



Nina Mikhailenko. *Last Tram*. Oil on canvas, 16" × 20".

The prognosis and treatment of primary orbital optic nerve sheath meningioma are reviewed.

Diagnosis and Treatment of Orbital Optic Nerve Sheath Meningioma

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Background: Primary and secondary optic nerve sheath meningiomas (ONSMs) are neoplasms that account for a large proportion of optic nerve and orbital tumors. The diagnosis is not always straightforward and is based on the appropriate clinical findings and neuroimaging. Biopsy or surgical intervention may occasionally be necessary but is associated with significant morbidity.

Methods: Issues related to clinical signs and symptoms, diagnosis, natural history, and treatment strategies are reviewed based on a review of published literature.

Results: Diagnosis is usually based on radiographic and clinical findings. Biopsies are not obtained in most cases, thus adding further to the bias of possible misdiagnosis in all reported case series that do not have the benefit of histopathologic confirmation. Natural history typically shows inexorable progression in most cases, although long periods of stability are occasionally reported. Treatment options include observation, radiation alone, surgery alone, and combined radiation and surgery. The optimum timing of interventional therapy and radiation are evolving.

Conclusions: After serial examination documents new decline in acuity and/or visual field, fractionated radiotherapy appears most likely to preserve visual function and is a valid treatment approach for primary orbital ONSM. Tumor enlargement, as determined by serial imaging, may also provide an indication to begin radiotherapy.

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Abbreviations used in this paper: ONSM = optic nerve sheath meningioma, CT = computed tomography, MRI = magnetic resonance imaging.

Epidemiology

Primary and Secondary Nerve Sheath Meningioma

Optic nerve sheath meningioma (ONSM) is a term applied to primary and secondary meningiomas of the optic nerve. According to a recent meta-analysis and subsequently reported large bi-institutional series, primary ONSMs accounts for approximately one third of primary optic nerve tumors and 5% to 10% of orbital tumors.¹⁻³ Since many reports and series fail to distinguish between primary and secondary tumors, determining the incidence

of either with accuracy is difficult. The vast majority (90%) of optic nerve sheath tumors are secondary.⁴

Primary ONSM represents a neoplasia of meningeothelial cap cells of arachnoid villi and can develop anywhere along the course of the optic nerve, from globe to prechiasmal intracisternal optic nerve.⁵ Lesions may be unilateral, bilateral, or multifocal, with the latter two subgroups occurring most commonly in patients with type 2 neurofibromatosis. A meta-analysis of published cases indicated that tumors confined to the optic canal are more frequently (38%) bilateral compared with those in other locations (5%). Although rare, 65% of reported cases of bilateral ONSM are intracanalicular.¹ It is difficult to determine if these cases are truly multifocal in origin or if they represent midline or unilateral lesions, with dissemination across the clivus and planum sphenoidale.

Meningiomas extending from other locations and involving the optic nerve are secondary and may arise from the cavernous sinus, falciform ligament, clinoid, sphenoid wing, pituitary fossa, planum sphenoidale, frontal-parietal area, or olfactory groove.

Age and Sex Distribution

It is generally accepted that ONSM occurs more commonly in middle-aged women, but reports of the female:male distribution vary widely in the literature. Ratios as high as a 5:1 female predilection have been cited, but accurate estimates may be closer to a female preponderance of 61%, as determined by Dutton's meta-analysis.¹ The overall mean age at presentation is cited as 40.8 years, varying from 36.1 years in men to 42.5 years in women.¹ ONSM has been diagnosed in a child as young as 2.5 years of age.⁶ Moreover, bilateral cases appear to have an earlier mean age of onset of symptoms at 12.8 years.¹

Clinical Signs and Symptoms

Commonly reported clinical manifestations of ONSM include ipsilateral visual loss, afferent pupillary defect, color vision disturbance, visual field defect, proptosis,



Fig 1. — Tubular growth pattern of optic nerve sheath meningioma. A T1-weighted, contrast-enhanced axial MRI shows diffuse enlargement and enhancement along the length of the left intraorbital (IO) and intracanalicular (IC) optic nerve sheath.

optic disc edema, motility disturbance, pain, and lower eyelid edema.⁷ Patients have symptoms of gradual or rapid visual loss, diplopia, or gaze-evoked visual obscurations.^{1,8,9} Ophthalmoscopic examination may reveal optic nerve head swelling, contiguous macular edema, nerve pallor, or choroidal folds. Optic disc swelling, accompanied by optic nerve pallor, also may be observed. As visual function declines, the optic disc edema may resolve, leaving a pale nerve head. Optociliary shunt vessels occasionally develop that represent the secondary dilation of preexisting retinal-to-choroidal shunting veins. Chronic optic nerve compression, caused by a variety of sources, prevents venous return of blood through the central retinal veins. Blood travels via these anastomotic vestigial circuits into the choroidal circulation and leaves the globe via vortex veins rather than the central retinal vein. The triad of visual loss, optic atrophy, and optociliary shunt veins was thought to be most commonly caused by ONSM.¹⁰ The optic disc may be pale, with no prior swelling observed, when the meningioma is at the apex or within the optic canal.¹¹ However, posterior lesions may also present with optic disc swelling.¹²

Radiologic Findings

Growth Patterns

The diagnosis of ONSM relies heavily on imaging findings. A recent article describing a retrospective review of 88 patients with ONSM at two institutions illustrated several radiographic growth patterns: tubular (diffuse, apical expansion, anterior expansion), globular, fusiform, and focal.² The tubular patterns, marked by widening along the length of the nerve sheath (Fig 1), were subdivided into diffuse expansion, apical expansion towards the orbital apex, or anterior nerve expansion towards the globe. Globular growth patterns were caused by exophytic expansion outside of the nerve sheath. Fusiform patterns were spindle shaped with tapers at the proximal and distal ends. This type was most likely to be confused with optic nerve glioma.² Focal exophytic lesions occasionally have been successfully resected.^{2,13}

Plain Film and Ultrasound Diagnosis

Prior to the advent of computed tomography (CT) and magnetic resonance imaging (MRI), clinicians depended on hyperostosis or optic canal enlargement on plain radiographic film studies. A- and B-modes of orbital ultrasonography were also useful. While orbital ultrasonography currently maintains a less definitive role than in the past, it remains widely available and in use in ophthalmic practices. With ultrasonography, the tumor typically is revealed as an enlargement of the nerve sheath signal, with medium to high internal reflectivity. In addition, the nerve sheath diameter remains constant when measured from the perpendicular and 30° angle to the visual axis. In

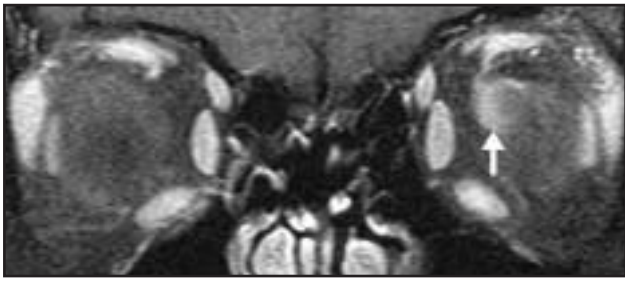


Fig 2. — Intradural optic nerve sheath meningioma. A T1-weighted, contrast-enhanced coronal MRI shows growth of the meningioma in a subdural pattern within the left nerve sheath partially surrounding the optic nerve within (arrow).

contrast, a difference in the nerve sheath diameter, or “positive 30° test,” is detected with an increased cerebrospinal fluid space, typically accompanied by increased intracranial pressure.¹⁴

Computed Tomography

Thin CT scans (1.5-mm sections) may reveal regular or irregular thickening of the nerve sheath meninges. Tumors typically create a well-defined moderate to marked homogenous enhancement after intravenous contrast infusion. On thin section axial CT images, hyperdense enhancement of the meninges surrounding a hypodense nerve is suggestive in appearance of a “tram track.” Similarly, on noncontrast CT images, linear calcification of the nerve is also suggestive of a tram track. The optic nerve itself appears normal in size or of a smaller diameter within an area of thickened meninges, compared to the contralateral nerve at the same level. The smaller nerve size is the result of circumferential compression or atrophy and is a useful differential point. An intrinsically expanded nerve is more commonly seen in optic nerve glioma or other inflammatory lesions. Calcification may (1) surround the nerve and cause a tram-track appearance on axial and coronal CT scan, (2) maintain a punctate diffuse location, or (3) cause an en plaque signal along the optic canal, which may be difficult to distinguish from normal bone. Calcification may be masked by contrast agents and should be sought on precontrast soft tissue and bone-windowed images.

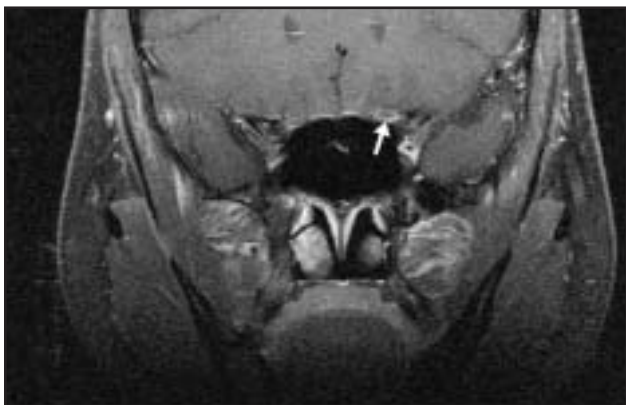


Fig 3. — Left intracanalicular optic nerve sheath meningioma. A T1-weighted, contrast-enhanced coronal MRI shows growth of the meningioma within the left optic canal (arrow).

Magnetic Resonance Imaging

Although MRI is less sensitive than CT in the recognition of calcification, it currently remains the procedure of choice for diagnosis of ONSM. High (≥ 1.5 tesla) field strength T1-weighted images of the brain and orbit, with fat suppression and gadolinium contrast, are used. ONSMs are typically isointense or slightly hypointense to brain and optic nerve tissue on T1-weighted images. ONSMs are typically hyperintense on T2-weighted images but may also be hypointense.

Radiographic Differential Diagnosis

It is not always possible to differentiate ONSM from other lesions involving the optic nerve and meninges. These other lesions include sarcoidosis, demyelinating optic neuritis or perineuritis, orbital inflammatory disease of the optic nerve, orbital schwannoma, lymphoma, hemangiopericytoma, and optic nerve metastasis. Accurate diagnosis depends on the clinical course and occasionally the biopsy.¹⁵

Histopathologic Analysis

Growth Pattern: Primary and Secondary ONSM

Meningiomas typically grow along paths of least resistance, remaining confined to the subarachnoid or intradural space of the intraorbital optic nerve (Fig 2). The tumor, however, may invade dura, adjacent orbital tissue, muscle, bone, and globe. The neoplastic growth typically intermingles with the optic nerve blood supply from the pial vasculature. Orbital tumors may traverse the optic canal and affect the intracranial segment of the optic nerve or adjacent structures. Similarly, principally intracanalicular lesions (Fig 3) may expand proximally to the intracranial nerve or distally to the orbital segment. Theoretically, lesions may affect adjacent intracranial tissues and spread



Fig 4. — Secondary optic nerve sheath meningioma. A T1-weighted, contrast-enhanced coronal MRI illustrates enhancing mass affecting the planum sphenoidale (ps), left inferior orbital fissures (iof), and left optic nerve (on) secondarily.

to the contralateral optic nerve.¹⁶ Such an unusual clinical scenario has not been described in any recently published long-term case series of primary ONSM.^{2,17,18}

Additional patterns of growth more commonly associated with intracranial meningioma have been described in the orbit associated with secondary orbital spread from intracranial lesions (Fig 4). Large intracranial tumors with secondary extension from adjacent structures (cavernous sinus, falciform ligament, clinoid, sphenoid wing, pituitary fossa, planum sphenoidale, frontal-parietal area, or olfactory groove) more commonly affect both anterior visual pathways.

Histology

Orbital meningiomas have histologic features similar to those of intracranial meningioma. The majority of primary ONSMs are of the transitional type: concentric whorl formations of spindle or ovoid cells, meningothelial (syncytium) with sheets of polygonal cells separated by vascular trabeculae, or a mixture. Psammoma bodies are more common in the transitional pattern. Mitoses, architectural disruption, calcification, and MIB-1 staining are also described.^{1,2} Primary orbital meningioma remote from the optic nerve is rare, and some authors maintain that there are diverse sites of origin (eg, ectopic orbital arachnoid, other intraorbital nerve sheaths, orbital mesenchymal cells, intracranial spread through orbital fissures, misdiagnosed lesions).^{5,19-22}

Prognosis

Clinical Course

Primary ONSMs are traditionally described as slow growing. Natural history has been extracted from various retrospective series and from published meta-analysis.^{1,2,17,18} Data are necessarily plagued by biases of reporting, data

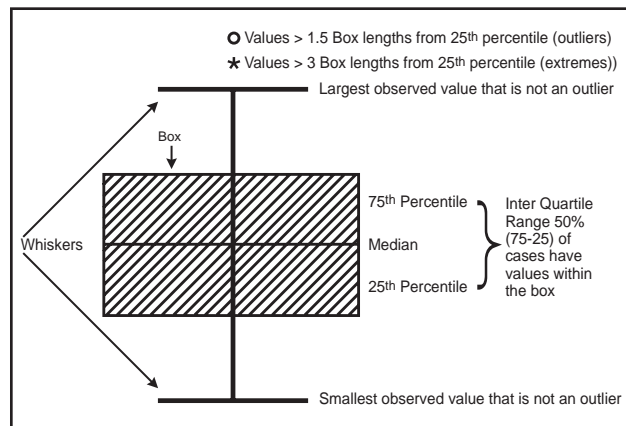


Fig 5. — Bar and whisker plots for nonparametric data. From Turbin RE, Thompson DR, Kennerdell JS, et al. A long-term visual outcome comparison in patients with optic nerve sheath meningioma managed with observation, surgery, radiotherapy, or surgery and radiotherapy. *Ophthalmology*. 2002; 109:890-899. Reprinted with permission from American Academy of Ophthalmology.

acquisition, and dissimilar evaluators, all of which are common to retrospective multicenter studies. In addition, meta-analyses from multiple centers may be based at least partially on the same patient population, and study results should be carefully considered concerning this factor.^{1,6,12,13,18,23,24} The location of ONSMs and the intricate relationship to the pial vascular supply have unavoidably contributed to the difficulty in histopathologic confirmation of ONSM. Since most lesions do not undergo biopsy, diagnosis is based solely on clinical findings. This may add further to the misdiagnosis bias in reported case series.

The worst clinical outcome with primary ONSM is ipsilateral blindness, severe proptosis necessitating enucleation or exenteration or, in rare cases, spread to the contralateral optic nerve or contiguous intracranial structures. Secondary ONSMs are more commonly associated with appreciable morbidity or mortality as correlated with the lesion and extent of the primary tumor. The morbidity and mortality related to treatment options must, of necessity, be weighed against these facts, especially in cases of primary ONSM.

Visual Outcome in Untreated and Treated Cases

The natural history of visual function in ONSM has only recently been elucidated. Most studies have documented slow, progressive visual loss and tumor growth over years in the affected eye. However, some patients remain stable for many years, while others develop

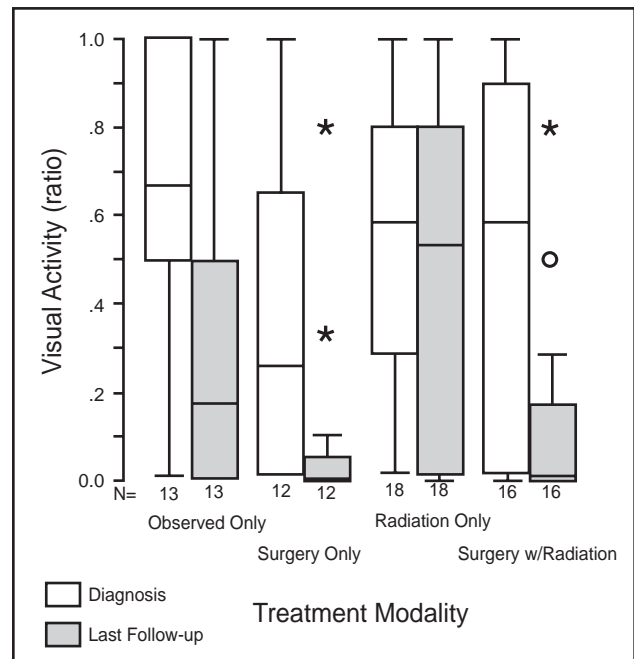


Fig 6. — Plot of visual acuity expressed as a decimal ratio according to treatment method at initial and final examination (20/20 = 1.0, 20/40 = 0.5, no light perception = 0). Bar and whisker plots for nonparametric data. From Turbin RE, Thompson DR, Kennerdell JS, et al. A long-term visual outcome comparison in patients with optic nerve sheath meningioma managed with observation, surgery, radiotherapy, or surgery and radiotherapy. *Ophthalmology*. 2002;109:890-899. Reprinted with permission from American Academy of Ophthalmology.

“aggressive” periods with rapid visual loss with or without tumor enlargement. Occasionally, spontaneous improvement in visual function may occur.¹⁷

Turbin et al¹⁸ studied 59 patients with better than no light perception at presentation. The patients were divided into 4 groups: 13 patients (group 1) were observed only, 12 patients (group 2) had surgery only (4 biopsy or partial resection, 8 total resection), 18 patients (group 3) received radiation alone, and 16 (group 4) had surgery and radiation (14 patients in this group had biopsy or partial resection and radiation, and 2 had total resection and radiation). Visual acuities at diagnosis were statistically similar across the 4 groups ($P=.186$), with overall distribution of visual acuities at diagnosis $\geq 20/40$ in 56.3%, 20/50 to 20/400 in 12.5%, and $<20/400$ in 31.3%. At last follow-up, acuities were $\geq 20/40$ in 28.1%, 20/50 to 20/400 in 15.6%, and $<20/400$ in 56.3% (mean of 150.2 months; range of 51 to 516 months), which worsened as a whole among the 4 groups ($P=.004$). Utilizing nonparametric analysis, all of the treatment groups showed statistically significant visual loss except the radiation-only group, which showed a trend for loss that was not statistically significant. Graphic representations of the results are presented in Figs 5 and 6.

Egan and Lessell¹⁷ reported the natural history of 16 untreated patients. At diagnosis, 12 (75%) of the 16 patients had a visual acuity of $\geq 20/100$. At last evaluation, 8 (50%) of the 16 patients had a visual acuity of $\geq 20/100$ in the affected eye. Two patients maintained “stable” visual acuity but developed new visual field defects. Three patients had spontaneous mild improvement of visual acuity of 3 or fewer Snellen lines. In this study, 11 patients (69%) developed visual loss over a mean of 10.2 years. In the meta-analysis of cases prior to 1992, Dutton¹ reported 14% of observed patients maintained “stable vision,” although further details were lacking. Saeed and colleagues² found that 71% of 92 untreated and treated eyes maintained a visual acuity of 20/50 or better at a mean follow-up of 5.2 years. However, 27% of the eyes worsened to no light perception prior to intervention.

Although visual loss will generally occur in untreated eyes with ONSM, it typically occurs slowly, is limited to the affected eye, and is not associated with mortality. Therefore, treatments must take these natural history data and side effects into account when visual preservation is the primary goal.

Growth

In the series by Turbin et al,¹⁸ radiographic progression was observed in 4 patients who were observed only, in 7 patients who had surgery alone, and in 8 patients who had surgery plus radiation. Two patients who had radiation alone demonstrated radiographic progression prior to treatment. Only 2 patients treated with radiation showed radiographic progression after radiotherapy, and both had had at least one surgical procedure prior to the radiotherapy.

Saeed et al² estimated annual growth in volume and length to be 3.38 mm³ and 0.12 mm, respectively, in calcified lesions and 23.45 mm³ and 0.6 mm in noncalcified lesions, respectively. However, other reported series have not confirmed risk factors for clinical progression.

Andrews et al²⁵ described a case series in which ¹¹¹Inoctreotide scintigraphy was used to track clinical activity following radiotherapy. These authors believed that octreotide single photon emission-computed tomography (SPECT) brain study provides a sensitive and quantitative assessment of tumor response after radiotherapy. They postulated that ¹¹¹In-octreotide scintigraphy “may also provide unambiguous data that allow investigators to differentiate treatment failure (eg, persistent or increasing tracer uptake with visual loss) from treatment-related morbidity (eg, decreasing tracer uptake with visual loss).”

Special Case Consideration

Although most meningiomas are histologically benign and slow-growing, aggressive behavior is seen occasionally, particularly in individuals 20 year of age or younger.²⁶ In some cases, a rapid growth phase is observed during pregnancy,^{7,18,27} probably mediated by estrogen, progesterone, or androgen receptors.^{28,29} As in other forms of meningioma, stable, known, or occult ONSMs may exhibit accelerated growth and cause visual decline during pregnancy. Finally, tumors documented by long-term clinical follow-up to be stable may occasionally cause rapid visual loss, even in the absence of radiographic enlargement.

Treatment

For most patients with primary ONSM, fractionated radiation therapy is currently the technique most likely to achieve long-term preservation of visual function.¹⁸ Although entrance criteria (minimum 50-month follow-up) selected for early strategies of fractionated radiotherapy, the study by Turbin and colleagues¹⁸ provides the most comprehensive and durable evidence reported to date detailing the long-term efficacy of radiation therapy compared to other treatment regimens.^{30,31} However, other extensive, modern case series provide strong evidence concerning the efficacy of more modern radiotherapy delivery techniques, albeit with shorter duration of follow-up (Table).^{24,30,31,33}

Radiation Timing and Modality

Radiation therapy is now accepted as the appropriate vision-preserving therapy for the management of ONSM in nondiabetic patients with progressive visual loss.^{2,18,23-25,31-39} Although there are no direct data concerning radiation therapy in diabetic patients with ONSM, there are theoretical concerns regarding increased susceptibility of diabetic patients to the vascular complications associated with radiation therapy.¹⁸ The optimum timing of therapy (before,

during, or after visual changes) and the optimum radiation strategy — lateral port external beam, fractionated conformal, fractionated stereotactic, intensity-modulated radiation therapy (IMRT) — remain evolving concepts.^{2,18,23-25,31-39}

Effects of Radiation Treatment

Most radiation side effects are transient and self-limited. They include nausea, vomiting, focal alopecia, swelling, pain, and mild erythematous skin changes. Other side effects that are less frequent and often delayed include visual complications (radiation retinopathy, optic neuropathy, cataract formation, dry eye), cranial nerve dysfunction, pituitary dysfunction, brain necrosis, hearing loss, and new tumor induction (after decades).^{31,40,41} Recent advances in achieving greater precision for delivering higher isodose levels to smaller volumes of tissue (thereby reducing the dosage to uninvolved tissue) should theoretically reduce side effects to surrounding tissues. In fact, in 94 patients presenting with vision better than no light perception treated with modern fractionated highly conformal techniques, few significant side effects have been reported (Table).^{23,25,37-39,42} These data are not strictly comparable to those presented by Turbin et al,¹⁸ which included patients treated with conventional external beam fractionated, conformal fractionated, or combination (fractionated radiotherapy and surgery). In that study, involving 34 patients treated with some form of fractionated radiotherapy with or without surgery, 6 eyes developed radiation retinopathy, 2 eyes developed persistent iritis, 1 patient developed temporal lobe atrophy, and none developed radiation optic neuropathy over a mean of 150 months (range 51 to 516 months). The duration of follow-up in published cases that detail new techniques is more limited, and the future

side-effect incidence rate should be determined with longitudinal follow-up.

Surgical Role

Given its technical difficulties and trend toward postoperative blindness, surgery for ONSM has largely been replaced by radiation therapy. However, surgery retains its role in biopsy of atypical cases as well as extirpation in those cases in which complete tumor removal is necessary and visual preservation is not possible. The latter circumstance occurs more commonly in aggressive tumors in young patients. At present, microsurgical dissection for primary ONSM is not considered feasible due to the involvement of the pial blood supply to the nerve via neoplasm. Even subtotal resections often lead to blindness.¹⁸ Furthermore, tumor may spread into the orbit and adjacent structures from a biopsy site.^{3,18,26} However, authors from two treatment centers have described single cases in which focal tumor was removed, with subsequent improvement in postoperative visual function.^{2,13} Although one tumor recurred after microscopic resection, the visual function remained stable.^{2,18}

Role for Nerve Sheath Surgery

While the majority of patients undergoing surgical manipulation of nerve sheath meningiomas fare poorly, a rare subset of carefully selected patients may benefit from nerve sheath decompressive surgery under special conditions. Saeed et al² reported that of 10 patients who underwent nerve sheath decompression, only 1 sustained improved postoperative vision. However, none of these patients received adjuvant therapy. Wladis and colleagues⁴³ described 2 carefully selected patients, both with unilateral ONSM, who suffered progressive visual loss (20/200 and

Summary of Treatment Statistics of Modern Conformal Fractionated Techniques

Author	Eyes	Period	Maximum Total Dose	Fraction	Mode	Stable	Improved	Worse	Complications
Augspurger ³⁸	14	1994-98	50.4 Gy	1.7-2.0 Gy	IMRT	7	5	2	RTOG grade 2 (2) RTOG grade 3 (1)
Tsao ³⁹	15	1989-97	54.0 Gy	1.8 Gy	3D CRT	10	10	5	Radiation retinopathy (2)
Subramanian ⁴²	1	...	54.0 Gy	1.8 Gy	SRT	0	0	1	Radiation retinopathy (1)
Liu ³²	5	1994-2001	54.0 Gy	1.8 Gy	SRT	1	4	0	None
Narayan ³⁷	14	1986-2001	56.0 Gy	1.8-2.0 Gy	3D CRT	7	5	2	Radiation retinopathy (1) Iritis (2) Dry eye (1)
Andrews ²⁵	24	1996-2001	54.0 Gy	1.8 Gy	Cf-SRT	12	10	2	Optic neuritis (1)
Pitz ²³	15	1989-2000	54.0 Gy	3.6 Gy	Cf-SRT	3	6	6	Pituitary abnormality (2)
Saeed ²	6	1976-1999	45.0 Gy	...	Cf-SRT	0	5	1	Cataract (1)

Eyes: The subset of total eyes studied with better than no light perception at the time of treatment. **Fraction:** The daily fractionated dose received at the isocenter. **Mode:** The delivery strategy (IMRT = intensity-modulated radiotherapy, 3D CRT = 3-dimensional conformal fractionated radiotherapy, SRT = fractionated stereotactic radiotherapy, Cf-SRT = highly conformal stereotactic radiotherapy). **Stable:** No significant worsening of visual parameters assessed by the study and no progression of the size of the tumor. **Improved:** Visual function improved as assessed by the author. **Worse:** Decline in visual function as defined by the author or as shown by tumor growth. (One case called "improved" by the author dropped acuity by 1 line and had radiographic progression despite improved visual fields. This case was included under "worse" for this discussion.) **Complications:** RTOG = scale defined by the Radiation Therapy Oncology Group. Transient complications not listed.

no light perception) and florid disc edema. The first had previously undergone stereotactic fractionated radiation therapy, and the second was subsequently treated with stereotactic fractionated radiotherapy after salvage surgery. After biopsy and fenestration, visual acuities in the first and second patients had improved to 20/25 and 20/200, respectively, coinciding with resolution of disc edema. Visual function for these patients remains stable at 6 and 2 years, respectively.⁴³ In these 2 cases, nerve sheath surgery was undertaken only after other treatment options were exhausted, and both required fractionated radiotherapy before or after surgery. However, this approach could ultimately result in orbital spread of the tumor from the fenestration site.^{3,18,26}

Future Direction

Discussions are limited regarding ONSM and alternative methods of management of progressive visual loss after radiation therapy, and no formal studies have been designed to deal with these sequelae. Radiation dose tolerance curves typically preclude radiation boost therapy to the optic nerves, if already treated with conventional radiotherapy doses. Some authors are proponents of chemotherapy (antimetabolites, receptor antagonists) to treat other forms of unresectable meningioma or to deal with failures of previous therapeutic attempts. Some agents (eg, hydroxyurea, mifepristone, interferon alpha, tamoxifen, cyclophosphamide, doxorubicin, vincristine, ifosfamide/mesna, and dacarbazine) may have a modest effect on other meningioma but have limited or no track record for ONSMs.⁴⁴⁻⁴⁶ Moreover, numerous other agents (eg, STI 571, cilengitide, temozolomide, antineoplaston, SCH 66336, octreotide, erlotinib, gefitinib, imatinib) are in various stages of phase II and III trials involving meningioma with various indications. We have limited experience with these agents and would limit their use to formal rigorous experimental protocol or compassionate therapy until further supportive data are available.

Conclusions

The diagnosis of ONSM is usually presumptive and based on the appropriate clinical picture supported by appropriate neuroimaging. Biopsy is not routinely advocated, as surgical intervention carries significant morbidity and mortality.

Historically, most patients were observed until vision was lost, tumor progression threatened intracranial invasion, involvement of the contralateral optic nerve occurred, or proptosis became unmanageable. At that point, the tumor would be completely excised, almost invariably leaving the patient blind in the affected eye. Our treatment strategy has included reassessment of patients with ONSM on a 3- to 6-month schedule, with serial neuroophthalmologic and visual field examination,

unless progressive symptoms or unusual disease activity indicates sooner and more frequent evaluation. Patients often undergo reimaging at 3 months, and they are followed radiographically at 6- to 12-month intervals after the disease has stabilized.

Although at our institute treatment strategy is individualized, we currently recommend fractionated, highly conformal radiotherapy to patients as soon as serial examination documents a new decline in acuity and/or visual field. Tumor enlargement without loss of visual function, as determined by serial imaging, may also provide an indication for radiotherapy.

The appropriate treatment strategy for radiotherapy is an evolving concept; however, radiotherapy should be delivered via fractionated, 3-dimensional stereotactic techniques that provide the most precise conformal application of the dose to affected tissues. Theoretically, this approach should reduce the risk of side effects to surrounding radiosensitive ocular and neural tissues.

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