Commentary: Current standards in retinoblastoma care

The authors present a review of the current standards in retinoblastoma care.[1] Notably, the goals of retinoblastoma care remain the same—save life, save eye, save sight. Orbital retinoblastoma is still a challenge in several parts of the world and deserves more attention.[2] Management of a pediatric malignancy such as retinoblastoma is complex not only because of its varied presentation but also due to the added emotional and psychological aspects when dealing with the affected family; perceptions being influenced by cultural beliefs. The role and protocol for sibling screening form an integral part of the extended care for the family.[3]

Authors have lucidly illustrated the different treatment modalities and their approach towards the management of retinoblastoma. The treatment is truly challenging and with the introduction of newer and promising techniques such as intra-arterial chemotherapy and intravitreal chemotherapy, the rates of enucleation have decreased and more eyes are being salvaged with better visual outcomes. Employing this approach of directly targeting the tumor has also helped in reducing systemic side effects. However, this enthusiasm is tempered by studies that have documented complications related to these newer treatment modalities.[4,5]

While it is quite endearing to have the latest techniques at a clinician's disposal, it's more often an in-depth understanding of the multi-faceted aspects of disease management that is more enduring a skill to acquire. In other words, retinoblastoma care still has a long learning curve. As mentioned by the authors, a well-elicited history, thorough examination under anesthesia, and evaluation of imaging with ocular ultrasound and magnetic resonance imaging of brain and orbits must be carried out for decision-making on the best management strategy. Ultrasound is a readily available, useful bedside screening tool that helps differentiate simulating conditions like Coats' disease which often exhibits mobile subretinal echoes—a good way of making use of dynamic sonography. It can also help confirm the diagnosis of an unsuspected diffuse infiltrating retinoblastoma, with thickened retinal leaves.

It appears that outcomes with intra-arterial chemotherapy and adjunctive intravitreal chemotherapy have shown an ascending trend and are comparable among centers across geographical locations. However, this could vary from region to region depending on the techniques, skill, and experience with the procedures. Some outstanding challenges in the analysis of outcomes continuing to confront the ocular oncologists are a lack of uniformity in employing classification systems, and the need to expand the one related to vitreous seeds.[6]

On a day-to-day basis, a caregiver also needs to marshal the available resources deftly—thus employing a more practical approach rather than knowledge distilled from the literature search. For example, cryotherapy can be used to treat localized vitreous seeds in close vicinity of tumors with scleral indentation and encompassing them within the ice-ball. Unilateral group B extramacular tumors can also be managed with primary plaque radiation. While following-up patients for possible second primary malignancies is important, the level of evidence for systemic intravenous chemotherapy resulting in the prevention of second primary malignancies is not very strong. Dwelling on weak evidence to formulate strong opinions remains undesirable. Future care might incorporate aqueous humor aspiration for the detection of molecular prognostic markers (viz chromosome 6p gain) for targeted, patient-centered therapies.[7]

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