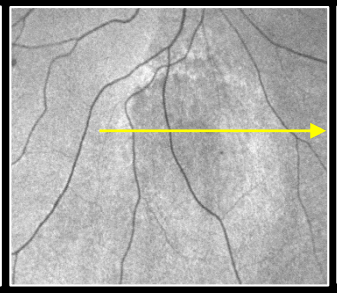
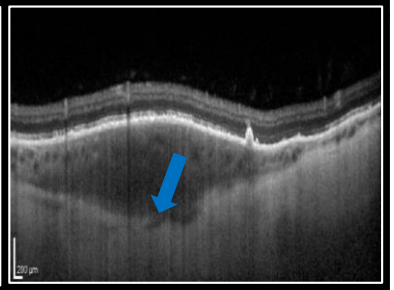
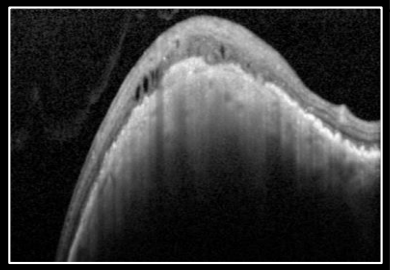
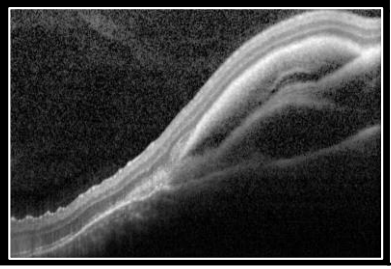
Centre for Eye Health

# CHAIR-SIDE REFERENCE: HYPO-PIGMENTED POSTERIOR LESIONS

## LESIONS OF THE CHOROID

| Optomap /retinal photo | Fundus Autofluorescence | Optical coherence tomography | Description |
|---|---|---|---|
| **Amelanotic choroidal naevus** |  |  | • Common, benign lesion with up to a 1% lifetime risk of malignant transformation.<br>• **Fundus appearance**: Up to 10% of naevi are de-pigmented. Appear as a flat lesion with detectable borders, round/oval in shape. Typically <2mm thick and <5mm in diameter, located posterior to the equator.<br>• **FAF:** appearance variable. Naevi have low intrinsic AF, however associated changes such as RPE hyperplasia (hypo-AF) impact the lesion's FAF properties.<br>• **OCT**: Flat/slightly elevated lesion with homogenous pattern of medium reflectivity without posterior shadowing. Choriocapillaris thinning overlying the naevus. Posterior margin (arrow) may be delineated.<br>• **Associated signs**: drusen, subretinal fluid (SRF), photoreceptor loss or attenuation, RPE atrophy/hyperplasia, RPE detachment and/or an RPE trough.<br><br>*Documentation and routine review required.* |
| **Amelanotic choroidal melanoma** | *FAF not available* |  | • 15% of choroidal melanomas may be non-pigmented and 30% mixed.<br>• **Fundus appearance**: Solitary raised hypo-pigmented mass > 2mm thick, often with associated sub-retinal fluid (SRF) and lipofuscin (orange pigment).<br>• **FAF**: Lipofuscin appears hyper-AF while RPE changes can appear hypo-AF.<br>• **OCT:** SRF is a significant risk-factor for malignancy (Shields et al.)<br>• **B-scan**: Typically mushroom/dome-shaped, indicating extension through Bruch's membrane. The lesion is acoustically hollow.<br>• **Associated signs:** SRF, sub-retinal haemorrhage, sentinel vessels, choroidal folds, retinal detachment or inflammation.<br><br>*Prompt referral to an Ophthalmologist is required.* |
| **Choroidal metastasis** | *FAF not available* |  | • Primary lesion elsewhere in the body (commonly lungs or breast).<br>• **Fundus appearance**: Ill-defined, mildly elevated, yellow lesions often with overlying pigmentary changes. Multifocal and/or bilateral in 25% of cases.<br>• **OCT**: Irregular (lumpy-bumpy) anterior tumour surface, photoreceptor elongation, presence of subretinal fluid often with "speckles" visible within the area of fluid; and thinning of the overlying choriocapillaris.<br>• **B-scan**: flat or slightly raised lesion typically with inhomogeneous reflectivity.<br><br>*Prompt referral to an Ophthalmologist is required.* |

A systematic approach to assessment of choroidal lesions involving careful consideration of its imaging characteristics may be helpful when forming a diagnosis. A useful mnemonic was developed by Shields and associates to explore risk factors for choroidal naevus transformation into melanoma. https://pubmed.ncbi.nlm.nih.gov/30844944/

***T**o **F**ind **S**mall **O**cular **M**elanoma **D**oing **Im**aging*

- Thickness>2mm
- Subretinal Fluid
- Symptoms
- Orange pigment (lipofuscin)
- Melanoma hollow with ultrasound
- Diameter greater than 5mm

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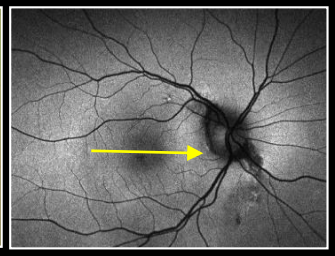
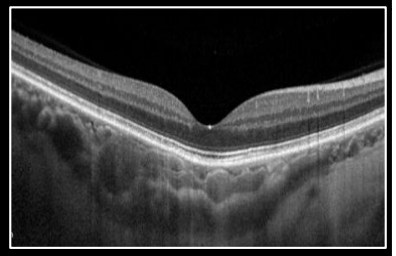
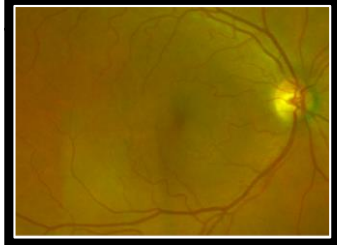
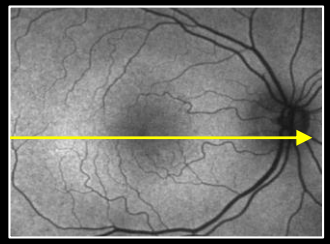
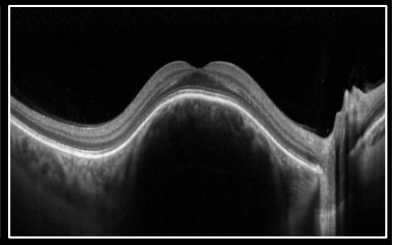
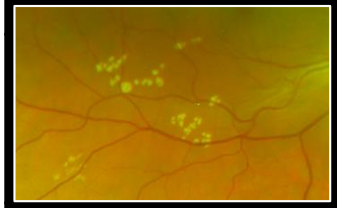
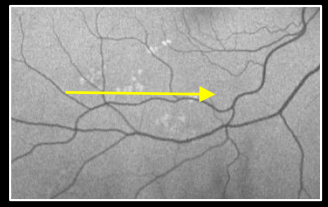
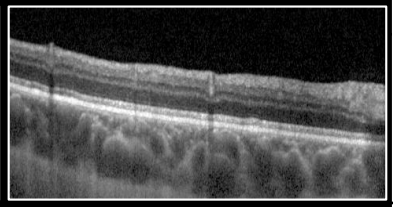
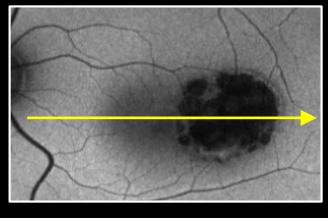
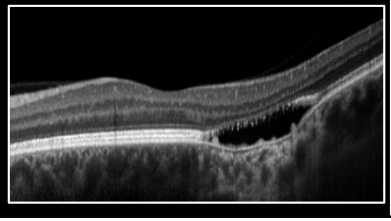
| **Choroidal osteoma** | | | • Rare benign ossifying tumour of the choroid.<br>• **Fundus appearance:** Irregular shape, slightly elevated, typically with a fine superficial vascular network. Over time, colour changes from yellow-orange to yellow-white. Typically unilateral in the juxtapapillary or macular areas.<br>• **FAF**: Appearance varies, depending on level of calcification, retinal atrophy and potential complications such as SRF and CNV.<br>• **OCT:** Change in choroidal architecture. Inner and outer retinal layers unaffected.<br>• **B-scan**: Characteristically strong acoustic shadow. May be required for diagnosis.<br>• **Complications**: RPE disruption, photoreceptor atrophy, SRF and CNV.<br><br>*Documentation and routine review.* |
|  |  |  | |
| **Choroidal haemangioma** | | | • Benign vascular tumours characterised by collections of vessels of different sizes within the choroid.<br>• **Fundus appearance:** *Circumscribed choroidal haemangioma (CCH)* is usually unilateral, solitary and not associated with systemic disease. Red-orange and round in shape, surrounded by a ring of slight pigmentation.<br>• *Diffuse choroidal haemangioma (DCH)* is associated with Sturge-Weber syndrome. Typically involve more than half the choroid and have poorly-defined borders.<br>• **OCT**: RPE and outer retinal layers overlying the lesion may be affected.<br>• **Complications:** May be associated with SRF or chronic macular oedema.<br><br>*Documentation and routine review with referral of SRF or macular oedema* |
|  |  |  | |
| **LESIONS OF THE RPE** | | | |
| **Grouped Congenital Albinotic Spots (Polar Bear Tracks)** | | | • **Fundus appearance**: Multiple small, flat, discrete, white lesions typically clustered in a single quadrant. Lesions commonly increase in size towards the periphery.<br>• **FAF**: May show hyper-AF (indicating RPE dysfunction) or hypo-AF (indicating RPE atrophy).<br>• **OCT:** Lesions lie at the level of the RPE. May note attenuation of the ellipsoid zone.<br><br>*Documentation and routine review.* |
|  |  |  | |
| **Torpedo maculopathy** | | | • **Fundus appearance**: A congenital, solitary spindle-shaped chorioretinal lesion typically located temporal to the fovea.<br>• **FAF**: Typically shows hypo-AF, sometimes with hyper-AF along lesion margins<br>• **OCT**: Lesions are either flat or excavated. Associated neurosensory detachment, photoreceptor loss, retinal thinning and RPE changes - atrophy or hyper-reflectivity. Inner retinal layers are typically unaffected.<br><br>*Documentation and routine review required.* |
|  |  |  | |

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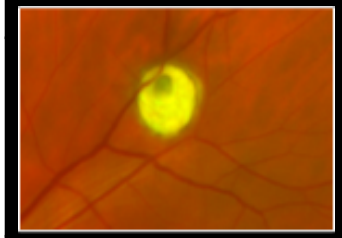
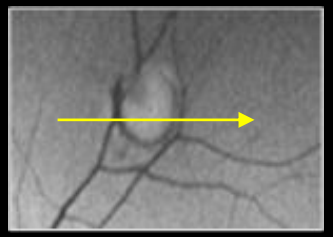
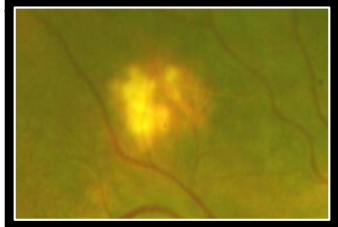
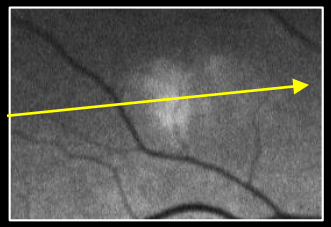
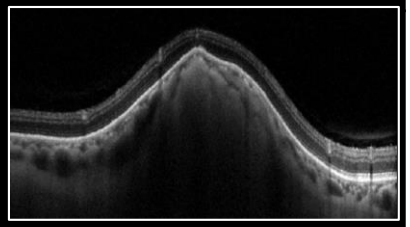
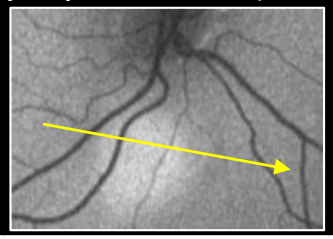
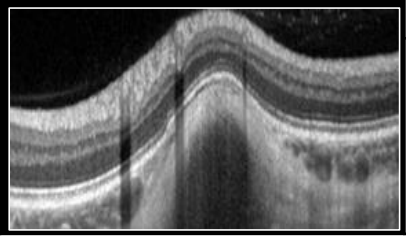
## LESIONS OF THE RETINA

| *Optomap /retinal photo* | Fundus Autofluorescence | Optical coherence tomography | Description |
|---|---|---|---|
| **Retinal Astrocytic Hamartoma (Retinal Astrocytoma)** | | | • Benign tumours arising from the glial cells of the retinal nerve fibre layer.<br>• **Fundus appearance:** Globular white elevated lesion with intrinsic blood vessels. Early semi-translucency increasing in calcification over time. Minimal growth.<br>• **OCT:** Optically empty adjacent cystic intra-retinal spaces may be seen.<br>• **FAF**: Calcification of the tumour is associated with hyper-AF.<br>• **Systemic associations:** Most commonly associated with neurofibromatosis type 1 or tuberous sclerosis but may occasionally be an isolated finding.<br><br>*Documentation and routine review. Refer lesions with evidence of progression* |
|  |  |  | |
| **Primary Intraocular Lymphoma (PIOL)** | | | • An extra-nodal non-Hodgkin, diffuse large B cell lymphoma with high morbidity.<br>• **Fundus appearance**: Flat creamy orange-yellow mass that may be single or multiple and usually associated with vitritis.<br>• **OCT:** Mass located deep in the sensory retina.<br>• **Systemic associations**: Between 56-80% of cases of PIOL subsequently develop brain lymphoma.<br><br>*Prompt referral to an ocular oncologist is required.* |
|  Image Courtesy of Dr N.Assaad | FAF not available | OCT not available | |

## LESIONS OF THE SCLERA

| | | | |
|---|---|---|---|
| **Sclero-choroidal calcification** | | | • Calcium deposition at the level of the sclera and choroid.<br>• **Fundus appearance:** Multiple discrete yellow placoid lesions typically found in the superotemporal post-equatorial retina.<br>• **OCT:** 'Mountain-like' scleral elevations including flat, rolling, rocky-rolling and table profiles. Compression of the overlying choroid and RPE abnormalities.<br>• **FAF**: Lesions can show iso- or hyper- autofluorescence.<br>• **Systemic associations**: Conditions altering calcium and phosphate metabolism.<br><br>*Documentation and routine review. Refer to GP for appropriate screening* |
|  |  |  | |
| **Focal scleral nodule (Solitary idiopathic choroiditis)** | | | • A rare, benign scleral lesion of unknown aetiology showing minimal growth.<br>• **Fundus appearance:** Discrete, yellow-white scleral lesion with surrounding orange halo. Active lesions have ill-defined margins, sub-retinal fluid and yellow intra-retinal exudative material. Focal haemorrhages may also be present.<br>• **FAF:** Typically show hyper-AF which may be speckled if the RPE is disturbed.<br>• **OCT:** Smooth and nodular or dome-shaped lesion with thinning of the overlying choroid. May be associated with SRF.<br><br>*Routine review of inactive lesions, refer active lesions to an Ophthalmologist.* |
|  |  |  | |