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OCULAR DISEASE

TUMOURS OF THE UVEAL TRACT



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CLASSIFICATION

I. *TUMOURS OF CHOROID*

- a) Benign
 - 1. Naevus
 - 2. Haemangioma
 - 3. Melanocytoma
 - 4. Choroidal osteoma
- b) Malignant
 - 1. Melanoma

II. *TUMOURS OF CILIARY BODY*

- a) Benign
 - 1. Hyperplasia
 - 2. Benign cyst
 - 3. Meduloepithelioma
- b) Malignant
 - 1. Melanoma

III. *TUMOURS OF IRIS*

- a) Benign
 - 1. Naevus
 - 2. Benign cyst
 - 3. Naevoxanthoend-
othelioma
- b) Malignant
 - 1. Melanoma



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Malignant melanoma of choroid

- It is the most common primary intraocular tumour of adults, usually seen between 40-70 years of age.
- It is rare in blacks and comparatively more common in whites.
- It arises from the neural crest derived pigment cells of the uvea as a solitary tumour and is usually unilateral.



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Gross pathology-Pathology

1. ***Circumscribed (pedunculated) tumour.*** Initially it appears as flat, slate-grey area, which becomes raised and pigmented with growth and eventually ruptures through the Bruch's membrane. Further, growth of the tumour produces exudative retinal detachment.
2. ***Diffuse (flat) malignant melanoma:*** It spreads slowly throughout the uvea, without forming a tumour mass. It accounts for only 5 percent cases.



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Histopathology

- 1. *Spindle cell melanomas. These are composed of spindle-shaped cells and make up 45% of all tumours.*
- 2. *Epithelioid cell melanomas. These consist of large, oval or round, pleomorphic cells with larger nuclei and abundant acidophilic cytoplasm.*
- 3. *Mixed cell melanomas. These are composed of both spindle and epithelioid cells and thus carry an intermediate prognosis.*
- 4. *Necrotic melanomas. These make up the remaining 5% of the all tumours.*



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Clinical picture

- **1. Quiescent stage.** *During this stage symptoms depend upon the location and size of tumour.*
- Small tumour located in the periphery may not produce any symptom, while tumours arising from the posterior pole present with early visual loss.
- **Signs:**
 - A small tumour limited to the choroid;** *appears as an elevated pigmented oval mass*



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Fundus photograph showing choroidal melanoma as raised pigmented subretinal mass.





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ii. A large tumour which penetrates through the Bruch's membrane and grows in the subretinal space is characterised by a large exudative retinal detachment.

2. *Glaucomatous stage.* *It develops when tumour is left untreated during the quiescent stage. Glaucoma may develop due to obstruction to the venous outflow by pressure on the vortex veins,*



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Malignant melanoma of the choroid causing exudative retinal detachment : A; Diagrammatic depiction in cut section ; B, Fundus photograph.

A



B





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- **Symptoms:** *The patient complains of severe pain, redness and watering in an already blind eye.*
- **Signs.**
 - Conjunctiva is chemosed and congested.*
 - Cornea may show oedema.*
 - Anterior chamber is usually shallow.*
 - Pupil is fixed and dilated.*
 - Lens is usually opaque, obstructing the back view.*
 - Intraocular pressure is raised, usually eye is stony hard*



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- 3. Stage of extraocular extension.** Due to progressive growth the tumour may burst through sclera, usually at the limbus.
- It is followed by rapid fungation and involvement of extraocular tissues resulting in marked proptosis



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Extensive malignant melanoma of the choroid involving orbit.





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- **4. *Stage of distant metastasis.***
Lymphatic spread is usually not known.
- Blood-borne metastasis usually occurs in the liver and is the commonest cause of death.



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Differential diagnosis

1. During quiescent stage

- i. A small tumour without an overlying exudative retinal detachment should be differentiated from a naevus, melanocytoma and hyperplasia of the pigment epithelium.
- ii. A tumour with overlying exudative retinal detachment should be differentiated from simple retinal detachment

2. During glaucomatous stage

differentiation is to be made from other causes of acute glaucoma.



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Investigations

- 1. Indirect ophthalmoscopic examination.*
- 2. Ultrasonography: Both A and B scan*
- 3. Fluorescein angiography*



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Treatment

1. **Conservative treatment to salvage the eyeball** should be tried unless the tumour is very large.
 - i. **Brachytherapy** is usually the treatment of choice in tumours less than 10mm in elevation and less than 20 mm in basal diameter.
 - ii. **External beam radiotherapy** with protons or helium ions is indicated in tumours unsuitable for brachytherapy either because of size or posterior location to within 4mm of disc.
 - iii. **Transpupillary thermotherapy (TTT)** with diode laser is indicated in selected small tumours, particularly if pigmented and located near the fovea or optic disc.



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2. ***Enucleation.*** It is indicated for for very larger tumours in which conservative methods to salvage the eyeball are not effective.
3. ***Exenteration with chemotherapy and radiotherapy*** is required in the stage of extraocular spread.
- 4. ***Palliative treatment with chemotherapy and immunotherapy*** may be of some use in prolonging life of the patients with distant metastasis.



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Malignant melanoma



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Introduction

- In the ciliary body it is usually diagnosed very late, due to its hidden location. It may extend anteriorly, posteriorly or grow circumferentially.



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Clinical features

1. ***Earliest features of a localised melanoma include*** slight hypotony, unaccountable defective vision and localised 'sentinel' dilated episcleral veins in the quadrant containing tumour.
2. ***An anterior spreading tumour:***
 - i. It may cause pressure on the lens resulting in anterior displacement, subluxation and cataract formation.
 - ii. It may involve iris and is visible immediately. Soon it may involve the angle of anterior chamber resulting in secondary glaucoma.
 - iii. It may extend out through sclera along the vessels, presenting as an epibulbar mass.



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- 3. *Posterior spreading tumour may involve choroid and present as exudative retinal detachment.*
- 4. *The tumour may extend circumferentially involving whole of the ciliary body.*



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Treatment

- 1. Enucleation.*** *It is required for large ciliary body tumours extending anteriorly, posteriorly or circumferentially.*
- 2. Local resection.*** *Cyclectomy or iridocyclectomy may be enough, if fortunately tumour is detected in early stage.*



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Malignant melanoma



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Introduction

- It presents as a single or multiple rapidly growing vascular nodules.
- It spreads in the angle producing secondary glaucoma.
- It may penetrate through limbus and present as epibulbar mass.
- Pathological features are similar to that of melanoma of the choroid.



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Treatment

- 1. *Wide iridectomy.*** *It is performed for a tumour limited to the iris.*
- 2. *Iridocyclectomy.*** *It is required for a tumour involving iris and ciliary body.*
- 3. *Enucleation.*** *It should be performed when iris melanoma is associated with secondary glaucoma.*