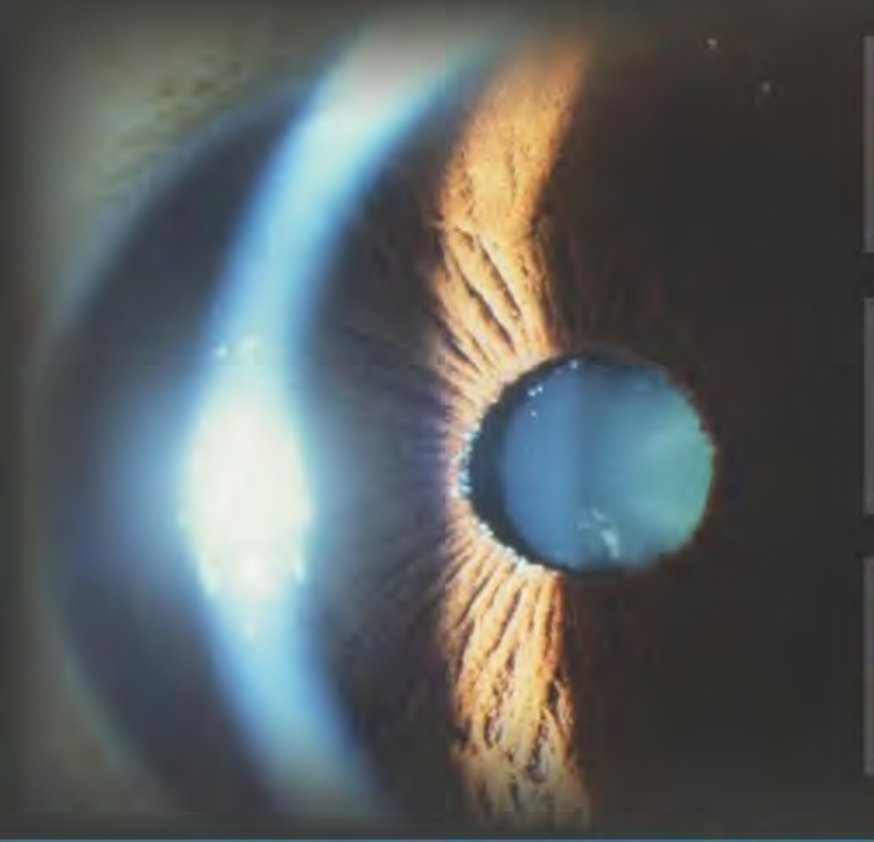
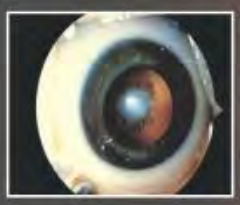
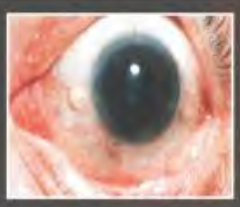
THIRD EDITION

OPHTHALMIC

Diagnosis & Treatment







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Ophthalmic Diagnosis and Treatment

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Third Edition

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Preface

Ophthalmic Diagnosis and Treatment, Third Edition, provides in a simplified format, recommendations and treatments of conditions most often encountered by eye care providers. Entities are in alphabetical order for easy, rapid access of information for the eye care provider and student. Each entity has Diagnosis (definition, symptoms, differential diagnosis, etc.) on the left page and Treatment (diet and lifestyle, treatment aims, prognosis, pertinent references, etc.) on the right page. We have added new illustrations and have updated the material.

The same format is used throughout the book. The information, not intended to be encyclopedic, highlights the salient facts of each entity. Full-color illustrations are used throughout the book. As much as possible, each entity is followed by its diagnostic code, which is most helpful for billing purposes and for organizing patients' records.

We anticipate that the book will be used by eye care professionals and students as a quick reference guide of the significant points most commonly encountered and important entities pertaining to the eyes.

Myron Yanoff

Acknowledgments

The editors would like to acknowledge Joe Rusko and the staff of Jaypee Brothers Medical Publishers for their outstanding help in producing this book.

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Contents by Specialty

Cataract and Systemic and Orbital Disorders

Corneal and External Diseases

Glaucoma

Neuro-ophthalmology

Pediatric Ophthalmology

Retinal and Vitreous Disorders

How to use this book

This book provides current expert recommendations in the form of tabular summaries on the diagnosis and treatment of all major disorders throughout ophthalmology. Essential guidelines on each of the topics have been condensed into two pages of vital information, summarizing the main procedures in diagnosis and management of each disorder to provide a quick and easy reference.

Each disorder is presented on facing pages: the main procedures in diagnosis on the left and treatment options on the right.

Listed in the main column of the **Diagnosis** page is the definition; other common names; the common symptoms, signs, and complications of the disorder; pearls and considerations; and referal information, with brief explaining their significance and probability of occurrence, together with details of investigations that can be used to aid diagnosis.

The **left shaded side column** contains information to help readers evaluate the probability that a patient has the disorder. It may also include other information that could be useful in making a diagnosis (e.g. classification or grading systems, comparison of different diagnostic methods).

The numbers that appear in parentheses next to disease names at the top of each page and scattered throughout the text are from *The International Classification of Diseases* (New York, 1995, McGraw-Hill). These numbers are used by physicians to organize their patients' medical records and to facilitate the timely reimbursement of their services.

On the **Treatment** page, the main column contains information on lifestyle management and nonspecialist medical therapy of the disorder, with general information on specialist management when this is the main treatment.

Whenever possible under "Pharmacologic treatment", guidelines are given on the standard dosage for commonly used drugs, with details of contraindications and precautions, main drug interactions, and main side effects. In each case, however, the manufacturer's drug data sheet should be consulted before any regimen is prescribed.

The main goals of treatment (e.g. to cure, to palliate, to prevent), prognosis after treatment, precautions that the physician should take during and after treatment, and any other information that could help the clinician to make treatment decisions (e.g. other nonpharmacologic treatment options, special situations or groups of patients) are given in the **right shaded side column**. The general references at the end of this column provide readers with further practical information.

1. Acute Posterior Multifocal Placoid Pigment Epitheliopathy (363.15)

DIAGNOSIS

Definition

An acquired, self-limited inflammatory disorder and vasculitis of the retina, retinal pigment epithelium (RPE), and choroid in otherwise healthy young adults.

Synonyms

None; often abbreviated AMPPE.

Symptoms

Rapid, painless loss of vision: in one or both eyes.

Signs

- Acute phase shows multiple circumscribed gray-white lesions at the level of the RPE located in the postequatorial retina (*see* Fig. 1)
- Late phase shows changes in the RPE similar to laser burns.
- Associated serous detachment: uncommon
- Perivenous exudation in the retina
- Slight dilation of the retinal veins
- Papilledema, papillitis, optic neuropathy
- Episcleritis
- Iridocyclitis.

Investigations

Fluorescein angiography, in the early phase, shows blockage of choroidal fluorescence (*see* Figs 2A to C), with mid-phase to late-phase diffuse staining of the acute lesions.

Complications

- *Choroidal neovascularization*: rare
- Cerebritis: has been reported.

Differential Diagnosis

Serpiginous choroidopathy

Cause

Unknown

Epidemiology

- Disease affects healthy young men and women
- One-third of patients give a history of a viral prodrome
- Recurrences are frequent.

Diagnosis continued on p. 4

TREATMENT

Diet and Lifestyle

No precautions are necessary.

Pharmacologic Treatment

Oral steroids should be instituted promptly if there are signs of associated cerebritis.

Treatment Aims

To observe a patient

Prognosis

Visual prognosis is good, and there is a low incidence of recurrence after treatment.

Follow-up and Management

Based on the severity of the symptoms, patients should be followed every few weeks until they are out of the acute phase.

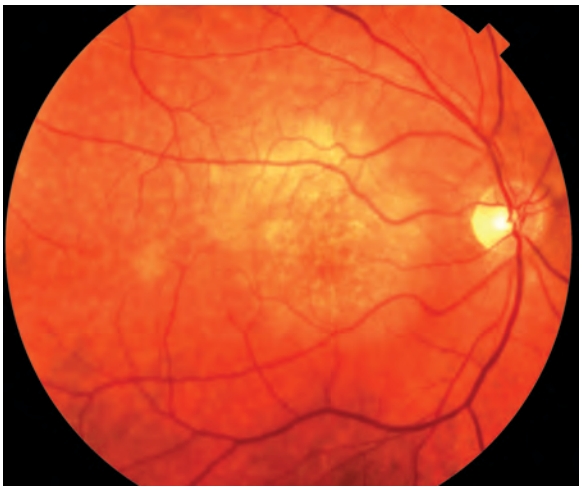


Fig. 1: Intense whitening of the outer retina and RPE in the macula of a patient with AMPPE.

Treatment continued on p. 5

DIAGNOSIS—cont'd

Pearls and Considerations

- Spontaneous visual recovery is expected with or without systemic therapy, with most eyes achieving 20/40 or better vision in 1–6 months from onset
- It is important to look for an underlying cause because the condition has been related to some antimicrobial agents, such as ampicillin and sulfonamides. If an antimicrobial agent is identified, it should be discontinued to prevent further recurrences.
- Associated cerebral vasculitis may occur as late as 3 months after presentation of AMPPE.

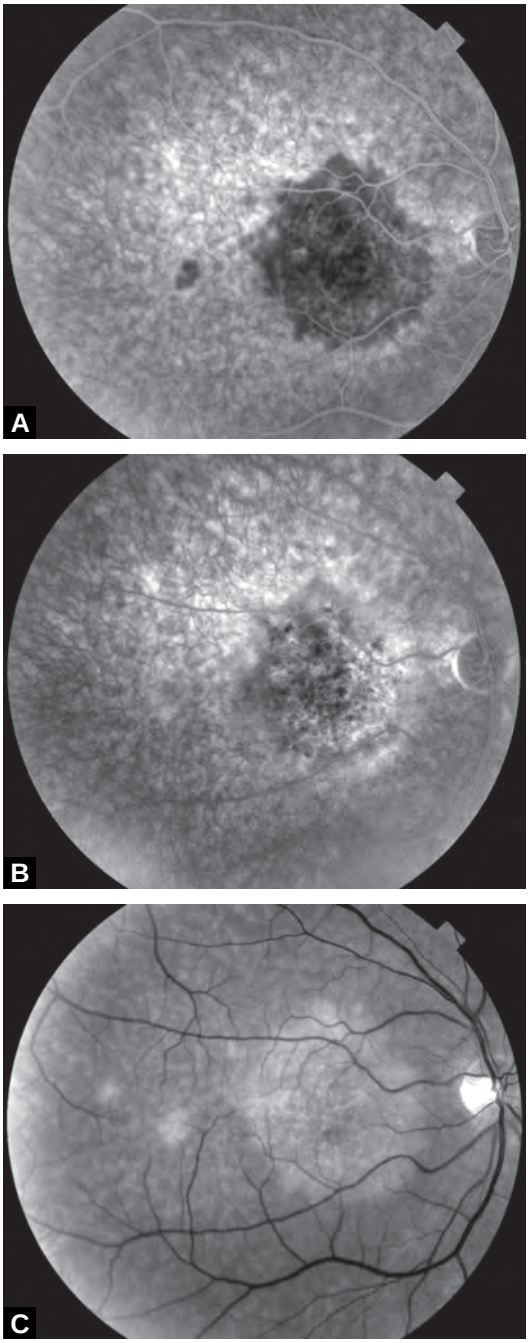
Referral Information

A computed tomography scan, magnetic resonance imaging or cerebral arteriogram is indicated in patients with severe headache to rule out cerebral vasculitis.

TREATMENT—cont'd

Nonpharmacologic Treatment

No nonpharmacologic treatment is recommended.



Figs 2A to C: (A) Red-free photograph as well as early and late phases of the fluorescein angiogram are seen; (B) There is early blockage of fluorescein in both the large central and the smaller temporal macular areas; (C) In the later frame, the temporal spot is no longer visible and there is hyperfluorescence of the central macular region. This early blockage and late hyperfluorescence is the hallmark of AMPPE.

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Puilido JS, Blake CR. Inflammatory choroiditis. In: Tasman W et al. (Eds). Duane's Clinical Ophthalmology (CD-ROM). Philadelphia: Lippincott Williams & Wilkins; 2004.

2. Acute Retinal Necrosis (363.13)

DIAGNOSIS

Definition

Severe inflammation leading to necrosis of the retina starting in the periphery and progressing circumferentially and centripetally caused by one of the herpes viruses. Often referred to in the abbreviated form: ARN (acute retinal necrosis).

Synonyms

Necrotizing herpetic retinitis

Symptoms

- Blurry vision
- Ocular pain
- Photophobia
- Floaters.

Signs (Fig. 1)

White, confluent, retinal necrotic lesions affecting a large portion, if not the entire, peripheral retina is the hallmark of the condition. Panuveitis may be present, i.e. conjunctival inflammation, scleritis, vitreous and anterior chamber cells, obliterative retinal vasculitis as well as tenderness of the globe. There is a paucity of retinal hemorrhages compared to other conditions such as CMV retinitis. Patients are typically not immunocompromised. Most patients present unilaterally but progression to bilateral disease is common.

Investigations

- Fluorescein angiography shows staining of the peripheral vessels. Loss of peripheral circulation due to occlusive vasculitis is also seen. The posterior pole usually spared initially. If papillitis develops, leakage from the disk will be seen.
- Serial color fundus photos can document the progression of the disease.
- Labwork must include herpes 1, herpes 2 and varicella zoster titers. A vitreous biopsy to obtain polymerase chain reaction (PCR) of these DNA viruses may be done, as well.
- Labwork to rule out other conditions includes complete blood count (CBC), Lyme titer, human immunodeficiency virus (HIV), angiotensin converting enzyme (ACE) titer, toxoplasma titer, rapid plasma reagin (RPR) and fluorescent treponemal antibody absorption (FTA-ABS), purified protein derivative (PPD) skin testing, chest X-ray, and magnetic resonance imaging (MRI) of the brain and orbits.

Complications

Development of large retinal tears is common. These often lead to retinal detachment. Due to extensive retinal involvement, proliferative vitreoretinopathy with total retinal detachment often develops. Severe or even complete loss of vision may be the end result even with aggressive medical and surgical treatment.

Pearls and Considerations

Progressive outer retinal necrosis (PORN) has similar white lesions affecting the outer retina (i.e. beneath the retinal vessels) but without vitreous inflammation and without vasculitis. The cause is the same herpes viruses implicated in ARN. This is seen in immunocompromised individuals and is almost always bilateral. It frequently leads to retinal detachment and blindness in both eyes, even with treatment.

Referral Information

Retinal specialist to manage ocular complications and infectious disease specialist to manage systemic antiviral treatment

Differential Diagnosis

- Cytomegalovirus (CMV) retinitis which is seen in AIDS patients. Intraretinal blood is prominent.
- *Syphilis*: this requires lumbar puncture to rule out central nervous system (CNS) disease.
- *Toxoplasmosis*: has a much better prognosis.
- *Lyme disease*: may have other findings such as cranial nerve palsy.
- *Behcet's disease*: mouth ulcers in 90% of patients. HLA-B51 test often positive.
- *Ocular lymphoma*: diagnosed with vitreous biopsy and associated with CNS lymphoma
- *Fungal endophthalmitis*: need to do fungal blood cultures.

TREATMENT

Diet and Lifestyle

No special precautions are necessary.

Pharmacologic Treatment

- Immediate antiviral treatment with the goal of preventing disease in the fellow eye and preventing progression to retinal detachment. Oral valacyclovir 1 gram PO TID for 6 weeks. May substitute with acyclovir 600 mg five times a day.
- Systemic steroids are also given, although it is still controversial whether to initiate immediately or wait a week to give time for the antiviral medicine to work. Prednisone 60 mg PO daily for 1 or 2 weeks followed by a slow taper over the next month.
- May need to admit to hospital for intravenous antiviral and steroid medications.
- Topical prednisolone acetate one drop up to every 2 hours with a slow taper over the next month depending upon anterior chamber inflammation. Topical atropine 1% one drop BID for cycloplegia and to decrease ocular pain.

Nonpharmacologic Treatment

Peripheral laser retinopexy is recommended in order to prevent the development of retinal detachment. When retinal detachment develops vitrectomy and further laser with or without peripheral scleral buckling is done. Long-acting gas or even silicone oil is necessary to repair the complex rhegmatogenous/necrotic detachment.

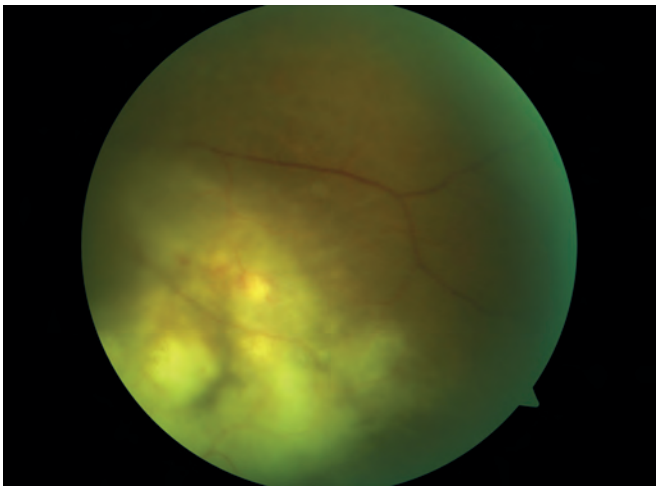

Fig. 1: Retina shows necrosis and detachment.

Treatment Aims

To prevent severe, irreversible visual loss by treating the underlying viral cause of the retinitis. In unilateral cases goal of antiviral medicine is to prevent bilateral disease.

Prognosis

If caught very early, may have good visual outcome. Most patients, however, suffer from some degree of both peripheral and central visual loss in spite of treatment.

Follow-up and Management

Must be followed daily while in hospital during acute phase of illness. As the retinitis stabilizes may be followed weekly, then monthly and as needed depending upon any surgery or complications.

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Luu KK, Missotten T, Salzmann J, et al. Acute retinal necrosis features, management and outcomes. Ophthalmology. 2007; 114:756-62.

Scott IU, Luu KM, Davis JL. Intravitreal antivirals in the management of patients with acquired immunodeficiency syndrome with progressive outer retinal necrosis. Arch Ophthalmol. 2002;120: 1219-22.

DIAGNOSIS

Definition

Any disease or condition of the eye or ocular adnexa that arises from a patient's underlying systemic acquired immunodeficiency syndrome (AIDS) infection or associated immunosuppression.

Synonyms

None

Symptoms

- Decreased vision, floaters, double vision
- Eye pain
- Facial rash
- Floaters
- Red eye
- Shadow.

Signs

Human immunodeficiency virus (HIV) retinopathy (noninfectious)

- Nerve fiber layer (NFL) and inner retinal hemorrhages
- Microaneurysms
- *Cotton-wool spot*: the most common manifestation of AIDS retinopathy.

Infectious Agents

Adnexa

- Herpes zoster ophthalmicus (HZO)
- Molluscum contagiosum.

Cryptococcus: papilledema, multiple choroiditis, meningitis, endophthalmitis.

Cytomegalovirus (CMV) retinitis: classic intraretinal hemorrhages with white infiltrates (*see* Fig. 1).

Granular form: "brush fire" appearance.

Hemorrhagic form: "tomato ketchup fundus".

Herpes retinitis [*acute retinal necrosis* (*ARN*) *or progressive outer retinal necrosis* (*PORN*)]: aggressive retinitis in which multiple white retinal infiltrates coalesce, leading to retinal detachment and visual loss (*see* retinal necrosis, acute).

Pneumocystis choroiditis: multiple, deep, yellow-orange lesions at the level of the choroid (*see* Fig. 2).

Toxoplasmosis: multifocal outer retinal lesions that spread quickly; normally, little vitreous inflammation is seen in immunocompetent patients; occurs in previously diagnosed patients (*see* Fig. 3).

Microsporidia: chronic keratoconjunctivitis.

Syphilis: plaque-like serous elevations in the posterior pole.

Differential Diagnosis

Cotton-wool spots: of diabetes, hypertension, ocular ischemia and vasculitis

Cause

HIV infection

Diagnosis continued on p. 10

TREATMENT

Diet and Lifestyle

Compliance with highly active antiretroviral therapy (HAART).

Pharmacologic Treatment

For Cytomegalovirus Infection

- Intravenous (IV) therapy with ganciclovir, foscarnet or cidofovir
- Intravitreal ganciclovir implant (sustained-release device) placed in the pars plana of the eye
- Intravitreal injections of ganciclovir or foscarnet.

For Pneumocystis sp. Infection

Bactrim, pentamidine.

For Herpes Simplex or Zoster Infection

Aggressive treatment with IV acyclovir.

For Toxoplasmosis

Pyrimethamine, sulfa drugs or clindamycin.

Treatment Aims

- To control the infection with appropriate antiviral and antimicrobial agents
- To stabilize or improve vision
- To treat secondary complications (e.g. retinal detachment) with laser or surgery.

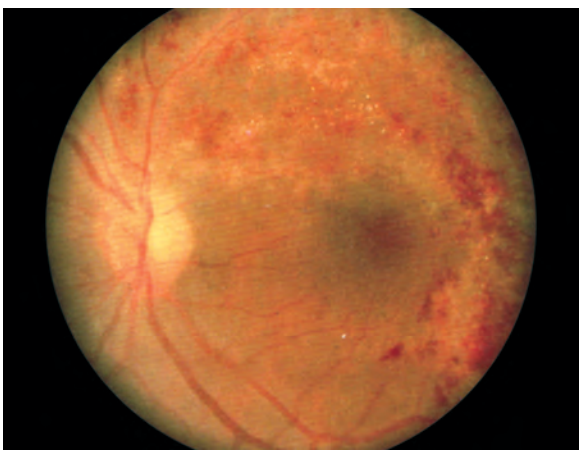

Fig. 1: Cytomegalovirus retinitis with the characteristic white perivascular retinal infiltrates and intraretinal hemorrhages or “pizza pie fundus”.

Treatment continued on p. 11

DIAGNOSIS—cont'd

Tumors

- *Kaposi's sarcoma*: flat purplish lesion on the eyelid or conjunctiva similar to lesions seen on the skin; may be mistaken for subconjunctival hemorrhage
- Central nervous system (CNS) lymphoma
- Lymphomas
- Squamous cell carcinoma of the eyelid.

Neurologic Manifestations

- Cranial nerve palsies
- Papilledema
- Nystagmus
- Optic neuritis
- Visual field defects.

Investigations

Careful history: reviewing the patient's past medical history often gives clues to the cause.

CD4+ count: usually less than 50 cells/mm^3 when patients develop CMV retinitis.

Cryptococcus: high risk of CNS infection.

Progressive outer retinal necrosis: CD4□+ count greater than 50 cells/mm^3.

Acute retinal necrosis: CD4□+ count less than 50 cells/mm^3.

Pneumocystis jiroveci (previously P. carinii): CD4□+ count less than 200 cells/mm^3.

Fluorescein angiography: helpful in differentiating retinal infections.

Computed tomography and magnetic resonance imaging scans: may be necessary for patients with CNS abnormalities.

Vitreous biopsy with analysis of polymerase chain reaction: helps distinguish herpes simplex from zoster and CMV when the clinical picture is not clear-cut.

Sequential fundus photographs: help monitor patients for response to therapy and evidence of disease progression.

Complications

- Visual loss
- Retinal detachment.

Medication-related

- *Cidofovir*: anterior uveitis and profound hypotony
- *Rifabutin (prophylaxis for atypical myobacterial infection)*: severe (sometimes bilateral) anterior uveitis.

Diagnosis continued on p. 12

TREATMENT—cont'd

For Syphilis

High-dose IV penicillin for 10–14 days.

Cryptococcus

Intravenous amphotericin, IV itraconazole, vitrectomy and intravitreal amphotericin for endophthalmitis.

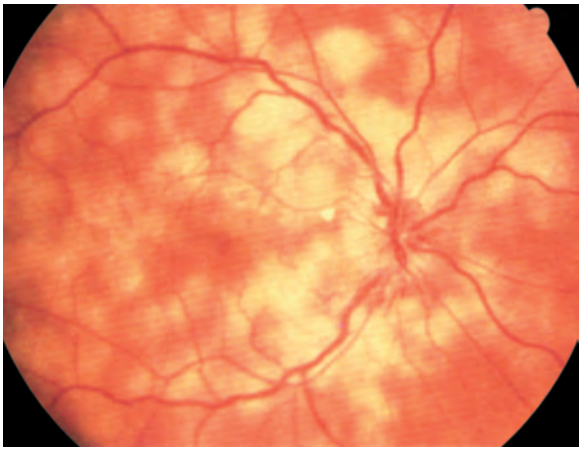

Fig. 2: *Pneumocystis* choroiditis showing deep choroidal, creamy yellow-orange infiltrates throughout the posterior pole.

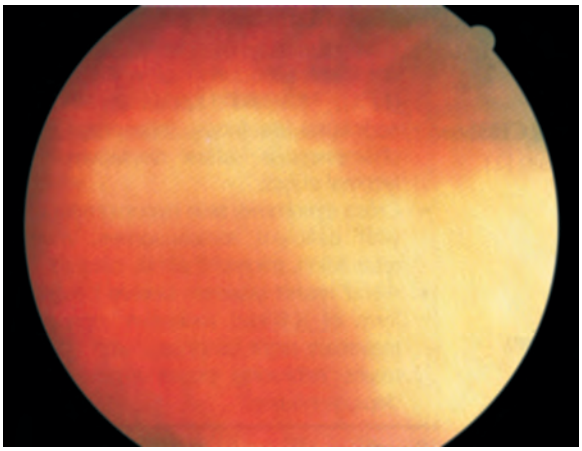

Fig. 3: Outer retinal toxoplasmosis in an AIDS patient showing the "brush fire" advancement of the infection.

Treatment continued on p. 13

DIAGNOSIS—cont'd

Pearls and Considerations

- Dilated fundus examination (DFE) should be performed every 3 months in patients with a CD4□+ count less than or equal to □50 cells/mm^3 because of the potential for asymptomatic CMV retinitis. Once the CD4+ count rises above 100 cells/mm^3, the risk of retinitis is small.
- Acquired immunodeficiency syndrome patients' complications should be treated cautiously with steroids because of the potential for further immunosuppression and infection
- Approximately 70–80% of AIDS patients will require treatment for an ocular complication at some time in their lives
- Cytomegalovirus is the most common severe and sight-threatening ocular infection in AIDS patients
- Infections of the cornea and adnexa are less common than intraocular infections in AIDS patients
- With HAART, many patients can reduce the detectable viral load to zero and can lead otherwise normal lives. Only when their immunity drops do they develop complications.

Referral Information

Variable and specific to the etiology of each complication.

TREATMENT—cont'd

Microsporidia

Topical fumagillin up to every 2 hours initially, and then taper slowly.

Nonpharmacologic Treatment

Regular periodic ocular examinations (e.g. CD4+ count < 50, every 3 months).

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4. Albinism (270.2)

DIAGNOSIS

Definition

The classification of a group of congenital diseases that result from defective pigment production (melanogenesis) can be ocular or oculocutaneous.

Synonyms

None; associated abbreviations: ocular albinism (OA) and oculocutaneous albinism (OCA), Chediak-Higashi syndrome (CHS), Hermansky-Pudlak syndrome (HPS), Griscelli syndrome (GS).

Symptoms

- *Painful photophobia*: In most patients
- *Decreased visual acuity (worse at distance than near)*: In most patients
- *Esthetic blemish from nystagmus*: In most patients
- *Poor binocular vision*: Because of near-total decussation of the optic nerves at the chiasm.

Signs

- *Horizontal nystagmus*: With possible null point and head posture to maximize visual acuity
- *Iris transillumination*: In patients with ocular albinism
- *Pink irides*: In patients with oculocutaneous albinism
- *Foveal hypoplasia or aplasia*: With retinal vascular presence in the foveal area
- *Decreased pigment in retinal pigment epithelium (RPE) and iris pigment epithelium (IPE)*: Figure 1
- Esotropia
- *Refractive errors*: Often myopia.

Investigations

- *Skin biopsy*: Patients with X-linked recessive ocular albinism have macroelanosomes
- *Testing for presence of tyrosinase*: Patients with tyrosinase-negative oculocutaneous albinism have no tyrosinase in hair bulbs; patients with tyrosinase-positive oculocutaneous albinism do
- Patients with Chediak-Higashi syndrome cannot opsonize certain bacteria
- Patients with Hermansky-Pudlak syndrome have platelet adherence abnormalities and capillary fragility.

Complications

- *Skin disorders, including melanomas*: Patients with oculocutaneous albinism are at risk for skin malignancies; protection is imperative
- *Photophobia*: It may disrupt outdoor activities
- *Chronic infections, including pneumonia*: In patients with Chediak-Higashi syndrome

Differential Diagnosis

Patients have depigmented skin or adnexal structures but no increased decussation of the optic nerves at the chiasm. Various albinoid forms exist:

- *Waardenburg syndrome*: Autosomal dominant association of white forelock (17%), piebald appearance, sensorineural deafness, synophrys, blepharophimosis, lateral displacement of lacrimal puncti.
- *Cross syndrome*: Skin hypopigmentation with deficient melanosomes, mental retardation, microphthalmia, cataracts.
- *Åland Island disease:* Believed to be a form of X-linked recessive, congenital stationary night blindness, with affected males exhibiting posterior-pole retinal depigmentation.

Cause

- Congenital, hereditary, stationary
- Oculocutaneous albinism segregates in most patients as an autosomal recessive; ocular albinism segregates in most affected patients as an X-linked recessive.

Associated Features

- *Tyrosinase negative*: White hair, pink skin.
- *Tyrosinase positive*: White hair as children; may become blond; may develop pigmented nevi.
- *Yellow mutant*: White hair and pink skin as infants; develop yellow hair at 6 months and normal skin pigmentation by 3 years.
- *Brown albinism*: Africans with reddish brown skin, red hair, freckles, brown irides.
- *Autosomal dominant oculocutaneous albinism*: White to cream skin, white hair, freckles, gray-blue irides.

Pathology

- Caucasian patients with ocular albinism have spotty areas of deficient melanin in the IPE melanosomes and generally no melanin in RPE melanosomes. Patients with oculocutaneous albinism have diffuse deficiency of IPE and RPE melanin; they also have varying amounts of absent dermal melanin.
- Patients with ocular and oculocutaneous albinism have absent foveal depressions, with vascular incursion into the area normally occupied by the fovea. They also have almost total decussation of the optic nerves at the chiasm.

Diagnosis continued on p. 16

TREATMENT

Diet and Lifestyle

Many patients will benefit from wearing sunglasses and sunscreen protection for the skin. Low-vision aids are recommended for elderly patients.

Pharmacologic Treatment

No pharmacologic treatment is recommended.

Treatment Aims

- To ensure patient comfort and safety
- To prevent bleeding diatheses in patients with Hermansky-Pudlak syndrome
- To treat infections early in patients with Chediak-Higashi syndrome.

Other Treatments

Glasses where appropriate; nystagmus prevents full benefit in many patients.

Prognosis

- Prognosis is stable in most patients; the nystagmus amplitude lessens with age, but visual acuity usually does not improve.
- Prognosis is guarded in patients with Hermansky-Pudlak or Chediak-Higashi syndrome.

Follow-up and Management

Yearly follow-up is sufficient for most patients. Those with Hermansky-Pudlak or Chediak-Higashi syndrome will require individualized follow-up and management of systemic problems. Many patients will want genetic counseling.

Treatment continued on p. 17

DIAGNOSIS—cont'd

- *Clotting abnormalities*: In patients with Hermansky-Pudlak syndrome
- Most patients with albinism and nystagmus cannot obtain a driver's license because their distance visual acuity is 20/100 to 20/200. Their near visual acuity, however, may be close to 20/20 if children are permitted to hold objects closely. Thus, children usually succeed in normal schooling environments if they are permitted to sit in front of the classroom and walk to the blackboard when necessary. Older children may require low-vision aids for near and distant work.

Classification

- Ocular albinism (X-linked recessive, rarely autosomal dominant or recessive)
- *Oculocutaneous albinism (autosomal recessive)*: Tyrosinase negative, brown albinism, tyrosinase positive, rufous albinism, yellow mutant, Hermansky-Pudlak, Chediak-Higashi.

Pearls and Considerations

- Ultraviolet-protective sunglasses are recommended for patients with any variant of ocular albinism or oculocutaneous albinism
- Patients affected by oculocutaneous albinism should be encouraged to be screened frequently for skin cancer
- Color vision remains normal in patients with albinism
- Clinicians should be mindful of the distinction between ocular albinism (with reduced acuity of 20/70 to 20/200) and *blond fundus,* associated with lightly pigmented individuals who have normal visual acuity.

Referral Information

Some variations of oculocutaneous albinism are potentially fatal; therefore, medical and hematologic referral should be considered to rule out these variants.

TREATMENT—cont'd

Nonpharmacologic Treatment

- Strabismus surgery
- *Eye muscle surgery*: To align the head in patients who adopt a posture resulting from the presence of a nystagmus-associated null point.

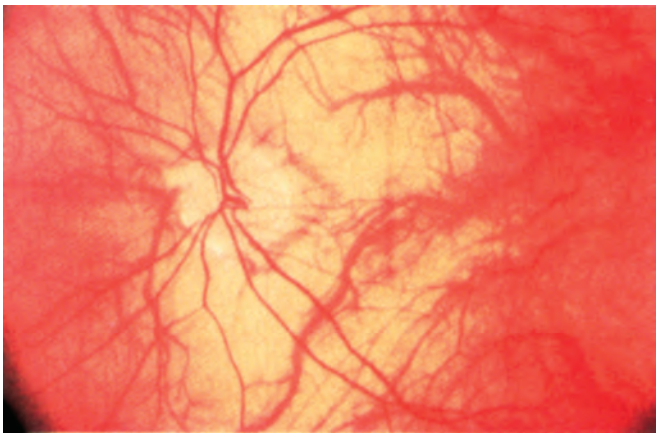

Fig. 1: Note pale fundus caused by lack of pigment in the retinal pigment epithelium and choroid.

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5. Amblyopia (368.0)

DIAGNOSIS

Definition

- Derived from Greek word meaning "dullness of vision"
- A functional reduction in the best-corrected visual acuity of an eye caused by "misuse" or "disuse" during the critical period of visual development not solely attributable to an organic abnormality.

Synonyms

"Lazy eye"

Symptoms

Decreased visual acuity from lack of use of an eye

Signs

- Decreased accommodative ability
- *Afferent papillary defect*: typically in patients of visual acuity less than 20/200
- Eccentric viewing may view with non-foveal retinal areas (abnormal retinal correspondence)
- *Enhanced crowding phenomenon*: single optotypes are discerned better than linear ones
- Loss of stereoscopic ability
- Strong fixation preferences in preverbal children.

Investigations

- *Visual acuity testing*: at least 2-line difference in visual acuity between eyes
- History of strabismus, anisometropia, or lens opacity
- Cycloplegic refraction
- Contrast sensitivity testing
- Electrodiagnostic testing and visual-evoked potentials.

Complications

- Permanent loss of visual acuity
- Loss of stereoscopic ability
- Loss of accommodative ability
- Occlusions or penalization may uncommonly cause visual loss (typically reversible) in the initially normal eye
- Patches can cause periocular skin rashes or abrasions.

Pearls and Considerations

- Prolonged use of atropine may lead to systemic reactions, hypersensitivity reactions of the lids, irritation, redness, edema, and follicular conjunctivitis or dermatitis
- Patients treated with atropine therapy should be monitored on a regular basis for these side effects
- Amblyopia should be suspected in any strabismic child who appears to have a preference for one eye over the other
- Amblyopia should be suspected in cases of reduced visual acuity in patients with greater than or equal to 2.00 diopters (D) of difference in hyperopic refractive error, or greater than or equal to 4.00 D difference in myopic refractive error.

Differential Diagnosis

- Incorrect refraction
- Central nervous system lesions
- Optic nerve dystrophies
- Keratoconus
- Subtle foveal lesions
- High refractive errors and dishabituation to clear retinal image.

Cause

- *Strabismus*: from foveal suppression in nonfixing eye
- *Anisometropia*: from persistently blurred image in eye with greater refractive error
- *Deprivation*: from media opacity (e.g. cataract, corneal lesion)
- *Isometropic (refractive)*: from persistent binocular image blurring caused by high refractive errors; controversial (many will respond to refractive correction alone).

Epidemiology

- Incidence 2–2.5% in general population
- No *de novo* development after 5.5 years of age. Earlier onset leads to more rapid development and deeper amblyopia.

Pathology

- Unequal visual competition causing: Atrophy of the lateral geniculate nucleus in the amblyopic eye. Loss of ability to respond to light within the primary visual cortex of either eye
- *Deprivation amblyopia*: axon bodies in lateral geniculate layers 2, 3, 5 (ipsilateral) decreased 18–25% in size; deprived-eye receptive bands in cortical layer IVc are narrower than normal-eye receptive bands
- *Anisometropic amblyopia*: no human specimens available; in monkeys, pathology is similar to deprivation amblyopia but at a slightly smaller effect
- *Strabismic amblyopia*: as with deprivation amblyopia, but in the lateral geniculate; body axons receiving input from the central 10° of fixation are affected.

Diagnosis continued on p. 20

TREATMENT

Diet and Lifestyle

No special precautions are necessary.

Pharmacologic Treatment

- No first line is indicated
- *Second line*: trials of oral levodopa and carbidopa suggest efficacy in temporarily improving acuity from an average of 20/121 to 20/96 in older amblyopic children and teenagers
- Penalization (atropinization of a highly hyperopic patient's better-sighted eye to switch fixation to the amblyopic eye) is recommended in some patients.

Treatment Aims

Equalizing and maintaining visual acuity between eyes without creating a contralateral amblyopia.

Other Treatments

- Anisometropic amblyopia patients will require refractive correction
- Patients who have media opacities will require surgery for these.

Prognosis

- Most children who have strabismic amblyopia will respond to treatment if initiated before 7 or 8 years of age; children as old as approximately 15 year deserve an attempt at treatment, and many will respond.
- Children who have anisometropic amblyopia are less responsive to treatment; those with media opacities are least responsive.

Follow-up and Management

- Close follow-up to ensure compliance and prevent occlusion amblyopia
- Check vision in both eyes during visits
- Patients who have aligned eyes and peripheral fusion should be patched no more than 5 hours/day. Patients who have constant strabismus can be patched all waking hours. Patients undergoing full-time occlusion should be re-examined weekly per year of age (e.g. q 3 week in a 3-year-old) until the acuity is equal.
- Taper occlusion following maximal improvement in visual acuity.

Treatment continued on p. 21

DIAGNOSIS—cont'd

- Patients with profound amblyopia are at increased risk for accidents because as much as one-fourth of their binocular visual field may be obscured or lost secondary to their reduced vision
- Microstrabismic amblyopia (or monofixation syndrome) is often detected later than other strabismic amblyopias because the small-angle esotropia is not obvious. Careful cover test analysis should be undertaken in all children to facilitate early detection
- Compliance with occlusive treatment is more important than the type of occlusion utilized.

Commonly Associated Conditions

- Anisometropia
- Strabismus
- High refractive error
- Asymmetric or unilateral lens opacities or visual pathway disruption.

Referral Information

- Refer for surgical correction of organic causes such as cataract (Fig. 1)
- Consider referring to appropriate pediatric specialists for implementation and monitoring of patching, orthoptic devices, and medical therapy.

TREATMENT—cont'd

Nonpharmacologic Treatment

- *Occlusion of the better-sighted eye*: either all waking hours or some fraction, followed by 1–2 hours/day occlusion until the patient's ninth birthday; this can be accomplished by a patch or occluding contact lens
- *Pleoptics*: dazzling of extrafoveal retina with bright light, followed by foveal stimulation; attempted in Europe on older patients with amblyopia
- *Game-format devices*: often used together with occlusion to stimulate amblyopic fovea
- *Red-glass treatment*: red filter placed over amblyopic eye to stimulate central fixation in patients with deep amblyopia having eccentric viewing; rarely used today.

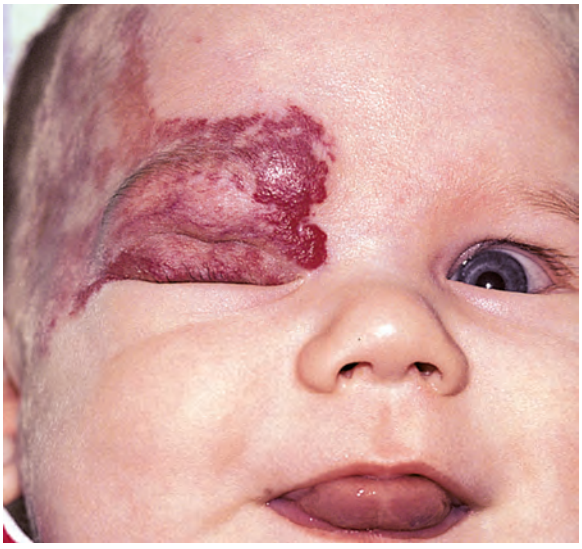

Fig. 1: Stimulus-deprivation amblyopia. A 6-month-old infant with infantile hemangioma of the right upper lid, completely covering the visual axis.

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6. Anatomically Narrow Angle (365.02)

DIAGNOSIS

Definition

A condition in which the anterior chamber angle is narrow or partially closed with an associated shallow anterior chamber, putting the patient at increased risk for angle-closure glaucoma.

Synonyms

None; associated abbreviations include ACG (angle-closure glaucoma) and CACG (chronic angle-closure glaucoma).

Symptoms

Patients are asymptomatic.

Signs

- *Hyperopia*: because these patients usually have smaller-than-average eyes, they will be hyperopic
- *Shallow peripheral anterior chamber*: generally with a van Herick grading of less than or equal to 2 when examined with the slit lamp (*see* Fig. 1)
- *Narrow angle on gonioscopy*: the anterior-chamber angle will appear quite narrow on gonioscopy, usually less than or equal to 20°; by definition, these patients do not have a closed angle or elevated intraocular pressure (IOP). However, the angle may appear optically closed, meaning that no angle structures are visible on gonioscopy, and the iris appears to be in contact with the peripheral cornea. An asymptomatic, anatomically narrow angle should open past the trabecular meshwork with pressure gonioscopy.

Investigations

Complete eye examination, including gonioscopy: in patients with critically narrow or optically closed angles, pupil dilation for funduscopy may provoke an attack of angle-closure glaucoma; such patients may be candidates for prophylactic peripheral iridectomy (*see* Nonpharmacologic Treatment).

A-scan ultrasound biometry: measurement of the anterior-chamber depth and axial length of the eye gives some indication of the risk of angle closure; an anterior-chamber depth of less than 2 mm is associated with a high risk of subsequent angle closure.

B-scan ultrasound biomicroscopy: this imaging technique allows a precise measurement of the width of the anterior-chamber angle and the relative positions of the iris root, angle structures and ciliary body.

Anterior segment OCT (optical coherence tomography): this is the most recently introduced option for imaging the anterior chamber and angle.

Complications

Angle-closure glaucoma: estimated lifetime risk is ~30% in patients with critically narrow angles.

Differential Diagnosis

Other conditions that may produce a narrow anterior-chamber angle on gonioscopy without elevated IOP or other signs of glaucoma include:

- Plateau iris configuration
- Phacomorphic angle narrowing: caused by a large lens
- Nanophthalmos
- Retinopathy of prematurity
- Subluxation of the lens
- Spherophakia.

Cause

Anatomically narrow angles result from the anatomy and configuration of the structures of the anterior segment of the eye. The eyes are smaller than average with a short axial length, shallow anterior chamber, small corneal diameter, anteriorly inserted iris root, and a normal or larger-than-normal crystalline lens. The result is an increase in the normal physiologic resistance to aqueous flow at the pupillary margin and a forward bowing and displacement of the peripheral iris toward the angle and peripheral cornea. The forward bowing of the iris may become great enough to cause the iris to adhere to the trabecular meshwork, obstruct the outflow of the aqueous, and cause angle-closure glaucoma (365.2).

Epidemiology

Population studies have found that anatomically narrow angles occur in 0.5–1.0% of the white and black American population. In some other ethnic groups (e.g. Koreans, North American Inuit), narrow angles are much more common.

Associated Features

- Hyperopia
- Small optic disks with very small or absent physiologic cups.

Diagnosis continued on p. 24

TREATMENT

Diet and Lifestyle

- Avoid drugs and activities that may dilate the pupil; such agents may provoke an attack of angle closure
- Many antihistamines and sympathomimetics found in proprietary cold and allergy medications should be avoided
- Phenothiazine and many other psychotropic drugs should be avoided
- Certain activities that cause the pupils to dilate, such as being in low-light situations (e.g. movie theater) or sexual intercourse, may cause pupillary dilation and angle-closure glaucoma in susceptible individuals.

Pharmacologic Treatment

Miotics

The use of miotics (e.g. pilocarpine) may reduce the risk of angle closure for a time. Studies have shown, however, that long-term use of pilocarpine does not prevent angle closure in high-risk individuals. With long-term use, miotics actually increase the likelihood of developing angle closure.

Treatment Aims

To widen the angle and eliminate the risk of angle closure

Prognosis

Studies have shown that the risk of angle closure in subjects with asymptomatic but very critically narrow angles is between 15% and 30%, a fairly high risk for a serious and potentially blinding disease. Many clinicians therefore advocate the use of prophylactic laser iridectomy in such patients, especially if pupil dilation is required regularly (e.g. diabetic patient at risk for retinopathy).

Follow-up and Management

Patients with critically narrowed angles should be advised of the risks and offered a laser iridectomy. Once an iridectomy has been done, the patient is no longer at risk for angle closure and may be followed as would any otherwise-normal individual. Patients who decline iridectomy should be advised of the symptoms of angle closure and instructed to seek medical attention immediately if any symptoms develop. In the absence of symptoms, patients declining iridectomy should be followed once or twice a year with gonioscopy and IOP measurements.

Treatment continued on p. 25

DIAGNOSIS—cont'd

Pearls and Considerations

- Anterior-chamber depth can be measured by ultrasound. A normal finding is generally ~3.5 mm. Anterior chambers less than or equal to 2.5 mm are considered at risk for angle closure
- Gonioscopy is contraindicated in patients with hyphema, compromised cornea or laceration of the globe
- Pilocarpine may result in severely blurred vision in patients with central lenticular opacities. A full ophthalmic examination should be performed to rule out such opacities before initiating pilocarpine therapy
- Pilocarpine should be used with caution in patients with cholelithiasis, biliary tract disease, cardiovascular disease, and pulmonary disease.

Referral Information

Referral should be considered for prophylactic peripheral iridotomy. Appropriate glaucoma workup should be obtained in patients with suspected acute or chronic angle-closure events; and in those with high IOP, referral for trabeculectomy or filtering surgery should be considered to lower pressure adequately.

TREATMENT—cont'd

Nonpharmacologic Treatment

Laser Iridectomy

Laser iridectomy is definitive treatment. By creating an alternative route for the aqueous from the posterior chamber to the anterior chamber, the relative pupil block and the forward bowing of the iris are eliminated. This allows the angle to widen and removes the risk of future angle closure. The treatment is almost 100% effective and very safe. Serious complications following laser iridectomy are extremely rare.

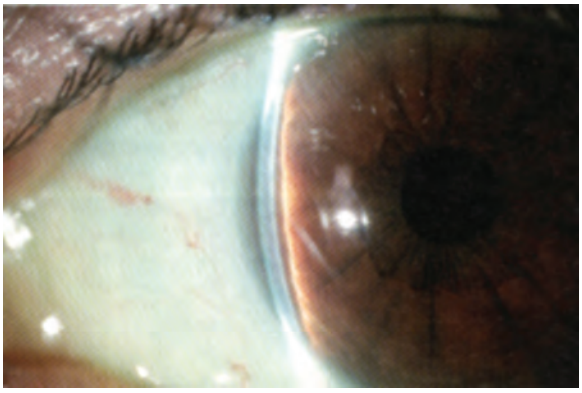

Fig. 1: Anatomically narrow angle. Van Herick test demonstrating shallow peripheral anterior chamber with the slit-lamp beam.

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7. Angioid Streaks (363.43)

DIAGNOSIS

Definition

Full-thickness breaks in calcified, thickened Bruch's membrane with disruption of overlying retinal pigment epithelium (RPE).

Synonyms

None

Symptoms

- Patients are usually asymptomatic in the early course
- Gradual decrease in vision: common with increase in age
- Loss of vision to legal blindness: seen in ~50% of patients
- Scotoma
- Metamorphosia.

Signs

- Radially oriented cracks in the pigment layer emanating from the optic nerve (*see* Fig. 1)
- "Peau d'orange" or mottled appearance of the RPE (*see* Fig. 2)
- Peripheral atrophic spots
- Disk drusen
- Subretinal crystalline deposits
- Subretinal hemorrhage: if associated with trauma or choroidal neovascularization
- Choroidal neovascular membrane (CNVM).

Investigations

Physical examination: to rule out systemic disorders, such as pseudoxanthoma elasticum (PXE), Ehlers-Danlos syndrome, Paget's disease and sickle cell disease.

Fluorescein angiogram: if choroidal neovascularization is suspected.

Serum alkaline phosphatase and urine calcium: if considering Paget's disease.

Sickle cell prep and hemoglobin electrophoresis: if considering sickle cell disease.

Complications

- High risk of subretinal bleeding secondary to blunt trauma
- Retinal pigment epithelium detachment
- Choroidal neovascularization
- Disciform scarring.

Differential Diagnosis

- Senile angioid streaks
- Choroidal rupture secondary to trauma
- Senile macular degeneration
- Idiopathic choroidal neovascularization
- Myopia with lacquer cracks
- Presumed ocular histoplasmosis.

Cause

- Idiopathic, 50%
- Pseudoxanthoma elasticum, 34%
- Paget's disease, 10%
- Sickle cell hemoglobinopathies, 6%
- Ehlers-Danlos syndrome
- Greenblad-Stranberg syndrome.

Epidemiology

- Fifty percent of patients with angioid streaks will have systemic associations.
- Eighty-five percent of patients with PXE have evidence of angioid streaks.
- Eight to fifteen percent of patients with Paget's disease have angioid streaks.

Associated Features

- Peau d'orange appearance
- Pseudoxanthoma elasticum "plucked-chicken skin" gastrointestinal tract bleeding, cardiac abnormalities.

Pathology

- Thickening, elastic degeneration, and calcification of Bruch's membrane
- Breaks in the elastic and collagenous layers of Bruch's membrane
- Fibrovascular ingrowth
- Secondary RPE atrophy, choriocapillaris damage, photoreceptor loss, RPE hypertrophy, serous retinal detachment, disciform scarring.

Diagnosis continued on p. 28

TREATMENT

Diet and Lifestyle

Safety glasses should be worn, because trauma precipitates hemorrhage.

Pharmacologic Treatment

A series of intravitreal anti-VEGF injections (Avastin or Lucentis) are performed if choroidal neovascularization is present. Due to the natural history of the RPE/Bruch's membrane pathology, patients do not experience as much visual improvement as seen in macular degeneration patients.

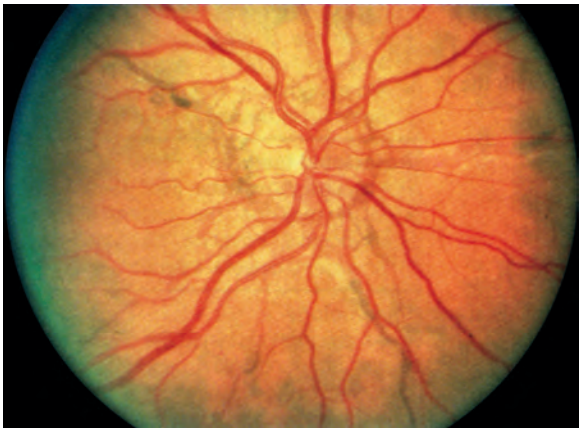

Fig. 1: An angioid streak is seen coursing radially from the disk into the inferior macula in a patient with pseudoxanthoma elasticum. The streak is grey and deep to the retina, in contrast to the normal red vessels within the inner retina.

Treatment Aims

- To educate patients and detect secondary choroidal neovascularization early
- Low-vision aids may be useful
- Genetic counseling should be considered for hereditary conditions.

Prognosis

Long-term prognosis is guarded. Patients can lose vision to the level of legal blindness if the streaks involve the fovea or if secondary choroidal neovascularization in the fovea occurs.

Follow-up and Management

- Amsler grid testing daily
- Dilated fundus examination every 6 months.

Treatment continued on p. 29

DIAGNOSIS—cont'd

Pearls and Considerations

- In patients with angioid streaks, minor trauma can result in rupture of Bruch's membrane with resultant hemorrhage or choroidal neovascularization
- Patients with angioid streaks benefit from regular Amsler grid testing to monitor for visual changes associated with formation of CNVM
- The color of angioid streaks depends on natural fundus coloration and the amount of overlying RPE atrophy. Streaks appear red in blond fundi or medium to dark brown in more pigmented fundi.

Referral Information

- Patients who present with associated choroidal neovascularization should be referred for anti-VEGF injections (e.g. Avastin or Lucentis) or PDT as appropriate
- Patients identified with angioid streaks should have an appropriate medical referral to rule out underlying systemic disease. Evaluation should include skin biopsy and photographs
- Asymptomatic family members with inherited diseases such as pseudoxanthoma elasticum, may benefit from testing as well as monitoring for any ocular manifestations.

TREATMENT—cont'd

Nonpharmacologic Treatment

Amsler grid testing: for early detection of choroidal neovascularization.

Laser photocoagulation: of choroidal neovascularization.

- Thermal laser often causes further damage to the Bruch's/RPE complex, so it is no longer the treatment of choice even for extrafoveal lesions
- Prophylactic laser treatment should not be performed because it may induce choroidal neovascularization
- Photodynamic therapy (PDT) may be an option in certain cases, but it, too, can further damage the pigment epithelium and Bruch's membrane.

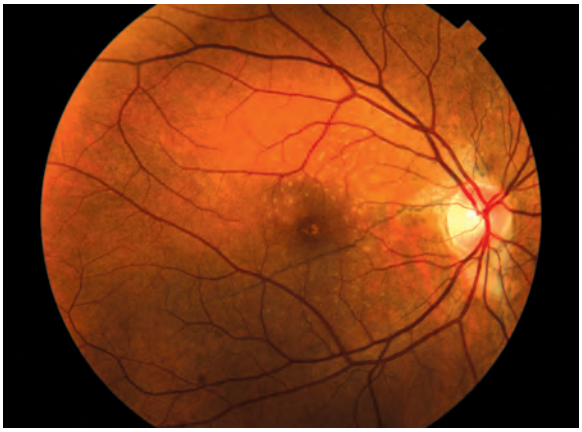

Fig. 2: Patient with pseudoxanthoma elasticum and "peau d'orange" appearance to the retinal pigment epithelium.

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8. Anterior Ischemic Optic Neuropathy, Arteritic (Giant Cell Arteritis, Temporal Arteritis) (377.30)

DIAGNOSIS

Definition

Ischemia of the anterior optic nerve head secondary to underlying systemic granulomatous vasculitis of the large- and medium-sized blood vessels; a sight-threatening medical emergency.

Synonyms

Giant cell arteritis (GCA), temporal arteritis; abbreviated as AAION.

Symptoms

Acute unilateral or bilateral blindness: 2–9 weeks after onset of headaches.
Premonitory amaurosis fugax: May present in 10% of patients.
Amaurosis: Induced by bright light.
Painful diplopia: May present in 6% of patients.

Systemic Symptoms

- Recent headache in temple, occipital region, neck, eye, or ear; jaw claudication; tongue pain and numbness; toothache; intermittent fevers; night sweats; anorexia, weight loss; malaise; depression
- Proximal muscle pain, myelopathy, polymyalgia rheumatica.

Note: Many patients may have no constitutional complaints and will present only with the eye sign (occult GCA).

Headache

- Headache is the initial manifestation of GCA in 50–90% of patients
- Head pain is a "different kind of headache" that can be severe and boring
- Headache is worse at night and with exposure to cold
- There is tenderness of the scalp overlying the greater superficial temporal arteries. These arteries may appear enlarged, nodular, and erythematous. Patients may complain of sensitivity when brushing hair or placing head on pillow because of scalp and temple tenderness.
- Pain may involve neck, face, jaw, tongue, ear, or throat. Jaw claudication and neck pain have a high specificity.

Signs

- Afferent pupillary defect (APD), if unilateral
- Anterior ischemic optic neuropathy (AION) with pallid disk swelling (Figs 1A and B), bilateral AION, central retinal artery occlusion (CRAO), branch retinal artery occlusion (rare), combined CRAO and AlON, cilioretinal artery occlusion (combined with any of the above) (Figs 1A and B).
- Choroidal ischemia, infarcts of the nerve fiber layer, pupil-sparing third-nerve paresis, sixth- and seventh-nerve palsies, ischemic ocular syndrome.

Differential Diagnosis

- Idiopathic anterior ischemic optic neuropathy
- Acute angle-closure glaucoma
- Herpes zoster sine eruptione
- Aneurysm
- Temporomandibular joint syndrome
- Pituitary apoplexy
- Carotid artery dissection
- Wegener's granulomatosis
- Systemic cholesterol microembolization syndrome
- Carotid-cavernous fistula.

Cause

Autoimmune vasculitis of the elderly of unknown cause.

Epidemiology

- Overall incidence, 2.9:100,000; 50–59 years of age, 1.7:100,000; more than 80 years of age, 55.5:100,000.
- Mean age of presentation is 75 years with lower limit of 50 years.
- More common in white women; less common in black and Asian populations.

Associated Features

- Polymyalgia rheumatica. (Ret. 4)
- Giant cell arteritis may be the cause of death (myocardial infarction, dissecting aortic aneurysm, cerebral infarction).

Immunology

- An autoimmune syndrome develops from the immunologic response directed toward antigens residing in the wall of medium-sized arteries
- Genetic risk is supported by a sequence motif in the *HLA-DRBI* gene
- T lymphocytes undergoing clonal expansion have been demonstrated infiltrating the temporal artery wall
- A small number of tissue-infiltrating T cells produce interferon-γ, an important cytokine governing the disease process
- Circulating macrophages secrete interleukin-6, the major inducer of acute-phase reactants.

Pathology

- Active disease is characterized by fragmentation and destruction of the internal elastic lamina and inflammatory infiltrate in the vessel wall.
- Healed vasculitis shows a diffuse intimal thickening, intimal and medial fibrosis and fragmentation or loss of internal elastic lamina.

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