Original Article

# Profile of serpiginous choroiditis in a tertiary eye care centre in eastern India

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**Purpose:** To study the clinical profile of serpiginous choroiditis in eastern India. **Materials and Methods:** Ninety-one eyes of 54 patients with serpiginous choroiditis presenting to a tertiary care centre in eastern India between January 2006 and December 2010 were included in the study. Clinical presentation, treatment given, and visual outcome of the eyes were studied. **Results:** Thirty-five (64.8%) patients were male and 19 (35.2%) were female in the age group of 13-62 years (mean age: 34.1 ± 18.7 years). Blurring of vision (71; 78%) and floaters (36; 39.5%) were commonest symptoms. In 75 (82.4%) eyes, choroiditis started from optic nerve head and spreading centrifugally. Overall, 38 (41.75%) eyes had macular involvement at first visit. Mantoux test reading was 10 mm or more (Group A) in 12 (22.22%) patients and less than 10 mm (Group B) in 42 (77.77%) patients. Difference between Groups A and B in macular involvement at first visit (10; 50% vs. 28; 39.4%) and rate of recurrence (3; 15% vs. 14; 19.7%) was not statistically significant ($P$ = 0.37 and 0.68). Oral steroid (51; 94.4%) was the commonest mode of treatment. Fifty-one (56%) eyes had two lines or more improvement in vision. **Conclusions:** The present study details the clinical presentation, treatment, and visual outcome of serpiginous choroiditis. Mantoux test reading does not affect the clinical presentation or the treatment outcome in these eyes.

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Serpiginous choroiditis is a chronic, bilateral, and recurrent inflammatory disease of the choroid and retinal pigment epithelium.[1-4] It derives its name from the typical clinical appearance of centrifugally spreading lesion with serpentine borders in the active stage of the disease.[1] Macular involvement in the form of choroiditis in active stage and scarring, fibrosis, and choroidal neovascular membrane as the sequelae are the major causes of visual loss in this disease.[5] The disease follows a chronic course with recurrences and has predilection to start from the peripapillary region.[1-5] However, isolated macular and peripheral diseases have been well documented.[1,5-7]

Myriad of studies have tried to explain the etiology of serpiginous choroiditis, yet the exact etiology for this condition remains elusive.[1,8-19] In the middle of 20th century, tuberculosis was thought to be the cause, but later, it was described to the present as serpiginous-like choroiditis (SLC).[8-10] Francisella tularensis and Bartonella henselae have also been reported to cause serpiginous choroiditis.[11] Positive serology for Herpes simplex virus in patients with serpiginous choroiditis had fueled the speculation of viral etiology, but histopathology study with polymerase chain reaction (PCR) on choroidal tissue refuted the hypothesis.[12,13] Serpiginous choroiditis has also been seen as unusual presentation in patients with ocular sarcoidosis, Crohn's disease, lung carcinoma, uterine cervix carcinoma, systemic lupus erythematous, non-Hodgkin's lymphoma, and autoimmune hepatitis.[14-17] Added to the elusive etiology, the similar clinical picture of acute posterior multifocal placoid pigment epitheliopathy, posterior scleritis further complicates the correct diagnosis and management of a case of serpiginous choroiditis.[10,18,19]

Serpiginous choroiditis has varied clinical presentations in different geographical regions.[1,2,4] Abrez *et al*. from south India have reported series of patients with serpiginous choroiditis treated with a combination of oral steroids, immunosuppresses, and antituberculous treatment.[1] Gupta *et al*. from north India have reported it to be present at an earlier age than Caucasian population and also as more likely to present as unilateral disease.[2,4,18] In endemic region like India, management of serpiginous choroiditis starts first probably with exclusion of ocular tuberculosis, followed by administration of systemic steroids and immunosuppression. The present study intends to present the clinical profile, management, and outcome of serpiginous choroiditis in eastern India.

## Materials and Methods

It was a retrospective case analysis of patients with serpiginous choroiditis visiting a tertiary referral care eye hospital in eastern India between January 2006 and December 2010. The data was obtained from the medical records that included clinical features, investigations, and treatment. Acute yellow-white lesion at the level of retinal pigment epithelium and choroid with serpiginous projections spreading centrifugally was considered diagnostic of serpiginous choroiditis. All patients needed to have active serpiginous choroiditis in at least one eye and a minimum follow-up of 6 months to be included in the study.

Demographic data, presenting symptoms, and laterality were noted from the medical records. Visual acuity measurement with Snellen's chart, anterior segment examination with slit lamp, fundus examination with indirect ophthalmoscope, and slit lamp biomicroscope was performed at all visits. The

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site of serpiginous choroiditis, macular involvement, and vision-threatening complications like choroidal neovascular membrane and subretinal macular fibrosis were looked for at all the visits. The number and timing of recurrence of disease was noted for all the eyes. Other associated features like retinal vasculitis, pars planitis, and retinal vascular occlusion were also noted.

All patients underwent fundus fluorescein angiogram (FFA) at least at the first visit. FFA was repeated if there was a new lesion in same or the other eye or if there was a suspicion of recurrence of the disease. Active choroiditis was defined as grayish-yellow cream-colored lesion at the level of retinal pigment epithelium showing early central hypofluorescence and late fuzzy hyperfluorescence at the expanding margins on FFA. Healed choroiditis was defined as variable retinochoroidal scarring with distinct hyperflorescent margins without fuzzing on FFA. Mantoux test and chest X-ray were performed in all patients, while sputum examination for acid-fast bacilli was performed in patients with positive Mantoux test and radiological evidence of pulmonary tuberculosis. The patients were divided into two groups (A and B) based on Mantoux test reading of more than and less than 10 mm, respectively. Treatment details with drugs used and route of administration were noted. Favorable anatomical outcome was defined as completely healed lesion with no activity at final follow-up. Favorable functional outcome was defined as two lines or more of improvement in visual acuity on Snellen's chart at final follow-up. The statistical software SPSS, version 11, USA was used to perform Chi-square test. $P < 0.05$ was considered significant.

## Results

Ninety-one eyes of 54 patients with serpiginous choroiditis were included in this study. Out of these, 52 patients belonged to eastern region of India and 2 patients were from Bangladesh. There were 35 (64.8%) males and 19 (35.2%) females in the age group of 13-62 years (mean: 34.12 ± 18.72 years). The mean follow-up was 36.53 months (range: 6-133 months, standard deviation 29.3 months). A total of 37 (68.5%) patients had bilateral disease and 17 (31.5%) had unilateral disease. Out of total 91 eyes, 50 (54.94%) had 20/60 or better visual acuity at presentation [Table 1]. Blurring of vision (71; 78.02%) and floaters (36; 39.50%) were the commonest symptoms.

None of the eyes had active or sequelae of anterior uveitis. At presentation, 69 (75.82%) eyes had exclusively active serpiginous choroiditis, 12 (13.18%) eyes had both active and healed lesions, and 10 (10.98%) eyes had only healed choroiditis lesions. In 75 (82.41%) eyes, choroiditis started around optic nerve head (ONH) and extended centrifugally. Isolated macular involvement without origin near ONH margin was seen in 10 (11%) eyes, while peripheral lesions without involvement of macula or ONH margin were seen in 6 (6.59%) eyes [Fig. 1]. Overall, macular involvement at initial presentation was noted in 38 (41.75%) eyes. Retinal vasculitis was noted in 2 (2.19%) eyes and branch retinal vein occlusion was seen in 1 (1.09%) eye. None of the eyes had localized or diffuse vitreitis.

Mantoux test reading was 10 mm or more (Group A) in 12 (22.22%) patients and less than 10 mm (Group B) in 42 (77.77%) patients [Table 2]. There was no significant difference between

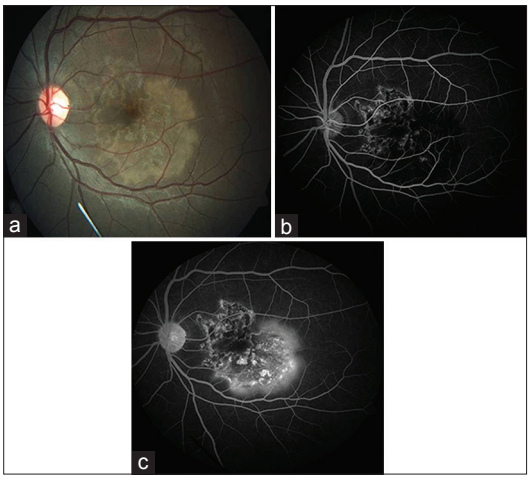


**Figure 1:** (a) Colour fundus photograph showing macular serpiginous choroiditis with activity at temporal edge. (b) Fundus fluorescein angiogram (FFA) image showing distinct hyperfluorescence at the inactive nasal edge of the lesion. (c) Late FFA image showing fuzzy hyperfluorescence of active temporal edge, while hyperfluorescence at healed edge remain distinct

Group A and B with respect to laterality ($P = 0.84$), macular involvement at first visit ($P = 0.37$), recurrence rate ($P = 0.68$), and outcome ($P = 0.44$, 0.26) [Table 2]. None of the patients had active pulmonary tuberculosis after clinical and laboratory investigations (sputum for acid-fast bacilli, X-ray chest, computerized tomography of chest). Three (5.55%) patients had calcified hilar opacities on chest X-ray and were suspected to have old healed pulmonary tuberculosis.

Thirty-five (64.81%) patients were treated with oral steroids (1 mg/kg) alone. Sixteen (29.62%) patients received oral steroids in combination with immunosuppressives. Azathioprine was used in 14 (25.92%) patients in a dose of 2-5 mg/kg in three divided doses, while cyclosporine was used in two (3.70%) patients in a dose of 5 mg/kg in two divided doses. Remaining three (5.55%) patients received immunosuppressive (azatioprine) alone. The two patients in cyclosporine was use were intolerant to azathioprine. Overall, high-dose intravenous steroid therapy (HDIST) was needed due to fovea-threatening choroiditis in 20 (37.03%) patients. Posterior sub-Tenon injection of triamcinolone acetonide was used in 21 (23.07%) eyes. Periocular steroid was used when patients who had systemic steroid-related side effects, prohibiting its use or had deranged liver function test after immunosuppressive. Combination of oral steroid and immunosuppressive as initial therapy was employed in case of eyes that had fovea-threatening serpiginous choroiditis or in whom oral steroid had to be stopped because of comorbid conditions like uncontrolled hypertension or diabetes mellitus.

Seventy-five (82.41%) eyes did not have a recurrence after initial treatment, while 12 (13.18%) eyes had one recurrence and four (4.39%) eyes had two recurrences. All these 16 eyes with recurrence were unilateral disease in patients with earlier bilateral involvement. Ten (28.57%) of these patients were initially treated with oral steroids alone ($n = 35$). Six (42.85%) of these patient were initially treated with oral steroids with azatioprine ($n = 14$). There was no significant difference in the rate of recurrence with respect to initial treatment ($P = 0.34$).

Fifteen (93.75%) eyes with recurrence had initial choroiditis around ONH margin, and one (6.25%) eye had isolated macular involvement earlier.

At final follow-up, 83 (91.20%) eyes had completely healed serpiginous choroiditis, while eight (8.79%) had still active lesions. Sixty-six (72.52%) eyes had final visual acuity of 20/60 or better at final follow-up [Table 1]. Favorable anatomical outcome was achieved in 76 (83.51%) eyes. Favorable functional outcome was achieved in 52 (57.14%) eyes. Macular scar and fibrosis (22; 56.41%), cataract (16; 41.02%), and neovascular glaucoma (1; 2.56%) were the cause of non-achievement of favorable functional outcome in the remaining 39 (42.85%) eyes.

## Discussion

Elusive etiology of serpiginous choroiditis makes the diagnosis and management of this condition a daunting task.[9,20,21] Although it causes painless progressive loss of vision, acute vision-threatening choroiditis needs timely institution of HDIST to salvage the vision.[22] Similar clinical presentation of ocular tuberculosis that requires antituberculous treatment for control of the disease further complicates the management of serpiginous choroiditis.[9] Study of clinical characteristics of serpiginous choroiditis is essential for the in-depth understanding of this disease, which would help in proper management.

Preponderance of case in men with a mean age of presentation of 34 years noted in our study was similar to those noted in northern and southern India.[1,18] However, it was different from other studies with Caucasian population, which reported later onset of disease.[4,23] Bilateral involvement of eyes was more common in our study population unlike the report from north India.[18] Majority of eyes with serpiginous choroiditis had initial lesion around the ONH in the present study, a feature shared by other Indian and Caucasian series.[1,4,18,23] However, isolated macular involvement was less common in our group of patients than that reported by Abrez *et al*. from south India (11% vs. 21%). Overall, more than one-third patients had macular involvement at initial presentation, necessitating early diagnosis and institution of therapy.

**Table 1: Initial and final visual acuity (*n*=91)**

| BCVA | Initial | Final |
|---|---|---|
| 20/20-20/60 | 50 (54.94) | 66 (72.52) |
| <20/60-20/200 | 27 (29.67) | 13 (14.28) |
| <20/200 | 14 (15.38) | 12 (13.18) |

BCVA: Best corrected visual acuity, Values in parenthesis denote percentage

**Table 2: Relation of mantoux test findings with disease characteristic**

| Disease characteristics | Findings | Mantoux test | | *P* value |
|---|---|---|---|---|
| | | Less than 10 mm 42 persons 71 eyes | 10 mm or more 12 persons 20 eyes | |
| Laterality | Unilateral | 13 (31) | 4 (33.33) | 0.843 |
| | Bilateral | 29 (69) | 8 (66.66) | |
| Macular involvement at first visit | Yes | 28 (39.43) | 10 (50) | 0.377 |
| | No | 43 (60.56) | 10 (50) | |
| Recurrence | Yes | 14 (19.71) | 3 (15) | 0.681 |
| | No | 57 (80.28) | 17 (85) | |
| Favorable anatomical outcome | Yes | 59 (83) | 18 (90) | 0.444 |
| | No | 12 (17) | 02 (10) | |
| Favorable functional outcome | Yes | 42 (59.15) | 09 (45) | 0.265 |
| | No | 29 (40.84) | 11 (55) | |

Values in parenthesis denote percentage

Classification of patients based on Mantoux test reading did not reveal any significant difference in disease characteristics across the groups. Since none of the patients with Mantoux test positivity had any evidence of systemic tuberculosis, none of them had anterior segment inflammation or vitritis and had serpiginous instead of multifocal arrangement of lesions; antituberculous treatment was not instituted for the management in our patients. Redundancy of Mantoux test in the management of retinal vasculitis has been reported in past, and the same may be applicable to serpiginous choroiditis.[2,24] Interferon-gamma assays has shown promising results in diagnosis of latent tuberculosis in patients with uveitis.[25] However, it lacks the specificity to distinguish latent tuberculosis from active tuberculosis.[26] We had not performed interferon gamma assay in our group of patients and relied solely on Mantoux test and systemic evaluation (X-ray chest, sputum examination for acid-fast bacilli) for diagnosis of tuberculosis. Tuberculosis is known to be an ocular masquerade and can affect both anterior and posterior segment.[26] Differentiation of serpiginous-like choroiditis, which is a less common manifestation of ocular tuberculosis from serpiginous choroiditis can pose a diagnostic challenge for the ophthalmologist.[10,26] This distinction is more important as the treatment protocol of the two entity differ, as the former needs antituberculous treatment, while the later is treated solely with immunosuppressive agents. Bilateral involvement more commonly in the peripapillary region with clear vitreous has been differentiating features of serpiginous choroiditis from serpiginous-like choroiditis, which is seen more commonly in multifocal arrangement with significant vitritis.[9,26] Although serpiginous-like choroiditis itself and tuberculous association in patients with serpiginous choroiditis are presumed to be more common in endemic region like India, our study did not reveal any tuberculous association with serpiginous choroiditis.

Oral steroid was the mainstay of treatment in patients with serpiginous choroiditis. In our group of patients, there was no difference in the rate of recurrence among patients initially treated with oral steroid alone and those treated with combination therapy. This is in keeping with the study by Abrez *et al*. from South India, but, in contrast to the study by Christmas *et al*. from United States of America, who had noted a lower number of recurrence in patients treated initially with combination of oral steroids and immunosuppressives.[1,5] Similarly, neither of the anatomical location of the initial choroiditis had any bearing on the rate of recurrence. The presentation of serpiginous choroiditis is noted to vary across different study populations, and there is a need for increased reporting of cases for better understanding of this disease.[18]

The current treatment protocol for serpiginous choroiditis

also remains controversial.[5,27] Although oral steroid remain the mainstay, there is often a need for steroid-sparing agent (immunosuppressive) in view of non-response, recurrence, or steroid-induced side effects.[1,5,18,26-28] Antiviral and antituberculous therapy has been used in selected cases, but their use is still debatable until we find a firm etiological association.[1,26-28] Our study presents a series of patients who were treated with corticosteroids and immunosuppressives without antituberculous or antiviral therapy. This finding is significant as the present study comes from a region endemic for tuberculosis. The fact that we did not perform PCR for *Mycobacterium tuberculosis* or *H. simplex* can be cited as a drawback of this study. Such investigations are usually advised on positive leads on history and clinical examination, which we did not find in our patients, and hence these tests were not advised. The present study also stresses that Mantoux test postivity alone may not influence the clinical presentation, treatment, and outcome of serpiginous choroiditis. It adds to the existing knowledge about the clinical presentation and outcome of serpiginous choroiditis.

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