LECTURE BY

Dr.Ramakant Thakur

MBBS,M.S(Ophthalmology)

Assistant professor

Department of Ophthalmology

S K M C H Muzaffarpur Bihar India

Ph. No. -9939676146

Email-dr.ramakant0014@gmail.com

Under unit of Dr. Rajiv Kumar Singh

Associate professor and head of the department

Department of Ophthalmology

S K M C H Muzaffarpur Bihar India

TOPIC –

INTROCULAR TUMORS

Intraocular Tumors

Tumors of the uvea and retina are rare. They are of great importance, since most of them are malignant and endanger the life of the patient.

IRIS TUMORS

Iris Nevus (Fig. 20.1)

- It consists of slightly elevated, localized, discrete, small pigmented mass and is composed of proliferated melanocytes.
- The nevi are not progressive, and do not distort the pupil.
- As a rule, this is benign, but occasionally it takes on malignant proliferation.

Such a development is suggested by an increase in size, vascularization and pigmentation, which sometimes distorts and everts the pupillary margin *(ectropion uveae)*.

Fig. 20.1: Iris nevus

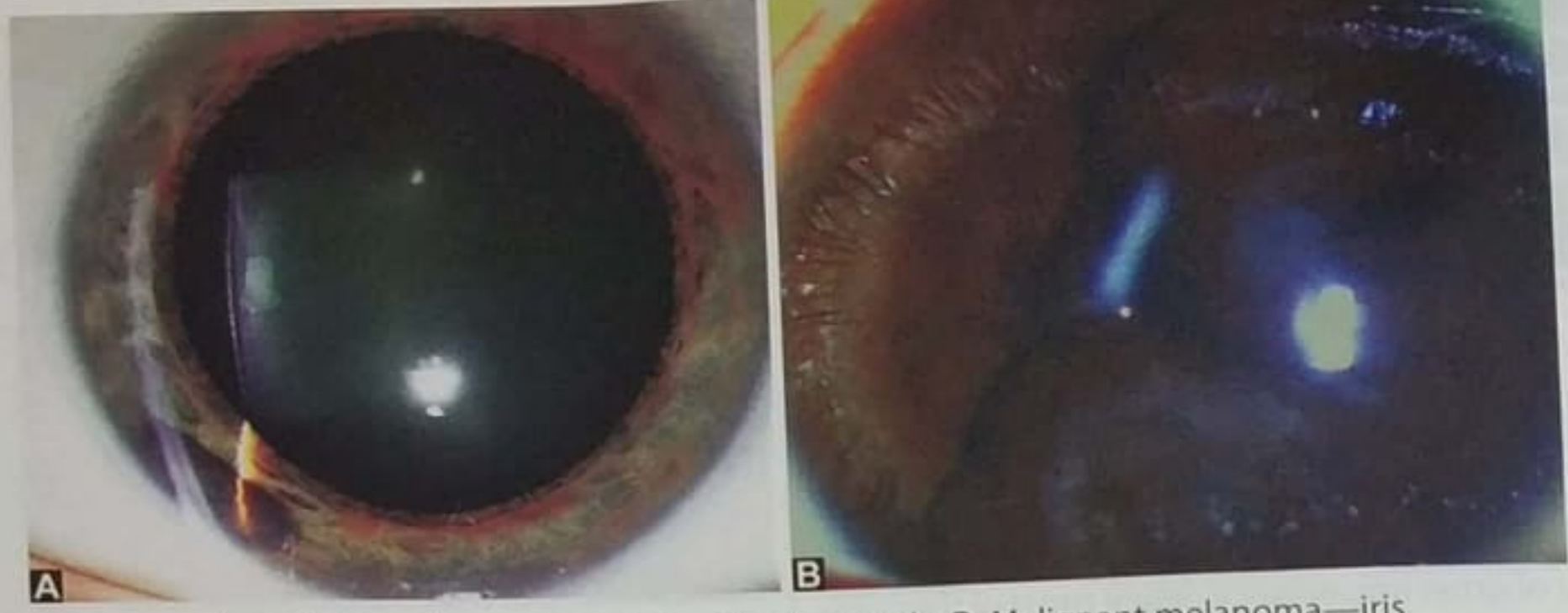
Malignant Melanoma (Figs 20.2A and B)

- It often arises from pre-existing nevus.
- It is usually symptomless, and the tumor is noticed by the relatives or friends as a brown mass on the iris surface.
- It has blood vessels growing in it and may be associated with seeding.
- Pupil is distorted and there may be glaucoma when it extends into the anterior chamber angle.
- The growth should be watched carefully and is best treated by broad iridectomy or iridocyclectomy if the angle is involved.

CILIARY BODY TUMORS

Medulloepithelioma (Diktyoma)

- Intraocular medulloepithelioma arises in the nonpigmented epithelium of the ciliary body and resembles embryonic tissue of the primitive retina and ciliary body.
- It is thought to be congenital, though it occurs between 4 years and 12 years of age.
- The chief symptoms are pain and poor vision.
- A white pupil (leukocoria), secondary cataract and secondary glaucoma are often present.
- Enucleation is often delayed due to failure to diagnose the condition.



Figs 20.2A and B: **A.** Iris melanoma at anterior chamber angle; **B.** Malignant melanoma—iris

Malignant Melanoma

Ciliary body melanoma is more common than iris melanoma, but less common than the choroidal melanoma.

- It tends to involve either the choroid or the iris, or both.
- It cannot usually be visible unless the pupil is widely dilated.
- Clinically, it may present in a variety of ways:
  - As subluxation of the lens with secondary glaucoma.
  - As dilated episcleral blood vessels in the same quadrant as with the tumor ***(sentinel vessels)***.
  - As retinal detachment.
  - As diffuse mass in a ring around the ciliary body (Fig. 20.3).
- Because of its location and minimal symptoms, the diagnosis may be often delayed.
- Indirect ophthalmoscopy, three-mirror gonioscopy, transillumination test and USG B-scan or ultrasonic biomicroscopy (UBM) are helpful for early diagnosis.
- Treatment is by enucleation (for large tumors), or by local resection (for small tumors).
- Since, ciliary body melanomas have a low degree of malignancy, the prognosis is usually good.

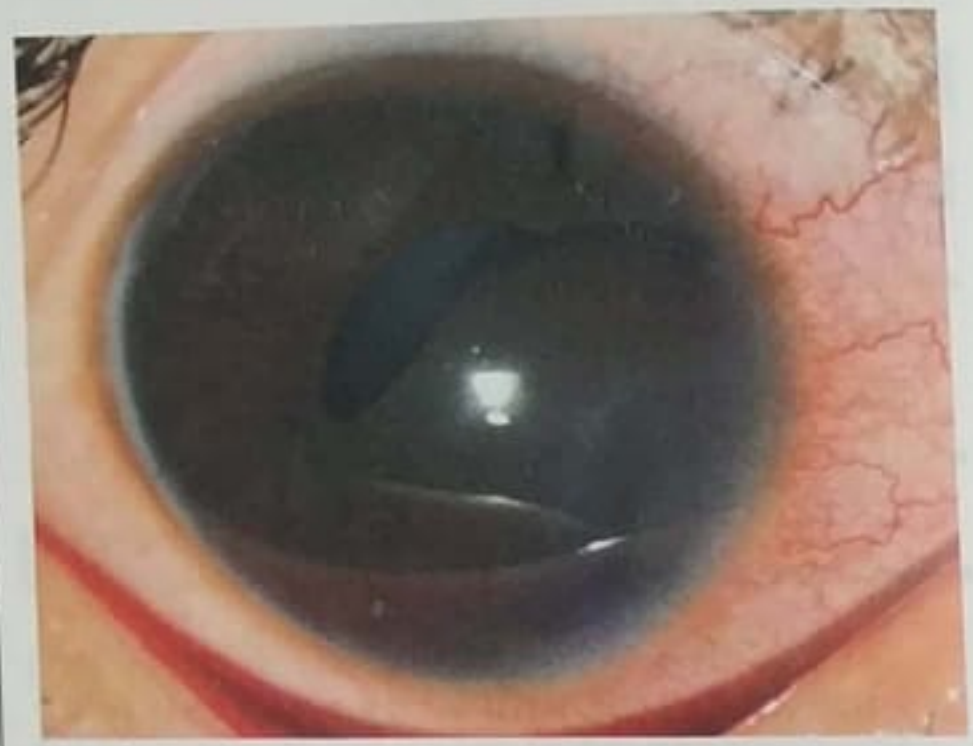


Fig. 20.3: Ciliary body melanoma

CHOROIDAL TUMORS

Benign Melanoma (Nevus) (Fig. 20.4)

- Typical benign melanomas of the choroid are flat or slightly elevated, oval or circular state-gray lesions.
- They are usually less than three disc diameters in size and occur most frequently at the posterior half of the fundus.
- They are usually present at birth and maximum growth occurs during puberty.
- Most of the benign melanomas are asymptomatic, although those near the macula may present with impairment of central vision.

Fig. 20.4: Benign melanocytoma of the choroid

- Benign melanomas may become malignant. An increase in size, elevation of the mass, irregular pigmentation over the tumor and visual field loss suggest malignancy.
- Treatment is not indicated except the patient should be followed up regularly with serial fundus photographs.

MALIGNANT MELANOMA OF THE CHOROID

The most common primary intraocular tumor in adults is malignant melanoma of the choroid (Fig. 20.5).

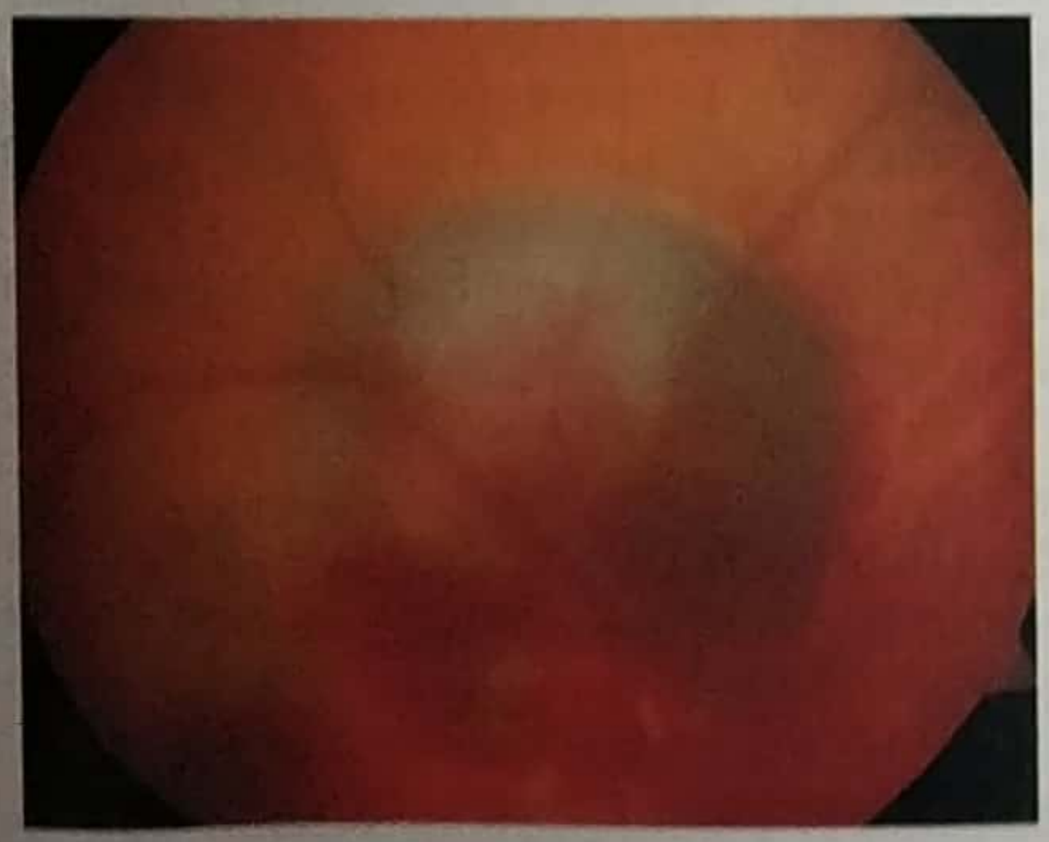


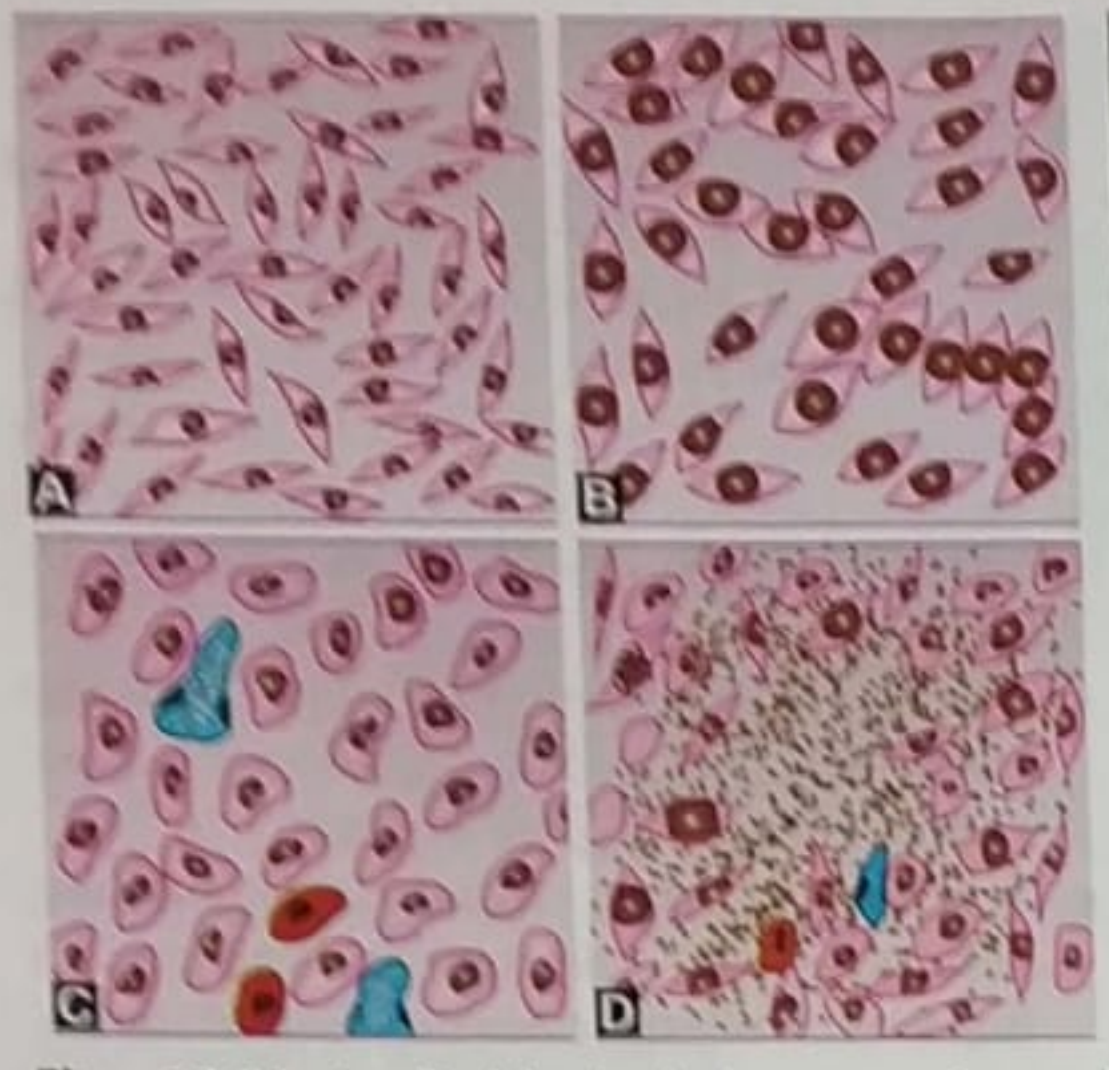
Fig. 20.5: Choroidal melanoma

- The average age of the patients with choroidal melanoma is 50 years.
- They are extremely rare among blacks, and slightly more common in males.
- Most malignant melanomas develop from pre-existing benign melanomas (nevi).
- They originate most frequently in the outer layers of the choroid, and may spread like a carpet between the sclera and the Bruch's membrane.
- Tumors may remain quiescent for many years, and then without apparent reason, suddenly begin to grow rapidly.
- Eventually, the Bruch's membrane perforates and the growth appears as mushroom or collar-button shaped mass.

Histological Classification (Callender's)

Uveal malignant melanomas can be divided into six types (Figs 20.6A to D) according to cellular features:

1. ***Spindle-A cell type:*** Tumors composed of this cell type have the best prognosis.
2. ***Spindle-B cell type:*** It is having the second best prognosis.
3. ***Fascicular type:*** The cells in a palisading arrangement are composed of either spindle-A or spindle-B cells and has the same prognosis as primary cell type.
4. ***Epithelioid cell type:*** This is the least common type and these cells carry the worst prognosis.
5. ***Mixed-cell type:*** This is composed of a combination of spindle and epithelioid cells with intermediate prognosis. This is the most common type of malignant melanoma.
6. ***Necrotic type:*** This is the tumor in which the specific cell type cannot be recognized. The necrotic process may initiate a severe inflammatory reaction which may be mistaken clinically, as endophthalmitis or uveitis.



Figs 20.6A to D: Histological types of uveal melanoma. **A.** Spindle-A type; **B.** Spindle-B type; **C.** Epitheloid type; **D.** Mixed type

Clinically, malignant melanoma of the choroid is commonly divided into four stages:

1. Quiescent stage
2. Glaucomatous stage
3. Stage of extraocular extension
4. Stage of metastasis.

The chief symptoms result from the exudative retinal detachment and secondary glaucoma caused by increase in the choroidal volume. The patient may present with decreased visual acuity, or a defect in the visual field, and ocular pain accordingly.

In some of the cases, the patient is ***asymptomatic,*** and the tumor is detected by a routine ophthalmoscopic examination.

Ophthalmoscopically

- The tumor is invariably unilateral and solitary.
- A typical melanoma appears as a pigmented and elevated oval mass.
- The color of the mass is frequently brown, although it may be mottled with black or dark-brown pigment or it may be amelanotic (Fig. 20.7).
- As the tumor grows a brown exudative detachment results owing to break in the Bruch's membrane.

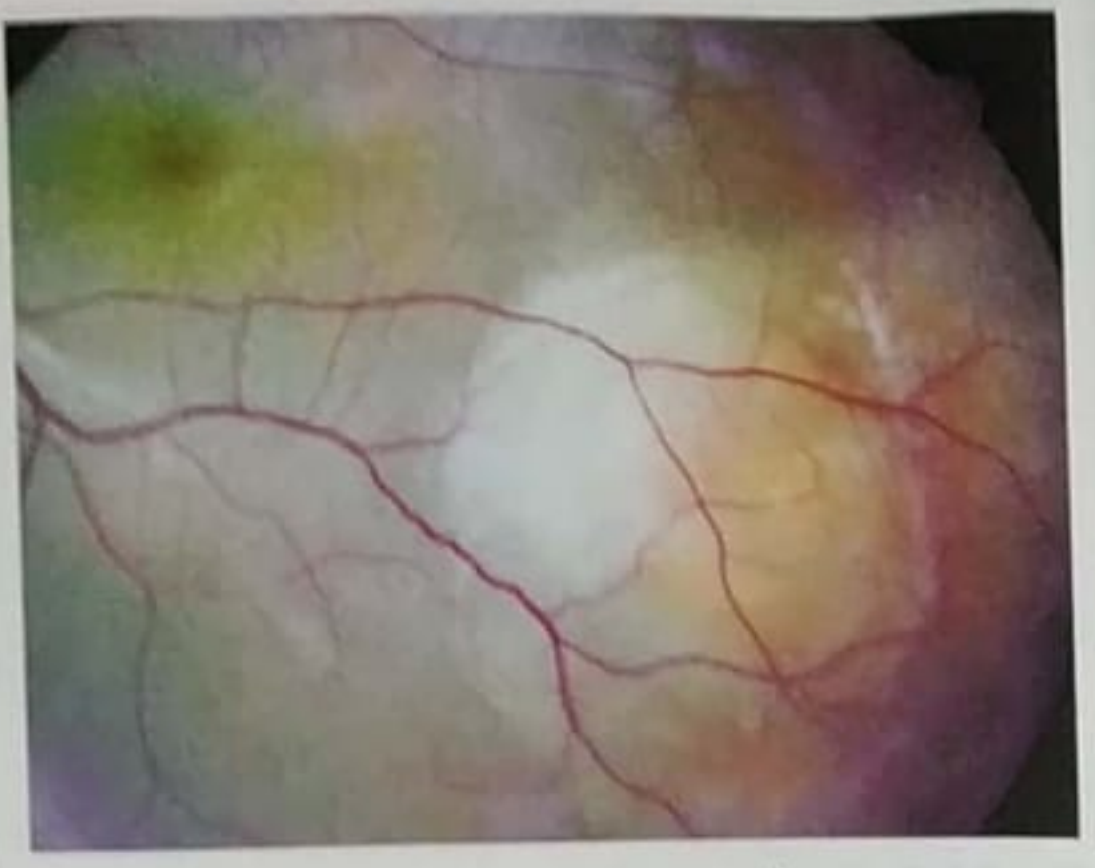


Fig. 20.7: Choroidal amelanotic melanoma

- An accumulation of orange pigment ***(lipofuscin)*** in the retinal pigment epithelium is commonly seen over the detached area but is not diagnostic.
- ***Other ocular features include*** choroidal folds, subretinal hemorrhage, vitreous hemorrhage, secondary glaucoma cataract and posterior uveitis.
- The lymph nodes are not affected, but distant metastasis in the liver and elsewhere are the common cause of death.

Investigations

- ***Medical evaluation:*** It is to exclude the possibility of a metastatic tumor of the choroid and to check for distant metastasis from the primary ocular growth. Liver function tests and X-ray chest are most valuable.
- ***Indirect ophthalmoscopy*** for better depth assessment.
- Three-mirror and contact lens examinations of the fundus.
- ***Transillumination*** test is useful in differentiating a pigmented tumor from a non-pigmented lesion.
- Fundus photography.
- ***Fluorescein angiography:*** In general, most melanomas show a mottled fluorescence during the arteriovenous phase of the angiogram with a progressive staining

of the lesion and prolonged retention of the dye.

- ***Ultrasonography:*** It is especially useful in detecting the presence of a tumor in an eye with hazy media. Both A-scan and B-scan are helpful to detect the solid nature of the mass. It shows a solid mass with homogenous appearance arising from the choroid with collar-button appearance, acoustic hollow and choroidal excavation.
- ***P32-uptake:*** Malignant melanoma has an increased rate of phosphate uptake and retains ***P32-isotope*** longer than other nonmalignant lesions.
- ***Visual field examination:*** It is of limited value.
- ***CT scan and MRI:*** They are valuable in detecting the extraocular extension.
- ***USG-guided tissue biopsy:*** It is to obtain cellular aspirates for analysis.

Differential Diagnosis

Rhegmatogenous retinal detachment (RD), metastatic tumor of the choroid, choroidal hemangioma, large choroidal nevus, and choroidal detachment.

Management

The correct management at present is controversial as the traditional treatment of enucleation has been questioned by many ophthalmologists, especially for small tumors.

- ***Observation:*** It is indicated for small tumors, where the diagnosis is not certain; patients over the age of 65 years; patients with liver metastasis; and with a slow growing tumor with normal vision.
- ***Enucleation:*** It is indicated for very large melanomas, especially, if all useful vision has been irreversibly lost. The overall 5 years survival rate is 75% after enucleation for all types of melanoma.
- ***Radioactive scleral plaques (Cobalt-60 or Iodine-125):*** They are suitable for small to medium size tumors.
- ***Heavy-charged particle irradiation (protons or helium):*** It is a new approach to radiotherapy and may be better than enucleation in eyes with large tumors. It is not available widely.
- ***Photocoagulation:*** Xenon arc photocoagulation is better than argon laser and indicated for small tumors away from the fovea.
- ***Choroidectomy (local resection):*** This difficult and complicated operation may be useful for the selected peripheral tumors.
- ***Palliative therapy:*** Chemotherapy and immunotherapy may be useful in cases with distant metastases.

METASTATIC CARCINOMA OF THE CHOROID (FIG. 20.8)

Metastatic choroidal tumors are probably the most common type of intraocular malignancy (which are often undetected).

- The most frequent primary sites are the bronchus (bronchogenic carcinoma) in males and breast (breast carcinoma) in females (Fig. 20.9).
- Other primary sites are kidney, testis and gastrointestinal tract. The prostate is an extremely rare primary site.

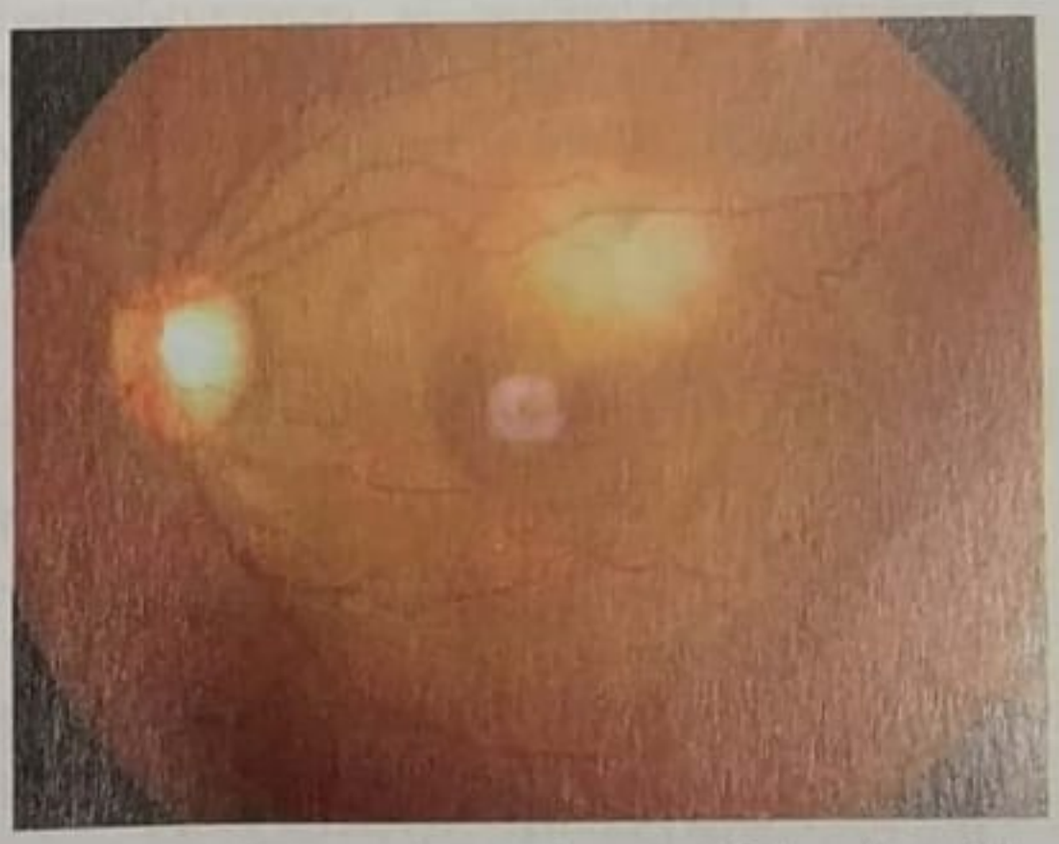


Fig. 20.8: Metastatic carcinoma of the choroid—from bronchogenic carcinoma

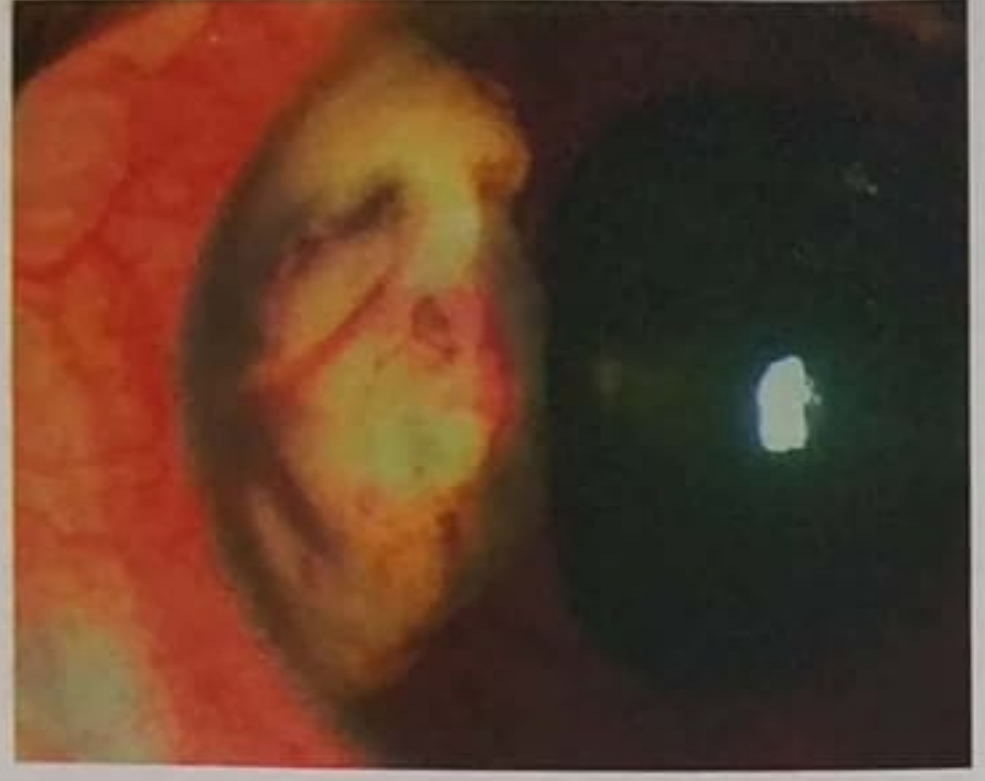


Fig. 20.9: Metastatic carcinoma of the iris from breast carcinoma

- Choroidal metastases have a definite predilection for the posterior pole of the fundus, with early loss of central vision.
- Typically, they appear as solitary or multiple, creamy-white, placoid or oval lesions which infiltrate laterally.
- They rarely become significantly elevated and have ill-defined borders.
- A careful examination of the opposite eye is important as bilateral metastases are common.
- ***Diagnosis*** depends mainly on accurate history and careful general physical examination.
- ***Treatment*** must be directed to the primary disease. Enucleation is contraindicated unless the eye is painful and blind. Palliative treatment is carried out with chemotherapy in conjunction with external-beam irradiation.

CHOROIDAL HEMANGIOMA

This is rare and is associated in about half of the patients with skin angioma (as in ***Sturge-Weber syndrome***).

- Typically, it appears as a dome-shaped or diffuse, reddish-orange lesion mostly at the posterior pole.
- Frequently, secondary changes, like exudative retinal detachment, pigment mottling and cystoid degeneration may occur.
- A useful clinical sign is the blanching of the lesion with pressure on the globe.
- P32-uptake test, ultrasonography (USG) and fluorescein angiography are helpful to differentiate it from other lesions (e.g. metastatic tumors and amelanotic melanoma).
- ***Treatment:*** Asymptomatic tumors do not require treatment. If the vision is threatened, extensive photocoagulation should be applied around the tumor.

RETINOBLASTOMA

It is the malignant intraocular tumor originating in the outer nuclear layer of the retina and strongly resembles fetal retina.

Etiology

- ***Incidence:*** 1 in 20,000 live births.
- ***Laterality:*** 25–30% of cases are bilateral.
- ***Age:*** The neoplasm is probably congenital, but the average age of diagnosis is 18 months. The vast majority of cases become clinically apparent before the age of 3 years. It is exceptional after the age of 7 years.
- ***Inheritance:*** A positive family history is present only in 6% of cases (mostly in bilateral cases). Mode of inheritance is autosomal dominant. Remaining 94% of cases are sporadic.
- ***Chromosomal abnormalities:*** In 5% of cases, it is associated with deletion of the long arm of chromosome-13 and with trisomy-21.

Other Facts

- Orbital sarcoma may occur in the irradiated field in children with enucleated retinoblastoma.
- Osteogenic sarcoma of femur and skull bones are more common in patients who have survived from bilateral retinoblastoma.

- Very rarely pinealoblastoma may be associated with some cases of bilateral retinoblastoma and then it is called ***trilateral retinoblastoma***.

Clinical Features

Clinical Presentation

- ***Leukocoria or amaurotic cat's eye reflex:*** The most common mode of presentation (60% of cases) and is due to reflection of light from the yellowish-white mass in the retrolental area (Fig. 20.10).
- ***Squinting of the eye:*** The second most common mode of presentation (20%).
- ***Secondary glaucoma*** which may be associated with buphthalmos.
- ***Proptosis*** (due to orbital involvement) (Fig. 20.11).
- ***Endophthalmitis or anterior uveitis*** may also be a presenting feature.
- ***Visual difficulties:*** Noticed by the parents.
- Nystagmus in bilateral cases if it occurs within 6 months.

Signs

- Unilateral dilated pupil
- White pupillary reflex
- Strabismus
- Intraocular tension is higher

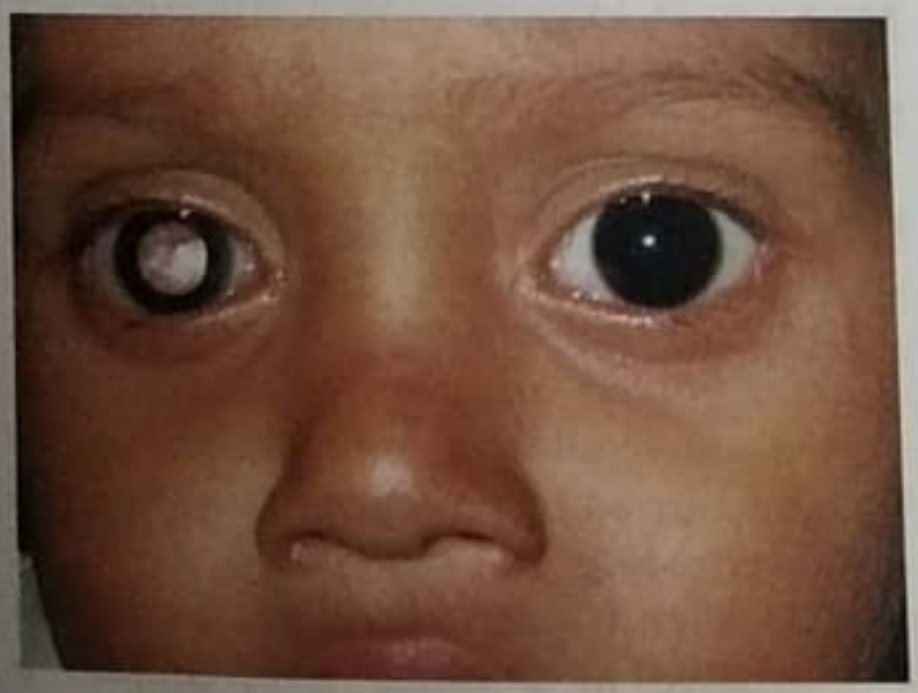


Fig. 20.10: Right-sided retinoblastoma—amaurotic cat's eye reflex

Fig. 20.11: Bilateral retinoblastoma with proptosis of the left eye due to orbital involvement

- ***Heterochromia of the iris:*** It is due to rubeosis irides owing to massive posterior segment ischemia.

Stages of Retinoblastoma

Stage 1: The quiescent stage (6–12 months).

Stage 2: The glaucomatous stage.

Stage 3: The stage of extraocular extension.

Stage 4: The stage of metastasis.

Spread of Retinoblastoma

- ***Direct spread***
  - Into the intraocular tissues.
  - Into the extraocular tissues:
    - Extension into the central nervous system (CNS) via optic nerve.
    - Extension into the orbit as a fungating mass (Fig. 20.12).
- ***Lymphatic spread:*** Along the orbital lymphatics into the preauricular and cervical lymph nodes.
- ***Spread by blood stream:*** Choroidal invasion is the main route of escape. Most common sites are the bones and liver, and lung is the least common site.

Diagnosis

Indirect Ophthalmoscopy

Indirect ophthalmoscopy with scleral indentation, following full dilatation of pupil

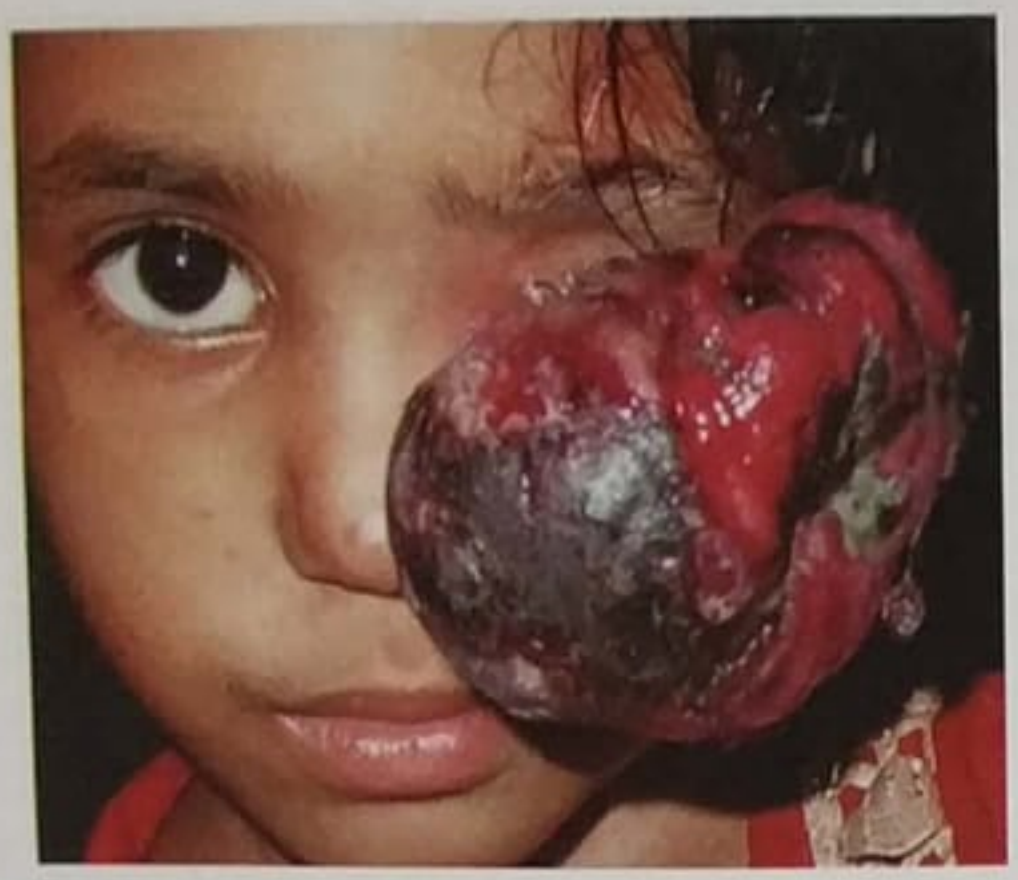


Fig. 20.12: Retinoblastoma—extensive direct extension into the orbit

should be performed in both eyes under general anesthesia. Tumors arising anterior to the equator are missed with direct ophthalmoscope. The appearance of the tumor may be:

Endophytic type (glioma endophytum) (Fig. 20.13)

- This projects from the retina into the vitreous cavity.
- White or pearly-pink colored mass with sharply demarcated margin.
- Presence of calcium deposits in most cases gives the appearance of ***cottage-cheese***.
- Multiple seeding of the tumor cells may be seen in the vitreous cavity.

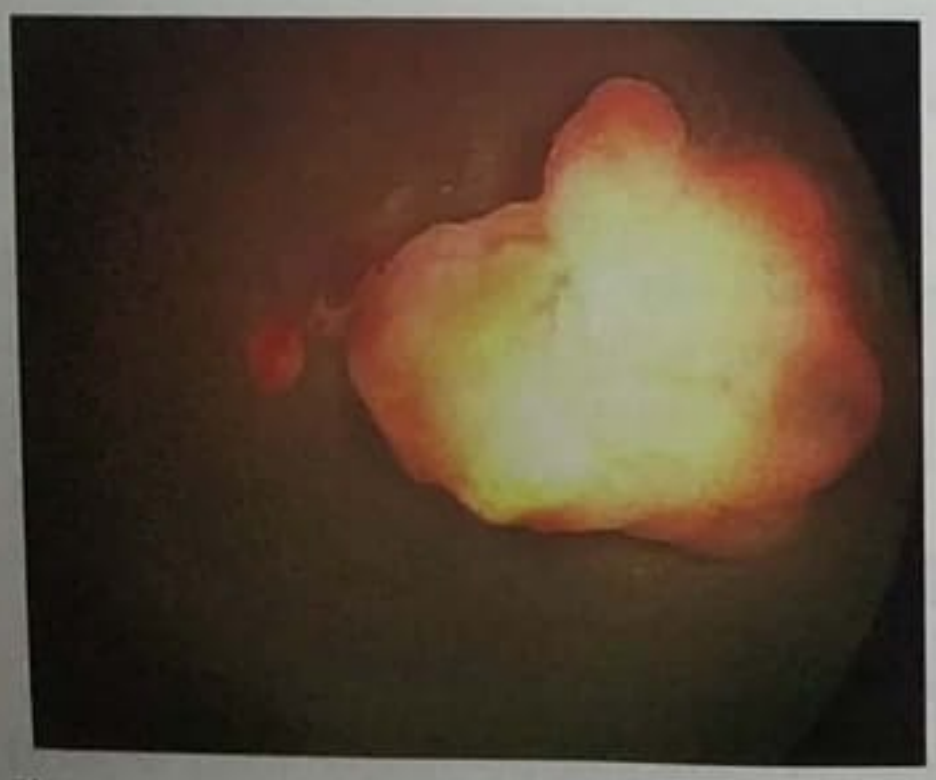


Fig. 20.13: Large endophytic retinoblastoma

Exophytic type (glioma exophytum)

- This grows in the subretinal space and gives rise to a total exudative retinal detachment.
- The tumor itself is difficult to visualize.

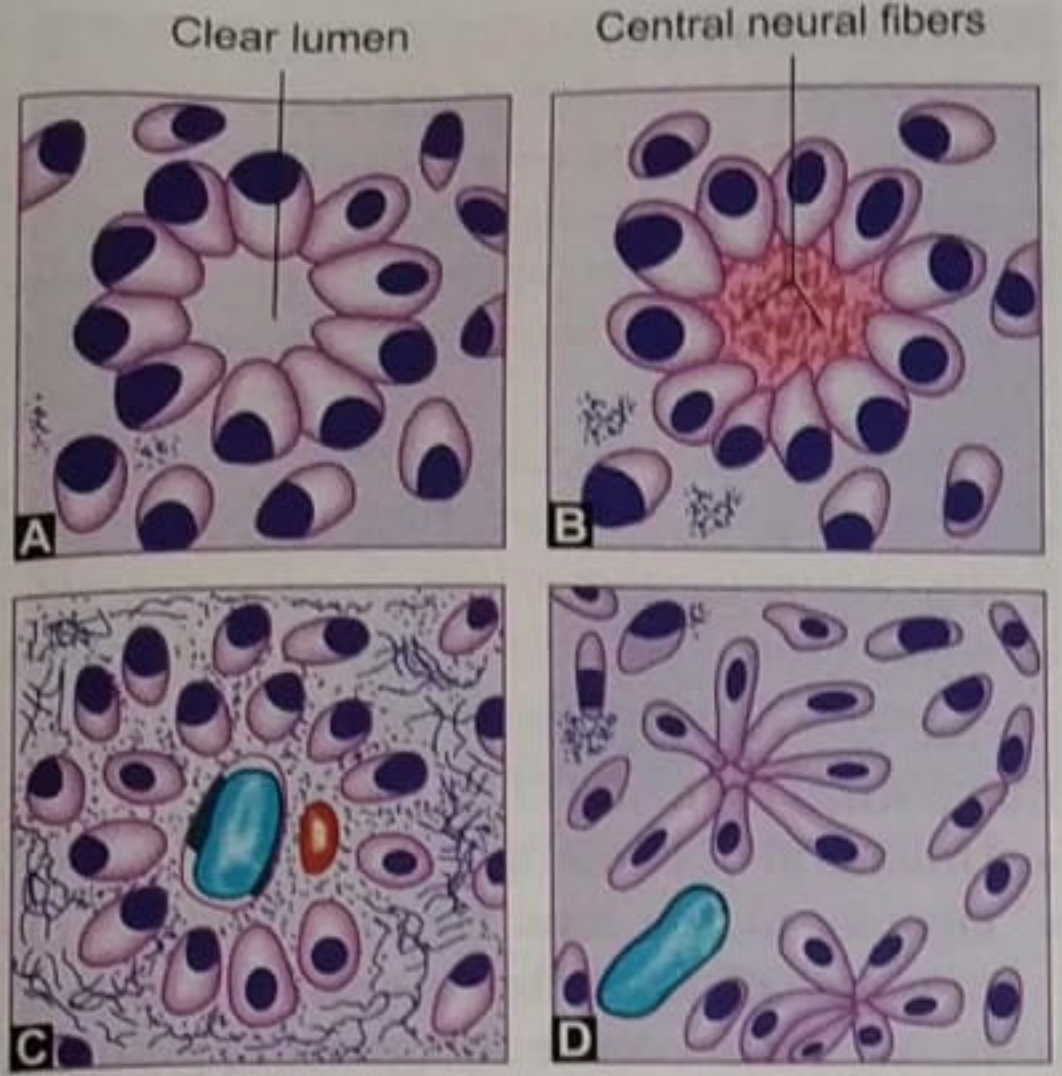
Plane type (glioma planum): It is difficult to visualize the tumor.

Special Investigations

- ***X-ray of the orbit:*** It is for the presence of calcification and erosion of the optic foramen.
- ***Ultrasonography:*** USG B-scan shows presence of tumor with calcification.
- ***CT scan of the orbit and brain*** to demonstrate calcification and CNS spread.
- ***Aqueous humor paracentesis***: It is for cytology and lactate dehydrogenase (LDH) enzyme assay. An aqueous to serum LDH ratio of greater than 1.0 is suggestive of retinoblastoma.
- ***Carcinoembryonic antigen (CEA)*** may be found in retinoblastoma patient.
- ***Enzyme-linked immunosorbent assay (ELISA)*** test to differentiate it from Toxocara endophthalmitis.
- ***Lumbar puncture*** and bone marrow aspiration for the evidence of metastases.
- ***Fine needle aspiration cytology (FNAC)*** when other diagnostic tests are inconclusive.
- ***Fluorescein angiography:*** It is not done routinely.

Histopathology

- ***Gross examination:*** A chalky-white, friable tumor with dense foci of calcification.
- ***Microscopic:*** Retinoblastoma consists of small, round, densely packed cells with large basophilic nuclei. It may be well differentiated or poorly differentiated. Well-differentiated retinoblastoma is characterized by presence of ***rosettes*** and ***fleurettes*** (Figs 20.14A to D).
  - ***Flexner–Wintersteiner rosette:*** Cells arrange in a single layer around a central clear lumen. This is true rosette and highly characteristic.



Figs 20.14A to D: Microscopic appearance of retinoblastoma. **A.** Flexner–Wintersteiner rosette area of necrosis; **B.** Homer–Wright rosette; **C.** Pseudorosette; **D.** Fleurettes

- ***Homer–Wright rosette:*** It is a radial arrangement of cells around a central triangle of neural fibers (rather than clear lumen) and is mainly found in neuroblastoma or medulloblastoma.
- ***Pseudorosette:*** In necrotic retinoblastoma, several layers of cells may be seen around a blood vessel, within the areas of extensive necrosis with the formation of pseudorosette.
- ***Flurette:*** It is composed of group of tumor cells and contains pear-shaped eosinophilic processes which project through a fenestrated membrane.

- ***Histology of metastatic lesions*** outside the retina shows a change of character of the cells. They resemble sarcomatous cells and rosettes are in rarity.

Treatment

- ***Enucleation:*** Excision of the eye with a long optic nerve stump is the treatment of choice for most tumors affecting the first eye. The treatment of the second eye depends on the size and location of the tumor.
- ***Exenteration:*** Exenteration of the orbit is indicated in case of orbital involvement of the tumor.
- ***Radiotherapy:*** It is done by external irradiation or by a Cobalt-60 scleral plaque. Cataract, radiation retinopathy and scleral necrosis may occur. It is indicated in small to medium size tumors and for recurrent or residual tumor of the orbit.
- ***Photocoagulation:*** Xenon arc photocoagulation is useful for certain small retinoblastomas not involving the optic nerve or the macula.
- ***Cryotherapy:*** It is useful for small peripheral tumors. The tumor should be frozen by triple-freeze and thawing technique.
- ***Chemotherapy:*** It is indicated following enucleation in advanced cases and in presence of distant metastases. Cyclophosphamide and vincristine are used every 3 weeks for 12–15 months.
- ***Combination of therapy:*** To achieve best results.

Genetic counseling: This is important in bilateral cases and if two or more cases of retinoblastoma occur in a family.

- Normal parents with one affected child in sporadic case—the chance of transfer to next sibling is 1.3–6%.
- When an affected person survives to maturity and procreates—chance of having retinoblastoma in offspring is about 25–30% in unilateral case, and the risk is more in bilateral cases.

Prognosis: The overall mortalities from retinoblastoma are between 15 and 20%. Mortality rate is higher with optic nerve involvement, large size tumor, poorly differentiated tumor and when there is choroidal invasion.

Follow-up

Follow-up is extremely important in case of treated retinoblastoma.

In general, the rule is every 4 months until 3 years of age, every 6 months until 4 years of age and yearly for next 3 years.

Examination under anesthesia (EUA) is done in each visit. The examination includes:

- Inspection and palpation of the socket after removing the prosthesis.
- Indirect ophthalmoscopy of the fellow eye.
- USG B-scan of the fellow eye to know the status of the tumor (if treated with other methods).

DIFFERENTIAL DIAGNOSIS OF LEUKOCORIA IN CHILDREN

The common causes of leukocoria or white pupillary (Figs 20.15 and 20.16) (***amaurotic cat's eye***) reflex in children are:

- Retinoblastoma
- Pseudoretinoblastoma (pseudoglioma)
- Congenital cataract
- Retinopathy of prematurity
- Toxocara endophthalmitis
- Persistent hyperplastic primary vitreous
- Retinal dysplasia
- Coat's disease
- Choroidal coloboma.

Retinoblastoma

- Usually unilateral (Figs 20.10 and 20.15)
- Usual age at diagnosis is 18 months
- No inflammatory sign in anterior segment
- Ophthalmoscopy shows, a pearly-white mass with presence of secondary calcification
- Lens is usually transparent
- Intraocular pressure is high
- The lesion is progressive
- X-ray orbit, USG and enzyme assay of the aqueous confirm the diagnosis.

Congenital cataract (Fig. 20.16)

- Unilateral or bilateral
- Opacity in the lens clearly indicates the presence of cataract
- It does not cause diagnostic problem, but a cataract does not exclude other causes of leukocoria especially persistent hyperplastic primary vitreous (PHPV).

Retinopathy of prematurity

- History of prematurity and low birth weight are present.
- History of prolonged exposure to oxygen.
- Bilateral in 100% of cases.
- First noted in neonatal period.
- Presence of tractional retinal detachment.
- Intraocular pressure is normal.

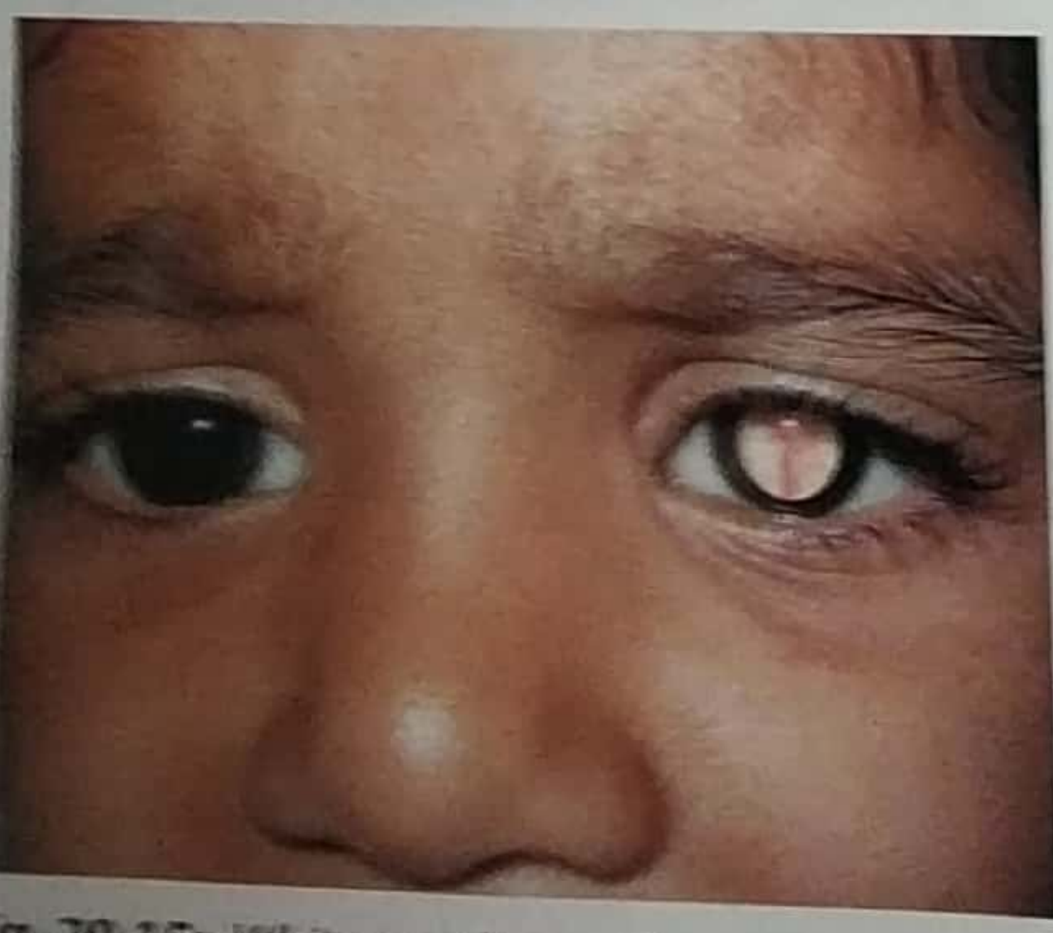


Fig. 20.15: White pupillary reflex—retinoblastoma in left eye

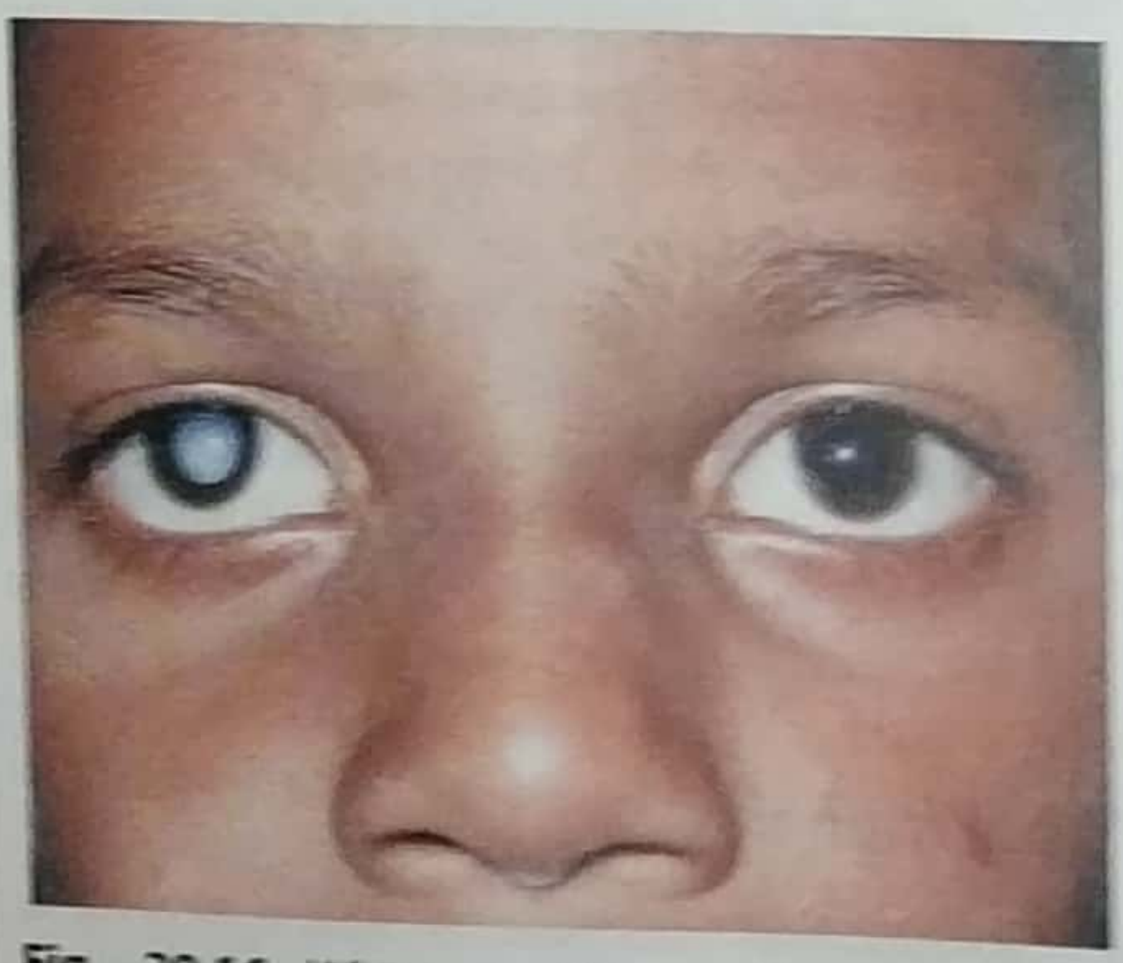


Fig. 20.16: White pupillary reflex—congenital cataract

Fig. 20.17: White pupillary reflex—toxocariasis

Toxocara endophthalmitis (Fig. 20.17)

- Usual presentation is between 2 years and 9 years of age.
- History of contact with pet cat or dog.
- Usually unilateral.
- Signs of inflammation in anterior segment and vitreous are present.
- Intraocular pressure is low and eventually the eye may be phthisical.
- ELISA test is very diagnostic.

Persistent hyperplastic primary vitreous (Fig. 20.18)

- Developmental disorder of the vitreous.
- Usually unilateral, and first noted in the neonatal period.

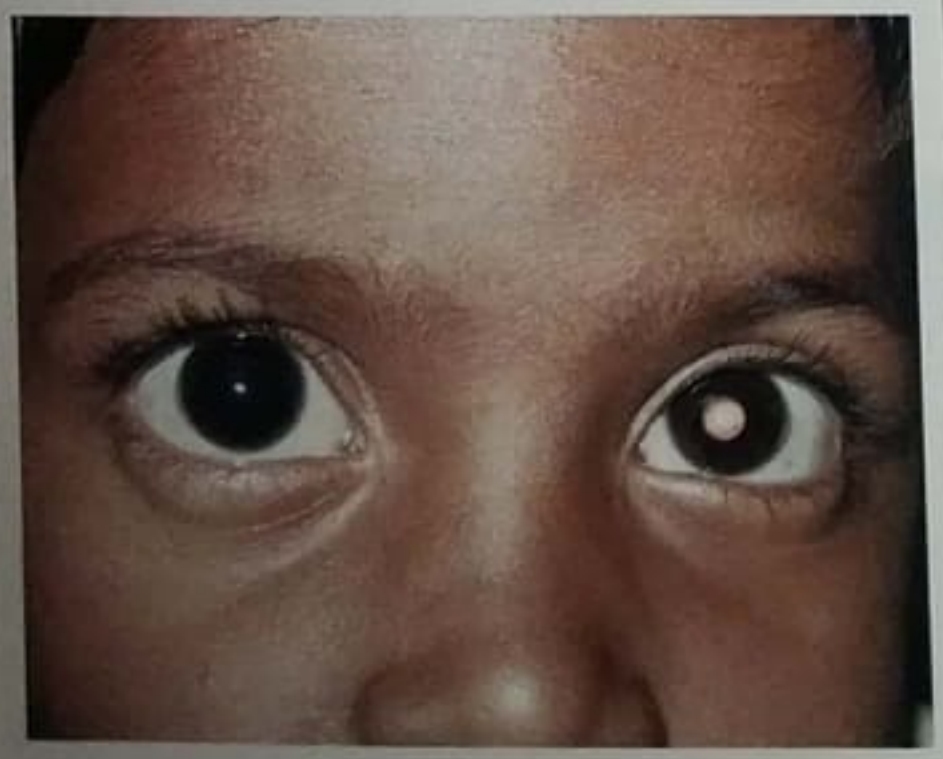


Fig. 20.18: White pupillary reflex—persistent hyperplastic primary vitreous

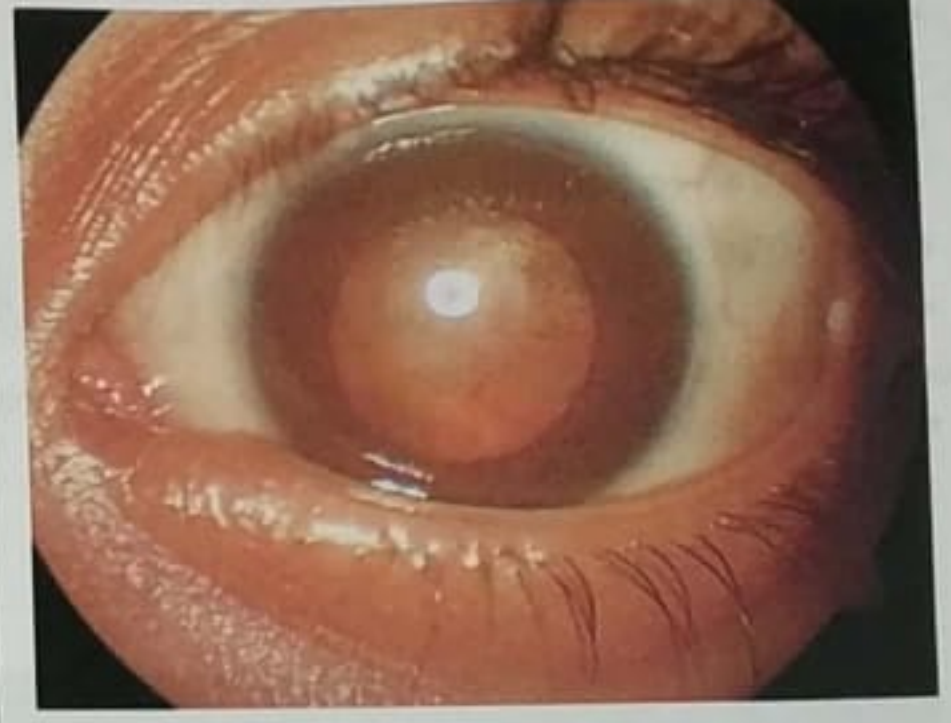


Fig. 20.19: White pupillary reflex—Coats' disease

- Associated with microphthalmos.
- Lens may be cataractous.
- Elongated ciliary processes are visible through the dilated pupil.
- Intraocular pressure may be high.
- USG B-scan confirms the diagnosis in presence of cataract.

Retinal dysplasia

- Unilateral or bilateral usually present at birth.
- Pink or white retrolental mass.
- The eye is microphthalmic with shallow anterior chamber and elongated ciliary processes.
- It is due to failure of the retina to develop normally.
- Usually associated with severe systemic abnormalities.
- USG B-scan helps to differentiate it from PHPV in unilateral cases.

Coats' disease (Fig. 20.19)

- It is almost always unilateral.
- It occurs primarily in older boys (although, it may occur at an early age).
- Large areas of retinal or subretinal exudates with cholesterol crystals.
- Dilated and tortuous retinal blood vessels at the posterior pole.
- Exudative detachment as a retrolental mass occurs over a period of years.

- It is a severe form of retinal vascular telangiectasia.
- Fundus fluorescein angiography may be required to confirm the diagnosis.

Choroidal coloboma

- Leukocoria is only with a large complete choroidal coloboma.
- Usual presentation is at birth.
- Unilateral or bilateral.
- They are always located inferonasally.
- The eye is usually microphthalmic in case of large coloboma.
- Ophthalmoscopy shows the defect is at the embryonic position of choroidal fissure with shiny-white sclera.

1. **Most common orbital tumor in children-**
   a) Rhabdomyosarcoma *(NEET Dec.16 Pattern)*
   b) Retinoblastoma
   c) Melanoma
   d) Chloroma

2. **Most common malignant intraorbital tumor in adult is-**
   a) Lymphoma *(All India Dec.15 Pattern)*
   b) Rhabdomyosarcoma
   c) Dermoid cyst
   d) Sarcoma

3. **Most common tumor to extend from intracranial to orbit is -** *(NEET Dec.12 Pattern)*
   a) Astrocytoma
   b) Pituitary adenoma
   c) Craniopharyngioma
   d) Sphenoidal wing meningioma

4. **The most common primary cause of intraocular tumor in children -** *(All India Dec.15 Pattern)*
   a) Retinoblastoma
   b) Rhabdomyosarcoma
   c) Neuroblastoma
   d) Melanoma

5. **MC orbital tumor -** *(NEET Dec.12 Pattern)*
   a) Nerve sheath tumor
   b) Hemangioma
   c) Lymphoma
   d) Meningioma

6. **Most common orbital tumor has its origin from -**
   a) Blood vessels *(NEET Dec.12 Pattern)*
   b) Nerves
   c) Muscle
   d) Lymph node

7. **Most common carcinoma of conjunctiva -** *(NEET Dec.12 Pattern)*
   a) Squamous cell Ca
   b) Basal cell ca
   c) Melanoma
   d) Lymphoma

RETINOBLASTOMA

8. **Retinoblastoma can occur bilaterally in how many percentage of the cases?** *(All India Dec.15 Pattern)*
   a) 10 - 15%
   b) 15 - 20%
   c) 20 - 25%
   d) 25- 30%

9. **Knudson's two stage hypothesis is for -** *(Jipmer May 18)*
   a) Glaucoma
   b) Retinoblastoma
   c) Optic glioma
   d) Meningioma

10. **Children with germline retinoblastoma are more likely to develop other primary malignancies in their later lifetime course. Which of the following malignancies can occur in such patients?**
    a) Osteosarcoma of lower limbs *(AIIMS Nov 13)*
    b) Thyroid carcinoma
    c) Seminoma
    d) Renal cell carcinoma

11. **Retinoblastomas show all of the following except-** *(NEET Dec. 16 Pattern)*
    a) Small round cells
    b) Necrosis
    c) Pseudorosettes
    d) Fleurettes

12. **Intraocular calcification in eye in child -** *(DNB July 15 Pattern)*
    a) Toxocara
    b) Retinoblastoma
    c) Angiomatosis retinae
    d) Malignant melanoma of choroid

13. **Most common presenting feature of retinoblastoma-** *(All India Dec.13 Pattern)*
    a) White reflex
    b) Proptosis
    c) Photophobia
    d) Pain

14. **Increased LDH in Aqueous Humor suggest a diagnosis of -** *(AI 09)*
    a) Galactosemia
    b) Retinoblstoma
    c) Glaucoma
    d) Gyrate atrophy

15. **The most common mode of spread of Retinoblstoma is -**
    a) Hematogenous *(AIIMS May 15, AI 12)*
    b) Lymphatogenous
    c) Optic nerve
    d) Trans-scleral

16. **Most common site of bony metastases in retinoblastoma-** *(DNB June 17 Pattern)*
    a) Skull bones
    b) Hip bones
    c) Ribs
    d) Vertebrae

TREATMENT

17. **A one year old child having leucocoria was detected to be having a unilateral, large retinoblastoma filling half the globe. Current therapy would involve -** *(AI 03)*
    a) Enucleation
    b) Chemotherapy followed by local dyes
    c) Direct Laser ablation using photodynamic cryotherapy
    d) Scleral radiotherapy followed by chemotherapy

18. **Most common chemotherapeutic agents combination used for treatment of retinoblastoma -** *(NEET Jan. 19 Pattern)*
    a) Vinblastine, Carboplatin, Etoposide
    b) Vincristine, Carboplatin, Etoposide
    c) Vinblastine, Vincristine, Etoposide
    d) Vincristine, Cisplatin, Etoposide

19. **Ideal treatment of B/L retinoblastoma-** *(PGI June 04, Jipmer 02)*
    a) Enucleation
    b) Radiation
    c) Chemotherapy
    d) Cyclophotocoagulation

20. **In retinoblastoma, after enucleation, which tissue is sectioned to find out systemic metastasis -**
a) Central retinal artery *(AIIMS June 99)*
b) Sclera and episclera
c) Optic nerve
d) Vortex vein

21. **Treatment of metastatic disease in retinobastoma is -** *(All India Dec.14 Pattern)*
a) Chemotherapy
b) Enucleation
c) Radiotherapy
d) Cryo

22. **A 5 yr old boy presented with leukocoria in right eyeball diagnosed to be retinoblastoma involving full eyeball, while other eye had 2-3 small lesions in the periphery. What will be the ideal management for this patient?**
a) Enucleation of both eyes *(AI 11)*
b) Enucleation of right eye & conservative management of the other eye
c) Enucleation of right eye and focal therapy of the other eye
d) 6 cycles of chemotherapy

MISCELLANEOUS

23. **Most common site of distant metastasis in intraorbital malignant melanoma is -** *(All India Dec.13 Pattern)*
a) Brain
b) Lung
c) Liver
d) Lymph nodes

24. **Vortex Vein invasion is commonly seen in -** *(AI 03)*
a) Retinoblastoma
b) Malignant melanoma
c) Optic nerve gliomas
d) Medullo- epitheliomas

25. **Most common type of optic nerve glioma is-**
a) Gemistocytic *(All India Dec.13 Pattern)*
b) Fibrous
c) Protoplasmic
d) Pilocytic

26. **Optic glioma associated -** *(NEET Dec 16 Pattern)*
a) Neurofibromatosis 1
b) Neurofibromatosis 2
c) Sturge Weber syndrome
d) Von Hippel Lindau syndrome

■■■