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Differentiating sight threatening from non-sight threatening disease: Retinal detachment and macular disorders

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Anatomical and functional continuity between the macula and other areas of the retina means that differentiation between sight-threatening and non-sight threatening disease often requires attention to both. Some diseases arise, and remain limited exclusively, to the macular area, such as age-related macular degeneration (AMD), others originate elsewhere in the retina but may involve the macula, for example retinal detachment (RD) and branch retinal vein occlusion. To aid understanding, RD and macular disorders will be covered separately in this article.

Retinal detachment

The retina is made up of several layers of neural elements, but for the purpose of understanding RD, two main layers to consider are the retinal pigment epithelium (RPE) and the neural retina (NR). It is essentially the separation between these two layers that causes a RD, while retinal tears occur in the NR.

Normal peripheral retina and benign degenerations

The ophthalmoscopic appearance of the peripheral retina often varies.¹ It is of critical importance to distinguish normal as opposed to pathological variations. The following more common lesions are not considered directly to confer an added risk of RD but require differentiation from true retinal breaks or detachments and also from more serious retinal degenerations which can predispose to RD.

Meridional folds

These are radial retinal folds which extend backwards from the ora serrata and are present in 20% of normal eyes. They may be mistaken for retinal tears and are more common in the upper nasal quadrant.

Oral pearls

Oral pearls usually form over dentate processes, varying from pinpoint to pinhead size. They are more common in elderly people. Oral pearls may be confused with tiny retinal tears during examination by indentation.

Pars plana cysts

The ora serrata often has small cystoid cavities which are normal. They extend towards

the pars plana and may extend along the entire length of the ora particularly in myopic eyes.

Retinal tufts

Retinal tufts are tiny projections into the vitreous base. They are innocent lesions but, when associated with cystic degeneration of the underlying retina, they may lead to retinal tear formation. They are more common in the nasal retina and it can be difficult to differentiate them from tiny retinal tears particularly during indentation examination. They do not need prophylaxis or referral to an ophthalmologist.

Paving stone degeneration (Fig. 1)

Paving stone degeneration is seen as a well circumscribed round pale or yellow flat area, which may be as large as two disc diameters. The lesions are usually grouped or confluent, and most frequently occur in the lower retina between the equator and ora serrata. Because of RPE atrophy, choroidal vessels are often visible through the degeneration as red lines. The lesions are usually surrounded by dark pigmentation. No prophylaxis or referral is required for paving stone degeneration.

Snowflake degeneration (Fig. 2)

As the name implies, snowflake degeneration appears as white areas which are not well delineated. They are usually found on the lower retina and they should be distinguished from lattice degeneration.

Dialysis

Dialyses are peripheral, usually well localised and solitary areas of large retinal disinsertions from the ora serrata. They are almost

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Module 9 Part 8 Differential Diagnosis of Ocular Disease

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always due to blunt trauma, but a clear history of trauma may not always be available. The examiner should look for signs such as phacodonesis (lens wobbling), iridodonesis, high intra-ocular pressure (IOP), iris transillumination and macular hole. These patients are usually young and are often diagnosed during a routine eye test. Dialysis most commonly involves the lower temporal quadrant of the retina. The presence of a water mark (a pigment line) indicates the long-standing nature of such detachments. Most cases require cryotherapy and a buckle procedure to reattach the retina. The treatment is usually non-urgent as this condition is chronic.

Retinoschisis

Retinoschisis is essentially a split in the NR, typically occurring in the lower temporal quadrant of the peripheral retina. Most retinoschisis are innocent lesions, usually picked up on routine examination and misdiagnosed as RD. Over time, large outer layer holes with rolled edges may appear, thus allowing fluid in the schisis cavity to gain access to the subretinal space, turning a retinoschisis into a RD.

Features and management of degenerations predisposing to RD

Lattice degeneration (Fig. 3)

Lattice degeneration is of much greater clinical significance than the lesions described above. While only 7-8% of normal adults show lattice degeneration, as many as 40% of high myopes (>5 dioptres) may have the condition.

Lattice degeneration appears as linear areas of retinal thinning with criss-cross white lines. Most lesions affect the retina between the vitreous base and the equator with the superior location being most commonly affected. For some reason, the horizontal meridian appears to be the least favoured site for lattice degeneration.

There is a higher risk (5-10%) of RD in people with lattice degeneration in the second, third and fourth decades of life, with no convincing evidence in the literature to indicate that prophylactic treatment reduces the risk of RD in eyes with lattice (or for that matter any) peripheral retinal degeneration.²

U-tears (Fig. 4)

Peripheral retinal tears of U-shaped configuration are almost always due to vitreoretinal traction. Presence of brown retinal pigment cells in the vitreous (referred to as tobacco dust or Shaffer's sign) is almost diagnostic of a retinal tear. Regardless of their association with a retinal degeneration, all U-tears should be considered for laser or cryotherapy treatment. An urgent referral to a local ophthalmologist, or ideally a vitreoretinal specialist, should be made.

U-tears involving the upper half of the retina require more urgent treatment because

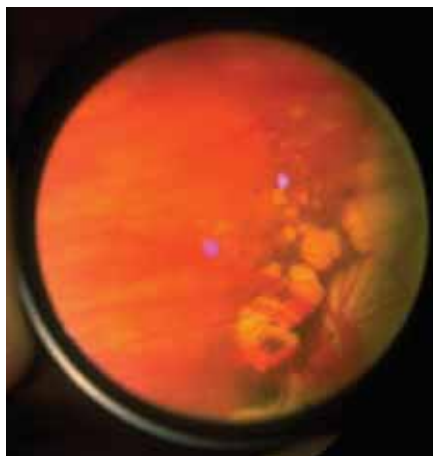


Fig 1: Paving stone degeneration

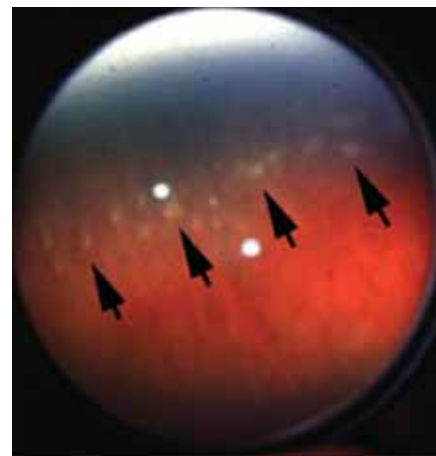


Fig 2: Snowflake degeneration

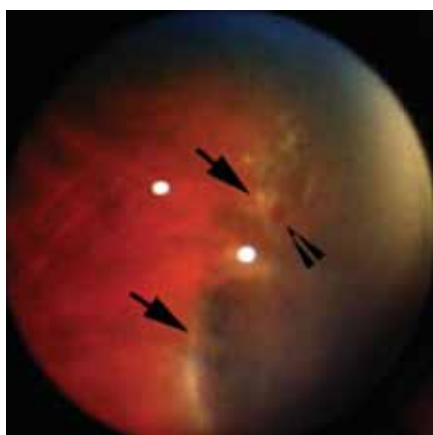


Fig 3: Lattice degeneration with round holes

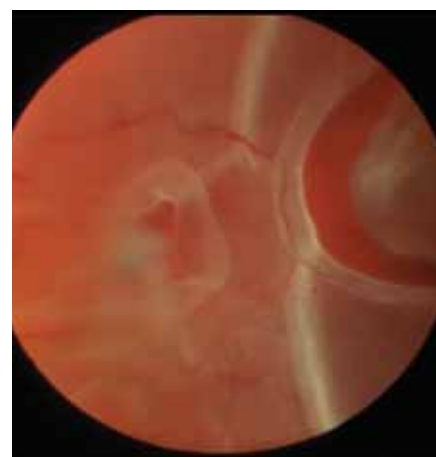


Fig 4: U-tear with retinal detachment

there is a higher risk of RD owing to gravity and eventual macular involvement. In the meantime, patients should be advised to limit their physical activity and watch out for RD symptoms. High-risk patients with a U-tear should ideally be treated or seen within 24 hours.

Snail track degeneration

As the name implies, lesions of snail track degeneration are broad linear areas that may appear to be slightly raised because of vitreous condensation. They are orientated along the posterior border of the vitreous base.

Choroidal coloboma

Choroidal coloboma is a congenital defect along the embryological lines of closure. This rare condition may be associated with a coloboma of the iris (oblong pupil).

Usually, a large excavation is visible on the lower retina; this may extend as far back as the optic nerve. White sclera is clearly visible through the defect thanks to the lack of normal choroid and atrophy of the overlying retina. Rarely, retinal tears develop along the edge of coloboma, leading to a RD.

Giant retinal tear (GRT)

This is probably one of the most dreaded

conditions in vitreoretinal surgery.

A break extending beyond two consecutive clock hours is classified as a giant retinal tear. Treatment can be quite challenging and usually involves a combination of vitrectomy, silicone oil and a scleral explant. Patients merit urgent referral and bed rest with advice on posture.

Chorioretinal scars

Any focus of infection or inflammation involving the choroid and the retina eventually leads to scarring, which looks more or less like a cryotherapy scar. The most common scars seen in clinical practice result from ocular toxoplasmosis; retinal tears can occasionally develop from contraction of the scar tissue. Prophylaxis is usually not required for chorioretinal scars, but some severe inflammations like acute retinal necrosis (ARN) may be associated with a high risk of RD.

Cryotherapy

A cryotherapy scar has a mottled black and white appearance because of mixed atrophy and hypertrophy of the RPE. Such scars are well known to develop new breaks which typically occur at the edges. Such patients should be referred preferably to a vitreoretinal surgeon, for assessment.

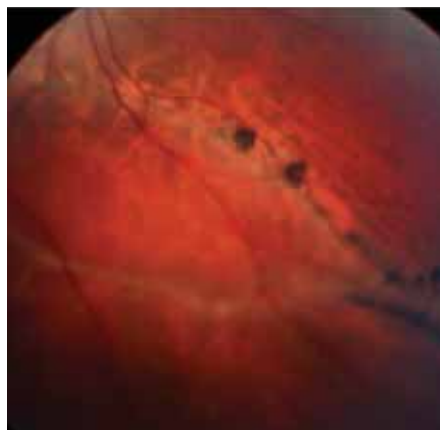


Fig 5: Demarcation line separating chronic RD below from normal retina above



Fig 6: Retinoschisis. Arrows marking the upper border.

Retinal detachment - features and management

If the retina can be visualised well, the diagnosis of RD made on the basis of clinical examination is not difficult in most cases. While the majority of RDs are due to the presence of one or more retinal breaks, certain diseases of the retina and choroid may give rise to a RD by other mechanisms. It is of paramount importance to distinguish such conditions from a rhegmatogenous retinal detachment (RRD), since there is a distinct difference in their management.

Rhegmatogenous retinal detachment

Most RDs seen in day to day clinical practice are rhegmatogenous in nature. In fact, almost all detachments are presumed to be secondary to one or more breaks, unless proved otherwise. However, it is common for a retinal break to be undetectable although the configuration of a RD usually helps in localising the break.³ A bullous RD has a causative break almost always above the horizontal meridian. In a detachment involving the lower part of the retina, one should look for a break on the higher side of the detachment.

In a total RD, the localising value of the RD configuration is usually lost but a careful

history may help. The examiner should look for a break opposite to the meridian where the visual field defect was first noted by the patient. Most cases of failure of RD surgery in the immediate post-operative period are due to missed breaks. Therefore, a thorough examination is mandatory prior to contemplating surgery. There is a definite association between cataract surgery and RD, as it occurs in up to 1% of pseudophakic eyes.⁴

The retina is usually fairly mobile in acute RRD. Presence of pigment (tobacco dust) in the vitreous is universal, and a PVD is present in most cases presenting with a RD due to a U-tear. Involvement of the macula carries a poor prognosis for vision. If the macula is still unaffected, urgent referral to a vitreoretinal surgeon should be arranged. The patient should be advised to limit his or her physical activity if possible. While waiting admission for surgery, they should be encouraged to assume a posture so that the RD is in the dependent position. For example, if there is a temporal detachment in the right eye, lying in bed with the right cheek resting on the pillow would help to minimise the spread of subretinal fluid (SRF). Likewise, in lower RDs an upright posture should be adequate. Such cases are less urgent than upper half detachments.

Chronic retinal detachment (Fig. 5)

A RRD acquires certain characteristics when left untreated for a period of time. Such cases usually arrive at diagnosis on routine examination or present with a long history of poor vision. Chronic RDs are usually found in the lower part of the retina. The retina shows thinning and cystic changes and is usually not very mobile. Wrinkling of the retina, membrane formation, rolled edges of the breaks and presence of pigment clumps in the vitreous suggest the onset of proliferative vitreoretinopathy which adversely affects the outcome of surgery. The fellow eye should be evaluated for prophylactic treatment. Such cases should be referred to a retinal surgeon on a non-urgent basis.

Retinoschisis (Fig. 6)

Retinoschisis is normally not a precursor to RD unless associated with both inner layer and outer layer breaks. According to long term follow up studies, only 2.5% of retinoschises eventually develop into RD.⁵ However, it can be quite difficult at times to distinguish a retinoschisis from RD, particularly a chronic RD in which the retina is usually thin. The presence of a pigment demarcation line is not a feature of schisis, but is commonly seen in a chronic RD.

Tractional retinal detachment

Contracture of the fibrovascular tissue in vasoproliferative conditions like advanced diabetic retinopathy and retinal vein occlusions may pull the retina off, giving rise to tractional retinal detachment (TRD). Such

detachments have a concave configuration and seldom progress rapidly. Surgical treatment is usually considered when the macula is involved or threatened. Occasionally, such detachments are complicated by formation of retinal breaks. Such combined rhegmatogenous and tractional detachments demonstrate signs of rapid progress and acquire a convex appearance. Generally, surgical intervention is considered necessary at this stage.

Solid detachments (Figs. 7 and 8)

Ocular tumours, like malignant melanoma of the choroid or choroidal metastases from a distant tumour, can sometimes present a puzzling clinical picture.⁶ Separation of retinal blood vessels from the underlying choroidal vessels should be used as a guide to distinguish a solid RD from subretinal fluid. A solid lesion is usually visible under the detached retina as an elevated area within the subretinal fluid. Occasionally, a choroidal mass pushes the retina towards the vitreous while the NR remains attached to the RPE. In other words, there is no obvious subretinal fluid. An ultrasound scan is an invaluable tool in differentiating a solid RD from one caused by SRF, and a detailed history is very important; a successfully treated breast cancer may present with secondaries in the eye years later.

Serous retinal detachment

A wide variety of local and systemic pathology can cause subretinal fluid accumulation. Absence of retinal breaks, lack of vitreous pigmentation and a smooth configuration are salient features of such detachments, which may be localised to the affected area of the retina or may be more generalised. The fluid typically tends to shift with a change in head position. Fundus fluorescein angiography (FFA) will demonstrate extensive leakage, while an ultrasound helps to exclude a solid lesion such as a tumour. Important underlying causes include a choroidal tumour, Coat's disease, central serous retinopathy (CSR), Vogt-Koyanagi-Harada Syndrome (VKHS), posterior scleritis and age related macular degeneration (AMD).

Choroidal detachment

Serous or haemorrhagic fluid in the supra-choroidal fluid can lead to choroidal detachment. A total choroidal detachment usually appears as four bullous elevations occupying the superior, inferior, nasal and temporal quadrants of the fundus. These detachments may be differentiated from RD by the absence of retinal breaks and pigment in the vitreous. In addition, they lack the mobility of a RD.

Serous detachments have a darker appearance than the rest of the fundus and haemorrhagic detachments appear very dark in colour. Serous fluid accumulation is usually seen in the postoperative setting where there is excessive hypotony, for

example a trabeculectomy which drains excessively.

Haemorrhage in the suprachoroidal space can occur spontaneously, during intraocular surgery, as a result of trauma or in association with intraocular vascular anomalies, such as choroidal haemangioma. Warfarin therapy is also a recognised cause of this condition. Choroidal haemorrhage is usually an acutely painful event and can be potentially blinding because of an enormous and sudden rise in intraocular pressure (IOP). In the operative setting, it can cause extrusion of all intraocular contents. Treatment is aimed at immediate reduction of IOP. A non-resolving choroidal haemorrhage sometimes requires surgical drainage and even vitrectomy.

Vitreous haemorrhage (VH)

Diagnosis of RD can be difficult in the presence of a dense cataract or vitreous haemorrhage. Some useful clues should help towards a reasonable judgement in such cases. A relative afferent pupillary defect (RAPD) is not a feature of VH, but when present, indicates a major retinal dysfunction like a central retinal vein occlusion or a major RD. An ultrasound B-scan is invaluable in making a diagnosis of RD. In a patient with a known history of diabetic retinopathy or retinal vein occlusion, a spontaneous vitreous haemorrhage is often due to neovascularisation. Often the treatment is conservative. In a high myope, on the other hand, a vitreous haemorrhage is usually secondary to a retinal tear or RD, hence should be considered sight threatening, unless proved otherwise. A vitrectomy is usually performed early to treat the underlying tear or RD. Vitreoretinal expertise may be required to make a distinction between the ultrasonographic appearance of a PVD, RD or a choroidal detachment.

The macula

Anatomy of the macula and its relevance to the disease

The term macula refers to the area of the retina confined within the temporal vascular arcades, about 5.5µm in diameter. The centre of the retina is called the fovea, with a central excavation termed the foveola. The precise centre of the foveola which has the highest visual acuity is called the umbo. For clinical purposes, the fovea is the most common term for the very centre of the macula. It measures about 1.5µm in diameter. In the macula, the ganglion cell layer is several cells thick, whereas elsewhere it is only one cell thick. The avascular centre of the fovea is surrounded by the vascular arcades which form a ring about 400µm in diameter.

Presentation of macular disease

The most common manifest symptom of any macular disease is loss of central vision, best judged by testing near vision. Distortion of vision, metamorphopsia, central or paracentral scotoma are other symptoms which



Fig 7: Malignant melanoma

the examiner should try to elicit. Binocular visualisation using a slit lamp and a condensing lens is paramount in the evaluation of macular lesions; it is quite possible to miss the diagnosis of macular oedema if a high resolution lens is not used for examination. The salient clinical features of individual macular conditions are described below.

Macular haemorrhage

The diagnosis of a macular haemorrhage is usually straightforward although the aetiology and hence the management can vary.

Choroidal neovascularisation (CNV)

Any condition leading to retinal scarring and a breach in Bruch's membrane can result in CNV. This usually presents with a sudden drop in visual acuity owing to macular haemorrhage. AMD is the most common cause in those over the age of 60.⁷ In younger patients, CNV may be due to myopia, as a result of retinal scarring, toxoplasmosis or ocular trauma leading to a choroidal rupture or idiopathic. An urgent FFA is mandatory in all cases particularly if the vision is still better than 6/60, as photodynamic therapy (PDT) may be considered.

Macroaneurysms

Macroaneurysms tend to occur in elderly people, mostly females. An association with systemic hypertension is observed in about 79% of cases.⁸ Macroaneurysms can cause retinal bleeding, exudation, macular oedema and even vitreous haemorrhage. Laser treatment is indicated only if there is sight-threatening leakage or a non-resolving vitreous haemorrhage. Systemic hypertension should be managed as appropriate.

Myopic maculopathy

Sometimes tiny splinter shaped macular haemorrhages appear in high myopes. They are believed to result from a split in Bruch's membrane and are called 'lacquer cracks'. CNV may grow through these

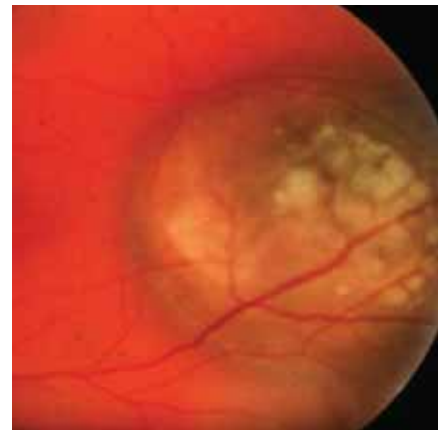


Fig 8: Malignant melanoma with retinal detachment

lacquer cracks and produce an abrupt macular haemorrhage.

Macular deposits

Most macular dystrophies start with some form of material deposition. They all share a common end-stage appearance of macular scarring which may be confused with AMD. Haemorrhage is not a feature of these conditions except when it is due to CNV formation in the scar.

Drusen

Drusen are lipofuscin deposits in Bruch's membrane. They can take various forms: well defined, with distinct borders, 'hard' drusen; or more diffuse or 'soft' drusen which may become confluent. It is this latter variety, particularly the confluent form, which carries the highest risk of exudative AMD.

In most individuals, drusen develop with advancing age. However, the autosomal dominant form of drusen appears in younger people; such cases have an increased risk of developing wet AMD later in life. The Macular Photocoagulation Study reported that the five-year risk of CNV in fellow eyes of individuals with unilateral neovascular AMD was 10% in people without large drusen and 30-46% in those with large drusen.⁹ Such patients can be managed in an optometry practice; they should be taught to monitor their vision. Any sign of haemorrhage or sudden distortion of vision should warrant urgent referral to an ophthalmologist with expertise in PDT.

According to a major study, the use of antioxidants and zinc reduced the risk of progression to 'wet' AMD in such cases from 28% in the placebo group to 20% in the treated group. However, the use of these agents is contraindicated in smokers.

Best's disease (Fig. 9)

This hereditary condition, also known as vitelliform dystrophy, is rare and appears during childhood. It is typified by a large yolk-like lesion which is usually bilateral and symmetrical in the centre of the macula. With age, the lesion breaks down, leaving

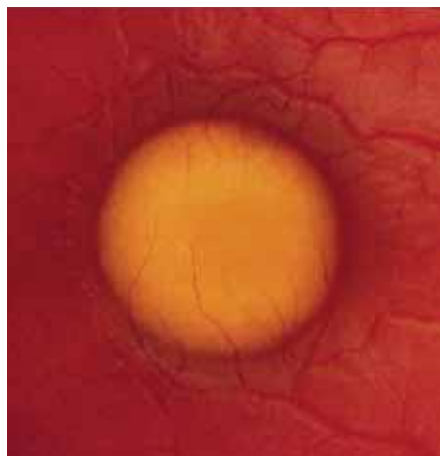


Fig 9: Best's disease

an atrophic area which may give rise to CNV formation. The diagnosis is confirmed by an absent response on the electro-oculogram (EOG) and a relatively normal ERG. Vision usually remains better than 6/60. As with any macular scar, these patients should be warned of the possibility of CNV, and genetic counselling should be offered.

Adult vitelliform foveomacular dystrophy

This adult variety of Best's disease is rather more important because of its resemblance to AMD. The lesions are smaller than those of the childhood form and usually appear in the fourth to sixth decades of life. Visual symptoms are often limited to mild blurring and metamorphopsia. A follow-up is not mandatory, but these cases should be referred to exclude conditions which mimic adult Best's disease.

Stargardt's disease (Figs. 10 and 11)

This hereditary condition presents early in life due to abnormal deposition at the level of the RPE. The characteristic flecks are seen in the macular area; they later coalesce to form large geographic areas of atrophy.

Exudates

Retinal exudates appear as sharply demarcated irregular areas. They may be scattered or clustered usually around a source of vascular leakage. They have a pale-white colour and are often associated with macular oedema. They usually disperse after successful laser treatment but sometimes persist indefinitely.

Exudates are the hallmark of leakage due from a vascular cause such as diabetic retinopathy (DR), CNV, macroaneurysms, retinal telangiectasia, Coat's disease, vascular tumours, severe acute hypertensive retinopathy and acute papilloedema.

Abnormal pigmentation Pattern dystrophy

This dystrophy can acquire various forms and shapes, for example 'butterfly dystrophy'. Common to all types is pigment

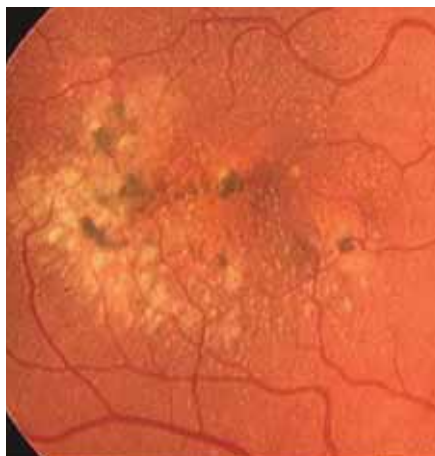


Fig 10: Stargardt's disease

disturbance in the central macular area. Visual symptoms are minimal and the prognosis for vision is usually excellent. Only a small number of cases develop extensive atrophy. There is no confirmatory test. The diagnosis is based on clinical assessment and somewhat subnormal electrophysiology test results.

Central areolar choroidal dystrophy (Fig. 12)

The affected patients usually present in their third or fourth decade of life with central visual loss. This condition reveals non-specific hyperpigmentation of the macula which later develops into an area of geographical atrophy of the RPE and scarring, usually resulting in severe visual loss. A few lucky individuals retain 6/6 vision thanks to sparing of the fovea.

Cone dystrophy

Cone dystrophy manifests during the first or second decades of life. The hallmark of cone dystrophy is progressive loss of visual acuity with deterioration of colour vision and photophobia. Clinical appearance may be highly variable, from a subtle diffuse macular granularity to a well demarcated circular atrophic area involving the macula. Vision can vary from 6/6 to hand movements.

Drugs

Drug-related maculopathies are rare. The most common recognised agents are chloroquine, tamoxifen, niacin and canthaxanthine (a skin tanning agent). They share a common feature of deposition in the macular area, with a variable effect on the vision. In some, the effect may not be reversible. Such cases should be monitored by serial visual acuity measurement, central visual fields, and electrophysiology tests when appropriate. Chloroquine has been almost totally replaced by hydroxychloroquine, a much safer alternative.

Resolved macular oedema

Even after complete resolution, macular oedema may leave behind some permanent

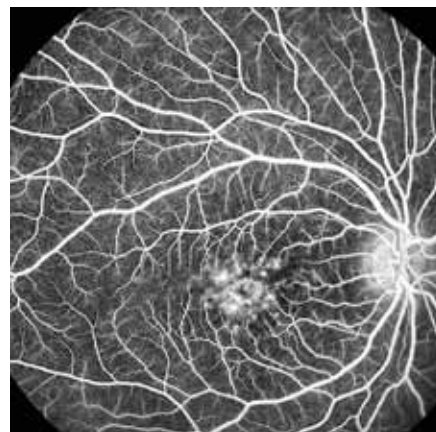


Fig 11: FFA showing silent choroid in Stargardt's disease

alteration in pigment at the macula. This may be associated with some loss of visual acuity.

Cystoid macular oedema (CMO)

Macular oedema is essentially the accumulation of fluid in the outer plexiform layer of the retina. It is usually visible to the trained eye as swelling or elevation of the central macula. The retina remains translucent; a good binocular visualisation and clear media are therefore required to make a clinical diagnosis. In a typical case, a FFA shows a 'petaloid' appearance as the dye fills the cystic spaces around the centre of the fovea, followed by frank leakage. The best way to diagnose and quantify CMO is an optical coherence tomography (OCT) scan. This also has the advantages of being non-invasive and quick. Various treatment strategies may be considered depending on the cause and duration of CMO and potential side-effects of the therapy. Patients affected by CMO should be referred quickly.

Vascular causes

Diabetic retinopathy remains the most common cause of CMO. Branch and central retinal vein occlusion may also be complicated by macular oedema. Idiopathic juxtafoveal microaneurysms are rare, but may present with CMO in younger patients.

The treatment of vascular macular oedema is usually with laser photocoagulation. The outcome of treatment usually depends upon the presence or absence of any underlying macular ischaemia. Conversely, CMO may deteriorate or appear after extensive panretinal photocoagulation for neovascularisation of the retina. In severe acute papilloedema, macular exudation may acquire a star shape (Fig. 13).

Post-operative causes

Subclinical CMO may be seen to be present in as many as 20% of cases after uncomplicated cataract surgery if one examines patients using FFA or OCT.¹⁰ Symptomatic CMO is more common after complicated

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cataract surgery or previous uveitis and in diabetic patients.

Inflammatory CMO is a common complication of all forms of chronic uveitis such as sarcoidosis, toxoplasmosis, birdshot choroidopathy, pars planitis, cytomegalovirus retinitis and posterior scleritis. The treatment is aimed at the underlying cause. More direct treatment for CMO in such cases is often by periocular steroid injection.

Hereditary

Some hereditary dystrophies are commonly associated with CMO, such as retinitis pigmentosa and autosomal-dominant cystoid macular oedema. Diamox and non-steroidal agents may be tried, but the condition usually remains refractory to treatment.

Tractional

Vitreomacular traction syndrome (VMTS) is characterised by persistent vitreous traction in the foveal area in association with macular oedema.¹¹

Vascular tortuosity and retinal folds may also be present. Like macular holes, it is seen more commonly in women, presenting with a slow deterioration in vision. CMO may also be caused by an epiretinal membrane (ERM).

Macular hole (Figs. 14 and 15)

The hallmark complaint of a macular hole is distortion of central vision. There is often a variable reduction in visual acuity. The condition most commonly affects individuals in their sixth or seventh decade of life; women are affected more commonly than men (a 2:1 ratio).

A full-thickness stage 3 (no PVD) or stage 4 (with PVD) macular hole is usually diagnosed without much difficulty. The centre of the macula has a punched-out appearance often surrounded by a cuff of fluid. The Watzke-Allen sign is usually helpful to test for a full thickness retinal defect. In this test, a thin slit lamp beam is focused on the fovea. In a full-thickness defect, a break or thinning is seen by the patient. An OCT scan is invaluable in establishing a diagnosis even in very subtle cases.¹²

Pseudohole

'Pseudohole' is the term used to define conditions which may be confused with a macular hole, for example, a hole inside an epiretinal membrane, and chronic CMO. Any sign of haemorrhage or sudden distortion of vision should warrant urgent referral to an ophthalmologist with expertise in PDT.

Visual acuity is usually not severely affected in a pseudohole, unless this is associated with underlying macular ischaemia. The Watzke-Allen test is usually negative, that is, the patient sees an intact beam of light. An OCT scan must be done if clinical differentiation proves inconclusive. An FFA may be considered to reveal the

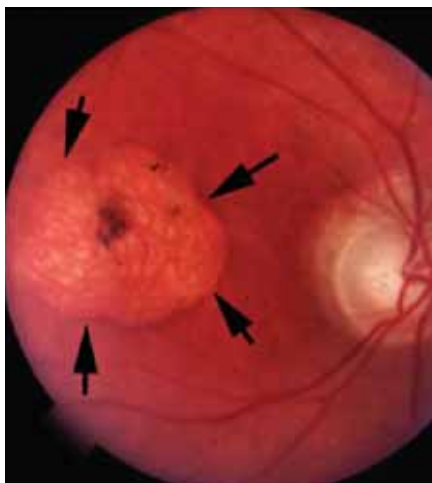


Fig 12: Central areolar maculopathy

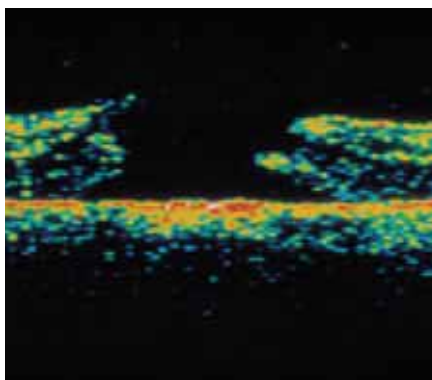


Fig 14: OCT of a full thickness macular hole before surgery. Note the missing neural retina with elevated edges and intact RPE

underlying pathology.

Central serous chorioretinopathy (CSCR)

This idiopathic condition typically affects otherwise healthy individuals, usually anxious men in their third to fourth decade of life. They usually present with a mild to moderate visual blur and metamorphopsia. A circular- or kidney-shaped smooth elevation of variable size is seen at the macula. A hypermetropic shift is common. As a rule, there is no haemorrhage. If haemorrhage is present, this excludes the diagnosis of CSCR and warrants an urgent FFA to exclude CNV.

Patients should be reassured that a great deal of improvement is to be expected, although recurrence is common. Treatment with laser or steroids is rarely of benefit.

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Fig 13: Macular star associated with papilloedema

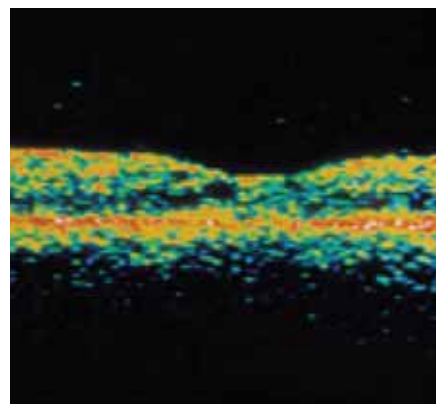


Fig 15: OCT of the same eye six months after vitrectomy and gas procedure. Normal foveal contour is restored.

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Module questions

Course code: C-2694

Please note, there is only one correct answer. Enter online or by form provided.

- Retinal tears occur in:
 - Neural retina
 - Retinal pigment epithelium
 - Bruch's membrane
 - Retinal pigment epithelium and neural retina
- A retinal detachment is most likely to arise from:
 - Oral pearls
 - Lattice degeneration
 - Paving stone degeneration
 - Snowflake degeneration
- Which one of the following is correct? Retinal:
 - Detachment is always a complication of pars plana cysts
 - Detachment is always a complication of retinoschisis
 - Detachment can arise from a cryotherapy scar
 - Breaks are usually easy to detect
- Which one of the following is correct regarding retinal detachments?
 - The presence of pigment in the vitreous is universal
 - There is no proven association with previous cataract surgery
 - Chronic retinal detachments are usually found on the upper part of the retina
 - A demarcation line is a feature of an acute, recent detachment
- Which one of the following is incorrect regarding retinal detachments?
 - Involvement of the macula carries a poor prognosis for vision
 - Upper half retinal detachments are less urgent than lower half
 - A relative afferent pupillary defect is a feature of a large retinal detachment
 - A break extending beyond two consecutive clock hours is classified as a giant retinal tear
- Which statement is incorrect regarding vitreous haemorrhage?
 - An ultrasound B-scan is very helpful in making a diagnosis of retinal detachment
 - Diagnosis is difficult in the presence of dense cataract
 - A vitrectomy is usually performed soon after presentation if required
 - A relative afferent pupillary defect is always to be expected in dense vitreous haemorrhage
- Which one of the following is correct?
 - The foveal avascular zone is approximately 400µm in diameter
 - Direct ophthalmoscopy is the most appropriate tool in the diagnosis of macular disorders
 - Macroaneurysms affect mostly males
 - Macroaneurysms affect mostly young people
- Which one of the following is correct regarding macular haemorrhage?
 - Choroidal neovascularisation is associated with hypermetropia
 - Fundus fluorescein angiography is rarely required
 - Photodynamic therapy is appropriate in all cases
 - Blood pressure measurement may be relevant
- Which statement is correct regarding drusen?
 - Drusen are located in the RPE
 - Fundus fluorescein angiography should be performed in all cases
 - All cases of drusen should be referred to an ophthalmologist
 - Appearance of haemorrhage warrants an urgent referral
- Which one of the following is correct? Visual acuity:
 - Is usually worse than 6/60 in Best's disease
 - Always returns to normal in macular oedema
 - Is usually more severely affected in pseudohole than in macular hole
 - May be 6/6 in cone dystrophy
- Which one of the following is incorrect regarding cystoid macular oedema (CMO)?
 - Macular oedema is the accumulation of fluid in the outer plexiform layer
 - A fluorescein angiogram reveals a petaloid appearance
 - Optical coherence tomography is the easiest way to diagnose the condition
 - Patients with CMO should be monitored in practice
- Which one of the following is incorrect regarding macular hole?
 - The hallmark is distortion of central vision
 - It most commonly affects people in the sixth or seventh decade of life
 - It affects women more than men
 - A stage 3 classification is associated with a posterior vitreous detachment

An answer return form is included in this issue. It should be completed and returned to: CET initiatives (C-2694), OT, McMillan Scott, 9 Savoy Street London WC2E 7HR to arrive by 6 September, 2006. Under no circumstances will forms received after this date be marked – the answers to the module will have been published in our 8 September, 2006 issue.

CET answers

Course code: C-2693

Here are the correct answers to Module 9 Part 7 which appeared in our June 30, 2006 issue.

- Correct answer is B**
The retina has a dual supply from branches of the ophthalmic artery, a branch of the internal carotid artery. Branches of the central retinal artery supply the inner two thirds of the retina, and the choroidal circulation supplies the outer third of the retina.
- Correct answer is C**
Endothelial cells are responsible for maintaining the blood-retinal barrier, and damage results in increased vascular permeability.
- Correct answer is A**
In patients presenting with bilateral CRVO, an evaluation to rule out a hyperviscosity syndrome should be considered.
- Correct answer is D**
Retinal ischaemia stimulates a pathological neovascularisation of the retina, which is driven by angiogenic factors, such as vascular endothelial growth factor (VEGF). The rationale of PRP for PDR is to convert the hypoxic environment of the peripheral retina into an anoxic one, thereby reducing the induction of angiogenic growth factors such as VEGF.
- Correct answer is A**
Malignant hypertension refers to a syndrome characterised by severe hypertension (eg systolic >200, diastolic >130mmHg) accompanied by encephalopathy or nephropathy, and/or optic disc swelling, retinal haemorrhages and exudates. It is a medical emergency and requires urgent referral.
- Correct answer is B**
Widespread haemorrhages are seen in central retinal vein occlusion. A cherry red spot is typically seen in central retinal artery occlusion.
- Correct answer is C**
Age related macular degeneration typically affects Caucasians.
- Correct answer is D**
Retinitis pigmentosa is a heterogeneous group of disorders characterised by a triad of night blindness, progressive visual field loss from photoreceptor and retinal pigment epithelium dysfunction and abnormal ERG findings. The abnormality occurs in both rods and cones, with the rod dysfunction predominating.
- Correct answer is B**
Unlike patients with fundus flavimaculatus who typically present as adults, patients with Stargardt's disease present in childhood with reduced visual acuity. The prognosis in fundus flavimaculatus is relatively good, the opposite can be said of Stargardt's disease, where many individuals suffer with the consequences of macular atrophy.
- Correct answer is C**
In Best's disease, the visual acuity normally starts to drop around the fifth decade.
- Correct answer is C**
Mutations have been identified in tissue inhibitor of metalloproteinases-3 (TIMP-3). The proteins encoded by this gene family are natural inhibitors of the matrix metalloproteinases, a group of peptidases involved in degradation of the extracellular matrix. Mutations in this gene cause the autosomal dominant disorder Sorsby's fundus dystrophy.
- Correct answer is D**
The opacities seen in spherule degeneration are composed of cholesterol crystals and are seen in eyes with a history of chronic vitreous haemorrhage or trauma.