A CASE OF BILATERAL RETINOBLASTOMA

Pathologic Findings

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Clinical Presentation

- 4 year old girl arrived to our service for inclusion in phase 1 clinical trial of gene mediated therapy of vitreous seeds (May 2002)
- Referral diagnosis of bilateral retinoblastoma treated elsewhere with:
  - external beam radiation OU
  - 6-9 courses of chemotherapy
  - cryotherapy in the right eye
  - multiple laser treatments, OU
Clinical Presentation

- Right Eye at time of presentation to us
- Very good visual acuity
Vitreous seeds

Retinal lesions

Vitreous seeds

OD 5/15/02
Retinal lesions  inf.
nasal
Pre-treatment. OD 5/15/02.
Sup nasal tumors with overlying vitreous seeds
Clinical Presentation

- Left Eye at time of presentation to us
- Poor visual acuity of probable hand motion
- Areas of posterior synechia of the iris
- Incipient posterior subcapsular cataract
Pre-treatment: OS 5/15/02
Superior temporal (1-3:00) lesion with vitreous bands and diffuse "seeds" over mass
Suicide Gene Therapy

- Transfer of HSV-TK gene into tumor cells
- Phosphorylation of antiviral prodrug ganciclovir (GCV)
- Inhibition of DNA replication
Eligibility Criteria

- All patients had bilateral Rb with only one functional eye containing measurable Rb that was refractory to standard therapies; enucleation was the imminent treatment.

- The patient must have had no evidence of metastases (no invasion of the ocular coats or the optic nerve)
Injection of vector

Cryotherapy at site of injection

Copious irrigation

Systemic I.V. Gancyclovir
q 12 hrs X 7 days
Post- gene tx and Rx. plaque

O.D.

Tumor and seeds regressed
Summary of Treatments

OD
- Gene tx x 2
- Rx. plaque
- C.R. (Sept 2002)
- Pt returned to country of origin
- Retina tumor recurred (12 months after)
- Neovascular glaucoma and hemorrhage
- Enucleation 2003 (Dec)

OS
- Cryo tx only
- MRI showed thickening of optic nerve suspicious for tumor involvement
- Partial RD
- Visual acuity very poor
- Enucleation 2002 (July)
• Changes consistent with parasitic endophthalmitis, most likely Toxocariasis
• Associated changes in retina, vitreous and optic nerve (eosinophilic infiltrate, gliovascular membranes, exudates)
• Changes associated with previous external beam radiation (cataract and neovascularization)
• No tumor found
More history...

- an episode of low fever
- 20% peripheral eosinophilia associated to moderately elevated liver enzymes when she first arrived to Houston
- Cleared in a month.
- No pictures of OS before arriving to us
- Retrospectively the serum available showed elevated Toxocara IgG levels when she arrived that decreased in ~ 4 months
Histopathologic Findings

OD
• Retinoblastoma, recurrent intraretinal
• Changes associated with previous treatment effect (neovascularization, hemorrhage, gliosis, inflammation in retinal vessels and atrophy)
• Bilateral retinal and vitreous lesions in a patient with retinoblastoma may have different etiologies
• Inflammatory signs and systemic manifestations of infection may be present in cases of infectious etiology
• Extensive treatment effect is seen in eyes that have received multimodality therapy
• Toxocariasis should be included in differential diagnosis of bilateral retinoblastoma with atypical presentation
Thank you!