MENINGIOMA OF THE OPTIC FORAMEN AS A CAUSE OF SLOWLY PROGRESSIVE BLINDNESS

REPORT OF 3 CASES

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MENINGIOMAS arising from within the orbital cavity make up only a very small segment of intracranial meningiomas. Primary intra-orbital meningiomas have been removed surgically and verified pathologically in 17 cases at the Mayo Clinic. Included in this group of intra-orbital meningiomas are those tumors that arise from the meninges within the confines of the optic foramen and that subsequently tend to extend both intra-orbitally and intracranially. These latter form an interesting subgroup in that, aside from their extreme rarity, they grow at an unusually slow rate and demonstrate a remarkable tendency toward bilaterality. The only symptom associated with the foraminal meningiomas is a slowly progressive blindness not accompanied by headache or pain.

The first reported instance of the "synchronous occurrence" of meningeal tumors involving both optic nerves was that described by Schott in 1877 as an incidental finding on postmortem examination in the case of a 55-year-old blind charwoman who died of hepatic cirrhosis. The tumors were the size of a bean and were intimately attached to the arachnoid sheath of each optic nerve.

To our knowledge, the only other case of such bilateral tumors is that reported by Dandy in 1923. The neoplasms were diagnosed clinically, and an intracranial operative procedure was used for the first time in the treatment of such lesions. Dandy particularly stressed the need for distinguishing these tumors from optic neuritis because he felt that unnecessary blindness could be prevented if the true situation were recognized sufficiently early. His case was that of a 13-year-old girl who had had progressive bilateral loss of vision for a period of 7 years. At operation he discovered collar-like growths that encircled the optic nerves and extended 0.75 cm. intracranially and 1.5 cm. intra-orbitally from their attachment at the point where the dura is "reflected" as it passes through the optic foramen. Dandy also reported the case of an 8-year-old boy who had noted bilateral loss of vision but in whom he found only a single neoplasm.

These foraminal meningiomas should not be confused with the small tumors arising along the mesial part of the sphenoid ridge and producing visual symptoms secondarily as a result of their encroachment upon the optic nerve.

Meningiomas arising from within the optic foramen have been removed

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surgically in 3 cases at the Mayo Clinic. In 2 instances the tumors were bilateral and in the 3rd it was unilateral.

REPORT OF CASES

Case 1.—A 46-year-old white man noted the onset of progressive loss of vision in the left eye 10 years prior to examination. Within 3 years he was completely blind in that eye. Four years later vision in the right eye began failing, slowly and progressively.

Ophthalmologic examination disclosed that vision was reduced to 6/15 and was limited to the nasal field of the right eye. The right optic disc was pale and was elevated about 2 D. The left disc appeared even paler and the disc margins were blurred. Roentgen examination of the skull and the optic foramina gave normal results.

The visual loss was attributed to a chiasmal lesion, one that originally had been prechiasmal on the left. A left transfrontal craniotomy was carried out and 3 distinct tumors were encountered (Fig. 1). The first was situated along the outer third of the sphenoid ridge and measured 2.5 by 2.5 by 2.0 cm. It was removed without difficulty. When the optic chiasm was exposed, a small, cufflike, pinkish-purple mass was seen encircling the left optic nerve at the margin of the optic foramen. The entire tumor was removed. A similar, but slightly larger tumor measuring 1 cm. in diameter was found encircling the right optic nerve. One small lobule of this mass extended down into the sella and another to the right of the optic nerve. Only partial removal was possible because of its firm attachment to the optic nerve at the foramen.

The patient's impaired vision subsequently progressed gradually to complete blindness.

Case 2.—A 30-year-old housewife first became aware of failing vision in the left eye at the age of 12 years, and over a period of several years gradually became completely blind in that eye. The cause was never determined and the patient felt well otherwise. Eighteen months prior to examination, she noted the onset of progressive diminution of vision in the right eye. There were no other symptoms.

Physical findings were normal with the exception of those relating to the eyes. There was no proptosis and the eyes rotated normally. Vision was reduced to perception of moving objects in the temporal field of the right eye. Ophthalmoscopic examination disclosed marked pallor of the right disc with residual edema of 2 D, and similar extreme pallor of the left disc but with moderate loss of disc substance.
temporally. Roentgen examination of the head and of the optic foramina gave normal results.

The lesion was suspected to be located in the region of the optic chiasm, and a right transfrontal craniotomy was performed. Upon elevation of the right frontal lobe, small, dual, cufflike tumors were encountered encircling and compressing the optic nerves at the optic foramina (Fig. 2). The tumors were dissected free from their attachments within the rims of the optic foramina with a blunt hook, and the dorsum of the optic canal on that side was decompressed.

The patient convalesced without incident and a few months after dismissal reported that she had noted some subjective improvement in the vision of her right eye.

Case 3.—A 41-year-old housewife experienced progressively diminishing visual acuity in the left eye over a period of 12 months. Examination revealed 6/20 vision in the right eye and 6/60 in the left eye. A large, dense cecocentral scotoma was found in the field of the left eye. The left optic disc was pale. Roentgen examination of the head revealed early pressure erosion of the floor of the sella turcica. Pneumoencephalograms were normal and further observation was recommended.

When examined 10 months later, vision in the left eye was so impaired that movements of the hand were perceptible only at a distance of 2 m. and the pallor of the left optic disc had progressed. The progression of symptoms was suspected to be due to a small tumor impinging upon the left optic nerve. The optic chiasm was explored through a left transfrontal craniotomy. A small reddish, granular tumor was encountered lying along the inferomedian surface of the optic nerve within the optic foramen (Fig. 3). The proximal third of the orbital roof was removed and the
tumor was found to measure not more than 4 mm. in diameter. The atrophic optic nerve was sacrificed and the neoplasm was removed in its entirety with a curet.

The patient recovered without incident. On recent re-examination she was asymptomatic and the vision in the right eye had remained unchanged.

**COMMENT**

All the tumors encountered in these 3 cases were meningiomas of the psammomatous variety and all had exhibited an amazingly slow rate of growth. The presence of meningiomas attached to the sheath of the optic nerve may be explained adequately by the clusters or "caps" of arachnoidal cells occurring so abundantly along the intra-orbital course of the optic nerve as suggested by previous investigators. In all probability, the cufflike growths arising within the optic foramen are but "sheath" meningiomas situated within a specific location. However, it is difficult to explain why this special predilection should exist and why these growths should demonstrate a tendency toward bilaterality. Microscopic examination of normal optic nerves and their sheaths (Fig. 4) does not disclose the clumping of arachnoidal cells to be any more frequent or more exaggerated within the optic foramina than anywhere else along the intra-orbital course of the optic nerve. Yet, 2 of the 3 patients presented bilateral tumors, 1 of the growths appearing fully 16 years after its mate.

Review of these cases serves to emphasize the fact that slowly progressive blindness, unaccompanied by headache or pain, is the only symptom to be expected in the presence of these tumors. Unfortunately, in all 3 instances blindness had been allowed to advance to the point at which damage to the optic nerves had become irreparable before the probable nature of the lesion was suspected. Surgical intervention in all these cases served only to establish the diagnosis and probably had no therapeutic effect. There is every reason to believe that with early recognition of these lesions and their prompt removal, failing vision may be controlled and possibly even improved or
restored. With the aid of the ophthalmologist and the technics that he has at his disposal, including carefully performed studies of the visual fields, it is possible to distinguish these tumors from non-surgical lesions of the optic nerve. The possible presence of a surgical lesion such as a foraminal meningioma should be suspected in all instances in which the patient presents himself with progressive, painless loss of vision due to a lesion of the optic nerve.

SUMMARY

Meningiomas arising from within the confines of the optic foramen are extremely rare and are of interest in that they tend to occur bilaterally and grow at an amazingly slow rate. Three additional cases are reported in which meningiomas of the optic foramen were removed surgically. In 2 instances the tumors were bilateral.

Slowly progressive blindness, unaccompanied by headache or pain, is the only symptom presented with these lesions. Early recognition and prompt surgical removal is paramount in the treatment of such tumors.

REFERENCES