Untreated Bilateral Optic Nerve Sheath Meningiomas Observed for 27 Years

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Abstract: A 48-year-old woman noted progressive, painless visual loss in her left eye. She was diagnosed with bilateral optic nerve sheath meningiomas (ONSMs), extending across the planum sphenoidale. Radiation was offered, but the patient declined. She has been followed for more than 27 years with stable visual function and neuroimaging findings. Bilateral ONSMs, although usually described as aggressive in nature, may follow a stable clinical course.

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A 75-year-old woman noted progressive, painless visual loss in her left eye that began at age 48. At that time, visual acuity was 20/30 in the right eye and no light perception (NLP) in the left eye. Neuroimaging revealed bilateral optic nerve sheath meningiomas (ONSMs), left greater than right, extending across the planum sphenoidale. She was given the options of surgery and stereotactic radiotherapy, but declined both. Serial examinations revealed stable visual acuity, visual fields, and neuroimaging findings.

FIG. 1. A. At presentation, the right visual field (HVF: 24-2) shows an inferonasal defect (deviation, –6.49 dB; pattern standard deviation, 8.42 dB). B. Fourteen years later, the right visual field (HVF: 30-2) is unchanged (deviation, –6.58 dB; pattern standard deviation, 8.63 dB). HVF, Humphrey visual field.
Her medical history was significant for hypertension treated with clonidine. Family history was noncontributory. She had a 20 pack-year smoking history and quit at age 40. A review of systems was negative for headaches or associated neurological symptoms.

At age 61, she first presented to our institution with a visual acuity of 20/30 in the right eye and NLP in the left eye, and the left pupil was amaurotic. The patient identified 11/15 color plates with the right eye and there was 3 mm of left proptosis. Anterior segment examination was unremarkable. Both optic discs were pale, with an optic optociliary shunt vessels on the left. Automated visual field testing of the right eye showed inferonasal loss (Fig. 1A).

Contrast-enhanced magnetic resonance imaging (MRI) of the brain and orbits were consistent with a bilateral ONSMs, left greater than right, with contiguous spread across the planum sphenoidale (Fig. 2A, B). The optic chiasm was uninvolved.

After more than 27 years of follow-up, the patient’s vision is unchanged. The appearance of the optic discs (Fig. 3) and visual fields (Fig. 1B) remain stable, as do findings on MRI (Fig. 2C–E).

**Fig. 2.** Three years after presentation to our institution, contrast-enhanced T1 axial (A) and coronal (B) magnetic resonance imaging (MRI) demonstrates bilateral optic nerve sheath meningiomas (ONSMs). Ten years later, the appearance of the ONSMs is unchanged on contrasted fat-suppressed T1 axial MRI (C, D) and on coronal view (E).

**Fig. 3.** Bilateral optic disc pallor is present with an optociliary shunt vessel inferiorly on the left optic disc.
ONSM is a slow-growing benign neoplasm arising from the arachnoid cap cells of the optic nerve sheath. A primary ONSM arises directly from the dural sheath of the optic nerve and is almost always unilateral, whereas secondary ONSM may arise from the dura adjacent to the planum sphenoidal and extend into one or both optic canals (1,2). Cases of bilateral ONSMs typically present initially with monocular vision loss with subsequent extension of the tumor to the contralateral optic canal or optic chiasm. While the typical natural history of ONSM is gradual progressive visual loss over many years, some patients however exhibit rapid progression (3). To date, there is no means of predicting the clinical course in patients with ONSM.

In a review of 477 cases, Dutton (4) found approximately 5% of ONSMs are bilateral. It is unclear whether our case represents bilateral ONSMs with spread across the planum sphenoidal, or a secondary ONSM that originated from the dura on the planum and spread anteriorly to involve both nerves (5). In reviewing the literature, these types of ONSM usually are aggressive, leading to poor visual acuity and posterior extension, and should be treated (1–3,6,7). Management options include conventional or fractional stereotactic radiotherapy and surgical excision (4). Because of the aggressive appearance of our patient’s tumor on neuroimaging, and loss of vision in her left eye, she was offered radiation treatment, but elected observation.

There are limited data on the natural history of ONSM, particularly the bilateral form, and treatment remains controversial. Time intervals of up to 25 years from the onset of visual symptoms in one eye to involvement of the fellow eye have been reported in patients with bilateral ONSM (8–10). Our patient has been followed for more than 27 years, with stable clinical and neuroimaging findings.

Our case highlights the variable natural history of ONSM. While a single case cannot be generalized, observation of patients whose visual function and neuroimaging studies are stable may spare the patient from sight-threatening complications of treatments such as surgery or radiation therapy.

REFERENCES