Journal of Clinical & Experimental Ophthalmology

Wang et al., J Clinic Experiment Ophthalmol 2013, S2
DOI: 10.4172/2155-9570.S2-006

Case Report Open Access

Atypical Serpiginous Choroiditis

Wei Wang[1,2], Lynn Gordon[2] and Ling Chen[1]*

[1]Eye and ENT Hospital, Department of Ophthalmology and Vision Science, Shanghai Medical School, Fudan University, Shanghai, China
[2]Ophthalmology, Jules Stein Eye Institute, David Geffen School of Medicine at UCLA, 100 Stein Plaza, Los Angeles, CA, 90095, USA

Abstract

We present an atypical case of serpiginous choroiditis, which may represent another variant of SC. The management of SC remains challenging, and decision on therapeutic approach should be made case by case after weighing benefits and side effects of the long-term immunosuppressive therapy.

Case Report

A 45-year-old man presented with blurred vision in the left eye for one week. His past medical and ocular histories were noncontributory. On examination, his visual acuity was 20/20 in the right eye (RE) and 20/200 in the left eye (LE). The intraocular pressure was 16 mmHg and 13 mmHg in the RE and LE eye, respectively. The anterior segment appeared quiet, whereas funduscopic examination revealed cream-colored plaques in the posterior pole of the LE (Figure 1B) and normal fundus in the RE (Figure 1A). Fluorescein angiogram showed hyperfluorescence plaque with late leakage in the LE (Figures 1C and 1D). Indocyanine green angiograph revealed hypofluorescence and hyperfluorescence in the early phase, hypofluorescence in the late phase (Figures 1E and 1F). Optical coherence tomography through the lesions revealed disrupted retinal pigment epithelium (RPE) and loss of the junction between the inner and outer segments of the photoreceptors. Extensive laboratory investigation (including syphilis, HIV, HLA-B27 and ANA, IgMs for herpes simplex virus, Rubella, toxoplasmosis and CMV) was performed with negative results. Chest computed tomography scan, purified protein derivative skin test and TB-specific interferon gamma test were negative. One week after initial presentation, his visual acuity dramatically decreased to light perception (LP). Intravenous dexamethasone was started at the dose of 10 mg per day, followed by oral prednisone (1mg/kg/d), and slowly tapered off in 3 months. His visual acuity in the LE was improved to hand motion (HM) and remained stable.

Four months after the initial presentation of the LE, the patient was referred to our clinic for decreased vision in the fellow (right) eye for one week. On examination, his visual acuity was 20/200 in the RE and HM in the LE, respectively. The anterior segment appeared quiet. Funduscopic examination showed cream-colored plaques in the posterior pole in the RE (Figure 2A) and scar and RPE changes in the LE (Figure 2B). No vitreous cells were noted in either eye. Autofluorescence showed hypofluorescence surrounded by hyperfluorescence in the RE (Figure 2C) and predominant hypofluorescence in the LE (Figure 2D). Fluorescein angiogram showed hyperfluorescence plaque with late leakage in the RE (Figure 2E) and window defect in the LE (Figure 2F). Indocyanine green angiograph revealed hypofluorescence and hyperfluorescence in the early phase, hypofluorescence in the late phase in the RE (Figures 2G and 2H). Oral prednisone was started at the dose of 1mg/kg/day. A good clinical response was observed, and his visual acuity improved 20/80 in the right eye. However, the prednisone was discontinued by the patient after 3 weeks due to bilateral edema of his lower limbs. Alternative immunosuppressive therapy was offered but was refused. The patient was closely observed for 8 months, and both eyes remained stable without signs of active inflammation (Figures 3A and 3B) His visual acuity was 20/100 in the RE and HM in the LE. Secondary choroidal neovascularization was not observed.

The Management of SC is Challenging

Serpiginous choroiditis (SC) is a rare, chronic, usually bilateral, progressive recurrent inflammatory disease involving choroid and the retinal pigment epithelium [1]. The classic presentations are ill-defined creamy chorioretinal patches in the peripapillary region representing

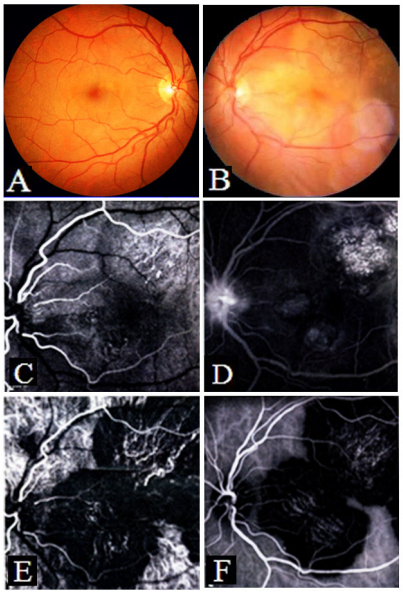

Figure 1: (A) Fundus photograph showing normal fundus in the RE. **(B)** Fundus photograph of the LE showing cream-colored plaques in the posterior pole, involving the temporal retina and the macular. **(C,D)** Fluorescein angiogram showing hyperfluorescence plaque with late leakage in the LE. **(E,F)** Indocyanine green angiograph of the LE showing hypofluorescence and hyperfluorescence in the early phase, hypofluorescence in the late phase.

***Corresponding author:** Ling Chen M.D., Ph.D., Fudan Affiliated Eye and ENT Hospital, Fenyang Road 83#, Shanghai 200031, China, Tel: +86-21-64377134; E-mail: linglingchen98@hotmail.com

Received January 29, 2013; **Accepted** February 16, 2013; **Published** February 23, 2013

Citation: Wang W, Gordon L, Chen L (2013) Atypical Serpiginous Choroiditis. J Clinic Experiment Ophthalmol S2: 006. doi:10.4172/2155-9570.S2-006

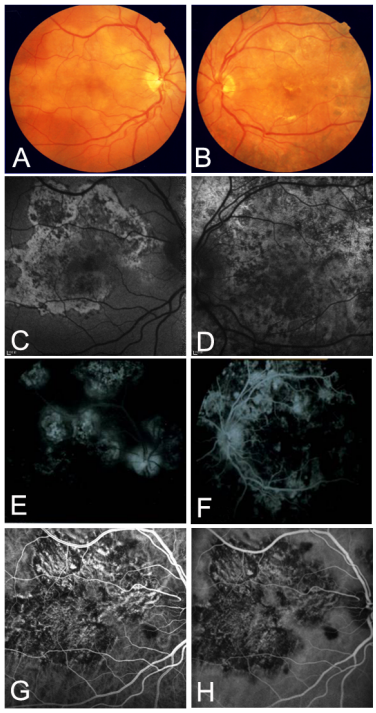

Figure 2: (A) Fundus photograph of the RE showing cream-colored plaques at level of RPE in the posterior pole. **(B)** Fundus photograph of the LE showing scar and RPE changes in the posterior pole. **(C,D)** Autofluorescence showing hypofluorescence surrounded by hyperfluorescence in the RE and predominant hypofluorescence in the LE. **(E,F)** Fluorescein angiogram showing hyperfluorescence plaque with late leakage in the RE and window defect in the LE. **(G,H)** Indocyanine green angiograph showing hypofluorescence and hyperfluorescence in the early phase, hypofluorescence in the late phase in the RE.

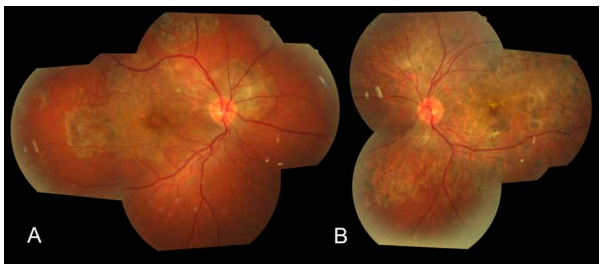

Figure 3: Eight month after initial presentation of the RE, the cream-colored plaques were disappeared and left with scar and RPE changes in the RE (A) and more pigment clusters in the posterior pole of the LE (B).

subretinal infiltration, progress in a centrifugal pattern [2] and hypofluorescent areas in both early and late phase of indocyanine green angiography (ICGA). The funduscopic and image findings in our case were quite similar to peripapillary SC, except for initial macular involvement. Although macular serpiginous choroiditis was reported in the literature, in that category the lesions in the macula are usually not continuous with disk [3], which is different from our case. The typical SC runs a chronic, relapsing and insidious course with many patients remaining asymptomatic until the macular involved later [1]. In contract, our case presented with early macular involvement and deteriorated rapidly with visual acuity dropped to LP in a week.

The main differential diagnoses include tuberculosis, APMPPE, outer retinal toxoplasmosis and multiple choroiditis. We did extensive workup to exclude tuberculosis in our case, since the management differs significantly. The acute lesions in APMMPE may be indistinguishable from SC, but APMMPE lesions usually resolve spontaneously with good visual prognosis and leave a mottled RPE without significant choroidal atrophy. Unlike APMMPE, poor visual outcome and significant choroidal atrophy were developed in our case. Multifocal choroiditis may also mimic SC, but usually have significant vitritis and anterior uveitis, which were absent in our case.

The management of SC is challenge. Corticosteroids may be efficacious in the treatment of the acute lesions, but have been reported to have no effect on the prevention of recurrences [4]. Immunosuppressive therapy has been reported to be effective in SC, but some cases relapsed despite triple immunosuppressive therapy [1,5]. Because of the potential life-threatening complications of immunosuppressive drugs, the patient needs to be informed and participate in the decision of long-term therapeutic approach. In our case, the patient had poor visual outcomes in the early stage due to macular involvement, and was informed the potential benefit of immunosuppressive therapy on disease activities but unlikely on visual acuity. Given the slim chance to improve the visual acuity in this case and potential severe side effects of long-term immunosuppressive therapy, the patient refused alternative immunosuppressive therapy and decided to be observed.

In summary, we present an atypical case of serpiginous choroiditis, which may represent another variant of SC. The management of SC remains challenging, and decision on therapeutic approach should be made case by case after weighing benefits and side effects of the long-term immunosuppressive therapy.

Funding

LC is sponsored by the Pujiang Talent Foundation (10PJ1401900), the National Science Foundation of China (81000381) and the Scientific Research Foundation for the Returned Overseas Chinese Scholars, State Education Ministry.

References

1. Lim WK, Buggage RR, Nussenblatt RB (2005) Serpiginous choroiditis. Surv Ophthalmol 50: 231-244.
2. Weiss H, Annesley WH Jr, Shields JA, Tomer T, Christopherson K (1979) The clinical course of serpiginous choroidopathy. Am J Ophthalmol 87: 133-142.
3. Mansour AM, Jampol LM, Packo KH, Hrisomalos NF (1988) Macular serpiginous choroiditis. Retina 8: 125-131.
4. Christmas NJ, Oh KT, Oh DM, Folk JC (2002) Long-term follow-up of patients with serpinginous choroiditis. Retina 22: 550-556.
5. Cordero-Coma M, Benito MF, Hernández AM, Antolín SC, Ruíz JM (2008) Serpiginous Choroiditis. Ophthalmology 115: 1633.

This article was originally published in a special issue, **Ocular Diseases: Immunological and Molecular Mechanisms** handled by Editor(s). Dr. C S De Paiva, Cullen Eye Institute, Baylor College of Medicine, USA