Cat-Scratch Disease

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Cindy Ung and Lucy H. Young

Introduction

Cat-scratch disease (CSD) was first recognized by Parinaud in 1889 in three patients with granulomatous conjunctivitis, preauricular lymphadenopathy, chronic fever, and contact with animals. This constellation of symptoms initially became known as Parinaud oculoglandular syndrome, and subsequent reports noted a history of cat exposure in some patients with this syndrome (Cassady and Culbertson 1953; Henry 1952). Sweeney and Drance (1970) first suggested an association between CSD and neuroretinitis in 1970, a finding later confirmed by Gass (Dreyer et al. 1984; Gass 1977). In 1994, Golnik and associates provided the first serologic evidence of systemic *Bartonella* infection in patients with neuroretinitis (Golnik et al. 1994). Cat-scratch disease is the most common cause of neuroretinitis with approximately two thirds of patients demonstrating seropositivity for gram-negative bacillus, *Bartonella henselae*.

The incidence of cat-scratch disease in the United States is estimated at 9.3 per 100,000 population (Jackson et al. 1993). Cat-scratch disease is almost always benign and self-limited, with the majority of cases reported in persons under 20 years of age (Carithers 1985). More than 90% of all cases of CSD are associated with a history of some type of contact with cats (Carithers 1985; Warwick 1967), and 57–83% have a history of a scratch from a cat (Margileth 1993; Spaulding and Hennessy 1960). It is the leading cause of regional lymphadenopathy in children and young adults.

Clinical Features

Cat-scratch disease is a systemic infectious disease and is often a self-limited condition. The transmission of *B. henselae* is usually from a cat scratch or bite, although it may occur from contact between cat saliva and an open wound. A small localized papule is usually first seen at the site of inoculation 3–10 days after inoculation. Mild local infection is followed by tender regional lymphadenopathy and then low-grade fever, malaise, headache, and myalgias several weeks after exposure (Dreyer et al. 1984). Ocular involvement has been estimated to occur in 5–10% of patients (Carithers 1985).

Posterior segment manifestations of CSD include neuroretinitis, intermediate uveitis, focal or multifocal chorioretinitis, peripapillary retinal edema, and vascular occlusions. Neuroretinitis appears to be the most common and is found in 1–2% of patients infected with *B. henselae* (Ormerod and Dailey 1999; Carithers 1985). It is characterized by optic disc edema and the presence of stellate macular exudates (Figs. 18.1a and 18.2a). The optic disc is the principal target in neuroretinitis with fluorescein leakage confined to the disc (Figs. 18.1b and 18.2b). Even in the presence of a full macular star, there is no fluorescein leakage in the macula on fluorescein angiography. The optic disc edema is often accompanied by peripapillary subretinal fluid (Fig. 18.3). Neuroretinitis is usually unilateral but can be asymmetrically bilateral as shown in Fig. 18.4 (Schlossberg et al. 1989). Disc edema usually occurs prior to the formation of a macular star by 1–4 weeks (Wade et al. 2000) (Figs. 18.3b and 18.5a). The macular star may be partial and is usually present in the nasal macula. The disc edema and macular star can take up to 6–12 months to resolve leaving the eye with mild disc pallor (Fig. 18.5). A relative afferent pupillary defect is seen in 90% of patients (Chi et al. 2012). Multifocal areas of retinitis or chorioretinitis can also occur in the absence of optic disc edema and macular exudates (Fig. 18.6). Other findings include a neurosensory detachment of the macula or peripapillary region as well as anterior and posterior chamber inflammation.

C. Ung · L. H. Young (✉)
Massachusetts Eye and Ear Infirmary, Boston, MA, USA
e-mail: lucy_young@meei.harvard.edu

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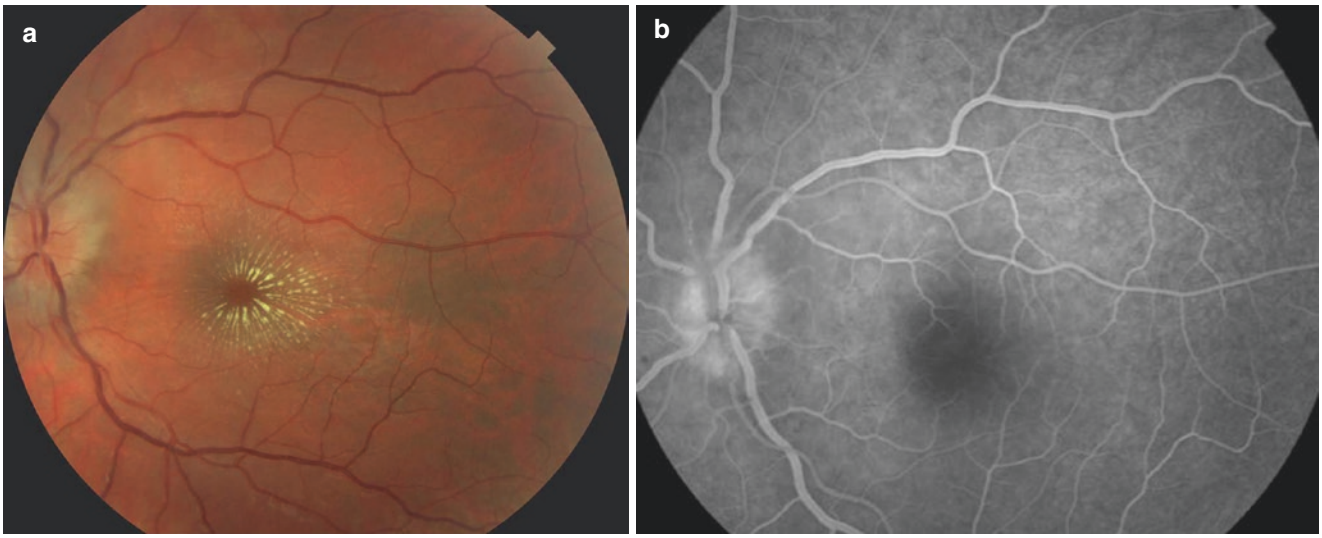

Fig. 18.1 (**a**) Fundus photograph shows disc edema and macular scar in the left eye of a 22-year-old healthy man with 1 week history of blurry vision OS (20/200). (**b**) Fluorescein angiography shows leakage confined to the optic nerve

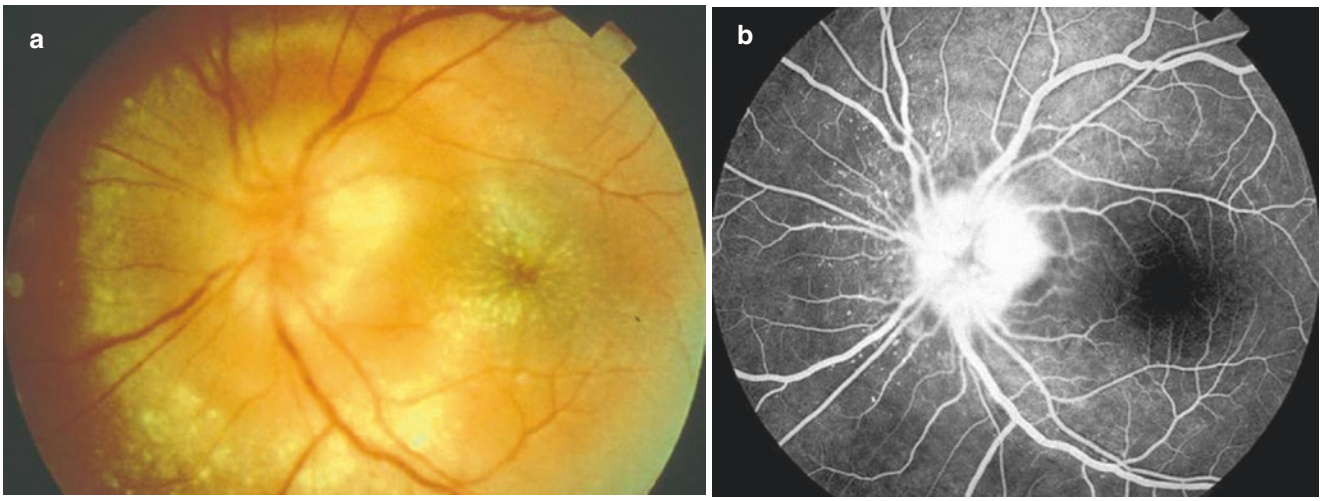

Fig. 18.2 (**a**) Fundus photograph shows unilateral disc edema. (**b**) Fluorescein angiography shows disc leakage without macular leakage

The more common ocular complication caused by *Bartonella* infection is Parinaud's oculoglandular syndrome, which affects approximately 2–5% of symptomatic patients (Carithers 1985). Parinaud's oculoglandular syndrome is a rare condition consisting of chronic low-grade fever, granulomatous follicular conjunctivitis, and ipsilateral regional lymphadenopathy involving either the preauricular or the submandibular of cervical lymph nodes. Transmission is generally from hand–eye contact from infected cat feces. Most patients experience localized disease that resolve within several months.

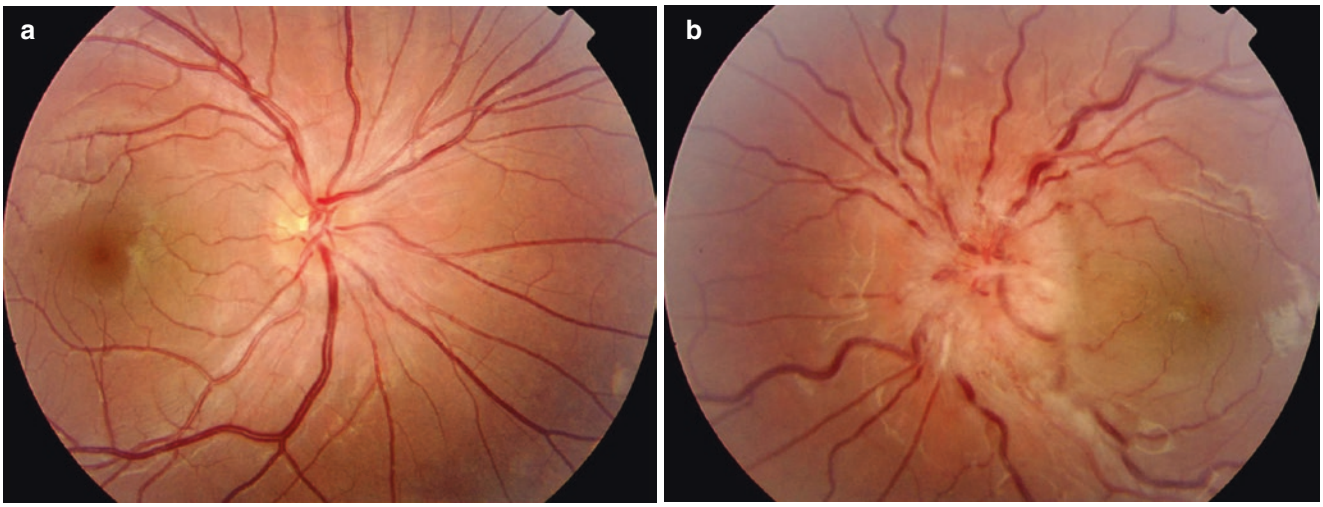

Fig. 18.3 Twelve-year-old healthy girl referred for a neuro-ophthalmologic consultation of disc edema OS > OD of unclear etiology. Sinus X-rays, head CT scan, MRI with contrast and lumbar puncture with opening pressure were ordered by the outside ophthalmologist, and these were all normal. Visual acuities were 16/13 OD and 16/200 OS (down from 20/60 2 weeks earlier). A positive left afferent pupillary defect was present. (**a**) Trace disc hyperemia with mild blurring of the inferonasal margin was noted in the right eye. (**b**) In the left eye, moderate disc edema with peripapillary subretinal fluid extending into the fovea was noted

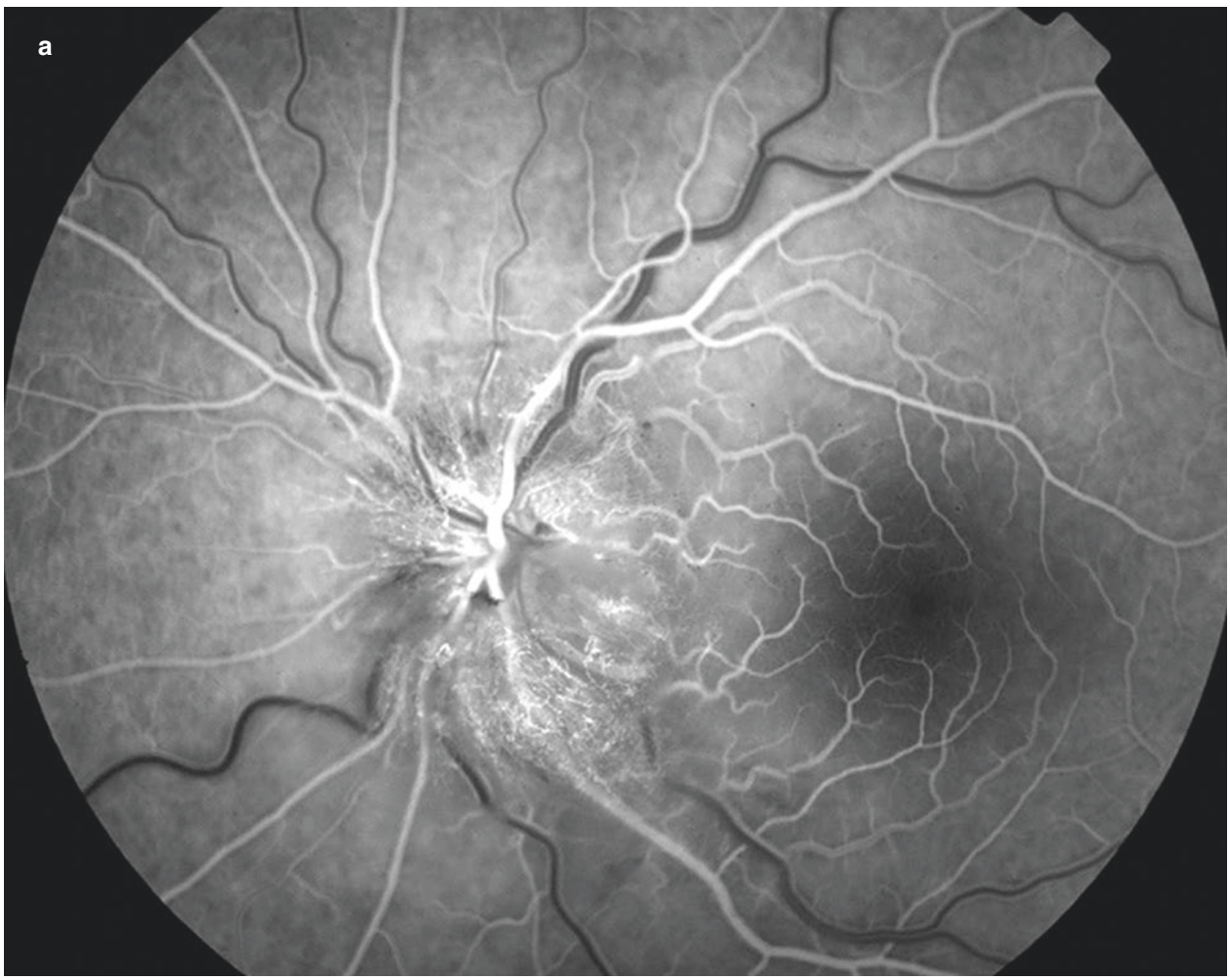

Fig. 18.4 Fluorescein angiography shows dilated disc capillary OS in the early phase (**a**) followed by increasing disc leakage with time (**b**). Late images show mild disc leakage and no macular leakage OD (**c**)

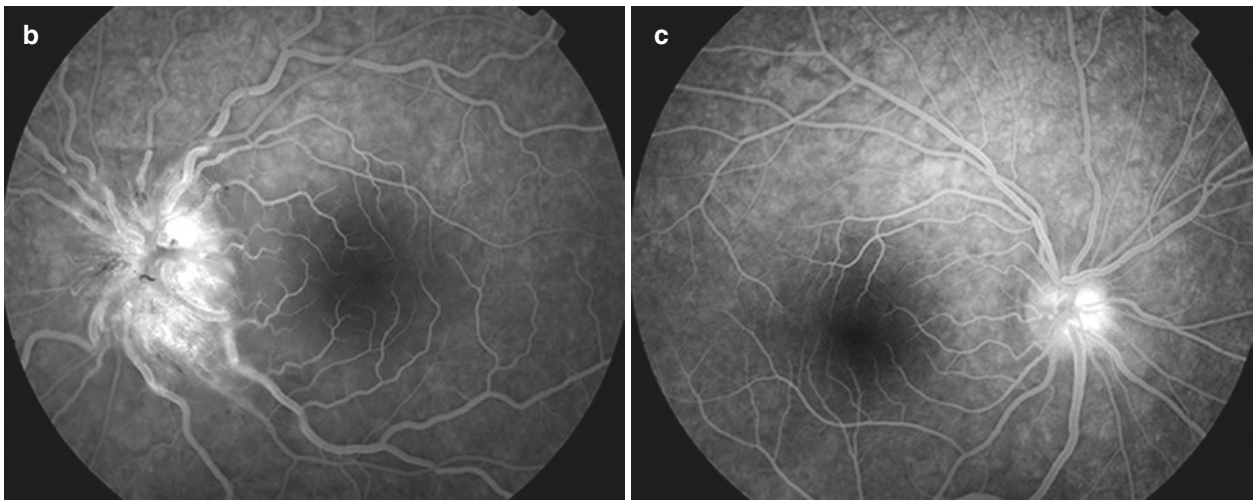

Fig. 18.4 (continued)

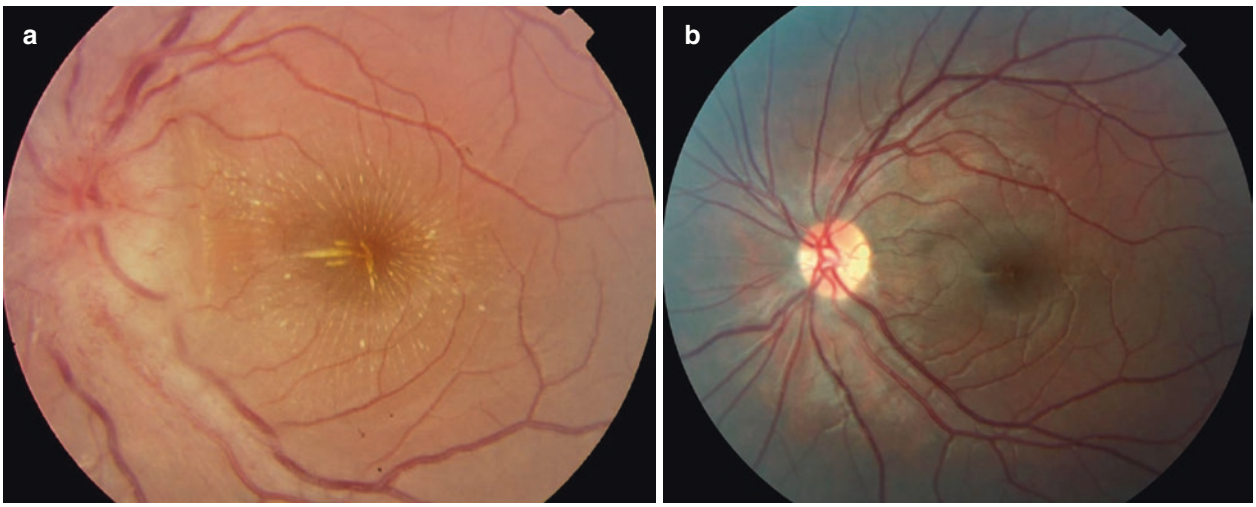

Fig. 18.5 In the same patient, fundus photography of the left eye at 8 days after initial consultation shows marked disc edema and a newly formed macular star (**a**). Fundus photography at 11 months later shows normal appearing macula and mild disc pallor (**b**). Visual acuity improved from 16/200 to 16/30 OS. (Courtesy of Joseph F. Rizzo, MD)

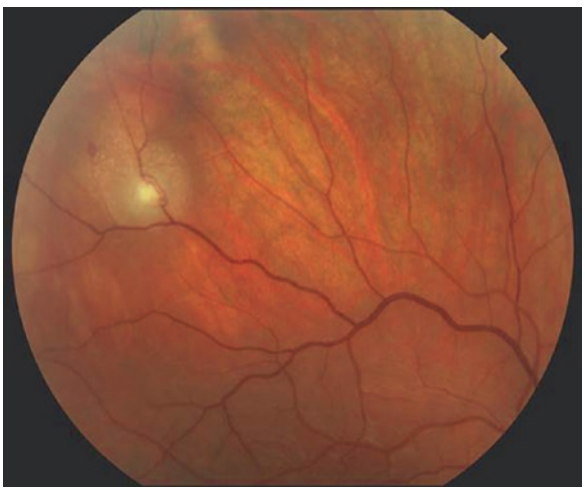

Fig. 18.6 A small chorioretinitis lesion in the contralateral eye of a patient with classic neuroretinitis

Differential Diagnosis

The differential diagnosis for neuroretinitis includes malignant hypertension, central retinal vein occlusion, diabetes mellitus, pseudotumor cerebri, sarcoidosis, syphilis, tuberculosis, toxoplasmosis, toxocariasis, Lyme disease, mumps, varicella, herpes simplex, and leptospirosis (Dreyer et al. 1984). It is important to remember that unilateral disc edema precedes the formation of a macular star by 1–4 weeks. As in the case in Fig. 18.2, this 12-year-old patient presented with asymmetrically bilateral disc edema and underwent extensive workup including a lumbar puncture prior to the development of a macular star and subsequent diagnosis of Bartonella. Invasive diagnostic procedures can be avoided by obtaining a thorough history and considering Bartonella in the differential for disc

edema even in the absence of a macular star. Causes of a macular star include hypertensive retinopathy, papilledema, anterior ischemic optic neuropathy, diabetic papillopathy, posterior vitreous traction, disc, and juxtapapillary tumors. Either by inflammatory or ischemic mechanisms, these entities can compromise the microvasculature of the optic disc and result in leakage of serum and lipids with macular star formation.

Bartonella henselae has been isolated as the etiologic agent, and serologic testing for *B. henselae* is available. There are two different serologic tests for the diagnosis of cat scratch. The first is an indirect fluorescent antibody test (IFA) for the detection of serum anti-*B. henselae* antibodies. This test was found to be 88% sensitive and 94% specific in the immunocompetent host and may fall to 70% or below in HIV-infected individuals (Dalton et al. 1995). The second is an enzyme immunoassay (EIA) with a sensitivity of 86–95% and specificity of 96% compared with IFA (Litwin et al. 1997; Barka et al. 1993). All tests have the potential for cross-reactivity with *B. quintana* as well as other species.

Management

There are no treatment guidelines for CSD as it is known to have a self-limited course in immunocompetent patients. The visual prognosis is good with or without treatment. Antibiotics are generally reserved for those with the most severe infections. Immunocompromised patients affected with CSD tend to require antibiotics such as erythromycin or doxycycline to control their infection (Schlossberg et al. 1989).

Given its superior intraocular penetration, doxycycline is preferred to erythromycin in patients older than 9 years of age. Doxycycline is contraindicated in younger children given its propensity for teeth discoloration. Both doxycycline and erythromycin can be given intravenously or combined with rifampin in more severe infections. Other antibiotics that can be used include azithromycin, ciprofloxacin, gentamycin, and trimethoprim-sulfamethoxazole (Relman et al. 1990). After resolution of the disease, final outcome can include residual visual field defect, decreased visual acuity, and contrast sensitivity and disc pallor on examination.

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