CHAPTER 9

Choroidal Disease

Highlights

- Pachychoroid is an entity with an evolving definition but is generally thought of as abnormally dilated choroidal vessels abutting the retinal pigment epithelium (with or without choroidal thickening).
- Fluorescein angiography can contribute to the diagnosis of arteritic anterior ischemic optic neuropathy.
- Hypertensive choroidopathy can be diagnosed by means of multimodal imaging because of its unique characteristics on optical coherence tomography and fundus autofluorescence.

Scope of Chapter

This chapter describes noninflammatory choroidal diseases that also involve the retina. Inflammatory disorders of the retina and choroid are discussed in Chapter 11. See also BCSC Section 9, *Uveitis and Ocular Inflammation*. Intraocular tumors such as melanoma are covered in BCSC Section 4, *Ophthalmic Pathology and Intraocular Tumors*.

Central Serous Chorioretinopathy

Central serous chorioretinopathy (CSC; also called *central serous retinopathy [CSR])* causes an idiopathic serous detachment of the retina related to leakage at the level of the retinal pigment epithelium (RPE) secondary to hyperpermeability of the choriocapillaris, as seen on indocyanine green angiography (ICGA). While the condition was originally described in 1866 by von Graefe as recurrent central retinitis, it was Maumenee who first performed fluorescein angioscopy on patients with CSC and found a leak at the level of the RPE, not from retinal vessels (the previously hypothesized source of leakage). Gass subsequently described the findings seen on fluorescein angiography (FA) and suggested that laser photocoagulation could be used to treat affected patients. Gass also stated that the disease was secondary to hyperpermeability of the choriocapillaris, a hypothesis that was confirmed decades later via ICGA.

Demographics and Features

Central serous chorioretinopathy occurs primarily in persons between the ages of 35 and 55 years, with a male-to-female ratio of 3:1; at present, there are no reliable statistics

suggesting any association with race. Patients describe a variety of symptoms, including sudden onset of blurred or dim vision, micropsia, metamorphopsia, paracentral scotomata, decreased color vision, and prolonged afterimages. Visual acuity ranges from 20/20 to 20/200, but in most patients, it is better than 20/30. Decreased visual acuity can often be improved with a small hyperopic correction.

CSC has several clinical variations. In an acute manifestation, the retina has a round or oval elevation in the macular region; it often involves the fovea. FA shows leaks from the RPE that may appear early in the angiographic sequence as a dot (the *dot* form) or as a tree-shaped movement of dye in the subretinal space (the *smokestack* form) (Fig 9-1). In some circumstances, vigorous leaks can cause deposition of a grayish-white, feathered-edge subretinal material that is generally believed to be fibrin. In chronic CSC, the RPE

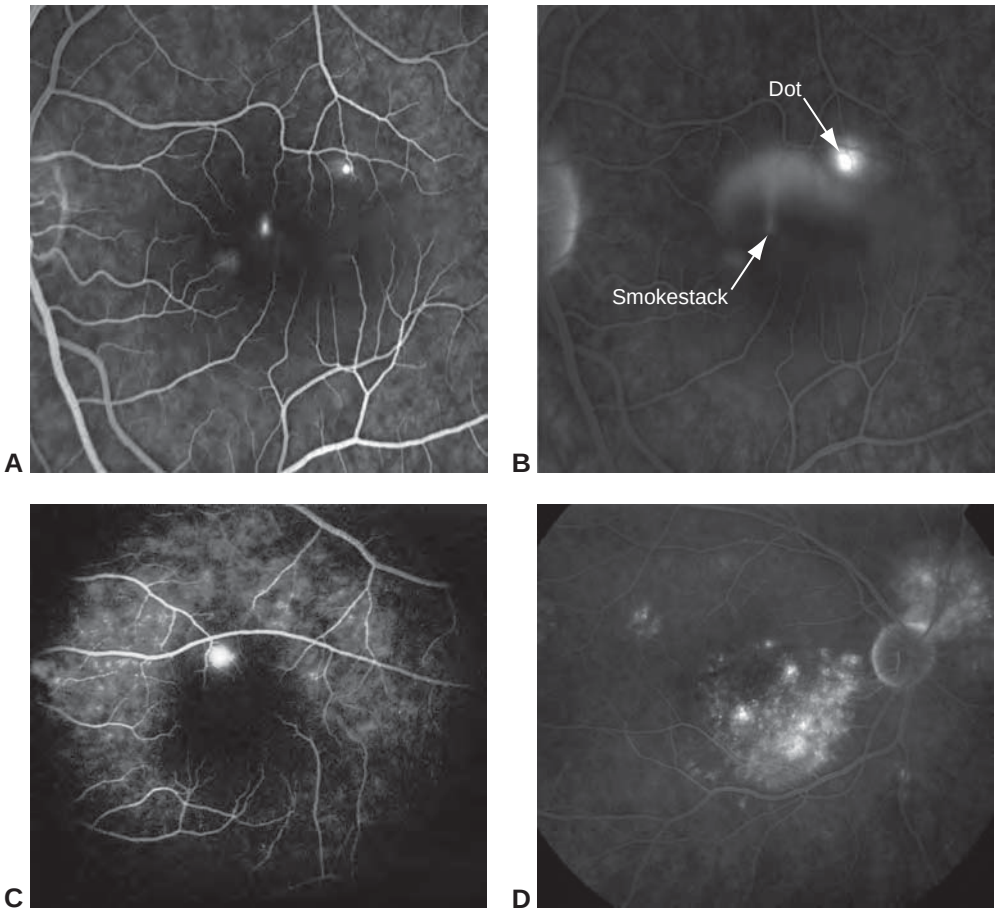

Figure 9-1 Fluorescein angiographic (FA) patterns of the leaks in central serous chorioretinopathy (CSC). **A,** In the early phase, the patient was seen to have 2 main leaks. **B,** Later in the angiogram, the leaks show 2 different morphologies: the *smokestack* and the *dot* varieties. **C,** Acute CSC generally presents with 1 leak or a few leaks. **D,** Chronic forms of CSC exhibit many small leaks, as demonstrated in this image from a different patient. *(Courtesy of Richard F. Spaide, MD.)*

shows granular pigmentation; FA reveals many small, sometimes inconspicuous leaks; and there is widespread shallow detachment with areas of atrophy of the photoreceptors (see Fig 9-1D).

CLINICAL PEARL

Retinal pigment epithelial changes on optical coherence tomography (OCT) or fundus autofluorescence (FAF) may suggest previous episodes of CSC and resolved subretinal fluid.

Systemic Associations

Central serous chorioretinopathy is associated with stress and with a tense, driven personality. Systemic associations include endogenous hypercortisolism (Cushing syndrome), hypertension, sleep apnea, use of psychopharmacologic medications, and pregnancy. Use of systemic corticosteroids, which may be administered through intramuscular, topical, inhalational, epidural, or even intra-articular routes, is associated with CSC; but, curiously, use of intraocular corticosteroids does not appear to be associated with the condition. Organ transplant recipients and patients with autoimmune disease requiring long-standing, high-dose steroids are particularly vulnerable to the more severe and chronic variants.

Fawzi AA, Holland GN, Kreiger AE, Heckenlively JR, Arroyo JG, Cunningham ET Jr. Central serous chorioretinopathy after solid organ transplantation. *Ophthalmology.* 2006;113(5): 805–813.e5.

Mrejen S, Balaratnasingam C, Kaden TR, et al. Long-term visual outcomes and causes of vision loss in chronic central serous chorioretinopathy. *Ophthalmology.* 2019;126(4): 576–588.

Imaging

The extent of the detachment can be documented with color fundus photographs. FAF imaging shows the accumulation of shed photoreceptor outer segments in the subretinal space, as well as distributed defects of the RPE. It has been theorized that the white dots seen under the retina are macrophages with fluorophores from phagocytized outer segments (Fig 9-2). Eyes with chronic CSC can display descending tracts during both FA and FAF imaging (Fig 9-3). Enhanced depth imaging OCT (EDI-OCT) shows thickening of the choroid and, in areas where thickening is most prominent, posterior loculation of fluid in the deep choroid. Figure 9-4 shows the internal structure of a healthy choroid, and Figure 9-5 shows the choroid in 1 healthy eye and in 3 eyes with CSC.

Although ICGA can reveal choroidal vascular hyperpermeability (Fig 9-6), it has largely been supplanted by OCT, even for detecting possible coexisting choroidal neovascularization (CNV), which may be present in up to 20% of cases in individuals older than 50 years. OCT angiography seems to be adept at detecting secondary CNV that may develop in these patients.

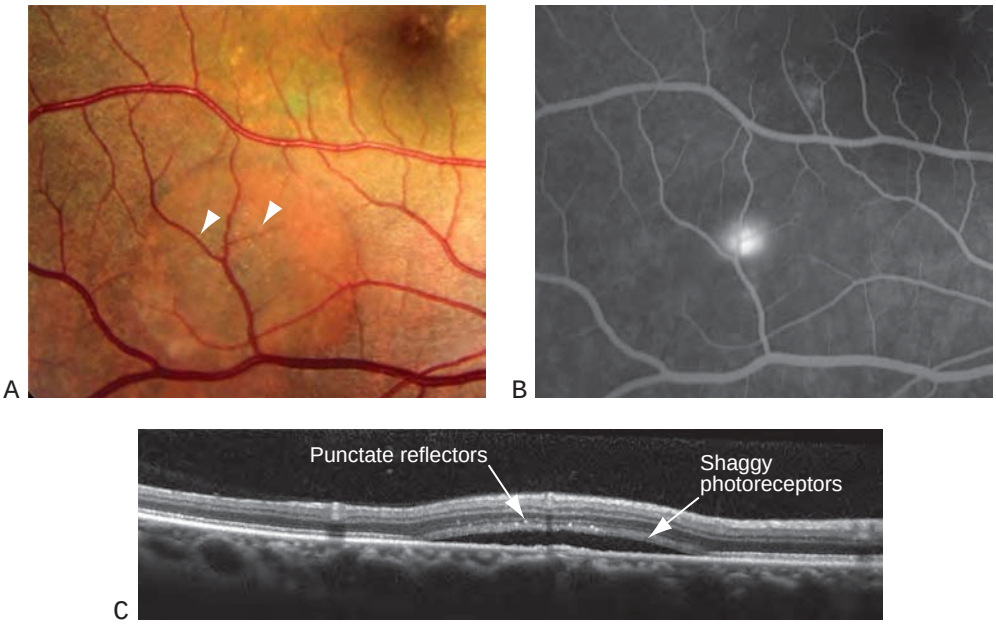

Figure 9-2 Central serous chorioretinopathy with white dots. **A,** Fundus photograph shows an ovoid elevation of the retina with white dots on the undersurface *(arrowheads)*. **B,** FA reveals a single leakage point. **C,** The elevated retina, seen in cross section, has a thick coat on its inner surface that has autofluorescent characteristics consistent with retinal outer segment–derived fluorophores. These fluorophores are therefore considered to be derived from the outer segments that could not be phagocytized by the retinal pigment epithelium (RPE) because of the physical separation, caused by the fluid, between the retina and RPE. The region of shaggy photoreceptors contains punctate dots that are highly reflective; it has been theorized that these dots are macrophages. *(Courtesy of Richard F. Spaide, MD.)*

Imamura Y, Fujiwara T, Margolis R, Spaide RF. Enhanced depth imaging optical coherence tomography of the choroid in central serous chorioretinopathy. *Retina.* 2009;29(10): 1469–1473.

Spaide RF, Klancnik JM Jr. Fundus autofluorescence and central serous chorioretinopathy. *Ophthalmology.* 2005;112(5):825–833.

Differential Diagnosis

Other entities that may be considered in the differential diagnosis of CSC include type 1 CNV and polypoidal choroidal vasculopathy (PCV; see Chapter 4), which is a variant of type 1 CNV. The FA findings can overlap significantly; both entities show leakage of fluorescein, and the visualization of the structures underlying the RPE is poor. If CNV is present, OCT demonstrates an irregular wavy, shallow elevation of the RPE by a layer of material with heterogeneous reflectivity. The neovascularization seen in association with CSC is generally easy to detect with OCT angiography. What complicates the issue, particularly regarding treatment, is that type 1 CNV and PCV appear to be associated with CSC; they may be its sequelae.

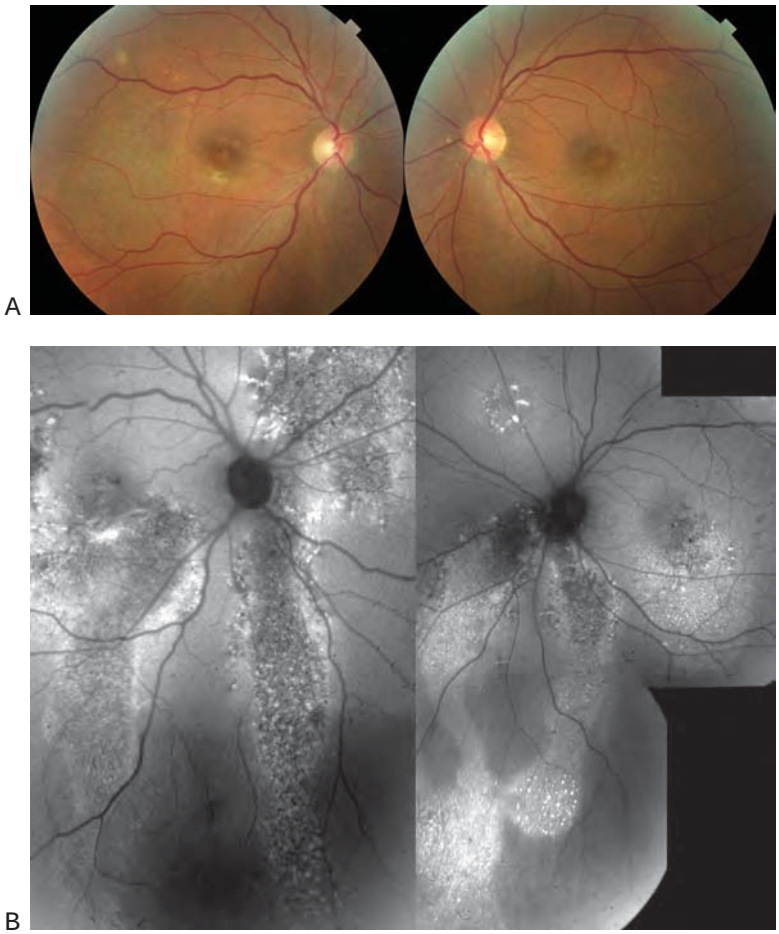

Figure 9-3 Autofluorescence abnormalities in CSC. **A,** Fundus photographs show the right and left eyes of a patient with CSC. Although subtle pigmentary changes are visible, it can be difficult to discern where fluid has accumulated. **B,** Autofluorescent images show widespread abnormalities induced by the presence of subretinal fluid, particularly the descending tracts created by the fluid. *(Reproduced with permission from Elsevier. Imamura Y, Fujiwara T, Spaide RF. Fundus autofluorescence and visual acuity in central serous chorioretinopathy.* Ophthalmology. *2011;118(4):700–705. Copyright 2011.)*

Pachychoroid spectrum and its evolving nomenclature

The widespread utilization of EDI-OCT has facilitated quantitative evaluation of the choroid and, with it, the recognition of a thick, or *pachy,* choroid phenotype, which is commonly seen in CSC. The spectrum of disease encompassed in this entity ranges from pigment epithelial changes in the setting of thick, poorly tessellated choroid *(pachychoroid pigment epitheliopathy)* to *pachyvessels,* which on cross-sectional OCT appear to compress and obliterate the overlying choriocapillaris, to PCV (see Chapter 4 for further discussion). Ophthalmologists have recently recognized that increased choroidal thickness may not be universally present in these eyes and have moved toward a more pathogenesis-based definition, with the understanding that these pachyvessels are actually dilated anastomotic

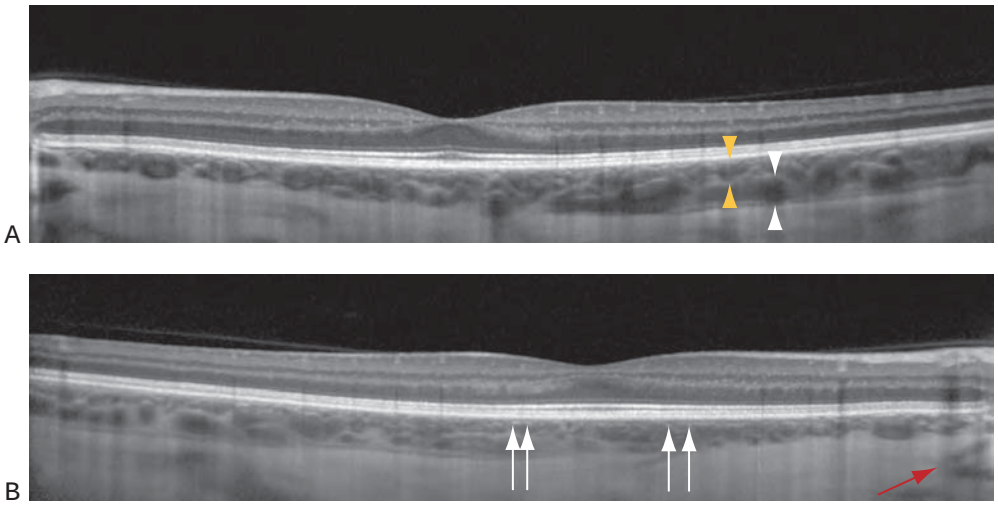

Figure 9-4 The internal structure of the healthy choroid, depicted on enhanced depth imaging optical coherence tomography (EDI-OCT). The choroidal vessels decrease in diameter from the outer to the inner choroid. **A,** The larger vessels *(white arrowheads)* are dark in the center with a thick hyperreflective wall. The medium-sized vessels *(yellow arrowheads)* have a smaller hyporeflective area in the center and a hyperreflective wall. **B,** As vessel diameter decreases, the central hyporeflective area decreases until it is not visible. At that size, the vessel appears as a white hyperreflective structure *(white arrows)*. Note the delineation of the hyporeflective line near the junction with the inner sclera, which appears to be in the suprachoroidal space. The *red arrow* points to a vessel coursing through the sclera. *(Reproduced with permission from Elsevier. Mrejen S, Spaide RF. Optical coherence tomography: imaging of the choroid and beyond.* Surv Ophthalmol. *2013;58(5):387–429. Copyright 2013.)*

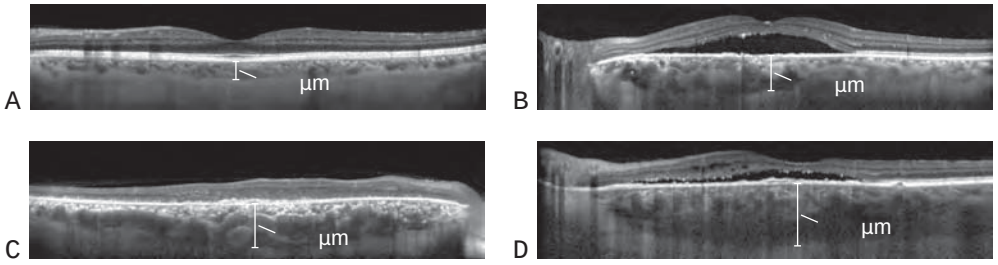

Figure 9-5 The choroid is seen in cross section using EDI-OCT. Subfoveal choroidal thickness was measured vertically from the outer border of the RPE to the inner border of the sclera *(brackets)* in a healthy eye in a 55-year-old man **(A)** and in 3 representative eyes with CSC: in a 44-year-old man **(B),** a 57-year-old man **(C),** and a 63-year-old man **(D).** *(Reproduced with permission from Imamura Y, Fujiwara T, Margolis R, Spaide RF. Enhanced depth imaging optical coherence tomography of the choroid in central serous chorioretinopathy.* Retina. *2009;29(10):1469–1473. doi:10.1097/IAE.0b013e3181be0a83)*

venous channels that run across the choroidal watershed zones, traversing the macula. These observations signal choroidal venous insufficiency as the potential unifying underlying pathogenesis of these entities.

Cheung CMG, Lee WK, Koizumi H, Dansingani K, Lai TYY, Freund KB. Pachychoroid disease. *Eye (Lond).* 2019;33(1):14–33.

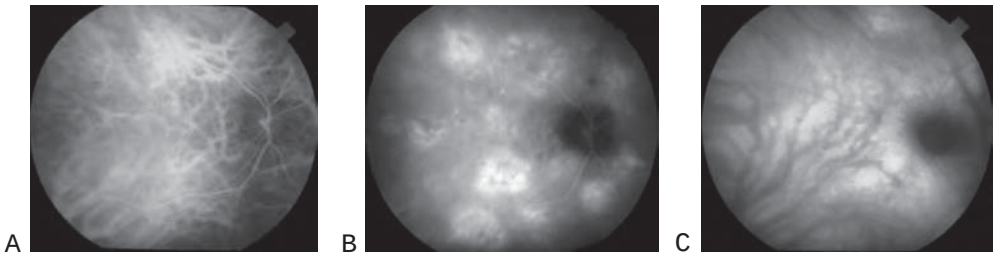

Figure 9-6 Stages of indocyanine green angiography (ICGA) in CSC. **A,** Early after injection, the dye can be seen within the choroidal vessels. **B,** During the middle phase of the angiogram, choriocapillaris hyperpermeability results in the appearance of multiple hyperfluorescent clouds. **C,** Later in the angiogram, the dye has largely been removed from the choroidal vessels. Dye that has leaked into the stroma has diffused posteriorly, silhouetting the larger choroidal vessels. *(Reproduced with permission from Spaide RF, Hall L, Haas A, et al. Indocyanine green videoangiography of older patients with central serous chorioretinopathy.* Retina. *1996;16(3):203–213. doi:10.1097/00006982-199616030-00004)*

Matsumoto H, Hoshino J, Mukai R, et al. Vortex vein anastomosis at the watershed in pachychoroid spectrum diseases. *Ophthalmol Retina.* 2020;4(9):938–945.

Warrow DJ, Hoang QV, Freund KB. Pachychoroid pigment epitheliopathy. *Retina.* 2013;33(8):1659–1672.

Treatment

Central serous chorioretinopathy is generally self-limited and resolves spontaneously, with the majority of patients attaining excellent visual results. However, it can be destructive in some chronic cases, causing visually significant scotomata. *Verteporfin photodynamic therapy (PDT),* whether at full fluence or, more commonly, reduced fluence, has been shown to decrease or eliminate subretinal fluid, decreases choroidal thickness, and reduces choroidal vascular hyperpermeability. Treatment is guided by either FA or ICGA, and a sufficiently large spot size is used to cover the main leakage point(s) with a surrounding safety margin of 1000 μm. This therapy (at full fluence, in studies of age-related macular degeneration) is associated with a 4% risk of vision decrease. CSC may recur after successful PDT, and therapy can be repeated safely, especially if lower fluence is used and the lesion is extrafoveal.

Laser photocoagulation therapy is no longer preferred for CSC, as secondary CNV occurred in the immediate postoperative period in up to 2% of eyes treated with this modality. Moreover, unlike PDT, photocoagulation has no effect on choroidal thickness.

Use of mineralocorticoid receptor antagonists (eplerenone or spironolactone) has shown some benefit in anecdotal reports. However, a recent randomized placebo-controlled trial using eplerenone in chronic CSC demonstrated no benefit with the medication (25 mg/day for 1 week, increasing to 50 mg/day for up to 12 months).

Lotery A, Sivaprasad S, O'Connell A, et al. Eplerenone for chronic central serous chorioretinopathy in patients with active, previously untreated disease for more than 4 months (VICI): a randomised, double-blind, placebo-controlled trial. *Lancet.* 2020;395(10220):294–303.

Maruko I, Iida T, Sugano Y, Ojima A, Ogasawara M, Spaide RF. Subfoveal choroidal thickness after treatment of central serous chorioretinopathy. *Ophthalmology.* 2010;117(9):1792–1799.

Choroidal Perfusion Abnormalities

The choroid receives its arterial supply from approximately 20 short posterior ciliary arteries and 2 anterior ciliary arteries. A network of branching arterioles distributes the blood throughout the choroid in a segmental fashion, ultimately leading to the choriocapillaris, and helps reduce the blood pressure as well. Although the vessels in the choriocapillaris exhibit relatively uniform patterns in any given region of the eye, the pressure gradients imposed by the feeding arterioles and draining venules establish a lobular perfusion pattern. Abnormalities in choroidal blood flow can be divided into several main categories based on the underlying disease process.

Arteritic Disease

In arteritic diseases such as *giant cell arteritis* (Figs 9-7, 9-8) or *granulomatosis with polyangiitis* (formerly called *Wegener granulomatosis;* Fig 9-9), inflammatory occlusion can cause sectorial areas of nonperfusion. FA or ICGA is typically performed in cases in which an arteritic cause of vision loss is suspected; flow defects in the choroid are often undetected by ophthalmoscopy alone. Patchy and delayed choroidal filling, especially around the optic nerve, is characteristic of arteritic ischemic optic neuropathy (see Fig 9-7).

Nonarteritic Disease

Nonarteritic problems with blood flow can occur as a result of embolic or systemic disease or as a manifestation of severe hypertension. Emboli from the heart, injection of corticosteroids or calcium hydroxylapatite, and intravascular coagulation all have the potential to occlude choroidal vessels. Vascular occlusion can also occur in patients with lupus anticoagulants.

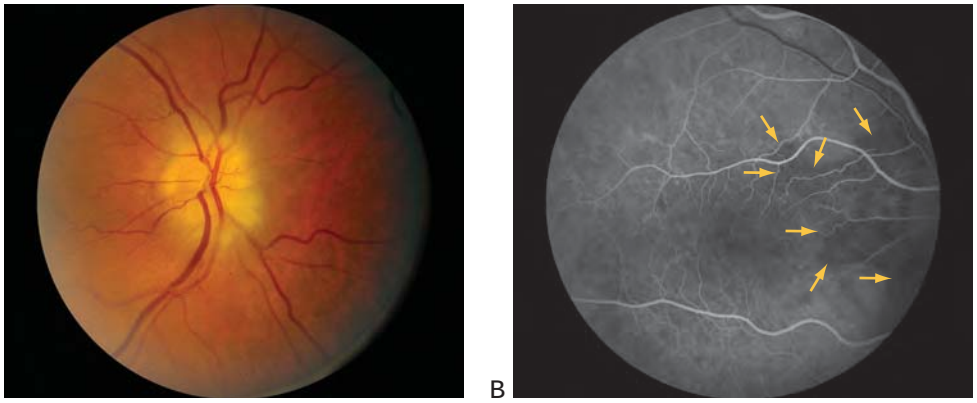

Figure 9-7 Arteritic anterior ischemic optic neuropathy with delayed choroidal filling. **A,** Color fundus photograph showing pallid edema of the right optic nerve head. **B,** Early venous phase of the angiogram showing incomplete filling of the choroidal lobule surrounding the optic nerve *(arrows)*. Normally, choroidal filling is complete by the early venous to mid-venous phase. *(Courtesy of Nicholas J. Volpe, MD.)*

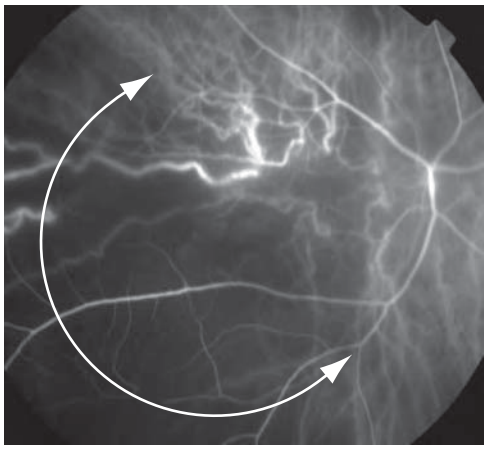

Figure 9-8 Giant cell arteritis. ICGA image taken 1 day after this patient had severe vision loss secondary to arteritic anterior ischemic optic neuropathy. A wedge-shaped area of choroidal nonperfusion is apparent *(curved arrow)*. The apex of the wedge of nonperfusion points toward the area of the occluded short posterior ciliary artery. *(Courtesy of Richard F. Spaide, MD.)*

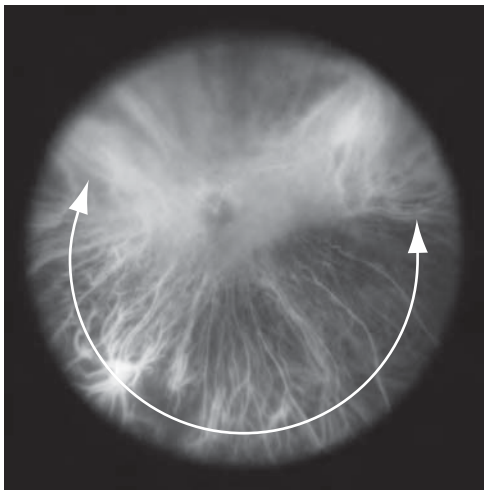

Figure 9-9 Granulomatosis with polyangiitis (formerly, Wegener granulomatosis). The early phase of wide-angle ICGA of the left eye reveals a widespread filling defect of the arterioles and choriocapillaris in the inferior fundus *(curved arrow)* and in a segmental area of the superior fundus *(asterisk)*. *(Reproduced with permission from Elsevier. Iida T, Spaide RF, Kantor J. Retinal and choroidal arterial occlusion in Wegener's granulomatosis.* Am J Ophthalmol. *2002;133(1):151–152. Copyright 2002.)*

Thrombotic thrombocytopenic purpura causes a classic pentad of findings: (1) microangiopathic hemolytic anemia, (2) thrombocytopenia, (3) fever, (4) neurologic dysfunction, and (5) renal dysfunction. Patients with this condition may have multifocal yellow placoid areas and associated serous detachment of the retina. Similar fundus findings may occur in patients with *disseminated intravascular coagulation,* in which consumption of coagulation proteins, involvement of cellular elements, and release of fibrin degradation products lead to hemorrhage from multiple sites and ischemia from microthrombi.

Similar fundus findings also occur in patients with acute hypertension, such as malignant hypertension or eclampsia. In addition to causing retinal and optic nerve head abnormalities, these disorders commonly lead to serous detachment of the retina associated with areas of yellow placoid discoloration of the RPE (Fig 9-10). The perfusion

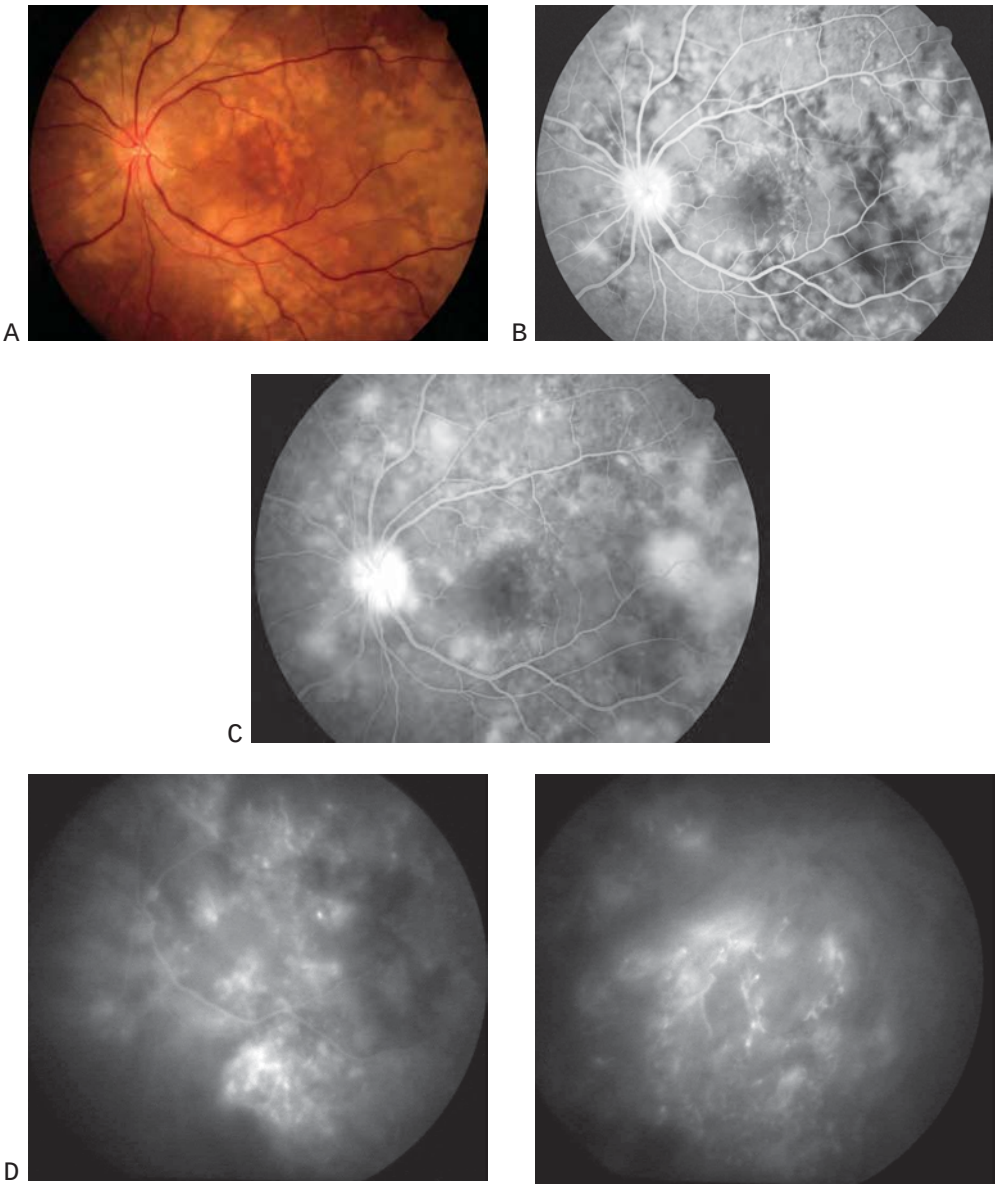

Figure 9-10 Preeclampsia with hemolysis, elevated liver enzymes, and low platelet count (HELLP) syndrome. **A,** Fundus photograph reveals a serous detachment of the retina and multiple yellowish placoid areas at the level of the RPE and inner choroid. **B,** Early-phase FA image shows reticular patterns of decreased choroidal perfusion bordering areas of hyperfluorescence. Early leakage from the level of the RPE is evident and becomes more apparent in the later phases of the study **(C).** There is also staining of and leakage from the optic nerve. **D,** ICGA image shows profound choroidal vascular filling defects alternating with areas of abnormal vessel leakage and staining, a rare finding. **E,** In the late phase, numerous arterioles show staining of their walls, indicating severe vascular damage. *(Reproduced with permission from Spaide RF, Goldbaum M, Wong DW, Tang KC, Iida T. Serous detachment of the retina.* Retina. *2003;23(6):820–846.)*

abnormalities may range from focal infarction of the choriocapillaris to fibrinoid necrosis of larger arterioles. Resolution of smaller infarcts, which initially appear tan in color, produces small patches of atrophy and pigmentary hyperplasia called *Elschnig spots* (Fig 9-11). Infarction of an arteriole can lead to *Siegrist streaks,* which are hyperpigmented flecks arranged linearly along choroidal vessels. Posterior ciliary artery occlusions can result in wedge-shaped zones of choroidal infarction called *Amalric triangles*.

Choriocapillaris Blood Flow Abnormalities

Choroidal blood flow defects affect lobule-sized areas of the choroid or areas supplied by arterioles and therefore affect 1 to several choroidal lobules. Many ocular diseases (eg, acute

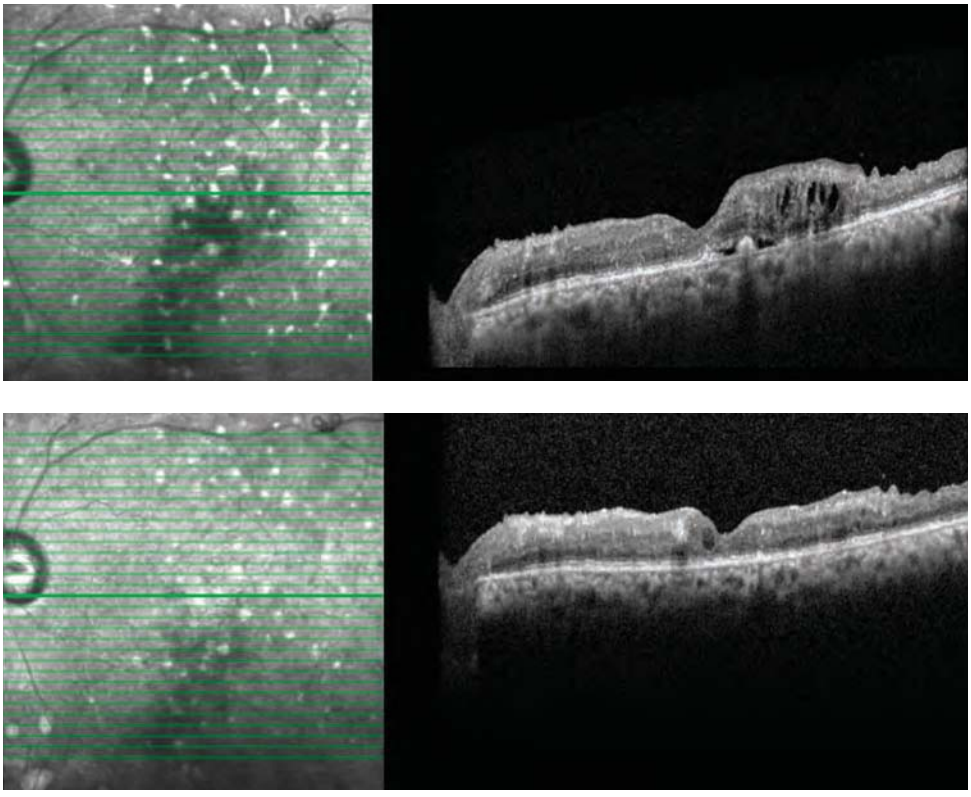

Figure 9-11 OCT images from a 43-year-old man with a history of hypertension, type 2 diabetes, chronic kidney disease, and proliferative diabetic retinopathy who had previously undergone vitrectomy for bilateral traction retinal detachment. He presented to the emergency department with a hypertensive urgency requiring admission and initiation of dialysis for acute renal failure. **A,** Imaging performed 2 months after these events. Notably, hyperreflective lesions on the infrared (IR) image *(to the right of the OCT image)* were new and not present at prior ophthalmologic visits. OCT shows sub-RPE deposits and subretinal and intraretinal fluid, consistent with hypertensive choroidopathy, and Elschnig spots. The patient was observed without retinal intervention while blood pressure and renal issues resolved. **B,** Vision stabilized and fluid resolved without further ocular intervention, with some fading of the IR hyperreflective lesions and inner retinal thinning over the following 18 months. *(Courtesy of Amani Fawzi, MD.)*

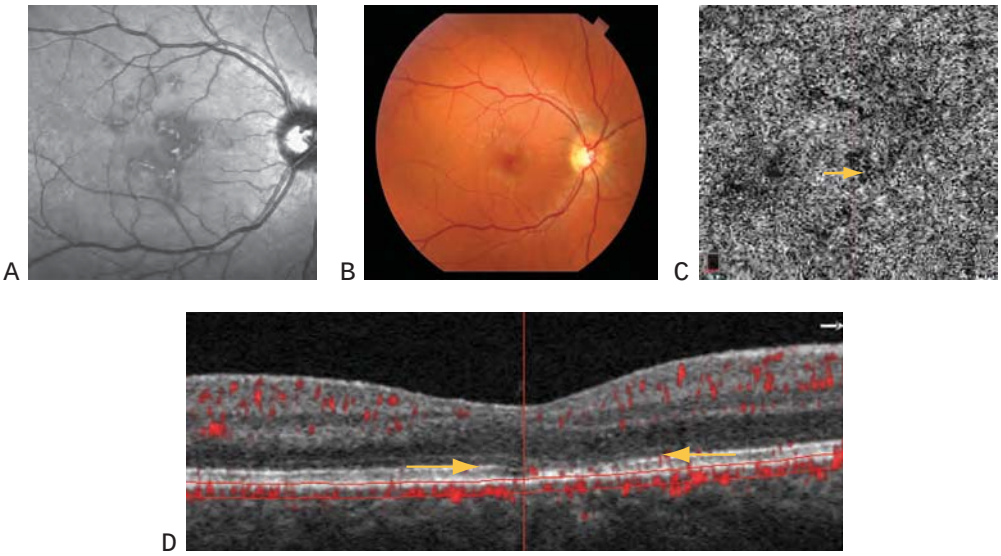

Figure 9-12 Resolving case of acute posterior multifocal placoid pigment epitheliopathy (APMPPE). **A,** Infrared image shows the hallmark jigsaw pattern pigmentary changes. **B,** These changes are visible as hyporeflective lesions on the fundus photograph. **C,** OCT angiography focused on the choriocapillaris shows focal flow deficit *(arrow)*. **D,** Corresponding outer retinal lesion on OCT is seen as thinning and disruption of the ellipsoid zone *(between arrows)*. *(Courtesy of Amani Fawzi, MD.)*

posterior multifocal placoid pigment epitheliopathy; Fig 9-12) characteristically produce lesions that are the putative size of a choroidal lobule. On OCT angiography at the level of the choriocapillaris, multiple areas of signal voids are frequently seen and are consistent with decreased perfusion, which is best depicted by this modality. These areas increase in size and number with age; they are also larger and more numerous in patients with hypertension, pseudodrusen, or, interestingly, late age-related macular degeneration in the *fellow* eye. These characteristic OCT angiographic findings are consistent with histologic studies showing a growing number of ghost vessels in the choriocapillaris (a sign of vessel death), basal linear deposits, and subretinal drusenoid deposits with increasing age. Some diseases are known to be associated with both RPE atrophy and geographic atrophy; they include pseudoxanthoma elasticum and maternally inherited diabetes mellitus and deafness. Even in the absence of RPE atrophy, patients with these diseases can exhibit remarkable loss of the choriocapillaris (Fig 9-13).

Hayreh SS. Posterior ciliary artery circulation in health and disease: the Weisenfeld lecture. *Invest Ophthalmol Vis Sci.* 2004;45(3):749–757; 748.

Increased Venous Pressure

In rare cases, choroidal blood flow abnormalities may be related to venous outflow problems, including those caused by *dural arteriovenous malformations* or *carotid-cavernous*

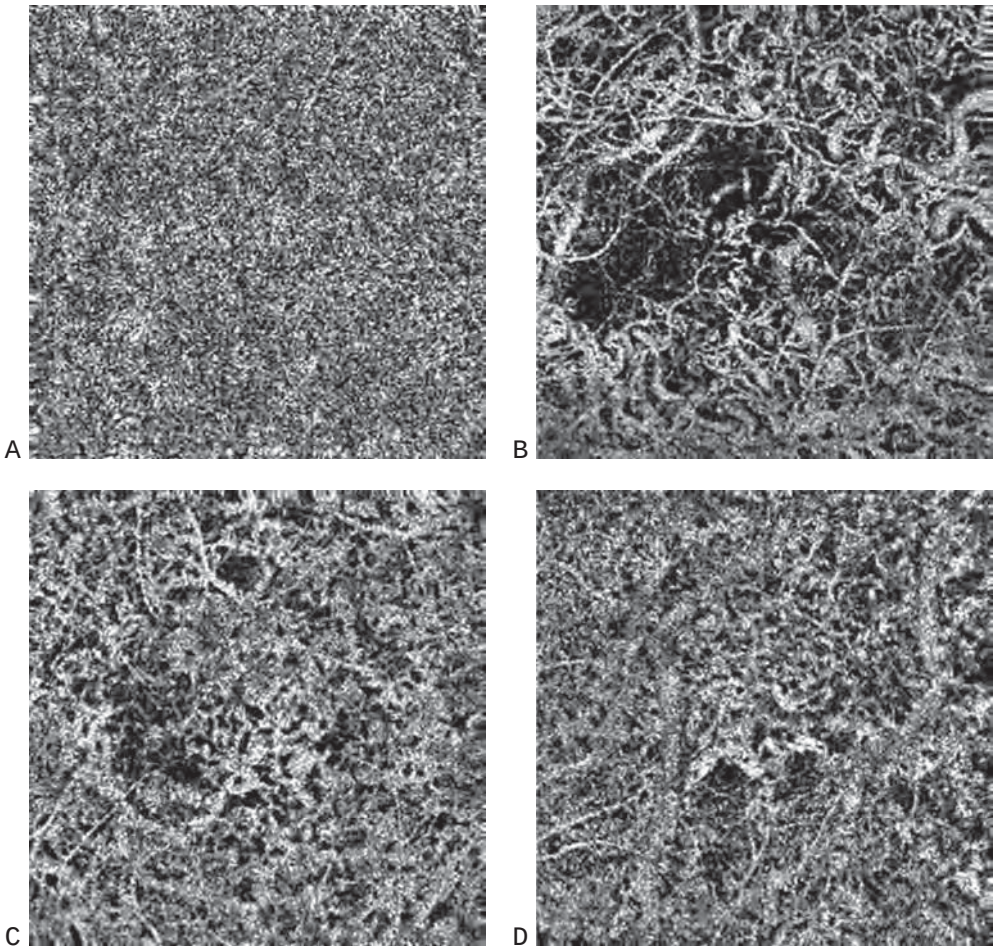

Figure 9-13 OCT angiography images of the choriocapillaris in a healthy patient and in patients with pseudoxanthoma elasticum (PXE). **A,** Healthy 63-year-old patient with no ocular disease. **B–D:** Three different patients with PXE: a 55-year-old patient **(B)** and two 63-year-old patients **(C, D).** The eyes with PXE show no evidence of RPE atrophy but have remarkable loss of the choriocapillaris. *(Reproduced with permission from Spaide RF. Choriocapillaris signal voids in maternally inherited diabetes and deafness and in pseudoxanthoma elasticum.* Retina. *2017;37(11):2008–2014. doi:10.1097/IAE.0000000000001497)*

fistulas (Fig 9-14). Diagnosis of choroidal blood flow abnormalities often requires dye-based angiography and occasionally a stethoscope (to detect a bruit). These patients should be referred for appropriate medical evaluation.

Age-Related Choroidal Atrophy

The thickness of the choroid decreases with higher levels of myopia and increasing age. In some older patients, the choroid is much thinner than expected. The eyes of these

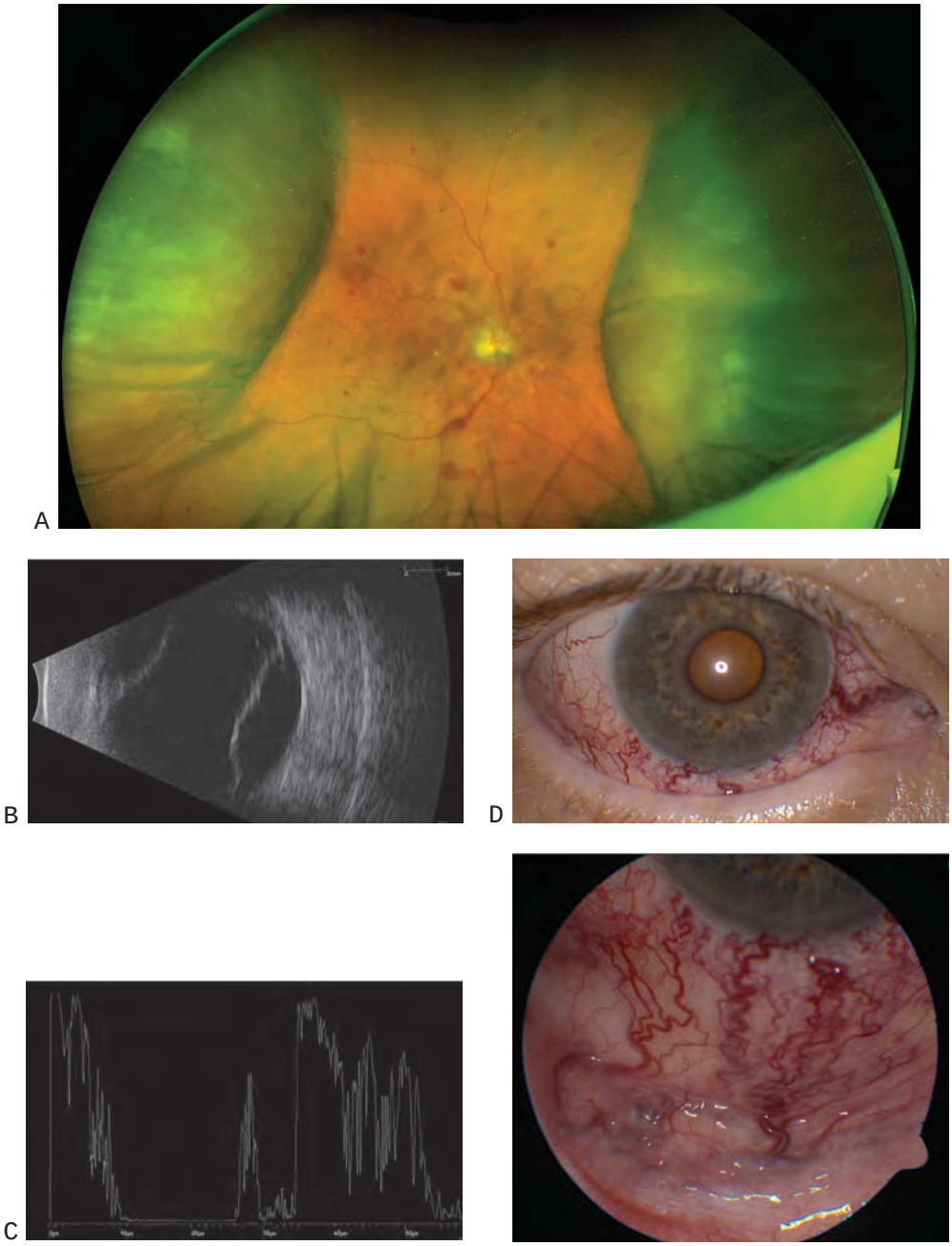

Figure 9-14 Constellation of classic findings with carotid-cavernous fistulas. **A,** Wide-angle color photograph from an older adult patient who was referred for "multiple choroidal melanomas" found incidentally on examination. **B,** B-scan shows large choroidals. **C,** Diagnostic A-scan shows very low internal reflectivity, consistent with serous choroidals rather than tumor masses. **D, E,** The patient was noted to have a red eye. On very specific questioning, he revealed that he had fallen and hit his head approximately 9 months earlier, and the eye began to turn red approximately 1–2 months after the fall. The patient was hypertensive. These images show classic corkscrew vessels. Patients with a carotid-cavernous fistula can also have a hyporeactive pupil from anterior segment ischemia, as was the case with this patient. *(Courtesy of Anthony B. Daniels, MD, MSc.)*

individuals tend to have *pseudodrusen,* which resemble drusen in appearance but are caused by collections of subretinal drusenoid deposits above the RPE. Like eyes with drusen, eyes with subretinal drusenoid deposits have an increased risk of CNV, particularly types 2 and 3, and geographic atrophy. More than 90% of eyes with geographic atrophy also have pseudodrusen. In contrast with patients with drusen, patients with subretinal drusenoid deposits perform more poorly on microperimetry, and dark adaptation is markedly prolonged in this group (Figs 9-15, 9-16).

Spaide RF. Disease expression in nonexudative age-related macular degeneration varies with choroidal thickness. *Retina.* 2018;38(4):708–716.

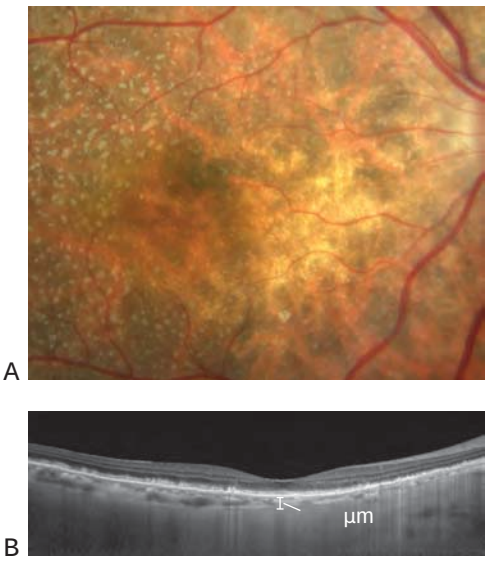

Figure 9-15 Pseudodrusen and the choroid. **A,** Fundus photograph shows an eye with prominent pseudodrusen. **B,** The true nature of the pseudodrusen is seen as subretinal drusenoid deposits. The subfoveal choroidal thickness is 121 µm. The choroid is thinner in eyes with pseudodrusen than in those with drusen. *(Reproduced with permission from Spaide RF. Disease expression in nonexudative age-related macular degeneration varies with choroidal thickness.* Retina. *2018;38(4):708–716. doi:10.1097/IAE.0000000000001689)*

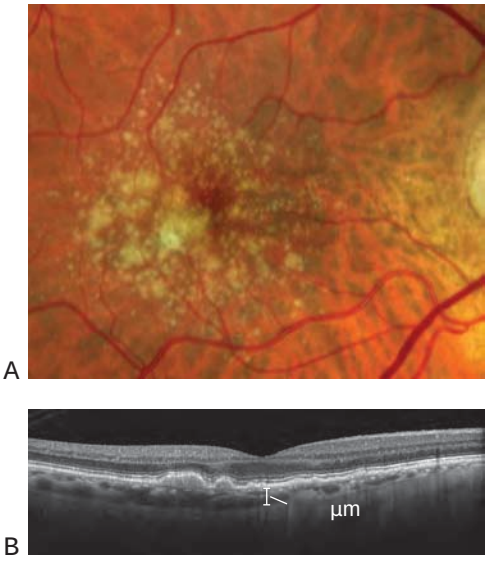

Figure 9-16 Drusen and the choroid. **A,** An eye with typical soft drusen. **B,** The subfoveal choroidal thickness is 162 µm. *(Reproduced with permission from Spaide RF. Disease expression in nonexudative age-related macular degeneration varies with choroidal thickness.* Retina. *2018;38(4):708–716. doi:10.1097/IAE.0000000000001689)*

Choroidal Folds

Folds in the choroid, sometimes called *chorioretinal folds,* occur for various reasons, including secondary to disease. Forces external to the eye, such as an indenting tumor or thyroid eye disease, can cause choroidal folds. The sclera may be thickened by posterior scleritis, thereby crowding the choroid (Fig 9-17). A relatively common phenomenon, which is poorly characterized, is the development of choroidal folds in middle-aged adults, with acquired hyperopia occurring in some of these individuals. Engorgement of the choroid causes an expansion of the tissue, which is limited by the sclera. Reduced intraocular pressure, occurring most commonly as a postoperative complication, can cause ciliochoroidal effusions and curvilinear choroidal folds in the posterior pole, a condition known as *hypotony maculopathy*. Use of medications such as topiramate can cause idiopathic swelling of the choroid resulting in chorioretinal folds and ciliochoroidal effusions without hypotony. Increased intracranial pressure can cause papilledema resulting in fine folds that course circumferentially around the optic nerve head; these folds are called *Patton lines*. Localized choroidal folds can be seen in association with CNV, choroidal neoplasms, and scleral buckles (Table 9-1).

Spaide RF, Goldbaum M, Wong DW, Tang KC, Iida T. Serous detachment of the retina. *Retina.* 2003;23(6):820–846.

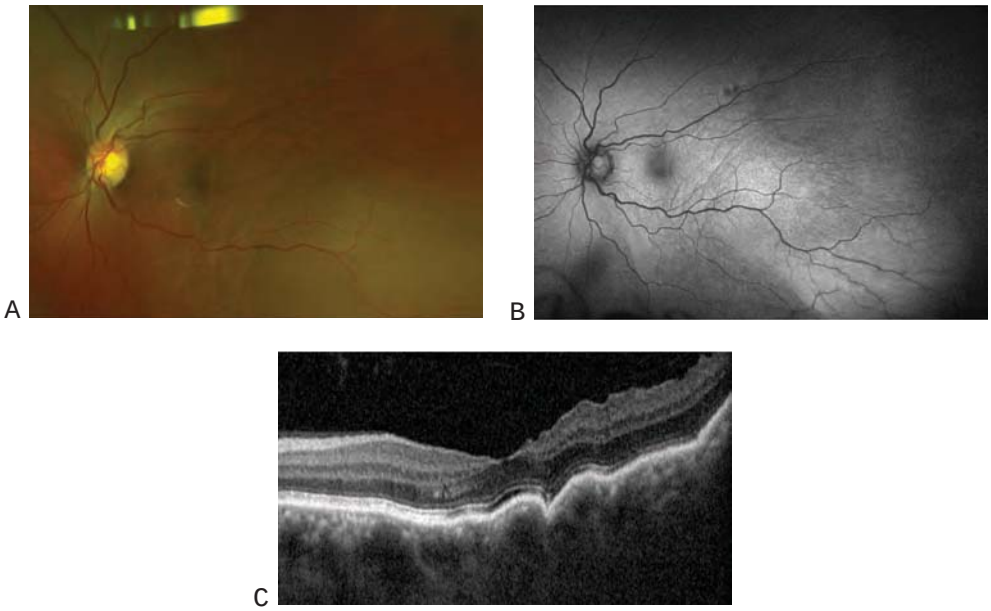

Figure 9-17 Choroidal folds in a patient with nodular posterior scleritis. **A,** Fundus photograph shows subtle striae associated with choroidal folds extending through the temporal macula. **B,** Fundus autofluorescence highlights the striae of folds with alternating hyper- and hypoautofluorescent changes emanating from the inferotemporal area of posterior nodular scleritis. **C,** Spectral-domain OCT shows characteristic undulations of RPE consistent with choroidal folds and a thickening choroid consistent with posterior scleritis. *(Courtesy of Avni P. Finn, MD, MBA.)*

Table 9-1 Differential Diagnosis of Choroidal Folds

Etiology of Choroidal Folds	Characteristic
Focal choroidal mass/neovascularization	Usually radiating from center of the lesion
Hyperopia	Usually horizontal in posterior pole/papillomacular bundle; extreme example in posterior microphthalmia
Hypotony	Ciliochoroidal effusions and curvilinear choroidal folds in the posterior pole
Idiopathic; theories include scleral calcification and/or inflammation	Oblique, along the insertion of the oblique muscles
Medications (eg, topiramate)	Ciliochoroidal effusions without hypotony
Papilledema	Patton lines, concentric with optic nerve head
Retrobulbar mass/orbital hardware	Varies according to indent location
Thyroid eye disease	Nonspecific

Choroidal Hemangiomas

Isolated choroidal hemangiomas are reddish-orange, well-circumscribed tumors of varying thickness that can affect the macula either directly or through subretinal fluid (Fig 9-18). Circumscribed hemangiomas transilluminate readily and exhibit highly echographic patterns on ultrasonography. During dye-based angiography, hemangiomas show very early filling of large vessels.

Sturge-Weber syndrome (encephalofacial hemangiomatosis) causes a diffuse hemangioma that, in children, may present first as glaucoma or amblyopia. The areas corresponding to the hemangioma typically appear reddish orange on ophthalmoscopy, a pattern referred to as "tomato ketchup fundus"; the underlying choroidal markings are not visible. The choroidal hemangiomas in Sturge-Weber syndrome are sometimes overlooked because they are diffuse and may blend imperceptibly into adjacent normal choroid. An ipsilateral facial nevus flammeus (port-wine birthmark) is also typically present in patients with this syndrome. See BCSC Section 4, *Ophthalmic Pathology and Intraocular Tumors,* and Section 6, *Pediatric Ophthalmology and Strabismus,* for further discussion.

Hemangiomas have been treated with laser photocoagulation, cryopexy, external beam and plaque radiation, and PDT.

Blasi MA, Tiberti AC, Scupola A, et al. Photodynamic therapy with verteporfin for symptomatic circumscribed choroidal hemangioma: five-year outcomes. *Ophthalmology.* 2010;117(8):1630–1637.

Madreperla SA, Hungerford JL, Plowman PN, Laganowski HC, Gregory PT. Choroidal hemangiomas: visual and anatomic results of treatment by photocoagulation or radiation therapy. *Ophthalmology.* 1997;104(11):1773–1778.

Uveal Effusion Syndrome

Uveal effusion syndrome is a rare condition in which abnormal scleral composition or thickness reduces transscleral aqueous outflow, inhibiting net fluid movement through

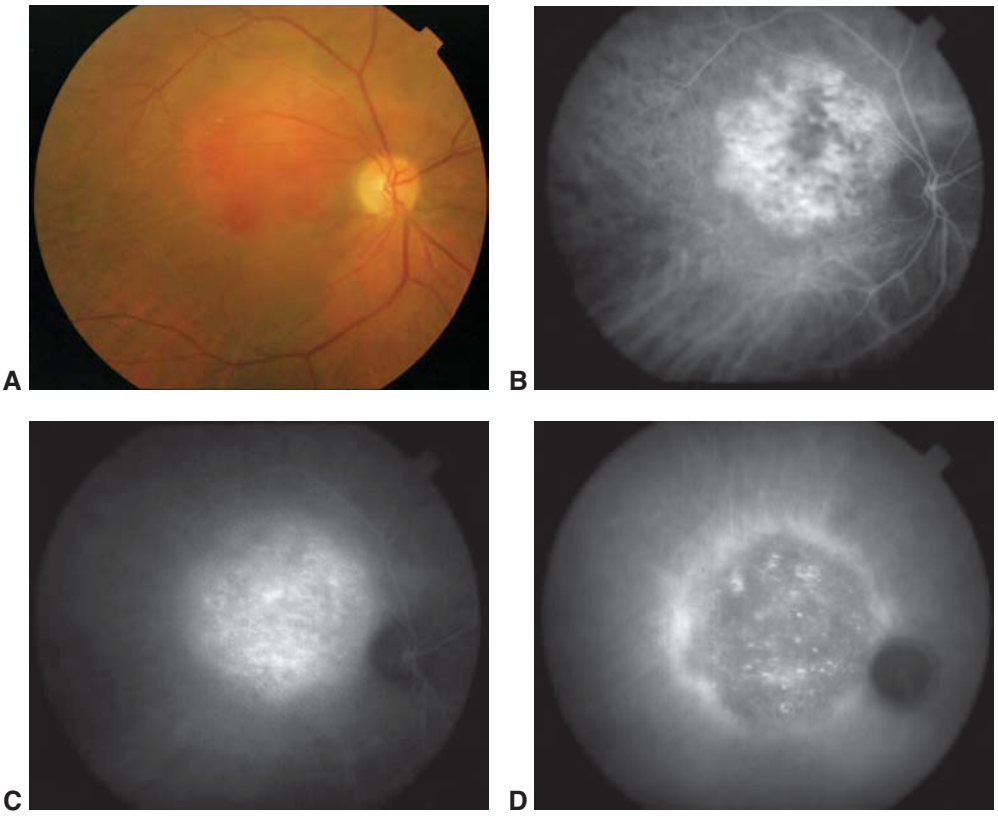

Figure 9-18 Choroidal hemangioma. **A,** Fundus photograph of the typical reddish-orange elevation of a circumscribed choroidal hemangioma. **B,** Soon after injection of ICG dye, the vascular composition of the hemangioma is revealed. **C,** Hyperfluorescence of the tumor occurs in the middle phase of the angiographic study from a combination of dye within and leakage from the vessels of the hemangioma. **D,** In the late phase of the study, the dye "washes out" of the lesion, leaving hyperfluorescent staining in the adjacent tissues. *(Reproduced with permission from Spaide RF, Goldbaum M, Wong DW, Tang KC, Iida T. Serous detachment of the retina.* Retina. *2003;23(6):820–846.)*

the posterior eye wall. Choroidal and ciliary body thickening, RPE alterations, and exudative retinal detachment may occur. The choroid is often so thick that OCT imaging is not possible, but the gross thickening can be imaged with ultrasonography. FA usually shows a leopard-spot pattern of hypofluorescence without focal leakage (Fig 9-19). Uveal effusion syndrome is a diagnosis of exclusion, usually made after ruling out all of the more common entities, such as posterior scleritis, and other etiologies of exudative detachments. A high index of suspicion for uveal effusion syndrome should be maintained for young patients with hyperopia. Scleral window surgery may yield anatomical restoration.

Elagouz M, Stanescu-Segall D, Jackson TL. Uveal effusion syndrome. *Surv Ophthalmol.* 2010;55(2):134–145.

Johnson MW, Gass JD. Surgical management of the idiopathic uveal effusion syndrome. *Ophthalmology.* 1990;97(6):778–785.

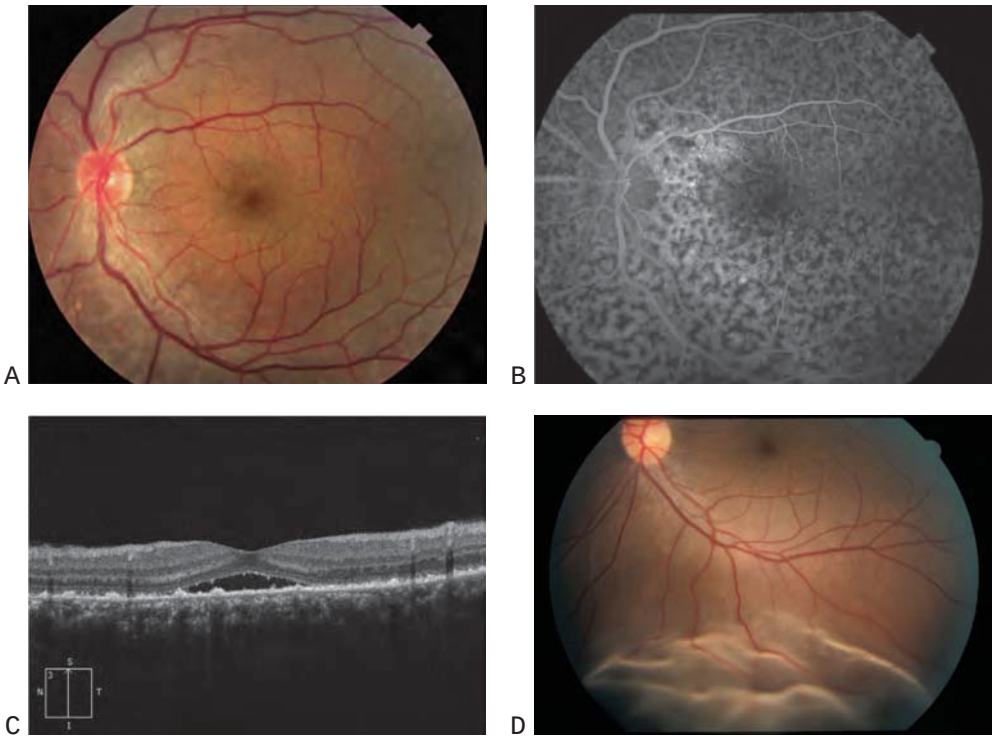

Figure 9-19 Idiopathic uveal effusion syndrome. **A–C:** In this patient's left eye, visual acuity was reduced to 20/70, and results of the systemic workup were negative. **A,** Fundus photograph demonstrates a blunted foveal reflex and irregular, subtle subretinal deposits. **B,** Corresponding FA image reveals a diffuse leopard-spot pattern of blocking with intervening window defects involving the entire posterior pole. **C,** OCT scan reveals a small amount of subfoveal fluid and outer retinal deposits. Not shown is a peripheral serous retinal detachment. **D,** Fundus photograph from a different patient with recent-onset uveal effusion shows the typical appearance of serous retinal detachment syndrome as well as an underlying choroidal detachment, which is common for this condition. *(Parts A–C courtesy of Ronald C. Gentile, MD; part D courtesy of Colin A. McCannel, MD.)*

Bilateral Diffuse Uveal Melanocytic Proliferation

A rare paraneoplastic disorder affecting the choroid, *bilateral diffuse uveal melanocytic proliferation (BDUMP)* causes diffuse thickening of the choroid, "giraffe skin" reddish or brownish choroidal discoloration, serous retinal detachment, and cataracts (Fig 9-20). The bilateral proliferation of benign melanocytes is usually associated with or often heralds systemic cancer. These proliferations can look like large nevi.

Most patients with BDUMP also exhibit nummular loss of the RPE, an anatomical change that differs distinctly from large nevi or thickening of the choroid. These areas of RPE loss are hypoautofluorescent during FAF but hyperfluorescent during FA. OCT shows mounds of residual material, presumed to be persistent RPE cells, between areas of loss. Tumors commonly associated with BDUMP are cancers of the ovary, uterus, and