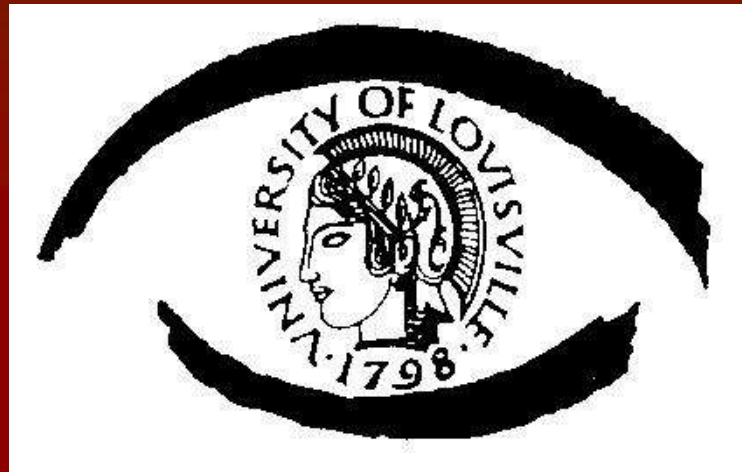


# Grand Round



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**08/19/10**

# Presentation

CC: “bulging right eye since a year”

HPI: 50 yo AAF c/o progressive proptosis of her R eye over 1 year. No other visual complaints.

POH: refractive error

PMH: DM, HTN

Family history: non-contributory

Meds: Lisinopril, Insulin

Allergies: sulfa drugs

ROS: negative

# Physical Exam

VA cc  $\left\{ \begin{array}{l} 20/20^{-1} \\ 20/20 \end{array} \right.$

P  $\left\{ \begin{array}{l} 3 \rightarrow 2 \\ 3 \rightarrow 2 \end{array} \right.$   
+ RAPD OD

T<sub>GAT</sub>  $\left\{ \begin{array}{l} 23 \\ 23 \end{array} \right.$

Light brightness: 50% dimmer in OD relative to OS

CVF: full OU

EOM: full OU. Orthophoric in primary gaze

Ishihara color plates: 9/14 OD, 13/14 OS

Hertel Exophthalmometry: 23 mm OD, 21 mm OS

# SLE

Ant segment: WNL OU

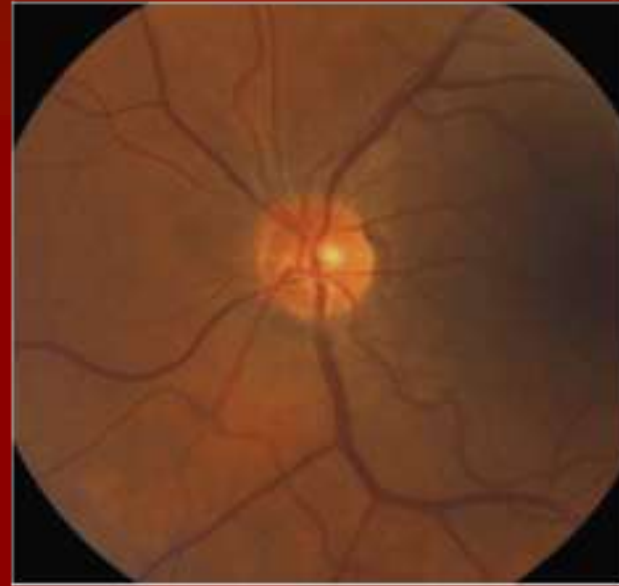
DFE :

- OD: optic nerve edema, otherwise WNL
- OS: WNL

# Fundus Photos

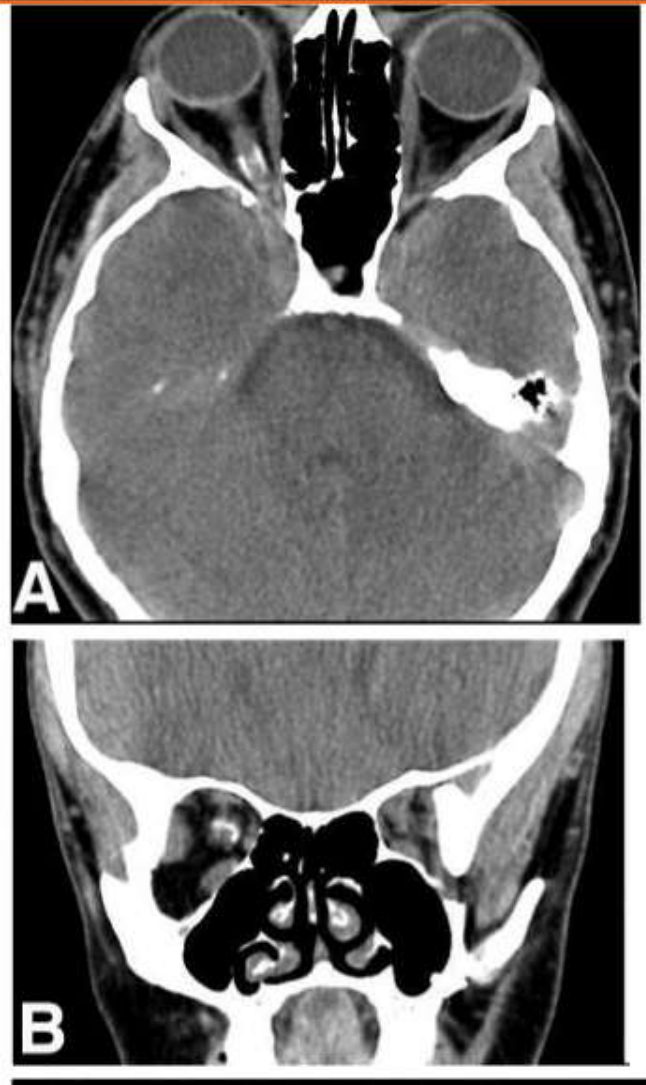


Optic disk swelling OD



WNL OS

# Imaging



Source: Neurosurg Focus © 2007 American Association of Neurological Surgeons

(C) A T1-weighted fat-suppressed Gd-enhanced MR image showing the abnormal enhancement of the optic nerve sheath surrounding nonenhancing optic nerve.

Axial CT (A) and coronal reconstruction (B) demonstrating circumferential calcification around the optic nerve.

“Tram tracking” can be seen on the CT images as well as the MR image.

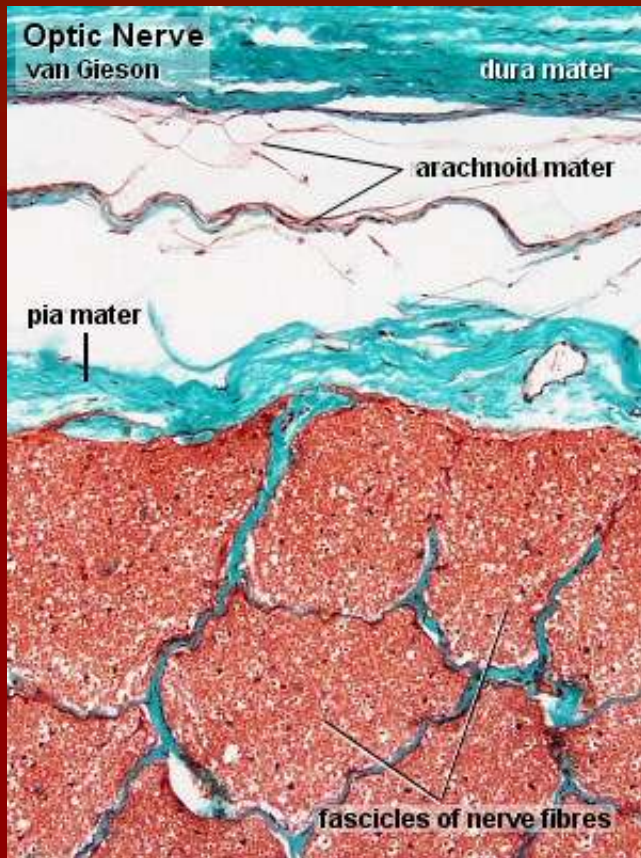
# Assessment

## Optic nerve sheath meningioma

### Differential Diagnosis

- Optic nerve glioma
- Meningocele
- Optic nerve choristoma
- metastasis
- Lymphoma
- Neurofibroma

# Optic Nerve Sheath Meningioma (ONSM)



- Primary ONSM (pONSM): arise within arachnoid of optic nerve
- Secondary ONSM: extend from intracranial primary site
- pONSM:
  - 1/3 of primary optic nerve tumors (2<sup>nd</sup> to glioma in frequency)
  - 5% to 10% of orbital tumors

# Epidemiology

- Female preponderance (3:1)
- Mean age at presentation 40.8 years
- Bilateral cases (5% of cases) have an earlier mean age of onset of symptoms at 12.8 years
- Childhood ONSM are generally associated with NF-2 and have been reported to be more aggressive than those in adults

# Symptoms

- ↓ VA in 96% of patients
- Color vision and contrast sensitivity may be affected early in the clinical course
- Proptosis in 59% to 90% of patients
- Diplopia
- Gaze-evoked visual obscurations
- Scotomas

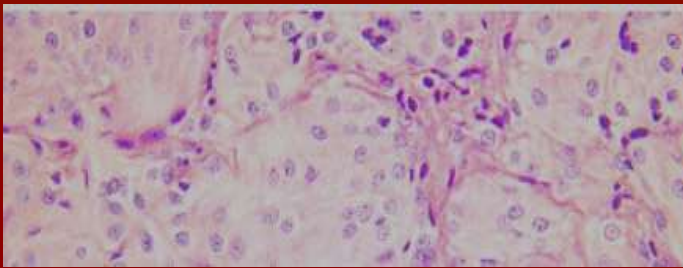
# Signs



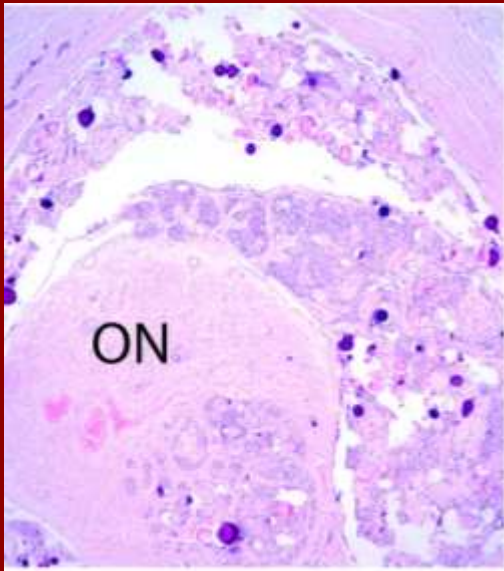
- Optic disk changes, either swelling or atrophy, ( 98% of patients). Optociliary shunt vessels may be seen on the disk surface  
(classic triad= progressive visual loss + optic disk atrophy + optociliary shunt vessels)
- Visual field defects, peripheral or central (83% of patients)
- Ocular motility limitation
- RAPD

# Pathology

Generally benign, low grade tumors, associated with prolonged, recurrence-free survival, but a subset are clinically more aggressive.



Typical meningothelial meningioma composed of relatively small, polygonal neoplastic cells arranged in small nests and indistinct whorls (H&E).



As ONSMs progress, it is thought that they compromise optic nerve function mainly by mass effect on the pial vascular supply, which induces ischemic changes as well as interfering with axonal transport

Intraorbital meningiomas can also invade the optic nerve but it is not clear whether that is equivalent to intracranial brain invasion in prognostic effect. <sup>1</sup>

# When to treat???

- Rarely leads to patient death, but usually leads to marked visual loss without intervention and can extend intracranially (15 to 23%)<sup>2</sup>
- Rate and course of progressive decline in vision are highly variable. Studies have shown a majority of patients still retain their baseline vision more than 5 years after diagnosis with observation alone (71% of eyes in their study that had an initial visual acuity of 20/50 or better retained this level of vision over 5.2 years)<sup>3</sup>
- Optimal timing of intervention for ONSM remains unclear. It has been suggested that treatment be reserved until visual acuity falls progressively below 20/40 or visual field is constricting <sup>4</sup>
- For VA of 20/40 or better, close follow-up, with frequent clinical examinations, including assessment of visual acuity, color vision, and visual fields, should be performed biannually for at least 2–3 years, after which they can be performed annually if visual function has stabilized

# How to treat ???

- The major goals of therapeutic intervention are:
  - to restore or preserve vision
  - to prevent intracranial spread and involvement of the opposite eye.
  
- Treatment modalities:
  - Surgery
  - Radiotherapy/ conformal fractionated radiotherapy
  - Combination

# A long-term visual outcome comparison in patients with optic nerve sheath meningioma managed with observation, surgery, radiotherapy, or surgery and radiotherapy<sup>5</sup>

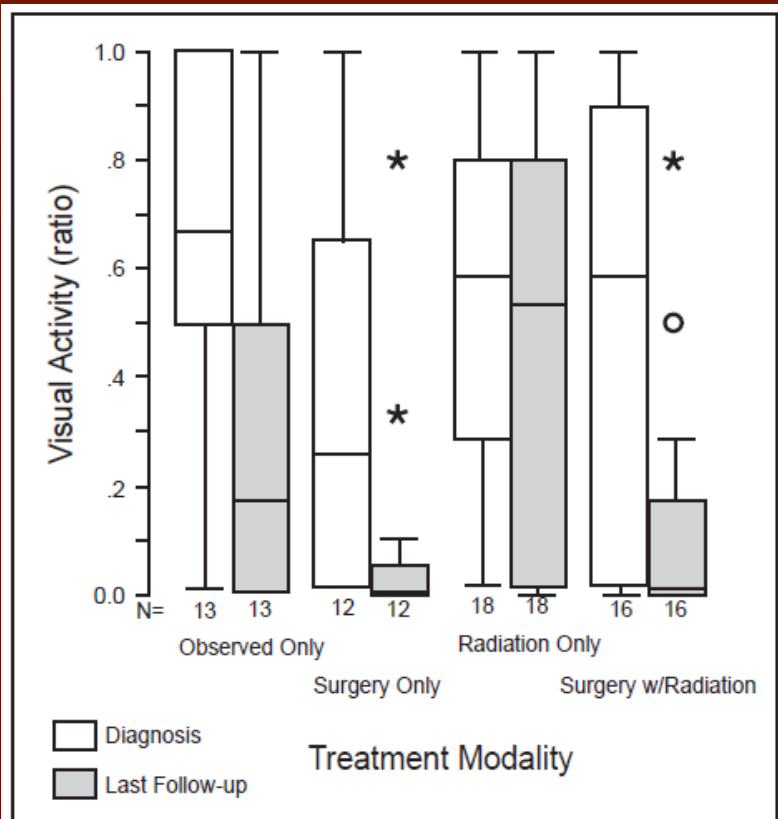


Fig 6. — Plot of visual acuity expressed as a decimal ratio according to treatment method at initial and final examination (20/20 = 1.0, 20/40 = 0.5, no light perception = 0). Bar and whisker plots for nonparametric data. From Turbin RE, Thompson DR, Kennerdell JS, et al. A long-term visual outcome comparison 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.

Design: retrospective comparative study

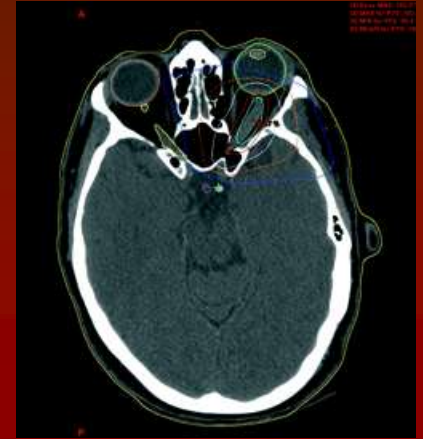
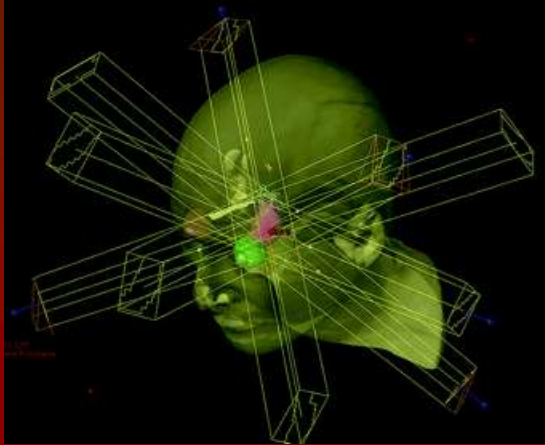
Participants: 59 patients with better than no light perception at presentation, at least 50 months of f/u

Surgery included biopsy or partial resection or total resection

Visual acuities at diagnosis were statistically similar across the 4 groups

Results: 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.

# Conformal fractionated radiotherapy



- Treatment of choice
- Most of the studies were limited by size or short duration of follow-up but they showed stable or improved vision in the majority of patients, often 90% of patients or more with progressive visual loss
- Acute toxicity was mild in these studies, and late toxicity ranged from 0% to 35% and included radiation retinopathy, iritis, dry eye, and orbital pain. <sup>5,6</sup>

# Surgery

- Pial blood vessels of the intraorbital and intracanalicular segments of the optic nerve are incorporated into the growth pattern of ONSM
- High risk of infarction of the nerve and visual loss<sup>2,5</sup>
- Surgery is indicated for large extensive neoplasms, for cosmetically disfiguring proptosis, and for the prevention of intracranial or transchiasmal spread

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4. J.W. Kim, J.F. Rizzo and S. Lessell, Controversies in the management of optic nerve sheath meningiomas, *Int Ophthalmol Clin* **45** (2005), pp. 15–23.
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6. D.W. Andrews, R. Faroozan and B.P. Yang *et al.*, Fractionated stereotactic radiotherapy for the treatment of optic nerve sheath meningiomas: Preliminary observations of 33 optic nerves in 30 patients with historical comparison to observation with or without prior surgery, *Neurosurgery* **51** (2002), pp. 890–902

**Thank You !**