concern because their situation in the chiasmal region is such that pressure on the visual pathways, with consequent visual defects, occurs at an early stage, long before any other signs or symptoms appear.

Meningiomas which are typically suprasellar exert pressure on the chiasm causing bitemporal field defects and the picture of primary optic atrophy (Cushing's Syndrome). This syndrome has so long been associated with suprasellar meningiomas that we are apt to lose sight of the fact that it applies only to typical cases. But many of these tumors are situated more anteriorly, so that pressure is primarily exerted on the optic nerves immediately in front of the chiasm rather than on the chiasm itself. This presents quite a different ophthalmologic picture. In these prechiasmal meningiomas the visual function of the two eyes is attacked successively rather than simultaneously. As a consequence vision in one eye may be seriously impaired while that of the other eye remains normal or shows only a small temporal field defect. Then as the tumor gradually involves the optic nerve of the second eye its vision becomes affected. This may take weeks, months, or even years.

The field defects in these cases are usually bizarre, consisting of a combination of central and peripheral involvement. One of the characteristics is the rapid development of central scotomas which may lead to a mistaken diagnosis of retrobulbar neuritis. In other cases the resultant optic atrophy is sometimes ascribed to such causes as vascular disease and syphilis.

The presence of an unexplained visual defect should never be ignored. Careful and repeated perimetric studies may yield the first clinical sign of a meningioma at the base of the brain.

Since not only vision but also life itself may be at stake, early diagnosis is essential. This can usually be accomplished by angiography. In such cases, the close relationship of the visual pathways to the arteries at the base of the brain assumes particular importance. In this congested area even a small lesion impinging on the pathways tends to displace the adjacent arteries. Their conversion into radiopaque landmarks provides visualization of the displacement, and sometimes of the vascular tree of the tumor itself. Hence, suprasellar and presellar meningiomas lend themselves especially well to angiographic study.

DR. DAVID O. HARRINGTON. I should like to make a brief comment on the specificity of the visual field defect in meningioma. Dr. Newell has mentioned the visual field defects, and he has pointed out the fact that they occurred in 75 out of 143 cases. They were present, for example, in 100 percent of meningiomas of the tuberculum sellae, in 10 out of 15 tumors of the sphenoidal ridge, in 7 out of 10 olfactory groove tumors, and so on. They are an important ophthalmological observation in these cases.

Dr. Newell also mentioned that very frequently in the prechiasmal type of meningioma the visual defects were not the usual bitemporal type of defect

which one might expect, and this is partially true. If the visual field defects in these prechiasmal types of meningiomas are analyzed carefully and if the perimetry is quantitative, a fairly large percentage of these defects will be found to show certain bitemporal characteristics which are of diagnostic value. For example, the patient who can see only hand movements in one eye and has an apparently normal field in the opposite eye may, when quantitatively analyzed on the tangent screen and perimeter with gross stimuli, show only a temporal loss in the field of the "blind" eye and, with very small stimuli, the beginning of a temporal quadrant loss in the "normal" eye. The field loss, then, is a bitemporal hemianopia, although at first glance it appeared to be a unilateral field loss.

The loss of central vision in these cases, when quantitatively analyzed on the tangent screen, will often develop the so-called junction scotoma of Traquair. In other words, an apparent central scotoma when carefully analyzed will resolve itself into a temporal hemianoptic scotoma for very large targets, while the peripheral field when tested with smaller test objects may show a temporal loss.

The same thing may be said of lesions in the posterior portion of the visual pathway, such as the meningiomas of the falx which lie between the occipital lobes. These tumors produce field defects having certain specific characteristics which are diagnostically valuable. Those which occur in the anterior portion of the falx and involve the anterior portion of the calcarine fissure may show such large areas of macular sparing that the field defect cannot be demonstrated on the tangent screen but requires the use of the perimeter. In one such case the area of macular sparing was more than 30 degrees. These field defects in lesions of the occipital lobe are also congruous, completely symmetrical even to minor degrees of deviation from the horizontal and vertical meridian. This symmetry of the field loss is of importance in the localization of the tumor and may even make possible an intelligent guess as to its pathology.

So, in conclusion, I should like to urge careful quantitative analysis of the visual field defects which are so frequently found in these meningiomata and which may be of great importance in their diagnosis and localization.

DR. C. WILBUR RUCKER. Dr. Newell has done us a fine service, I think, in calling attention to the frequency with which meningiomas cause disturbances in the visual system. You recall that he has a simple classification, based on whether the tumor is shaped like a globe or shaped like a carpet.

He points out that the globe-shaped tumors are recognized early because they produce symptoms that can be recognized, and that they are often removable. The carpet-like kind are more difficult to diagnose. The early signs are not very localizing and the late signs may be quite confusing.

One case I saw just before I left for this meeting is relevant. It concerns a woman who had headaches for two years and loss of sense of smell for one year. For two years she had also some loss of vision. During the two years in which she suffered visual impairment she had had her glasses changed twice without securing relief. Her fundi were normal. There were central scotomas in the fields of both eyes. As far as I could tell, there was involvement of both optic nerves. I did not know whether this was caused by an inflamma-

tion or a tumor. The neurologist found that she had an odd personality, that she was silly and forgetful. He confirmed the loss of sense of smell. These findings would indicate anterior fossa involvement. She had ataxia, and one cornea was anesthetic. From time to time she had a little nystagmus. So she had symptoms also in her posterior fossa.

The neurologist made a diagnosis of meningioma en plaque, arising in the anterior fossa and extending back to the brain stem. The neurosurgeon found such a tumor. He said it had arisen between the frontal lobes, encircled both optic nerves, and grown down into the ethmoid sinuses, across the floor of the middle fossa back to the brain stem. Of course, he was not able to remove it. Here, although the first symptoms concerned the eyes, the patient was not incapacitated, and the localizing symptoms appeared only after the condition had become inoperable. Perhaps if one or two of the ophthalmologists, instead of changing the glasses, had taken the trouble to chart visual fields, the diagnosis might have been made a bit earlier.

DR. ALAN C. WOODS. Dr. Joy has called attention to the fact that frequently in the so-called postfixation of the chiasm, the pressure point from the suprasellar tumor may be on a nerve, and Dr. Harrington has called attention to the bitemporal defects which are characteristic of pressure from below on the nasal side. In a very small percentage of people there is prefixation of the chiasm, and in these cases a suprasellar tumor may press on a tract. Dr. Talbot Daly had such a case some years ago. Instead of having a bitemporal defect or a unilateral amblyopia, there was a homonymous hemianopsia. Fortunately, other aspects of the case indicated a chiasmal lesion, and Dr. Daly used the transfrontal route and found a meningioma right along the tract.

DR. FRANK W. NEWELL. I wish to thank the discussers for their valuable comments, particularly their emphasis upon early diagnosis and early surgery.

The exophthalmos seen with meningiomas may arise from venous congestion, from hyperostosis of the orbit, or from actual invasion of the orbit by the tumor.

The homonymous defect mentioned by Dr. Woods has been observed in 3 of the 14 patients with tuberculum sellae tumors.

I agree with Dr. Benedict that the exact diagnosis of a meningioma depends upon histologic appearance. However, because of their slow growth, their involvement of certain areas of the cranium, and their occurrence at certain well-defined ages, one can be highly suspicious of a meningioma when certain symptoms are observed.