·Letter to the Editor·

Different outcomes of serpiginous choroiditis with or without ocular and systemic treatment

Zhong-Shan Chen, Qin Ding, Yan-Ping Song, Li Zhu, Zhi-Jian Huang, Ming Yan

Department of Ophthalmology, Wuhan General Hospital of Guangzhou Military Command, Wuhan 430070, Hubei Province, China

Correspondence to: Yan-Ping Song. Department of Ophthalmology, Wuhan General Hospital of Guangzhou Military Command, Wuhan 430070, Hubei Province, China. songyanping@medmail.com.cn

Received: 2012-02-16 Accepted: 2012-06-27

DOI:10.3980/j.issn.2222-3959.2012.04.26

Chen ZS, Ding Q, Song YP, Zhu L, Huang ZJ, Yan M. Different outcomes of serpiginous choroiditis with or without ocular and systemic treatment. *Int J Ophthalmol* 2012;5(4):535-538

Dear Sir,

I am Dr. Zhong-Shan Chen from the Department of Ophthalmology, Wuhan General Hospital of Guangzhou Military Command, Hubei Province, China. I write to present two cases with serpiginous choroiditis (SC) which is a rare, usually bilateral, chronic, progressive inflammation of the choroid, choriocapillaris and retinal pigment epithelium. In this paper we presented the different outcomes of SC patients with or without systemic and ocular treatments after long-term follow-up.

SC, also named geographic choroiditis, helicoid peripapillary choroidtis, is a rare, usually bilateral, chronic, progressive inflammation of the choroid, choriocapillaris and retinal pigment epithelium. The etiology is unknown and the onset usually occurs at peripapillary region, then the lesions progress gradually to macular and periphery regions. The lesions appear special geographic helicoid shape with successive serpentine edges and final massive geographic atrophy [1,2]. The impairment occurs deeply and infiltrates into retinal pigment epithelium and choroid layers. It is readily recurrent and progressively aggravate, finally leads to severe visual loss [3,4]. Because the etiology is very complicated the treatment was not sure. Some histopathologic studies showed diffuse and local infiltration of lymphocytes in the choroids indicating an immunologic inflammation in SC [5]. Therefore, anti-inflammatory and immunosuppressive therapies may be useful for SC. In this paper we presented the different outcomes of two cases of SC patients with different outcomes after long-term follow-up, one treated with systemic antituberculous and immunosuppressive drugs and other with no systemic and ocular drugs.

CASE SERIES

Case 1 A 37-year-old male complained progressive vision impairment for half a year. The patient felt vision fuzzy without headache, ocular pain, nausea, cough and so on. He had a history of systemic lupus erythematosus (SLE) which had been cured, with unclear therapies. His father had tuberculosis. Ocular examination: Best-corrected visual acuities (BCVA) were 4/20 of right eye and 4/4 of left eye. Anterior segment was normal, and right fundus appeared choroidal atrophy around optic papilla and macula; the lesion was located under retinal vessels with distinct border from normal tissue. Ocular fundus of left eye appears normal. Indocyanine green angiography (ICGA) showed hypofluorescence with distinct border in right eye (Figure 1A). Tuberculin test (purified protein derivative, PPD) showed strong positive and blood test for anti-tuberculosis antibody was positive, rheumatic and immunologic tests were all negative. Chest radiograph revealed normal. The diagnosis was peripapillary chorioiditis in right eye. In view of positive PPD and blood test his father's tuberculosis history, the patient was administered with anti-tuberculosis therapy initially (Rifampicin 450mg/d, Isoniazid 300mg/d, Pyrazinamide 750mg/d). After 6 months treatment the patient quited the drug by himself, and 5 months later he came for review. Fundus photos showed gray-white retinochoroidal lesions in right eye extending irregularly to midperiphery. The lesions in left eye infiltrated from the edge of optic papilla to nasal side of macula appearing light creamy-yellow (Figure 1B). Because there was no improvement in visual acuity the patient refused treatment. After drug withdrawal for 9 months he felt VA decreased in left eye. The BCVA was 4/25 in right eye and 4/5 in left eye. Anterior segment in both eyes was still normal. Fundus showed gray-white lesion extended to macular region in the left eye (Figure 1C). Physical examination showed negative PPD, blood test for anti-tuberculous antibody was negative, and chest radiography was normal. The diagnosis was SC. Respecting immunologic inflammation oral immuno-suppressive agents cyclosporine A (CSA, 3-5mg/kg per day)

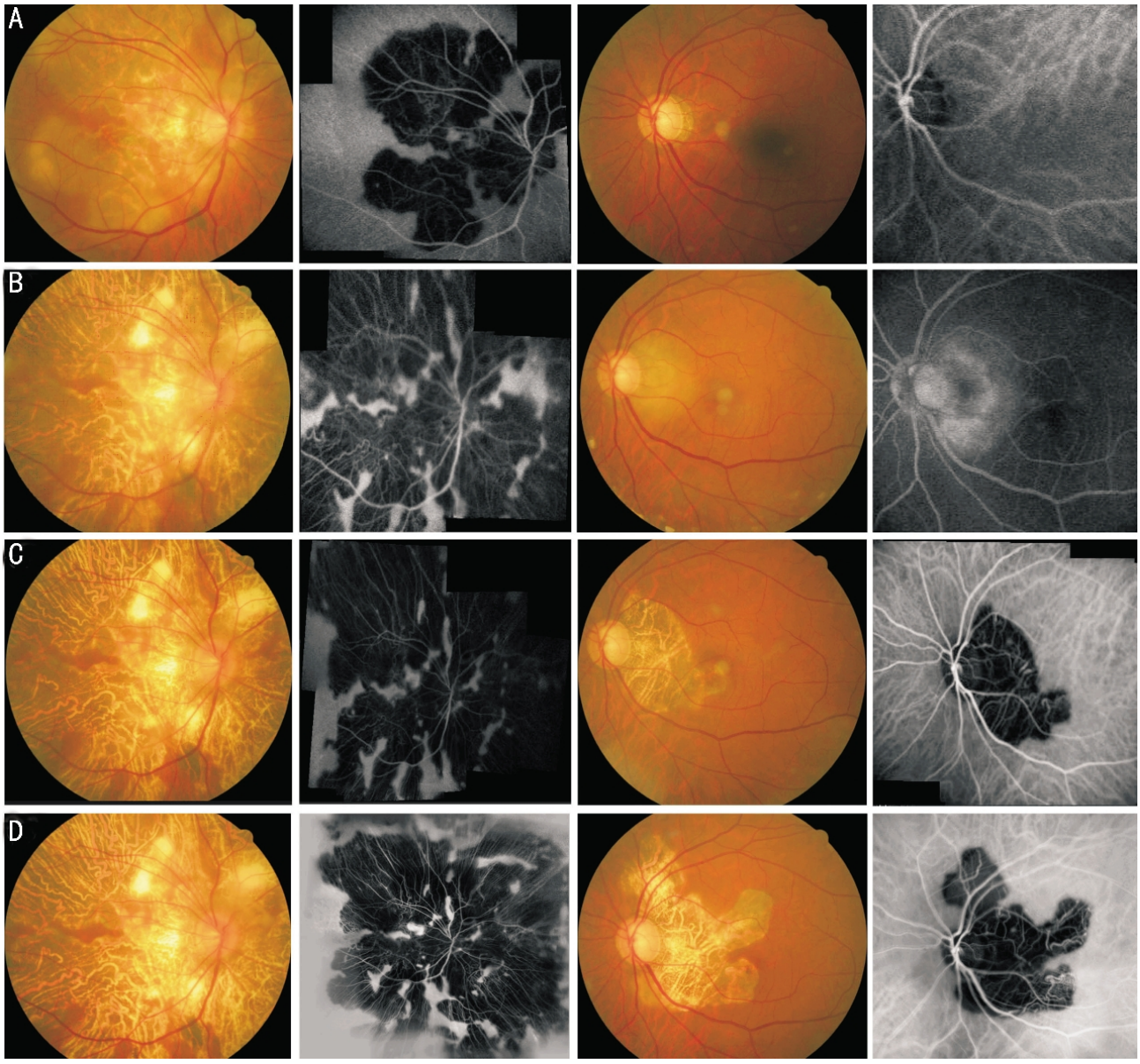

Figure 1 Case 1 A: Ocular fundus of right eye showed choroidal atrophy from the temporal edge of optic papilla, and appeared gray-white edema. The lesion locates under retinal vessels with distinct border from normal tissue. Ocular fundus of left eye appears normal. ICGA showed hypofluorescence with distinct border in right; B: Six months after anti-tuberculosis, gray-white geographic lesions in right eye extended to central and periphery regions. In left eye creamy yellow lesions developed from optic disc edge to macula C: Drug withdrawal for 9 months geographic lesions in right eye continuously developed and in left eye the gray-white lesions infiltrated to macula; D: One year after oral cyclosporin a geographic lesions in right eye progressed to periphery region, and lesions developed from optic disc edge to superior part of macula and central-periphery region in the left eye.

was started and retrobulbar injection of triamcinolone acetonide (TA, 20mg) was given in left eye for one time. Two months later BCVA was 4/20 in right eye and 4/5 in left eye. CSA was continuously administered for one year and visual acuity remained stable. Finally, five years after the first visit BCVA of this patient was 4/20 in right eye and 4/8 in left eye. Fundus showed geographic lesions in right eye progressed to periphery region, and lesions developed from optic disc edge to macula and central-periphery region in left eye (Figure 1D).

Case 2 A 65-year-od male attended the eye clinic for more detailed fundus examination without any symptoms. He was found fundus lesions in right eye by accident at a routine physical examination. Fundus showed choroidal atrophy at nasal edge of optic papilla in right eye (Figure 2A). The BCVA of both eyes were 4/5. All the laboratory examinations were negative. The diagnosis was chorioiditis in right eye. The patient refused treatment because of no visual impairment in the eyes. He was suggested to come back for follow-up per six months, but he never came back for fundus examination till three years later. The patient found visual acuity decreased obviously in right eye. Ocular examination showed BCVA was 4/25 in right eye and 4/8 in left eye. Anterior segment was normal, ocular fundus in

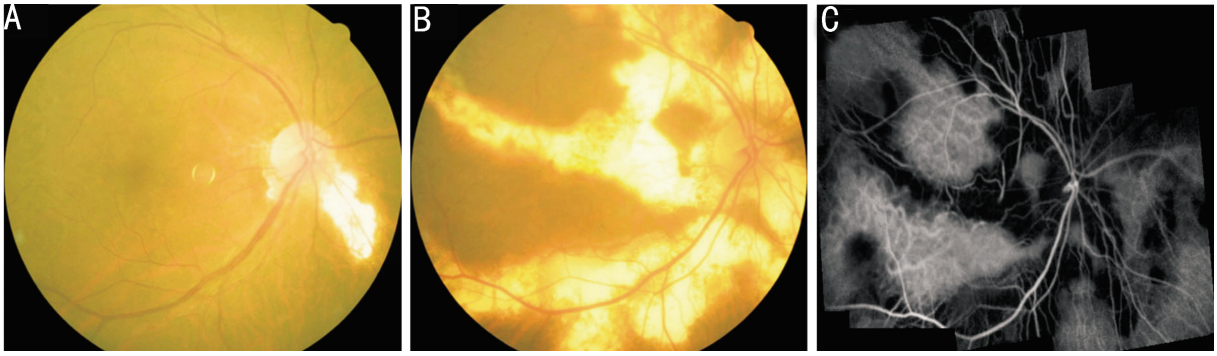

Figure 2 Case 2 A: Ocular fundus of right eye showed choroidal atrophy at nasal edge of optic papilla; B: Three years later choroidal atrophy developed to macula, posterior pole and central-periphery regions with helical appearance; C: ICGA of right eye showing helical hypofluorescence centered on optic papilla.

right eye showed choroidal atrophy developed to macula, posterior pole and central-periphery regions with helical appearance (Figure 2B), and ICGA showed helical hypofluorescence centered on optic papilla in right eye (Figure 2C). The patient rejected treatment again because he was afraid of the complications of immunosuppressive agents. He was followed up and one year later BCVA of right eye 4/40 and 4/16 in left eye.

DISCUSSION

Two cases characterized the clinic features and outcomes of SC, which is a kind of chronic progressive recurrent inflammation in choroid and retinal pigment epithelium. SC is a rare ocular disease, which tends to affect healthy young to middle aged adults aged from 20to 70 years old. Most studies reported a higher prevalence in male than in female, and susceptible in White [6], but were also found in Asians, African-Americans, and Hispanics. It clinically constituted less than 5% of posterior uveitis in most uveitis epidemiological reports. There is no familial predisposition. In a Finnish study, HLA B7 was found to be more prevalent in patients with SC (54.5%) than the general population (24.3%) [7]. Some reports of SC indicated it is associated with some systemic disease, such as Crohn disease, celiac disease, extrapyramidal dystonia, polyarteritis nodosa, and sarcoidosis. These probably represent coincidence rather than association.

Inactive lesions usually present geographic atrophies in retinal pigment epithelium (RPE) and choroicapillaris, and active lesions appear geographic or round cyano-gray or creamy-yellow impairment, sometimes the edges of atrophies appear cyano-gray. Active lesions may accompany venous white sheath, optic disc and macular edema, subretinal choroidal neovascularization, pre- and subretinal hemorrhage, or vitreous hemorrhage. It was regarded as atrophic or degenerative diseases in the past, but recent researches indicated it was related with tubercle bacillus, streptococcus, fungus or influenza virus infection, or related with autoimmune response [6].

Fundus fluorescence angiography (FFA) showed hypofluorescence in focal zones and leakage in active lesions, but transmitted fluorescence in inactive lesions. Indocyanine green angiography (ICGA) presented hypofluorescence which indicated poor vascular perfusion in choroids, and fluorescence staining or transmitted fluorescence in inactive lesions, even choroidal large vessels could be seen. Cardillo Piccolino investigated the autofluorescence (AF) characteristics of serpiginous choroiditis [8]. Hyper-AF was detected 2 to 5 days after the appearance of the lesions, providing a clear delineation of the area of definitive retinal pigment epithelium (RPE) damage. A progressive decrease in AF was seen. AF seems to be a very sensitive imaging technique for detecting damage of the RPE in acute episodes of SC. A sequence of autofluorescence changes reflects the passage from activation to resolution of new lesions.

For the unclear or diverse etiologies proposed, there were many different treatments attempted for SC, including antibiotics, anti-virals, antimetabolites, corticosteroids, and immunosuppressive therapy which included chlorambucil, azathioprine, cyclophosphamide, cyclosporine A, and so on [9]. Nowadays some biological agents were used abroadly for SC, such as infliximab, ranibizumab, interferon alpha-2a, and so on [10-12].

In this report one patient was treated with anti-tuberculosis and immunosuppressive agents, finally the retinal and choroidal lesions developed progressively but visual acuity remained stable. The other patient rejected any form of therapy and finally the lesions extended and visual acuity was impaired severely. For the complications of SC, such as CNV and macular edema, some therapies including anti-VEGF, photodynamic therapy and intravitreal injection TA were performed [2,10]. But there are limited data on long term follow-up to evaluate efficacy of the different treatment strategies.

In conclusion, the clinical features, outcomes and therapies of SC are very complicated. For SC patients, the retinal and

choroidal lesions will develop progressively and visual acuity decrease gradually. In this paper the long term follow-up showed the rate of progress and final outcomes of SC with systemic and ocular treatments were different with the patients without any treatment.

REFERENCES

1 Araujo AA, Wells AP, Dick AD, Forrester JV. Early treatment with cyclosporin in serpiginous choroidopathy maintains remission and good visual outcome. *Br J Ophthalmol* 2000;84(9):979-982

2 Lim JI, Flaxel CJ, LaBree L. Photodynamic therapy for choroidal neovascularisation secondary to inflammatory chorioretinal disease. *Ann Acad Med Singapore* 2006;35(3):198-202

3 Lim WK, Buggage RR, Nussenblatt RB. Serpiginous choroiditis. *Surv Ophthalmol* 2005;50(3):231-244

4 Sahu DK, Rawoof A, Sujatha B. Macular serpiginous choroiditis. *Indian J Ophthalmol* 2002; 50(3): 189-196

5 Wu JS, Lewis H, Fine SL, Grover DA, Green WR. Clinicopathologic findings in a patient with serpiginous choroiditis and treated choroidal neovascularization. *Retina* 1989; 9(4): 292-301

6 Cordero-Coma M, Benito MF, Hernandez AM, Antolin SC, Ruiz JM. Serpiginous choroiditis. *Ophthalmology* 2008;115(9):1633, e1-2

7 Erkkila H, Laatikainen L, Jokinen E. Immunological studies on serpiginous choroiditis. *Graefes Arch Clin Exp Ophthalmol* 1982;219(3): 131-134

8 Cardillo Piccolino F, Grosso A, Savini E. Fundus autofluorescence in serpiginous choroiditis. *Graefes Arch Clin Exp Ophthalmol* 2009;247(2): 179-185

9 Abrez H, Biswas J, Sudharshan S. Clinical profile, treatment, and visual outcome of serpiginous choroiditis. *Ocul Immunol Inflamm* 2007;15(4): 325-335

10 Battaglia Parodi M, Iacono P, Verbraak FD, Bandello F. Antivascular endothelial growth factors for inflammatory chorioretinal disorders. *Dev Ophthalmol* 2010;46:84-95

11 Seve P, Mennesson E, Grange JD, Broussolle C, Kodjikian L. Infliximab in serpiginous choroiditis. *Acta Ophthalmol* 2010;88 (8): e342-343

12 Sobaci G, Bayraktar Z, Bayer A. Interferon alpha-2a treatment for serpiginous choroiditis. *Ocul Immunol Inflamm* 2005;13(1):59-66