Review Article :

RETINOBLASTOMA: AWARENESS MORE CHALLENGING THAN TREATMENT

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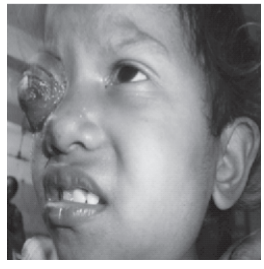
Introduction:

Retinoblastoma is the most common intra-ocular malignancy in children and is associated with high mortality. However, timely diagnosis and proper management has lead to improved survival rates. There has been a dramatic change in the management of retinoibalstoma in the last decade with introduction of chemotherapy and advances in focal therapy. Now there is more focus on improved eye and vision salvage.

However inspite of all these advances as a result of lack of awareness and health care facilities, a large number of children in our country still present with advance stage of retinopblastoma and extraocular spread.

Common Presenting Features Of Retinoblastoma:



1. Leucocoria – 50%
2. Strabismus – 20%
3. Red painful eye – 7%
4. Poor vision – 5%
5. Asymptomatic – 3%
6. Orbital cellulitis – 3%
7. Unilateral mydriasis – 2%
8. Hetrerochromia iridis – 1%
9. Hyphaema – 1%

Differential diagnosis of leucocoria :

- Retinoblastoma
- Persistent fetal vasculature
- Retionopathy of prematurity
- Cataract
- Coloboma of choroid or optic disc
- Toxocariasis
- Coat's disease
- Retinal dysplasia
- Norrie's disease

Basic work up in a child with leucocoria :

1) Detailed history: antenatal history, birth history of prematurity, exposure to oxigen, exposure to animals, positive family history pof retinoblastoma including siblings and parents, any death in the siblings and hostory of enucleation.

2) Clinical evaluation: include (a) recording of visual acuity, (b) examination under anaesthesia for anterior segment details, corneal diameter, intra-ocular pressure, fundus examinatio of both eyes.

3) Investigations : laboratory investigations: ELISA for Toxocara & TORCH infection Genetic study Classification of Retinoblastoma (International Intraocular Classification)

Group – A - Very low risk

Eyes with small intraretinal tumors away from critical structures.

1 - Tumors are 3mm or smaller
2 - Confined to retina
3 - Located 2DD from fovea and 1DD from optic nerve.
4 - No seeding

Group – B – Low risk Eyes with no seeding. Tumor(s) are discrete but any size or location

1 - Any size or location not in Group A
2 - No seeding
3 - Cuff of subretinal fluid not >5mm from tumor allowed

Group- C –Moderate risk

Eyes with focal vitreous and subretinal seeding only. Tumors are discrete but any size or location.

1 - Seeding must be local
2 - Up to one quadrant of retinal detachment allowed Group –D –High risk
Eyes with diffuse vitreous or subretinal seeding and/or massive, non-discrete endo or exophytic disease.
1 - More extensive seeding than Group-C
2 - Massive and/ or diffuse intraocular disease
3 - Include fine or greasy vitrous seeds or avascular masses
4 - Subretinal seeding may be plaque-like
5 - Up to total retinal detachment Group- E- Very high risk Eyes destroyed anatomically or functionally. One or more of the following :
1 - Irrereversible neovascular glaucoma
2 - Massive intraocular haemorrhage
3 - Aseptic orbital cellulitis
4 - Tumor anterior to anterior vitreous face
5 - Tumor in the ant segment
6 - Tumor touching the lens
7 - Diffuse infiltrating retinoblastoma
8 - Phthisis or prephthisis

TREATMENT :

1 - Eyes with Group-A- Treated with laser photocoagulation or cryotherapy
2 - Eyes with Group- B, C, D – Chemoreduction
3 - Eyes with Group-E- Is managed by enucleation or both eyes are equally advanced then chemoreduction and low dose radiotherapy is attempted
4 - Bilateral group C,D and E eyes often require the addition of subconjuctival carbopltin as alocal boost in chemotherapy
Which cases metastatic work up is needed and the investigations to be done

- Cases where optic nerve is not seen on clinical examination
- MRI showing gross optic nerve thickening
- Chilldren with abnormal complete blood count or those whose tumors show massive choroidal involvement and which extend beyond lamina cribrosa on pathological examination of the enucleated specimen
- All cases of orbital retinoblastoma Investigations include
- MRI Scan Head and orbits
- Hematologic evaluation
- Chest X-ray
- Abdominal Ultrasound
- CSF analysis
- Bone marrow biopsy Treatment Protocol in cases with extraocular spread
- 3- 4 Cycles of neoadjuvant chemotherapy (vincristine, carboplatin and etoposide) followed by orbital imaging, repeat systemic staging, investigations. EUA followed by local therapy followed by further cycles (3-4) of chemotherapy and/ or EBRT (36-40Gy).
- In patients with intracranial spread intrathecal methotrexate is added. EBRT is given to the affected part in case of metastasis.

Follow up :

First examination- 3-6 weeks after initial therapy
1-3 months during first year
3-4 months during 2nd year
4-6 months until age 4-6 years
Then annually

Improving awareness regarding retinoblastoma:

Awareness among all sections of popul;ation is absolutely necessary.

1) Regular CMEs involving ophthalmologists, pathologists, oncologists.
2) Training protocols for health workers of state level health centers, hospitals, medical colleges.
3) Puppet show in the villages regarding main signs and symptoms of retinoblastoma.
4) Educating parents and family members to detect leucocoria.

5) Screening prgrams for children with a positive family history.

6) Speading awareness among general ophthalmologists regarding importance of prompt referral and atypical presentation of retinoblastoma so that they are not mismanaged or delayed prior to referral.

CONCLUSION :

Measures should be taken to improve awareness about the most common presenting sign of retinoblastoma is leucocoria. The urgency to get an ophthalmologist's opinion in such cases needs to be emphasized through mass media.

References :

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