Acute Monocular Visual Loss
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Temporal arteritis

Temporal arteritis is a medium- and large-vessel vasculitis that affects the extracranial branches of the carotid artery. Its presentation can vary from that of a chronic headache to sudden monocular loss of vision. Confirmation of the diagnosis requires a temporal artery biopsy and the cornerstone of treatment is the administration of high-dose steroids. Sudden loss of vision attributable to temporal arteritis constitutes an ophthalmologic emergency; prompt recognition of the disorder and institution of therapy can prevent further vision loss in the affected eye or new visual deficits in the contralateral eye.

Epidemiology

Temporal arteritis is the most common primary vasculitis among the elderly with an annual incidence of approximately 18 per 100,000 in people older than 50 years. Peak incidence occurs between 70 and 80 years of age with a 2:1 female to male ratio. Of those affected, 88% are white [1]. Certain human leukocyte antigen (HLA) types have been found to entail an increased risk for developing temporal arteritis [2]. Although discussion of the relationship between temporal arteritis and polymyalgia rheumatica is beyond the scope of this article, individuals who have these same HLA types are at an increased risk for developing both of these disorders [2,3].

Etiology

Although the exact inciting event of temporal arteritis is poorly understood, and no causal relationship has been established, there is some evidence that infection may play a role, specifically parvovirus B19,
Mycoplasma pneumoniae, and Chlamydia pneumoniae [4,5]. CD4 cells, responding to an unknown antigen, migrate through all three layers of the affected artery and initiate an inflammatory cascade. This inflammation causes a reactive proliferation of the intimal layer of the artery, with resulting narrowing of the arterial lumen and ischemia distal to the lesion [6]. The occlusion of end-arteries to the eye, scalp, tongue, and muscles of mastication causes the blindness, jaw and tongue claudication, and scalp ischemia associated with the disease. The inflammation also produces cytokines, which are believed to be responsible for the frequent low-grade fever and constitutional symptoms, such as anorexia, malaise, and weight loss, that are often associated with the disease [7]. Thrombotic occlusion of the arteries, however, does not seem to play a role in the pathogenesis. Granulomatous inflammation forms giant cells in the classic pathologic lesion and gives the disease its alternate name, giant cell arteritis [6].

Clinical features

The most common clinical manifestation of temporal arteritis is headache, seen in two thirds of patients and generally located in the area of the temporal or occipital arteries [8,9]. Systemic symptoms, such as low grade fever, chills, malaise, anorexia, and weight loss, are present in one half of patients [8,10]. Other associated symptoms include jaw claudication, tongue pain, and mandibular pain [10]. Visual complaints are also common with partial or complete vision loss in one or both eyes occurring in up to 20% of patients as a presenting symptom of disease [9–12]. Amaurosis fugax precedes vision loss in 44% of affected individuals; diplopia and visual hallucinations are less common, but may also precede vision loss [11–13]. Thirty percent of patients have neurologic manifestations that may include mononeuropathies, peripheral polyneuropathies, and occasionally TIAs or strokes [14,15].

Physical examination should include temporal and occipital artery palpation searching for nodular or firm, tender arteries with overlying erythema. Joint examination may reveal polyarticular pain with movement or synovitis, especially of the shoulders and hip. This finding may suggest polymyalgia rheumatica, which is associated with temporal arteritis [16]. The eye examination should include visual acuity, assessment of pupils for an afferent pupillary defect, and a funduscopic examination to evaluate the retina and optic disks [17]. Typical funduscopic findings in those who have visual loss from temporal arteritis include pallor and edema of the optic disk with scattered cotton-wool patches and small hemorrhages (Fig. 1) [18]. Laboratory evaluation may reveal elevated C-reactive protein and erythrocyte sedimentation rate (ESR), a normocytic anemia, and a reactive thrombocytopenia. One may also see mild elevations in the liver enzymes [16].

A 2002 meta-analysis [19] that included 41 studies and more than 2600 patients evaluated the accuracy of history, physical examination, and ESR
in predicting a diagnosis of temporal arteritis. The only two historical features found to appreciably increase the likelihood of temporal arteritis were jaw claudication and diplopia. Other visual symptoms, including monocular vision loss, were not found to be helpful in distinguishing temporal arteritis from other causes of sudden vision loss.

On physical examination, the presence of any abnormality of the temporal artery (beading, prominence, tenderness) was most predictive of temporal arteritis. Conversely, the absence of any of these findings reduced the likelihood of the diagnosis substantially. Evidence of optic atrophy or any funduscopic abnormality was not helpful in establishing or eliminating the diagnosis, because the end result of retinal ischemia is common to many causes of sudden vision loss [19].

A normal ESR, defined as age in years divided by 2 for men, and age in years plus 10 divided by 2 for women, conferred a negative likelihood ration of 0.2. Any ESR of less than 50 conferred a reduced probability of having the disease. An ESR greater than 100, although increasing the likelihood of temporal arteritis, was less predictive than the previously described historical features and physical examination findings. Similarly, the mean ESR for patients who had biopsy-negative and biopsy-positive temporal arteries was not statistically different. CRP was not evaluated in the meta-analysis. Other laboratory abnormalities, such as anemia, were not helpful in distinguishing those who had temporal arteritis [19].

**Diagnosis and treatment**

Suspicion of temporal arteritis should trigger rheumatology and ophthalmology consultation, especially in the setting of acute vision loss. The criteria for diagnosis of temporal arteritis as defined by the American College of Rheumatology include: age greater than 50 years, new headache, temporal artery abnormality, ESR greater than 50, and abnormal findings on a temporal artery biopsy. The presence of three or more criteria conferred
a sensitivity of 93.5% and a specificity of 91.2% for the diagnosis [20]. Characteristic giant cell granulomatous inflammation on temporal artery biopsy is considered the gold standard to confirm the diagnosis, however. Doppler ultrasound of the temporal arteries has been studied as a noninvasive adjunct to biopsy [21]. A sensitivity of 95% has been reported in biopsy-confirmed temporal arteritis when abnormalities in the structure of the arterial wall were discovered on ultrasound. The specificity is low, however, and a negative result cannot exclude the need for biopsy [22].

High-dose methylprednisolone is the first-line therapy in patients who have temporal arteritis who present with ocular manifestations. Three days of intravenous steroids followed by 2 years of oral prednisone beginning at 40 to 60 mg/d is a common suggested treatment course [16]. Patients who do not have ocular manifestations are started on oral prednisone without the methylprednisolone burst. Prednisone is gradually tapered in conjunction with monitoring ESR and CRP levels for change. Any increase in either halts any further reduction in prednisone level [18,23]. CRP has been shown to be more sensitive than ESR at diagnosis and during monitoring of relapse [24]. Various dosing protocols exist, but none have been prospectively validated.

Unfortunately, the visual loss associated with temporal arteritis is often permanent. In one retrospective study in patients who had biopsy-proven disease, central vision improvement after the initiation of steroid therapy was minimal, and was present in only 4% of patients [23]. Corticosteroids, however, do seem to prevent further vision loss; only 13% of those who had vision loss before diagnosis developed further vision loss after initiation of steroid therapy. Similarly, of those who had no visual complaints before steroid treatment, only 1% subsequently developed vision loss [11].

Optic neuritis

Optic neuritis is an acute demyelinating disorder of the optic nerve that typically presents as painful, monocular vision loss. Although most affected patients regain vision even without treatment, a substantial number subsequently develop multiple sclerosis (MS). Consequently, emergency physicians often enlist the help of ophthalmologists and neurologists in the management of these patients.

Epidemiology

Optic neuritis has an incidence of 6.4 per 100,000 with two thirds of cases occurring in women, generally between 20 and 40 years of age [25]. It is most common in northern latitudes (the United States and Northern Europe) and is more often diagnosed in white Americans than African Americans [25,26]. It is the presenting symptom in 15% to 20% of patients subsequently diagnosed with MS and occurs in one half of MS patients during the course of
their illness [25,27,28]. In addition, 31% of patients who have optic neuritis have a recurrence within 10 years of their initial presentation [29].

**Etiology**

The inflammatory demyelination of the optic nerve is believed to be an autoimmune phenomenon characterized by systemic T-cell activation present at the onset of visual symptoms [30]. Although the specific mechanism is unknown, inflammatory cytokines are believed to play a role and B-cell activation against myelin basic protein may be seen in the cerebrospinal fluid of affected individuals [31]. The inflammatory response against the optic nerve results in edema and breakdown of the myelin sheaths and perivascular cuffing of the retinal vasculature [32]. Genetic susceptibility for optic neuritis is suspected based on higher incidences among certain HLA types [33].

**Clinical features and diagnosis**

Optic neuritis is a clinical diagnosis based on history and physical examination findings. The Optic Neuritis Treatment Trial [34] surveyed 448 patients who had optic neuritis about their visual symptoms and performed detailed visual assessments. Eye pain accompanied vision loss in 92% of patients. Vision loss was typically monocular and progressed rapidly over a period of hours to days. Even patients who had 20/20 vision at presentation had defects in their ability to perceive color and contrast; patients often described their vision as “blurry” or felt the color has been “washed out” [35].

Although physical examination findings may vary in optic neuritis, pain with eye movements and an afferent pupillary defect are almost universally present [34]. Even in those patients who have normal visual acuity, mild optic nerve dysfunction causes an asymmetry in the pupillary reflex that can be elicited by the swinging flashlight test [25]. Visual acuity in the affected eye can range from 20/20 to no light perception. Although central scotoma is the classic visual deficit, a wide variety of visual field cuts may occur [34]. Two thirds of patients have a normal funduscopic examination with retrobulbar optic neuritis. One third of patients have optic disk swelling, blurring of disk margins, and swollen veins caused by optic nerve inflammation as it terminates in the retina (Fig. 2) [34].

Although the diagnosis of optic neuritis is often made on clinical grounds, gadolinium-enhanced MRI can help confirm the diagnosis and risk-stratify patients who are likely to develop MS. Optic nerve inflammation can be demonstrated in 95% of patients who have optic neuritis on gadolinium-enhanced MRI [36–38]. The imaging may also demonstrate oval-shaped lesions in the periventricular white matter that suggest MS [39]. In one study, the risk for MS 10 years after the first episode of optic neuritis was 56% versus 22% in those who did not have lesions [40]. This information may be useful for consulting ophthalmologists and neurologists and may guide the initial therapy of affected patients.
Treatment

Visual acuity in optic neuritis generally improves without treatment over the course of several weeks [29]. Treatment is focused on hastening the return of vision, preventing recurrences, and reducing the incidence of MS. In a study that randomized affected patients to either high-dose intravenous methylprednisolone or oral prednisone (or placebo), those who were given the methylprednisolone demonstrated a more rapid return to normal vision [41] and a lower risk for recurrent optic neuritis. The differences in visual acuity were not significant at 2 years follow-up, however. Oral prednisone was associated with an increased risk for recurrent optic neuritis compared with placebo. Methylprednisolone delayed the onset of MS compared with placebo at 2 years, but this advantage did not persist beyond this time frame [41]. According to the American Academy of Neurology, although corticosteroids may hasten the return of vision after initial presentation, there is no compelling evidence for long-term benefit for patients who have optic neuritis [42].

Several randomized trials have demonstrated that interferon beta 1a and interferon beta 1b may reduce the development of MS in patients who have optic neuritis [25,39,43,44]. Although this is typically started at the onset of symptoms, the initiation of this therapy is probably beyond the scope of emergency physicians and should only be done in conjunction with an involved neurology consultant.

Central retinal artery occlusion

Central retinal artery occlusion (CRAO) generally causes abrupt, painless loss of vision that can be permanent unless blood flow to the retina is
restored before the onset of irreversible ischemic damage. The retina is perfused by the retinal artery, a branch of the ophthalmic artery that arises from the internal carotid artery. The ophthalmic artery enters the orbit along with the optic nerve at which point the retinal artery branches off, enters the cerebrospinal space, and travels within the optic nerve where it provides blood flow to the central retina and optic nerve. In some patients, additional perfusion of the central retina (including the macula) is provided by the cilioretinal artery, an anatomic variant that allows for the preservation of central vision in some who have CRAO.

**Epidemiology**

Although the true incidence of CRAO is unknown, most estimates are that it occurs in between 1 to 10 in 100,000 individuals [45]. Risk factors for the development of CRAO are similar to those of other cardiovascular diseases, namely increasing age, hypertension, hypercholesterolemia, diabetes, elevated homocysteine levels, and tobacco use. Some studies have described an increased incidence in males, whereas others have failed to demonstrate a difference in disease prevalence in one sex over the other [46–48].

**Etiology**

CRAO and other strokelike syndromes share a final common pathway: interruption of blood flow to distal tissues. Although several causes of CRAO are described later, the most common are atherosclerotic disease of the ipsilateral carotid artery [49] and propagation of cardiogenic emboli to the retinal artery [50]. Several studies have suggested factors such as patient age and ethnicity may play a role in the distribution of these causes. For example, whites are more likely to have carotid disease [49], whereas those less than 40 years of age are more likely to have a cardiogenic embolus as the source of their symptoms [50], and patients older than age 70 are more likely to have temporal arteritis as the basis of their CRAO [49].

**Carotid artery atherosclerosis**

Atherosclerotic disease of the carotid artery is believed to be the most common source of emboli resulting in the disruption of retinal artery blood flow. Although the data are largely from small case series, the prevalence of carotid artery disease in those who have CRAO is generally believed to be 10% to 25%, although there is literature suggesting rates as high as 70% [49,51–55]. Additionally, CRAO in the setting of carotid disease may portend an increased risk for stroke; for this reason, carotid endarterectomy is often recommended in this setting [55].
Cardiogenic embolism

Between 2% and 20% of patients who have CRAO have a cardiac source of embolus [54,56]. These patients tend to be younger in age, and there may be aspects of their history that should alert the clinician to consider this as a possible cause, including: a history of congenital heart disease, rheumatic heart disease, myocardial infarction, endocarditis, or the presence of a cardiac tumor or murmur [57]. Chronic anticoagulation may be warranted for patients who have a cardiogenic source of embolus.

Other causes

Although carotid artery atherosclerosis and cardiogenic embolism represent the cause of most cases of CRAO, there are several other causes reported in the literature. These can be divided broadly into other vascular disease processes (carotid artery dissection [58], radiation injury of the retinal artery [59], Moyamoya disease [60], arterial vasospasm, and migraine) [61], hematologic disorders (sickle cell disease) [62], disorders resulting in hypercoagulable states (antiphospholipid syndrome, factor V Leiden mutation [63], protein S deficiency [64], and protein C deficiency) [65], and autoimmune/inflammatory conditions (giant cell arteritis [66], lupus [67], polyarteritis nodosa [68], and Wegener granulomatosis) [69]. There are case reports of all of these causes of CRAO; no large cohort studies have been published.

Clinical features

The visual loss associated with central retinal artery occlusion is generally painless, monocular, and dramatic, often leaving the affected individual with only a small area of unaffected vision. The symptoms may be less severe, however, if the retina is also perfused by a cilioretinal artery (15% of patients in one series) [70]. Similarly, the blindness may be transient, or the patient may have “stuttering” symptoms if an embolus moves along the vascular tree and eventually dissolves before causing complete vessel occlusion [71].

On funduscopic examination, the ischemic retina initially appears white and the macula classically is described as having a “cherry red spot” where the retinal epithelium is thinner and the retinal pigment epithelium and choroidal vasculature appear more prominent and can be seen more easily (Fig. 3). In those who have a cilioretinal artery, there may be an area of normal-appearing retina surrounded by an ischemic, pale retina. The actual embolus may be visible in as many as 40% of patients who have CRAO [72]. The clinician may see shiny, iridescent cholesterol plaques, grayish platelet deposits, or bright white calcium fragments. There is generally a complete or relative afferent papillary defect. Finally, funduscopic examination may reveal a characteristic “boxcarring” in the retinal veins and arteries as serum separates from the Rouleau stacking of red blood cells.
The diagnosis of central retinal artery occlusion is made on clinical grounds in combination with characteristic eye ground findings. It is a true ocular emergency; retinal ischemia lasting longer than 240 minutes may lead to massive irreversible vision loss [73], whereas restoration of blood flow within 100 minutes may preserve a patient’s vision [74]. Although many of those who have occlusion of a branch retinal artery may regain normal vision, spontaneous visual recovery in those who have CRAO is rare and the literature of this population is largely limited to case reports. In one case series, only 35% of patients recovered vision that was better than 20/100 [75], whereas in another series of 73 CRAOs, there were only four instances in which final visual acuity was better than counting fingers [70]. Visual acuity at presentation is believed to be predictive of eventual acuity after the acute CRAO.

Although there is literature documenting and describing the debilitating effects of CRAO, there is a relative paucity of literature examining and comparing the effectiveness of several different treatments. Given the poor outcomes of those afflicted with CRAO, however, several potential therapies have been advocated. Although some of the more conservative treatments are within the practice scope of emergency medicine, patients who have suspected CRAO should receive emergent ophthalmologic consultation and evaluation.

Although many of the therapies vary in their invasiveness and risk, all share a common goal of restoring blood flow to an ischemic retina. As mentioned previously, spontaneous resolution (ie, the embolus dissolving or moving on from the ophthalmic circulation) is relatively uncommon, occurring in 1% to 8% of cases [45]. Initial therapies involved attempts by
patients or clinicians to manually dislodge the embolus by massaging the eyeball over a closed lid. It is believed that this may also augment aqueous outflow and retinal perfusion may increase with relief of the digital pressure [76,77].

Other therapies have aimed to increase ocular perfusion pressure by decreasing intraocular pressure, which may be accomplished by performing anterior chamber paracentesis [78]; giving intravenous diuretics, such as acetazolamide [79] or mannitol [45]; using enhanced external counterpulsation (a procedure in which air-filled cuffs are applied to the vascular bed of the lower extremities during diastole) [80]; or performing a trabeculectomy [81], in which a fistula is created between the anterior chamber and the subconjunctival space.

Some have suggested treating affected patients with vasodilators with the hope of increasing retinal blood flow or perhaps even dislodging the culprit embolus. The use of agents such as nitrates [45], rebreathing carbon dioxide [82], and inhaling carbogen (95% oxygen and 5% carbon dioxide) [78] have been met with mixed results because systemic vasodilatation may result, causing a reduction in systemic blood pressure and a subsequent decrease in retinal perfusion. Others have advocated for alternative forms of medical therapy, including various platelet inhibitors, anticoagulants, aqueous formation inhibitors, hyperosmotics, and corticosteroids.

A more aggressive approach involves the use of lytic agents in the treatment of CRAO. Using principles adapted from the use of thrombolytics in acute myocardial infarction and ischemic stroke, some have advocated for its use in CRAO. Any benefits, however, must be weighed against the risk for life-threatening complications (ie, hemorrhage, especially intracranial) in what is a debilitating, but not necessarily life-threatening, disease. Several reports involving various agents, including urokinase and recombinant tissue plasminogen activator (rt-PA), given both systemically and locally are reported in the literature. A 2000 meta-analysis provides the best summary of these data (although it examines only 16 studies, all of which were retrospective and nonrandomized and included a total of only 100 patients). The authors concluded that local intra-arterial fibrinolysis offered a “marginally better visual outcome than conservative forms of management” [83]. Similarly, a 2005 Cochrane review suggested that “there is currently not enough evidence to decide which, if any, interventions for acute nonarteritic central retinal artery occlusion would result in any beneficial or harmful effect. Well-designed randomized controlled trials are needed to establish the most effective treatment” [84].

In summary, although a wide variety of medical and surgical treatments for CRAO have been used, none have been prospectively shown to have more than a limited or marginal benefit. There are few, if any, randomized controlled trials of these therapies, and there has been little advancement in the treatment of this disorder in the last 20 years.
Central retinal vein occlusion

There is much overlap in the epidemiology, causes, clinical features, and diagnosis and treatment between central retinal artery and vein occlusion. Below, critical similarities and differences are highlighted.

Epidemiology

Retinal vein occlusion (RVO) is the second most common retinal vascular disorder behind diabetic retinopathy [85]. Prevalence reports vary, largely depending on whether the data are taken from a hospital- or population-based sample. The prevalence of RVO in hospital-based studies generally ranges from 0.3% to 1.6% [86,87], whereas a large Israeli population-based study reported a 4-year incidence of retinal vein occlusion of 2.14 cases per 1000 of general population older than 40 years and 5.36 cases per 1000 of general population older than 64 years [88]. An Australian population-based study reported a 1.6% 10-year incidence of RVO in those older than 60 years [89].

Etiology

Since the initial description of RVO in the medical literature in 1854, several different classification systems have been used. Typically, RVO is first divided into that affecting a central vein (CRVO) and that affecting a branch vein (BRVO). Central vein occlusion is further divided into ischemic and nonischemic types.

BRVO is believed to result in large part from factors related to the anatomic relationship between the retinal vein and artery as they pass through the region of the lamina cribrosa. In this area, the structures share an adventitial sheath and are in close proximity to each other. This factor, in combination with degenerative changes in the wall of the vein and artery, leads to a narrowing of the vein lumen and subsequently to stasis and thrombosis. Ischemic RVO can also occur in any other area of the retina where the arteries and veins cross [90]. Many of the risk factors for the development of BRVO are similar to those of CRAO, and include hypertension [87,91,92], diabetes [87,91,93], cerebrovascular disease [91,92], cardiovascular disease [94], increased body mass index [94], dyslipidemia [94], thyroid disease [91], peptic ulcer disease [91], glaucoma [95], and hyperhomocysteinuria [96]. On ophthalmologic examination, those who have focal arteriolar narrowing or arteriovenous nicking are also more likely to develop RVO [89].

CRVO, however, is believed to occur because of clots present in the main draining vein of the retina. Depending on clot burden, collateral circulation, and perfusion of the retina, it is divided into ischemic and nonischemic types. In nonischemic CRVO, the vascular lesion is generally more proximal, allowing for collateral circulation to provide some blood flow to the retina. Consequently, visual loss is by and large less severe and debilitating.
In addition, in those eyes that have nonischemic disease, examination may reveal stasis of blood within the veins as opposed to frank thrombosis [90].

**Clinical features**

In those who have ischemic RVO, the visual loss is often dramatic and is frequently noted on awakening one morning. The persistent blurring may be preceded by episodes of amaurosis fugax in some patients. Conversely, nonischemic RVO may be relatively asymptomatic and may only be noted on routine ophthalmologic examination. Visual symptoms, when present, may be limited to a vague blurring of central vision with sparing of the peripheral vision and are often worse in the morning and improve gradually over the course of the day [90]. In one study, the final visual acuity was 20/400 or worse in 87% of those who had ischemic RVO, whereas it was 20/30 or better in 57% of those who had nonischemic disease [97]. As with CRAO, long-term visual acuity in those who have ischemic RVO is largely determined by visual acuity at presentation [98].

**Diagnosis and treatment**

Although the formal diagnosis of RVO and the differentiation between the ischemic and nonischemic varieties is beyond the clinical scope of most emergency medicine physicians, one should be able to appreciate several of the clinical aspects of RVO mentioned earlier, along with some typical findings on funduscopic examination.

The funduscopic examination classically reveals retinal hemorrhages (which may be mild or severe), dilated and tortuous retinal veins, edema of the optic disc, and small areas of yellow-white discoloration of the retina (“cotton wool spots”), which are indicative of local retinal ischemia (Fig. 4). When retinal hemorrhages are severe, and cover much of the fundus, the

![Fig. 4. Branch retinal vein occlusion](From Kaiser PK, Friedman NJ, Pineda R, II. The Massachusetts Eye and Ear Infirmary illustrated manual of ophthalmology. 2nd edition. China: Saunders; 2004; with permission.)
clinician may see the classic “blood and thunder” appearance of the retina (Fig. 5).

RVO is generally a self-limited disease (the retinopathy may resolve over weeks to years), but the complications of the disorder can be severe. Although there is little that an emergency medicine physician can do acutely for affected patients, referral to a specialist is of paramount importance. In addition to making a formal diagnosis, ophthalmologists should differentiate between ischemic and nonischemic disease, a distinction that has important ramifications in the complications that may be anticipated. Specifically, those who have ischemic disease are at a higher risk for developing ocular neovascularization, which may result in severe vision loss. A detailed discussion of this complication is beyond the scope of this article.

As with CRAO, various therapeutic strategies have been used with varying levels of success. Interventions, such as anticoagulation, lytic agents, hemodilution, hyperbaric oxygen, surgical decompression, and systemic and intravitreal steroids, are all reported in the literature. A recent meta-analysis reported that although hemodilution seemed to be of some benefit in some trials, and panretinal photocoagulation may improve visual acuity in those who have neovascularization, there is “limited level I evidence for any intervention to improve visual acuity in patients with RVO” [99]. Most studies are case reports, retrospective, do not have a control group, or involve very small numbers of patients.

Retinal detachment

Epidemiology

Retinal detachment is a relatively uncommon affliction of the eye, affecting approximately 1 to 2 in 10,000 people per year [100,101], or about 1 in 100.
300 people over the course of their lifetime [102]. Risk factors for the development of retinal detachment include increasing age [103], previous cataract surgery [104], focal retinal atrophy, myopia [105], trauma [106], diabetic retinopathy, family history of retinal detachment, uveitis, and prematurity [107].

Etiology

Although the pathogenesis of retinal detachment can be divided into three subtypes, all share a common final pathway: separation of the retina from the underlying retinal epithelium. In the first type, exudative retinal detachment, there is accumulation of serous or hemorrhagic fluid in the subretinal space, generally as a result of systemic conditions, such as severe acute hypertension, sarcoid, or cancer. In the second type, tractional retinal detachment, previous trauma from infection, surgery, inflammation, or hemorrhage results in fibrotic tissue that can provide traction on the retina resulting in detachment [107–109].

The third type, rhegmatogenous detachment, is the most commonly encountered. In this form of retinal detachment, age-related changes in the vitreous humor cause it to liquefy and shrink away from the back of the eye (termed posterior vitreous detachment). In turn, the vitreous may pull the retina from its underlying epithelial layer. Although patients who have posterior vitreous detachment often only experience bothersome floaters in their peripheral vision, more dramatic vision loss can occur if a retinal tear or flap occurs. As the vitreous pulls away from the retina, the vitreous is able to enter the subretinal space and create a plane of dissection. As more vitreous enters this space, more retina is separated from its underlying epithelial layer. As the detachment grows, so does the patient’s visual field deficit. Most rhegmatogenous detachments, if left untreated, expand to eventually include the macula, impacting central visual acuity [107–109].

The aforementioned risk factors for the development of retinal detachment all result in either changes in the makeup of the vitreous or in alterations in the normal anatomy or shape of the eyeball, resulting in retinal traction and forces that tend to pull the retina from the epithelium layer it normally covers.

Clinical features

The hallmark complaint of those who have vitreous detachment is the presence of floaters. Characteristically described as dots, cobwebs, lines, or strings in the visual field, the floaters of vitreous detachment (especially with an associated retinal tear) tend to occur abruptly. As the vitreous detachment progresses to retinal detachment, patients may describe visual field loss. This loss generally begins at the periphery (where the retina is the thinnest), and over hours to weeks may spread toward the central visual field.
axis. Other ocular complaints such as pain, tearing, redness, and drainage are typically associated with conditions other than retinal detachment.

**Diagnosis and treatment**

Funduscopic examination with a direct ophthalmoscope is generally not sufficient to rule out the diagnosis of retinal detachment. The narrow field of view provided by the instrument does not allow one to see the peripheral aspects of the retina where tears are more frequently seen. Although the clinician may see an abnormal red reflex or occasionally the classic white billowing retinal separation, and indirect examination in a dilated eye by an ophthalmologist is often necessary (Fig. 6) [110].

A central tenet in the treatment of retinal detachment is its initial prevention. Risk factor modification and education of susceptible patients is crucial. For example, those who have severe myopia should be encouraged to wear protective lenses when playing sports, and those who undergo cataract surgery should receive specific instructions about worrisome symptoms and reasons to return to care. Similarly, those who have known vitreous detachment or are at risk for developing the finding should be followed closely. Fortunately, only 1% to 2% of such patients have a retinal break. If vitreous hemorrhage is present, however, this risk increases to 70% [111]. If identified, retinal tears can be treated with cryotherapy or laser therapy, which serve to create a scar at the site of the tear and prevent fluid from entering the subretinal space. Success rates (ie, the prevention of retinal detachment) are greater than 95% with this technique [112].

Surgical repair of retinal detachments, typically done by a retinal specialist, also has high success rates. More invasive therapies, such as scleral buckling and posterior vitrectomy, post success rates of nearly 90% [113,114], whereas less invasive therapies, such as pneumatic retinopexy, may be performed in an office setting in select cases [115]. Retinal detachment surgery

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**Fig. 6. Retinal detachment. (From Kanski JJ. Clinical diagnosis in ophthalmology. China: Mosby Elsevier; 2006; with permission.)**
generally fails because of the growth of scar tissue on the retina in the weeks following repair [116]. This phenomenon, termed proliferative vitreoretinopathy, is also amenable to surgery, although success rates are not as high and resulting visual acuity is often poor [117].

If repair is technically successful, visual acuity is often restored to predecrement levels. Critical in determining eventual visual outcome is the presence or absence of macular involvement in the tear, and if it is involved, the duration and extent of its involvement. For this reason, those who have preserved central acuity should be referred for immediate surgery [118,119].

**Retinal vasculitis**

Retinal vasculitis is characterized by inflammation of the blood vessels of the retina. It may be primarily ocular and limited to the orbit or may be associated with various autoimmune and infectious systemic diseases. Retinal vasculitis typically presents as a painless decrease in vision and is diagnosed clinically by funduscopic examination; ancillary fluorescein angiography and laboratory testing may be used to define the degree of involvement and diagnose associated conditions. Treatments include local and systemic immunosuppressive regimens and treatment of the underlying disorder.

**Epidemiology and etiology**

Retinal vasculitis may be caused by infectious or systemic inflammatory conditions or may be restricted solely to the retina. Retinal vasculitides associated with primary ocular disorders include idiopathic retinal vasculitis, birdshot retinochoroidopathy, Eales disease, and pars planitis syndrome [120].

Autoimmune causes of retinal vasculitis include systemic lupus erythematosus (SLE), Behçet disease, sarcoidosis, Wegener granulomatosis, polyarteritis nodosa (PAN), HLA-B27 associated conditions, and inflammatory bowel disease (IBD). Various inflammatory ocular conditions may occur as part of these diseases. In SLE, retinal involvement, including vasculitis, is seen in 3% to 30% of patients [121,122]. Severe decreases in vision correlate with decreased survival and are likely related to uncontrolled systemic inflammation [121,123–125]. Similarly, 80% of those who have Behçet disease have ocular involvement [126]. Those who have skin lesions and arthritis have an increased risk for visual loss, generally resulting from recurrent vasoocclusive disease from retinal vein vasculitis [127]. Of those patients who have sarcoidosis, 20% to 25% have ocular disease [128,129], and 7% of those have posterior segment involvement (a subset of which includes retinal vasculitis) [129]. In Wegener granulomatosis ocular involvement occurs in 30% to 60% with significant vision loss in 8%. Retinal vasculitis is rare, however [130,131]. There are ocular findings in 10% to 20% of patients who
have PAN. Anterior segment findings are most common but retinal veins may also be affected [132]. In HLA-B27 associated conditions, anterior uveitis is most common; posterior segment disease occurs in 4% to 17%. Of those who have posterior segment disease, 24% have retinal vasculitis [133–135]. Retinal vasculitis is uncommon in IBD and has only been observed in case reports with Crohn disease [136].

Infectious causes of retinal vasculitis include toxoplasmosis, syphilis, tuberculosis, and several viruses, including varicella (VZV), herpes simplex (HSV), and cytomegalovirus (CMV). Toxoplasmosis is responsible for 25% of cases of posterior uveitis in the United States. The resulting vitreous inflammation often induces localized retinal vasculitis [137]. Ocular tuberculosis is an uncommon manifestation of extrapulmonary TB seen in approximately 1% of patients [138,139]. Similarly, retinal vasculitis in syphilis is rare, but may be seen in the secondary or tertiary stages [140]. HSV and VZV retinal vasculitis are seen in immunocompromised chemotherapy patients, and those who are congenitally infected [141]. CMV retinal vasculitis is seen in those who have HIV and low CD4 counts.

Clinical characteristics

Those who have retinal vasculitis typically present with a painless decrease in vision but may also report blind spots or floaters. More subtle symptoms, such as changes in color vision and changes in the perceived shape of an object, may occur with involvement of the macula [141].

Slit lamp examination reveals cells in the vitreous body [141]. Funduscopic examination may reveal characteristic “vascular sheathing,” the accumulation of inflammatory cells on vessel walls manifested as an area of retinal pallor paralleling the vessel lumen. This finding is more common in peripheral vessels, and there may be segments of unaffected arterioles. The increased vessel permeability that leads to vascular sheathing predisposes patients to vitreous hemorrhage and macular edema that may also be seen [120].

Diagnosis and treatment

Retinal vasculitis is diagnosed clinically based on a dilated funduscopic examination in conjunction with consultation from an ophthalmologist. A history and physical should be performed with specific attention to associated findings seen in the various systemic inflammatory and infectious conditions described above and should direct laboratory and radiographic testing. A detailed review of the clinical presentation of the various disorders is beyond the scope of this article.

Regardless of the cause of the retinal vasculitis, fluorescein angiography (injection of fluorescein in the retinal artery percutaneously to delineate the retinal vasculature) is a commonly used ancillary test. It can help define the
extent of disease and aid in narrowing the differential diagnosis of causative disorders. Angiography typically demonstrates perivascular staining that results from increased vascular permeability and is the visual correlate of vascular sheathing on funduscopic examination. Areas of nonperfused retina may also be detected as dark patches on the angiogram [142].

In addition to angiography, focused laboratory and radiographic assessment is still indicated even in the absence of historical or examination findings that suggest an associated condition. According to a study performed by the National Eye Institute, only 1% of patients initially diagnosed with idiopathic retinal vasculitis developed subsequent systemic disease after 4 years of follow-up. The authors recommend a CBC, urinalysis, ESR, syphilis and HIV serologies, and a chest radiograph in the absence of indications for additional testing, but caution against low-yield indiscriminate laboratory testing [120].

Treatment of retinal vasculitis generally includes an immunosuppressive regimen that may be topical, intraorbital, or systemic. Systemic corticosteroids are often the cornerstone of treatment, but regimens may also include methotrexate, azathioprine, cyclosporine, and other immunosuppressive agents, depending on the cause of the vasculitis. Important exceptions are retinal vasculitides associated with infectious causes, wherein treatment is based on correcting the underlying infection. Corticosteroids are used adjunctively in CMV, VZV, HSV, and toxoplasmosis to reduce inflammation [140]. The decision to initiate treatment of retinal vasculitis should be made in conjunction with ophthalmology consultation, in addition to infectious disease and rheumatology consultation if appropriate.

Summary

In summary, there are several causes of acute monocular vision loss, some of which are discussed here, that require prompt recognition by the emergency physician. Although the initial diagnosis is made by the emergency department clinician, a prompt, and at times emergent, referral to an ophthalmologist may be necessary.

References


