



Orbit

The International Journal on Orbital Disorders, Oculoplastic and
Lacrimal Surgery



ISSN: 0167-6830 (Print) 1744-5108 (Online) Journal homepage: <http://www.tandfonline.com/loi/iorb20>

Protocol-based management in orbital retinoblastoma

Akshay G. Nair, Swathi Kaliki & Saurabh Kamal

To cite this article: Akshay G. Nair, Swathi Kaliki & Saurabh Kamal (2017): Protocol-based management in orbital retinoblastoma, Orbit, DOI: [10.1080/01676830.2017.1383449](https://doi.org/10.1080/01676830.2017.1383449)

To link to this article: <http://dx.doi.org/10.1080/01676830.2017.1383449>



Published online: 20 Oct 2017.



Submit your article to this journal [↗](#)



View related articles [↗](#)



View Crossmark data [↗](#)

Full Terms & Conditions of access and use can be found at
<http://www.tandfonline.com/action/journalInformation?journalCode=iorb20>



LETTER TO THE EDITOR

Protocol-based management in orbital retinoblastoma

Akshay G. Nair^a, Swathi Kaliki^b, and Saurabh Kamal^c

^aOphthalmic Plastic Surgery and Ocular Oncology Services, Advanced Eye Hospital & Institute, Navi Mumbai, Maharashtra, India; ^bThe Operation Eyesight Universal Institute for Eye Cancer, L V Prasad Eye Institute, Hyderabad, Telangana, India; ^cOculoplasty and Ocular Oncology Services, EyeHub Hospital, Faridabad, Haryana, India

ARTICLE HISTORY Received 1 March 2016; Accepted 19 September 2017

We read with interest the article, “Adult onset retinoblastoma: A diagnostic dilemma” by Raj et al. where they have presented a case of a previously undiagnosed retinoblastoma (RB) which was eviscerated and subsequently treated.¹ Adult-onset RB is indeed rare and is most often misdiagnosed.² We thank the authors for presenting this rare clinical scenario. While most previously reported cases have been about ocular surgery in previously undiagnosed *intraocular* RBs, Raj et al. have reported a case where intraocular surgery was performed in an eye with previously undiagnosed *orbital* RB. As is noticeable from Figure 1 of their report, in addition to the perforated cornea, the conjunctival bulging noted superiorly and superotemporally are suggestive of extraocular extension. However, hindsight is always 20/20, and the authors, upon diagnosing RB, have meticulously conducted a metastatic workup and treated the disease with surgery, chemotherapy, and radiotherapy. In this communication, we wish to put forth three important issues that are relevant to the case in point here:

- (1) Standard protocol for metastatic screening in orbital RB
- (2) High-dose chemotherapy (HDC) vs. standard chemotherapy
- (3) Extended high-dose adjuvant chemotherapy

Standard protocol for metastatic screening in orbital RB

The treating clinician can prognosticate the disease only if the correct stage is known. In RB, the presence of orbital invasion of the intraocular tumor increases the risk of systemic metastasis by 10–27-fold and is

associated with poor prognosis; furthermore, metastatic RB is also associated with poor prognosis.^{3–5} Therefore, in orbital RB, it is important to perform additional tests and procedures to stage the disease correctly.^{6,7} This includes the following:

- A thorough systemic physical evaluation
- Assessment of all regional lymph nodes – if enlarged, fine-needle aspiration cytology (FNAC) of the affected nodes
- Imaging of the orbit and brain (computed tomography/magnetic resonance imaging)
- Chest X-ray
- Ultrasonography of the abdomen
- Bone marrow biopsy
- Cerebrospinal fluid cytology

High-dose chemotherapy vs. standard chemotherapy

The authors have reported that the patient received chemotherapy with vincristine, etoposide and carboplatin and radiotherapy. We believe that this is an opportunity to emphasize on the importance of HDC which is effective in orbital and metastatic RB.⁸ Given the rarity of accidental orbital RB, it is difficult to establish a protocol for its treatment. However, it has been recommended that primary orbital RB should be treated with a high-dose triple drug regimen comprising of the same drugs used by Raj et al.: carboplatin, etoposide, and vincristine.⁷ Honavar and Singh reported the outcomes of 16 cases with unilateral orbital RB without intracranial extension and systemic metastasis. They reported that after following a multimodal treatment protocol for orbital RB that included HDC, 88%

CONTACT Akshay G. Nair ✉ akshaygn@gmail.com 📧 Ophthalmic Plastic Surgery & Ocular Oncology, Advanced Eye Hospital & Institute, 30, The Affaires, Palm Beach Road, Sanpada, Sector 17, Navi Mumbai 400 705, Maharashtra, India.

Dr. Nair bears full responsibility for the manuscript.

© 2017 Taylor & Francis

patients were free of local recurrence or systemic metastasis at a 3-year follow-up.^{8,9}

Extended high-dose adjuvant chemotherapy

As mentioned earlier, multimodal treatment protocol by combination of chemotherapy, radiotherapy, and surgery is recommended in orbital RB. Here, intravenous systemic HDC comprising of vincristine, etoposide, and carboplatin for 3–9 cycles facilitates enucleation/extended enucleation/exenteration. Subsequently, after surgery, 40–50 Gy external beam radiotherapy (EBRT) over 20–25 fractions is administered to the pretreatment extent of the disease and this is followed by extended HDC for a total of 12 cycles.⁵ While Honavar and Singh recommend this protocol for primary orbital RB, we believe that this can be extrapolated for cases of accidental orbital RB as well.^{9,10}

Finally, as the authors have emphasized, preoperative imaging of the posterior segment must be done in all cases of painful blind eyes prior to destructive surgery.¹¹ Previously undiagnosed intraocular tumors may masquerade as other clinical entities further underscoring the need for imaging in such cases.¹²

It is also important to highlight the biological differences between adult-onset RBs and pediatric RBs. Adult-onset RB could be due to activation of undiagnosed spontaneously regressed RB or retinoma/retinocytoma or delayed activation of persistent embryonal retinal cell with RB1 mutation. The biologic behavior of tumor in adults versus children may be different and needs further evaluation. In the future, the road ahead may see specific therapies based on these biological differences. With limited evidence of treatment of adult-onset RB in the literature, similar protocols as in children may be used for adult-onset RB. However, the prediction of effectiveness of the currently available treatment in adults may not be made accurately and is only speculative.

We congratulate the authors for having presented a unique case of adult-onset RB. With aggressive treatment, even advanced cases of orbital RBs can be treated and lives saved. However, this requires protocol-based management

and a concerted team-effort from the ophthalmologist, pathologist, and the oncologist.

Disclosure statement

No potential conflict of interest was reported by authors.

References

1. Raj A, Arya SK, Punia RS, Kohli P. Adult onset retinoblastoma: a diagnostic dilemma. *Orbit*. 2016;35(1):51–53.
2. Kaliki S, Shields CL, Gupta A, et al. Newly diagnosed active retinoblastoma in adults. *Retina*. 2015;35(12):2483–2488.
3. Leal-Leal CA, Rivera-Luna R, Flores-Rojo M, Juárez-Echenique JC, Ordaz JC, Amador-Zarco J. Survival in extra-orbital metastatic retinoblastoma: treatment results. *Clin Transl Oncol*. 2006;8:39–44.
4. Singh AD, Shields CL, Shields JA. Prognostic factors in retinoblastoma. *J Pediatr Ophthalmol Strabismus*. 2000;37:134–141.
5. Kaliki S, Shields CL. Retinoblastoma: achieving new standards with methods of chemotherapy. *Indian J Ophthalmol*. 2015;63:103–109.
6. Ali MJ, Honavar SG, Reddy VAP. Orbital retinoblastoma: present status and future challenges – a review. *Saudi J Ophthalmol*. 2011;25(2):159–167.
7. Honavar SG. Orbital retinoblastoma. In: Singh AD, Damato BE, Pe'er J, et al., eds. *Clinical Ophthalmic Oncology*. first ed. Philadelphia: Saunders Elsevier;2007:477–483.
8. Dunkel IJ, Aledo A, Kerman N, et al. Successful treatment of metastatic retinoblastoma. *Cancer*. 2000;89:2117–2121.
9. Honavar SG, Singh AD. Management of advanced retinoblastoma. *Ophthalmol Clin North Am*. 2005;18:65–73.
10. Honavar SG. Orbital retinoblastoma. In: Ramasubramanian A, Shields CL, eds. *Retinoblastoma*. New Delhi: Jaypee Brothers Medical Publishers;2012:167–178.
11. Eagle RC, Jr, Grossniklaus HE, Syed N, Hogan RN, Lloyd WC, 3rd, Folberg R. Inadvertent evisceration of eyes containing uveal melanoma. *Arch Ophthalmol*. 2009;127(2):141–145.
12. Nair AG, Kaliki S, Ali MJ, Naik MN, Vemuganti GK. Intraocular malignant melanoma of the choroid presenting as orbital cellulitis. *Int Ophthalmol*. 2014;34(3):647–650.