Unilateral D Retinoblastoma

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Introduction

Retinoblastoma is the most common primary, malignant intraocular tumor of childhood

Its either unilateral or bilateral disease

Some unilaterally diagnosed cases go on to develop tumors in the fellow eye and become bilateral.

Children with unilateral retinoblastoma are usually diagnosed at a later age than bilateral retinoblastoma and most commonly presents with advanced intraocular disease

International Classification

Group A:

- Small tumor(s) located only in the retina
- No tumor is larger than 3 millimeters (mm)
- No tumor is closer than 2 disc diameters (DD) from the fovea (the central "pit" of the retina) or 1 DD from the optic nerve
- No vitreous seeding, subretinal seeding or retinal detachment

Group B:

- Tumor(s) located in any location in the retina
- No vitreous seeding
- No retinal detachment more than 5 mm from tumor base

International Classification

Group C:

- Fine localized (located in 1 spot) vitreous seeding
- Subretinal fluid without seeding involving up to one quarter of the retina
- Localized subretinal seeding less than 2 disc diameter from the tumor

Group D:

- Massive vitreous/sub-retinal seeding
- Retinal detachment: more than Group C and up to total retinal detachment

International Classification

Group E:

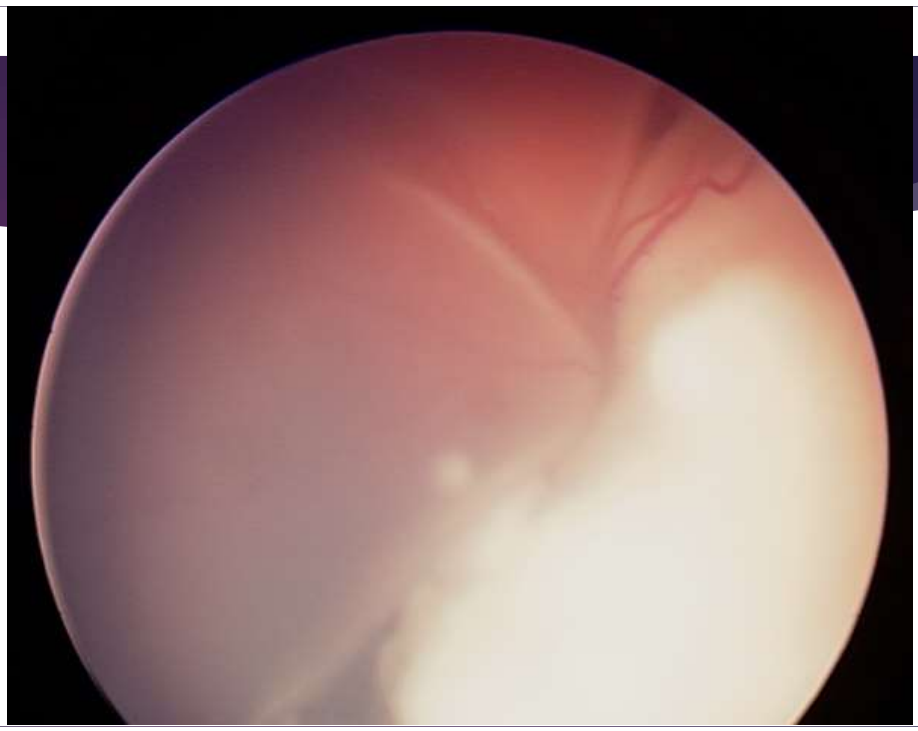
- **Presence of 1 or more:**
  - Tumor in CB/anterior segment
  - Neovascular glaucoma.
  - Vitreous hemorrhage.
  - Hyphema

Management

- **Group A:** Focal treatment, Laser or Cryotherapy
- **Group B & C:** Chemotherapy with focal treatment
- **Group D:** If the patient has good visual potential (according to macular affection) or patient age is less than 2 years → Chemotherapy with focal treatment
  - if the patient has poor visual potentials→ Enucleation
- **Group E** → Enucleation.

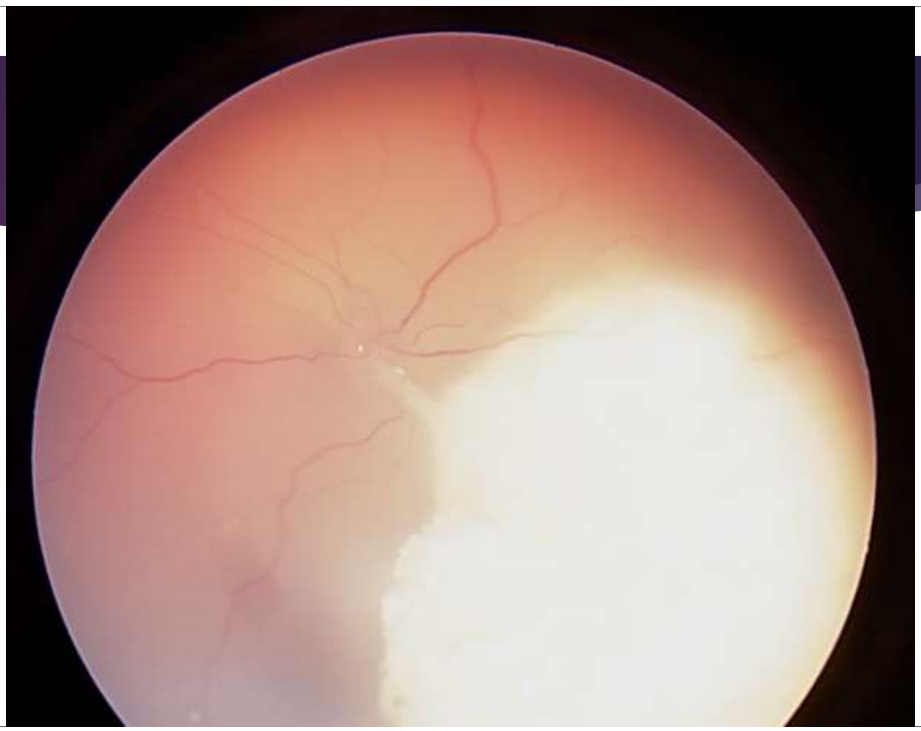
First case

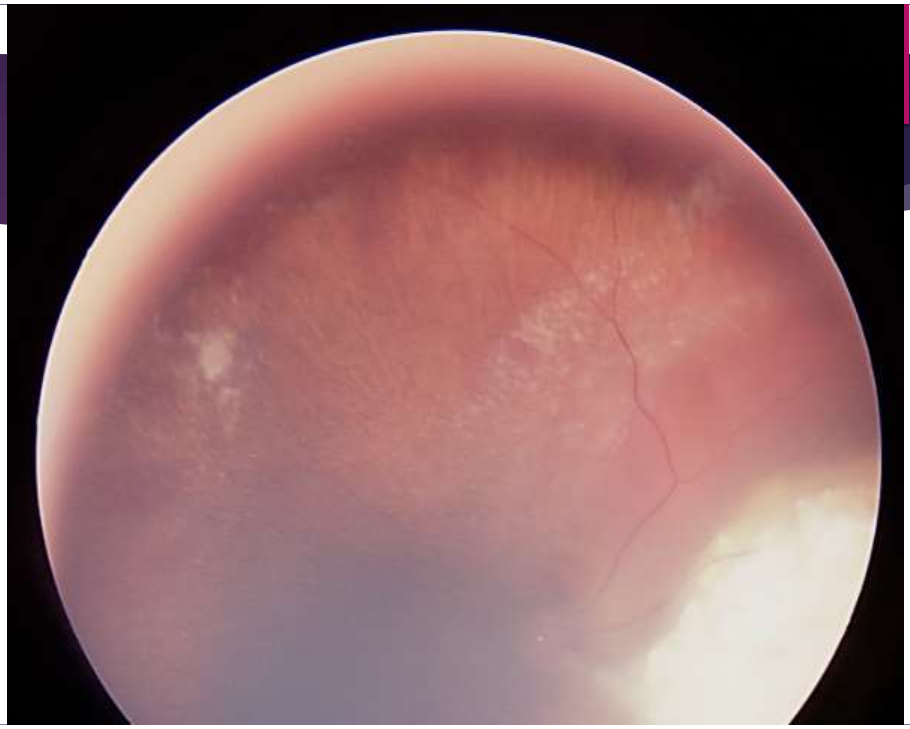
- Female child , 20 months
- presented with leukocoria in the left eye dated 1 month ago,
- no family history, no consanguinity.
- On examination, temporal mass filling the whole temporal retina associated with total exudative detachment and subretinal seeding it was classified as group D retinoblastoma.
- The other eye was free
- The decision was 6 cycles chemotherapy with transpupillary thermotherapy



First case

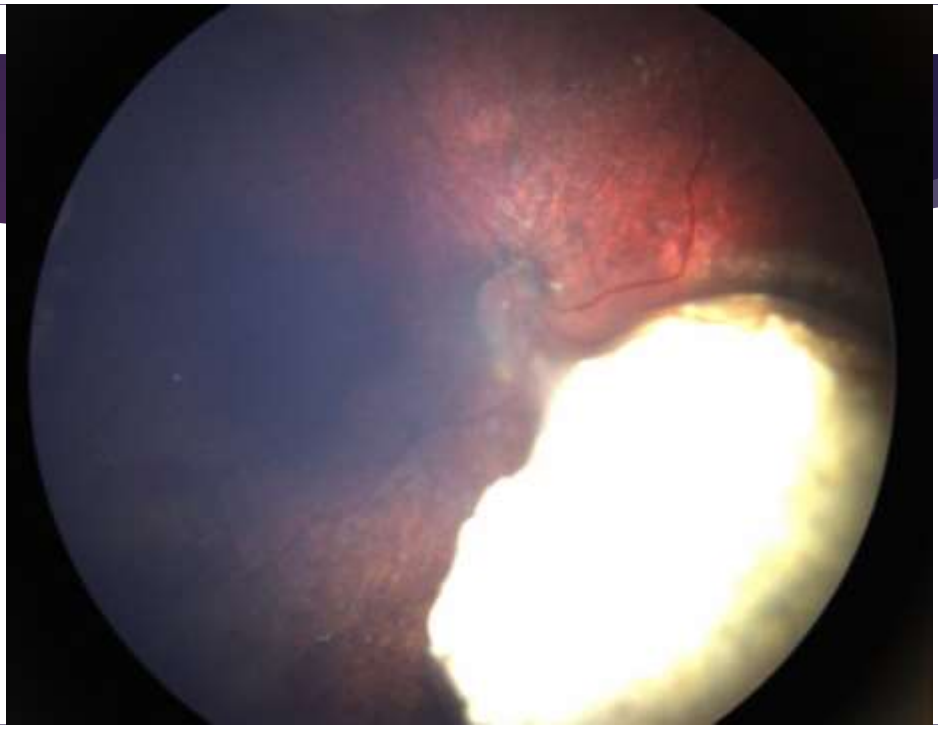
- After the second cycle chemotherapy, the tumor showed good initial regression
- The current tumor size is 8*8*4 DD with resolution of the subretinal fluid, only subretinal seeds
- We started TTT.

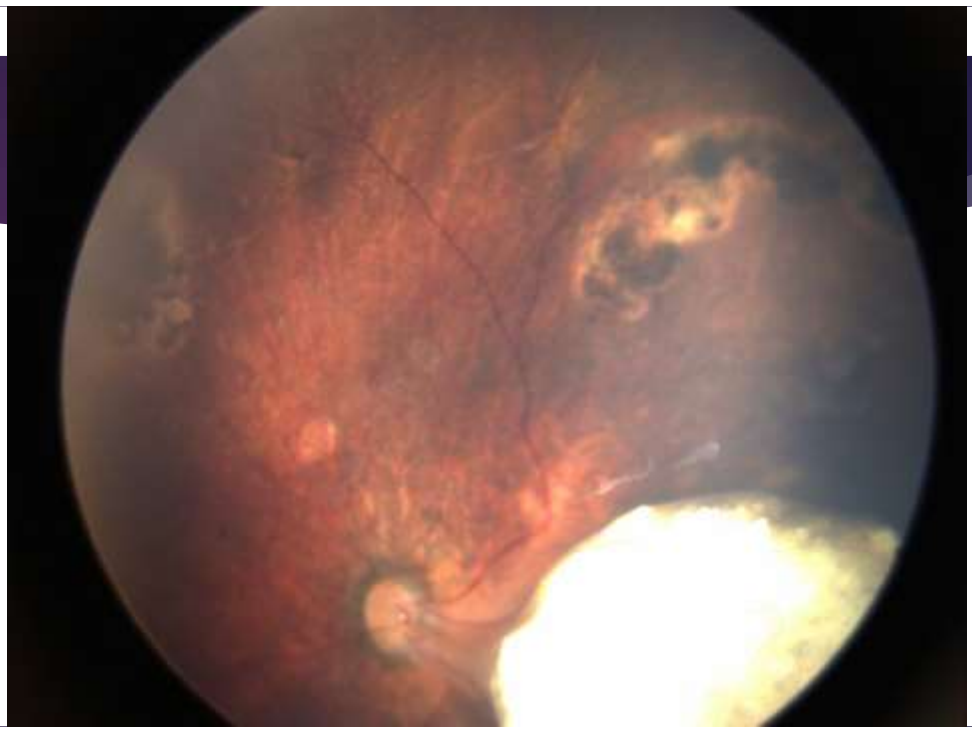


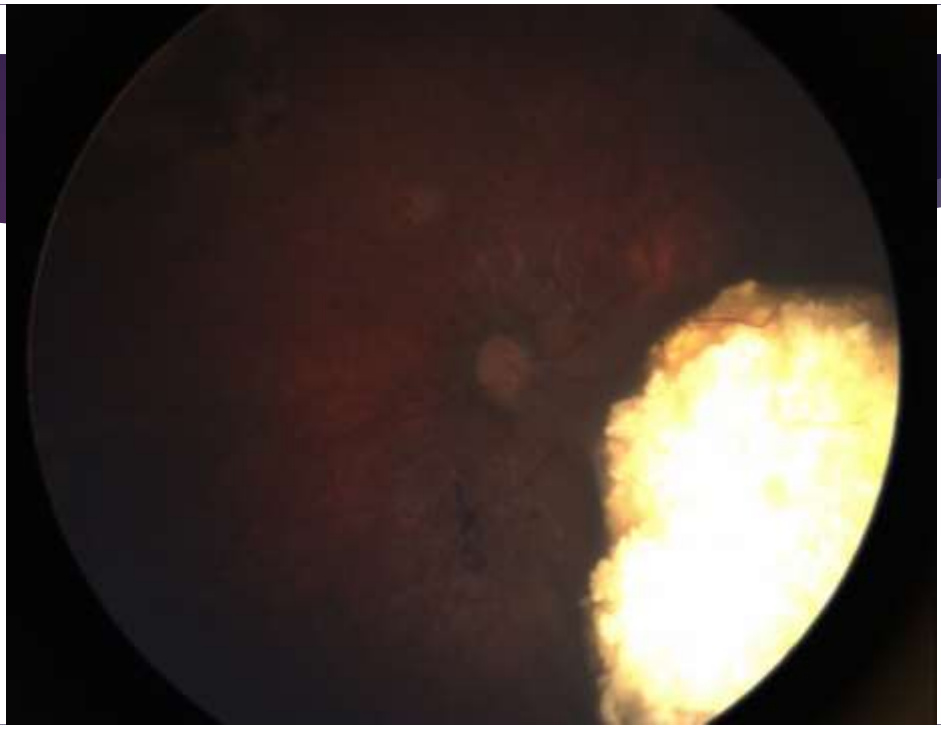


First case

- After the 6th cycle the tumor was same size with calcific changes
- It was still developing new retinal seeds for 6 months after end of chemotherapy, that was treated by TTT,
- it took seven TTT sessions, one per month.

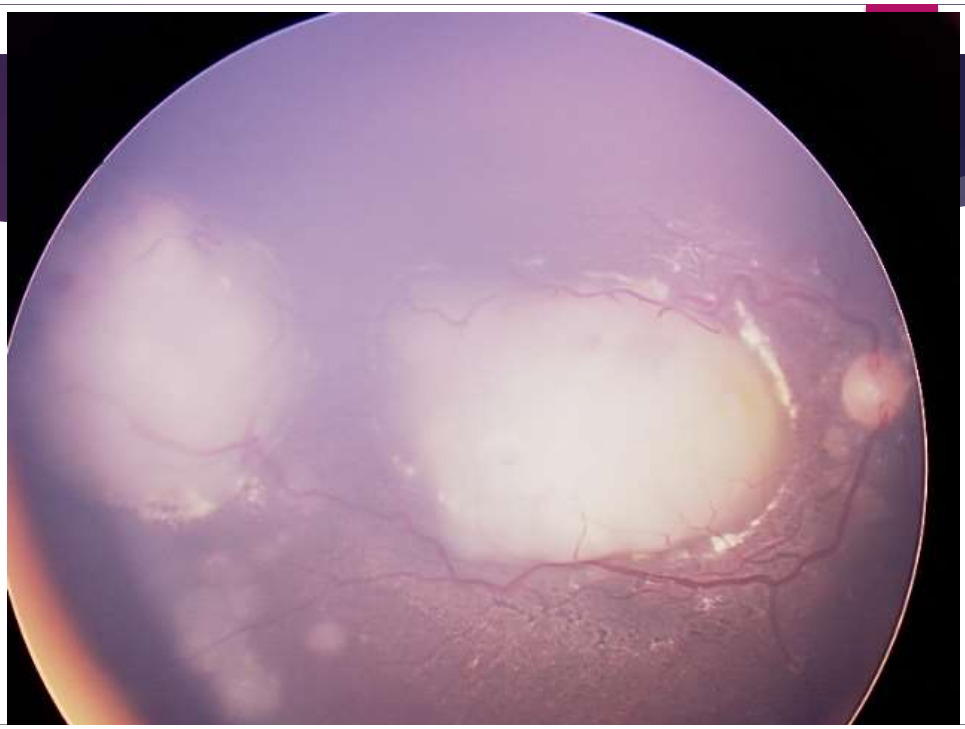






Second case

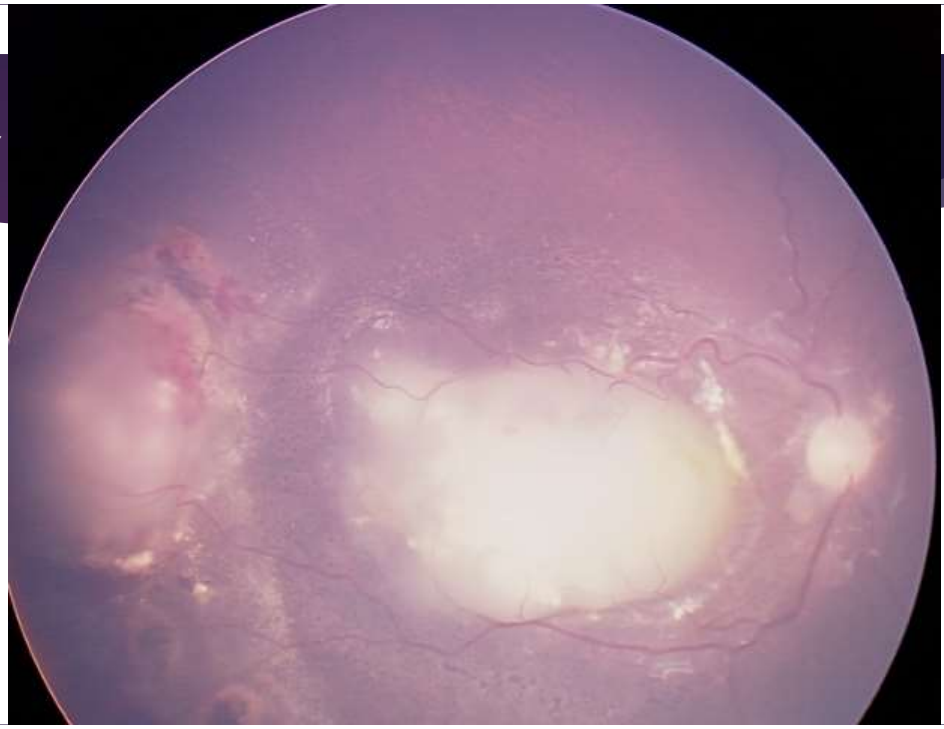
- An eighteen months old patient presented with leukocoria dated 3 months ago,
- there is no family history or consanguinity.
- On examination, there was 2 temporal tumors, the first on size is 6*4*2 DD, the other one size is 4*5*2 DD with minimal subretinal fluid around the edge of the two tumors, active subretinal seeds in more than one quadrant, it was classified as group D Retinoblastoma.
- The other eye was normal
- The decision was 6 cycles chemotherapy with transpupillary thermotherapy



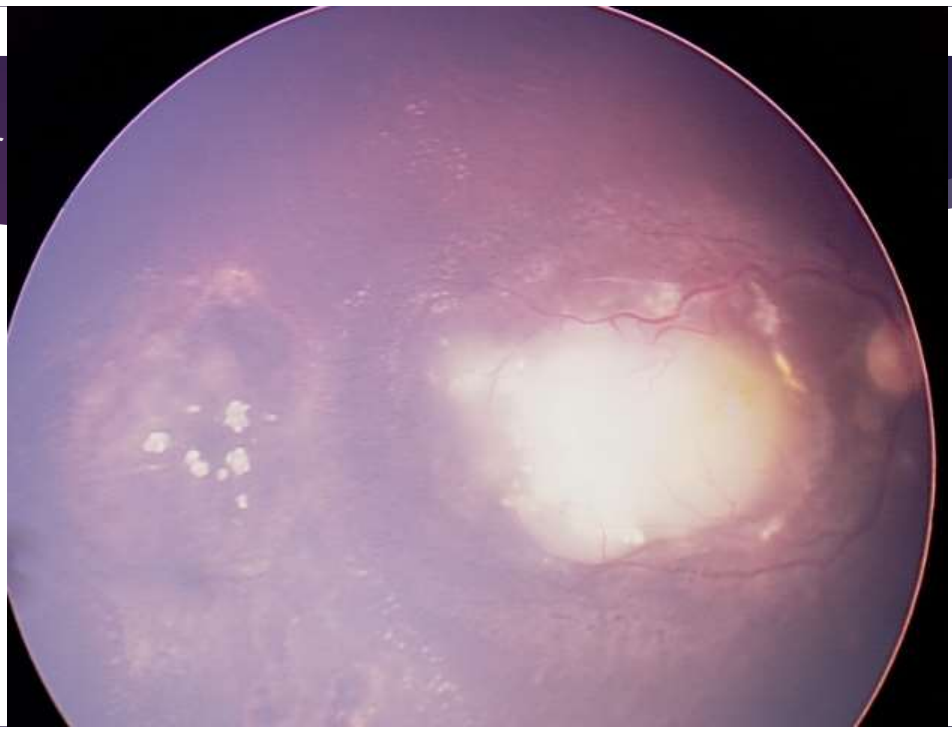
Second case

- after the second cycle, the tumor showed good initial regression,
- the larger tumor regressed to 5*3*1.5, the smaller one regressed to 3*4*1.
- TTT has been done to the surface of tumors and the active seeds

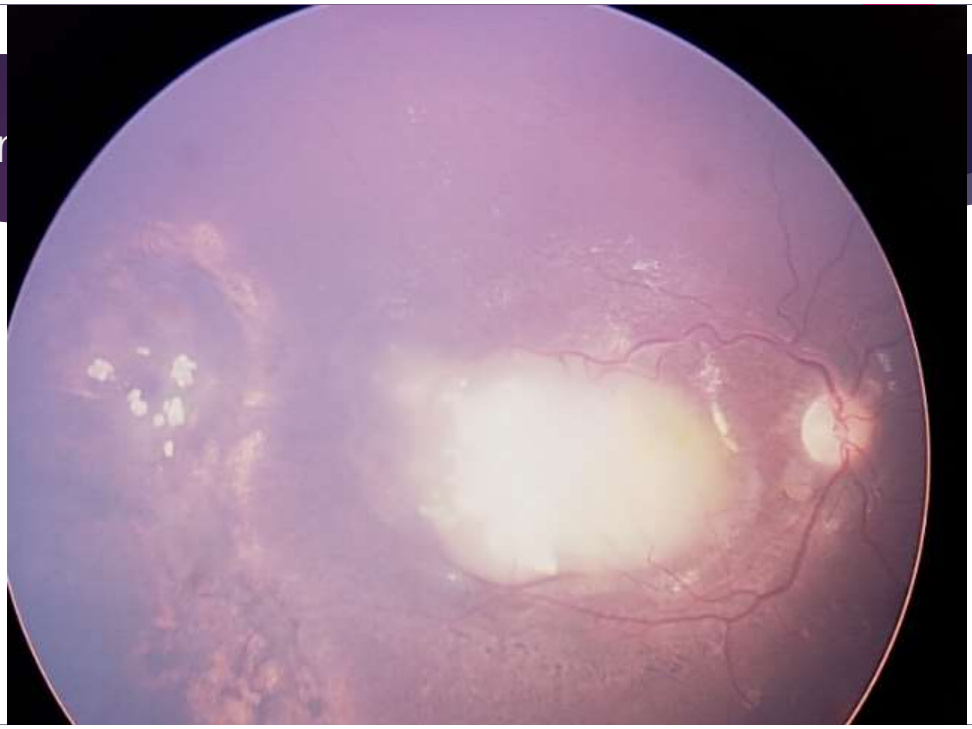
After



After



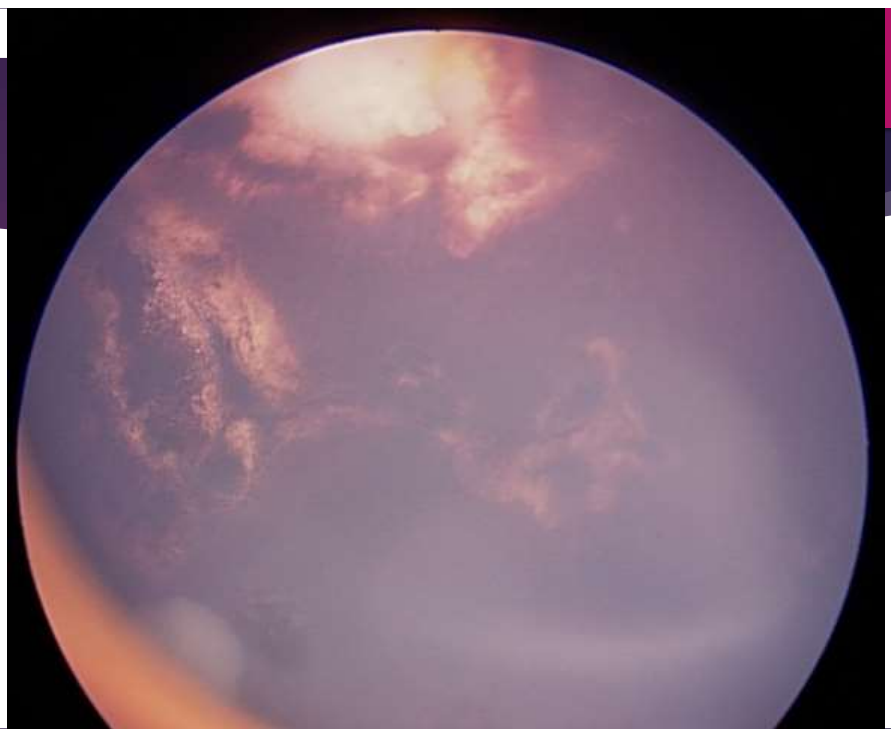
After



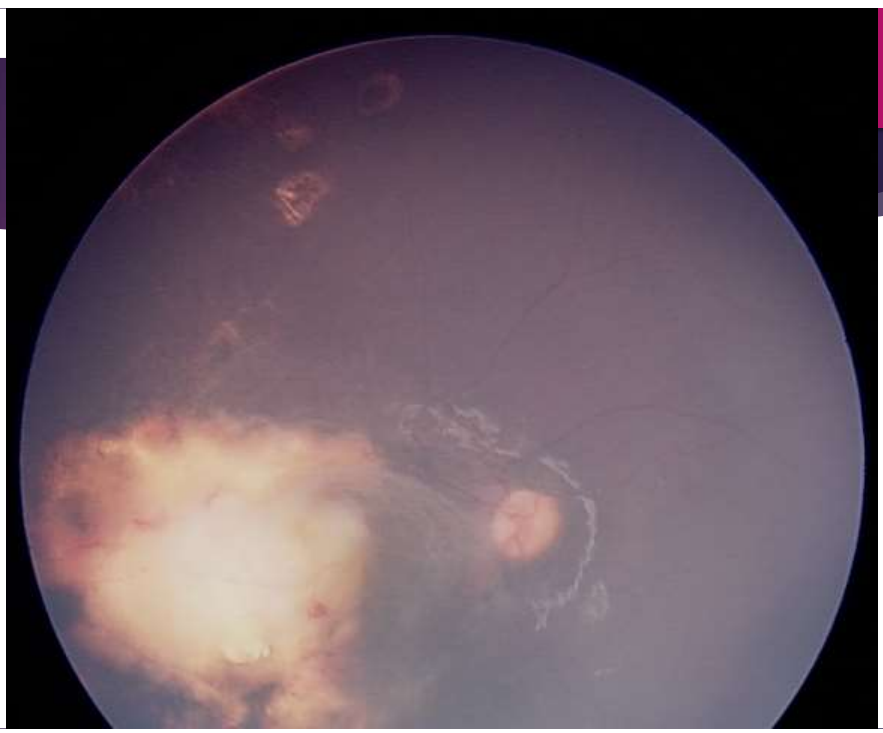
Second case

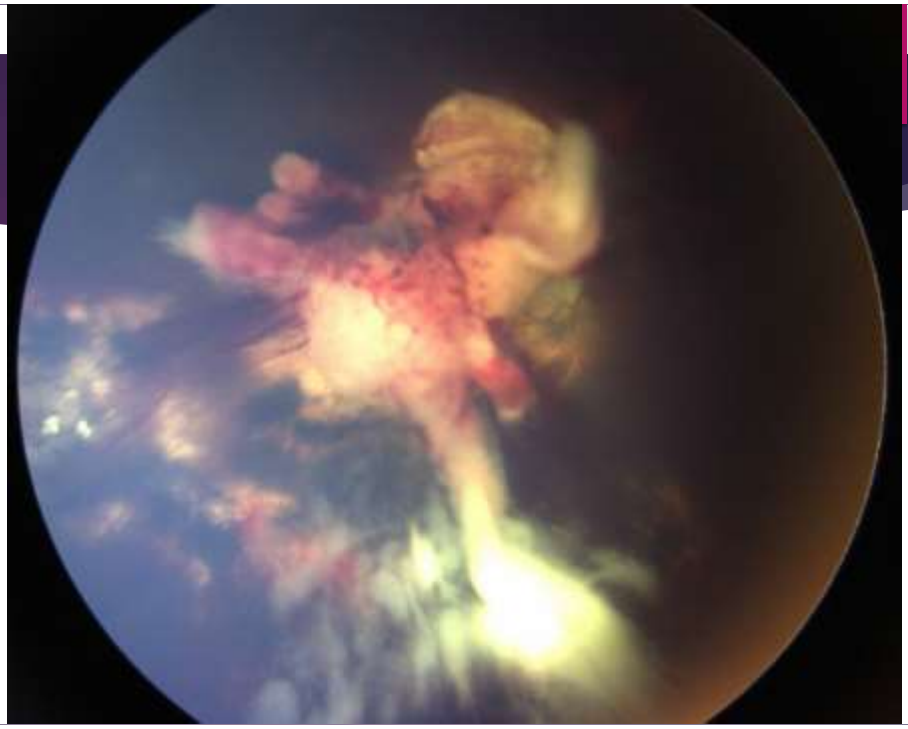
- After the end of chemotherapy, we continued regular follow up each month with focal therapy for the main tumor and the recurrent seeds that started to appear after the end of chemotherapy.

- Then after 9 months of the end of chemotherapy the patient developed peripheral new tumor we started to treat it with cyotherapy



- After 12 months from the end of chemotherapy, the patient started to develop vitreous seeds on the surface of the main tumor.
- We decided to treat the seeds with intravitreal melphalan injections.





Second case

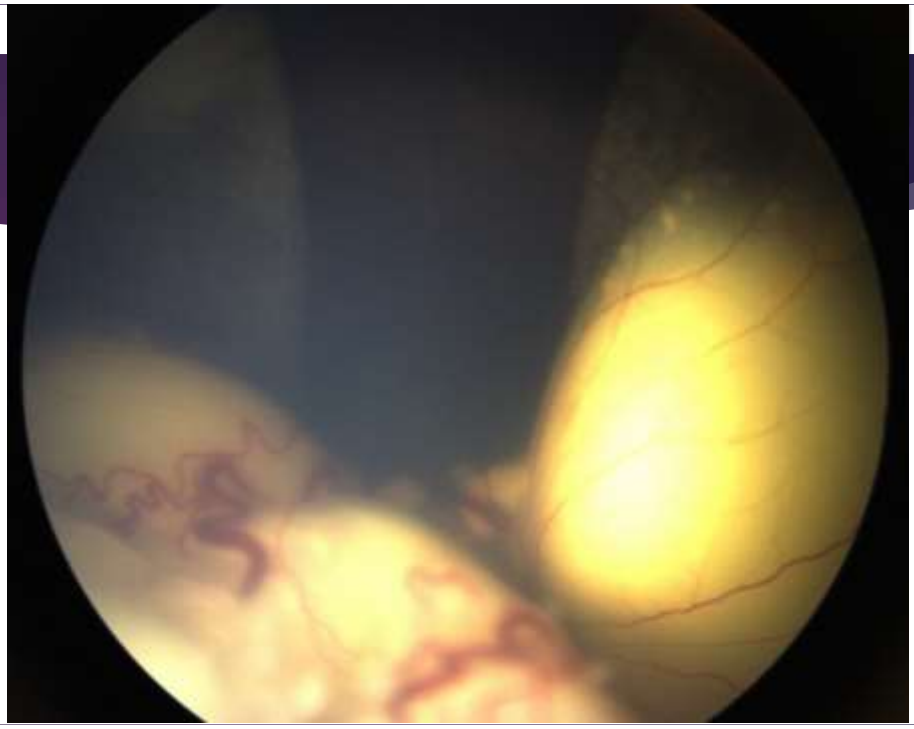
- After 3 successive intravitreal melphalan injection with a dose of 20 microgram /ml, the tumor was still progressing, with more dense vitreous seeds, active recurrence from the main tumor and multiple peripheral recurrences.
- We decided to go for enucleation

Second case

- histopathology of the enucleated eye revealed poorly differentiated tumor with massive choroidal invasion that required prophylactic adjuvant chemotherapy.

Third case

- Female child 34 months, presented with leukocoria 1 month ago,
- no family history or consanguinity.
- On examination 2 masses involving the inferior half of the retina with subretinal seeding and subretinal fluids in 2 quadrants, anterior chamber was free, it was classified as group D retinoblastoma.
- The other eye was normal



Third case

- We decided to go for enucleation.
- histopathology revealed well differentiated tumors, no optic nerve, no choroid or scleral invasion, adjuvant chemotherapy was not required.

Why there is a debate

- On one side primary enucleation offers a cancer free life , without chemotherapy side effects such as Ototoxicity , nephotoxicity and bone marrw suppression.
- On the other side , conservative therapy offers globe salvage and moreover a better cosmesis

The International Classification of Retinoblastoma Predicts Chemoreduction Success

- Of the 249 eyes, 23 (9%) were in group A, 96 (39%) were in group B, 21 (8%) were in group C, and 109 (44%) were in group D.
- Treatment success was achieved in 100% of group A, 93% of group B, 90% of group C, and 47% of group D eyes.

Unilateral retinoblastoma; natural history and an age-based protocol in 248 patients

104 cases with unilateral D retinoblastoma

58 cases managed with chemoreduction , the eye was saved in 23 cases (40%) , while 35 cases ended with secondary enulceation

4 cases out of them revealed high risk pathological features and received high risk protocol , one cases revealed a CNS metastasis and another one with bone marrow metastasis

Unilateral retinoblastoma; natural history and an age-based protocol in 248 patients

46 cases managed with primary enucleation , 23 cases (50%) revealed high risk pathological features , with subsequent high risk chemotherapy protocol

3 cases developed extraocular metastasis

Intra Arterial Chemotherapy

- Allowing delivery of more conentrations of chemotherapy to the eye with minimal systemic side effects.
- Disadvantages
  - Weak control over vitreous seeds
  - Local complications : Vascular occlusion , vitreous hemorrhage
  - CNS toxicity

Take Home message

- Chemoreduction success in group D 40% to 50%.
- Close regular follow up especially in the first year after chemotherapy with monitoring the decision whenever needed
- In Retinoblastoma , we are concerned first on saving life ,then saving the eye , then saving vision.


Thank
you