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CASE REPORT

Serpiginous choroidopathy

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ABSTRACT

Objective

To describe a case of serpiginous choroidopathy.

Methods

This is a case report.

Results

A 61-year-old female with a 22-year history of gradual blurring of vision in the left eye sought consultation. Ten months prior, her left vision worsened, described as central scotoma that progressed inferiorly. Best-corrected vision was 20/20 (right) and counting fingers at 1 foot (left). Inferior hemifield scotoma was documented on Amsler grid testing of the left eye. Anterior-segment findings were unremarkable. Retinal examination through a clear media revealed multiple contiguous hypopigmented patches radiating from the peripapillary area extending to the periphery in both eyes with extension to the superior fovea on the left. Fluorescein angiogram showed progressive faint hypofluorescence of the hypopigmented patches in both eyes with involvement of the superior fovea on the left. No active vessel leakage was noted. No treatment was given and regular Amsler monitoring was advised. Follow-up 3 and 6 months after revealed stable visual acuity and fluorescein angiogram (FA) findings.

Conclusions

This is a case of serpiginous choroidopathy with inactive pattern. There was unilateral decrease in central vision, scotoma, and retinal pigment epithelial atrophy in a serpentine pattern originating from the disc with macular involvement in one eye. FA aids in the diagnosis and monitoring of inflammatory activity as the presence of active leakage on the borders. Goals of management include monitoring, prevention of recurrences and progression, and rapid control of sequela with potential use of immunosuppressive therapy.

Keywords: *Serpiginous choroidopathy, Serpiginous choroiditis, Geographic peripapillary choroidopathy, White-dot syndromes, Uveitis*

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SERPIGINOUS choroidopathy is a rare disease first described in 1932 by Junius as peripapillary retinochoroiditis and known afterwards as a plethora of names until Gass in 1987 coined "serpiginous choroidopathy" because of its characteristic funduscopic pattern. The complete clinical features and fluorescein angiographic findings of serpiginous choroidopathy currently recognized was described as early as the 1970s. Variants of the disease have been reported but it is typically bilateral, chronic, progressive recurrent inflammation of the retinal pigment epithelium, choriocapillaris and choroid of unknown etiology. Because of the rarity and variable course of the disease, present understanding of this disease remains limited and long-term management a challenge.[1]

CASE REPORT

A 61-year-old female with a 22-year history of gradual blurring of vision in the left eye had sought multiple consultations but had unrecalled diagnosis. Ten months prior to consultation with us, her left vision worsened, described as a central scotoma that progressed inferiorly. Medical and ocular history and review of systems were unremarkable. Best-corrected vision was 20/20 (right) and counting fingers at 1 foot (left). Inferior hemifield scotoma was documented on Amsler grid testing of the left eye. Anterior-segment findings were unremarkable.

Retinal examination through a clear media revealed multiple contiguous hypopigmented patches radiating from the peripapillary area extending to the periphery in both eyes with extension to the superior fovea on the left (Figures 1, 2A, 3A). Fluorescein angiogram showed progressive faint hypofluorescence of the hypopigmented patches in both eyes with involvement of the superior fovea on the left. No active vessel leakage was noted (Figures 2B, 3B). No treatment was given and regular Amsler monitoring was advised. Follow-up at 3 months (Figures 2C, 3C) and 6 months (Figures 2D, 3D) revealed stable visual acuity and fluorescein angiogram findings.

DISCUSSION

Serpiginous choroidopathy is a rare disease constituting less than 5% of posterior uveitis and affecting healthy young to middle-aged adults (30 to 60 years of age) with no gender, race, or familial predilection or established systemic association. Although it is usually bilateral, the typical presentation is a unilateral decrease in central vision, metamorphopsia, or scotoma.[1] Our patient was a healthy 61-year-old female, with no comorbidities, whose blurring of central vision and scotoma in the left eye started when she was 31 years old.

As with our patient, anterior and posterior segments may be normal with no inflammatory cells or flares except for the peripapillary serpentine lesions in the fundus (Figures 1, 2A, 2B). Although the lesions are not multifocal, some authors have classified it as one of the white dot syndromes.

There are 3 distinct clinical presentations; namely, classic, macular, and atypical. About 80% of reported cases are of the classic or peripapillary geographic pattern, including our patient. The active disease begins as ill-defined patches of grayish or creamy yellow subretinal infiltrates originating in the peripapillary region and progressing in an irregular serpentine fashion centrifugally with an edematous overlying retina. These active lesions usually resolve after 6 to 8 weeks, with or without treatment, leaving areas of retinal pigment epithelial (RPE) and choriocapillary atrophy, surrounded by skip islands of normal retina. Multiple lesions in different stages of

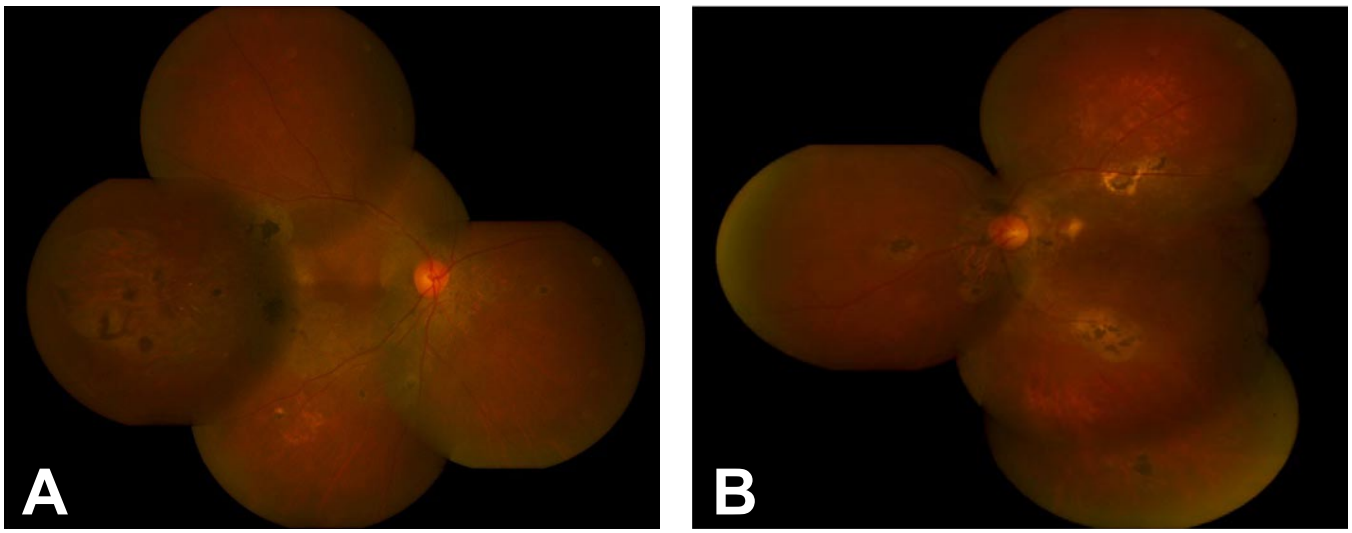

Figure 1. Fundus photo of the right eye (A) and left eye (B) showing multiple contiguous hypopigmented patches radiating from peripapillary area to the periphery.

resolution are usually present. Recurrences, usually at the edges of atrophic scars, occur at variable intervals, from months to years. The disease runs an insidious course with many patients asymptomatic until the macula is involved. Approximately two-thirds of patients have macular scars in one or both eyes at the time of presentation. Seventy-five percent of these patients develop visual loss in one or both eyes with final visual acuity of less than 20/200 despite treatment.[1] Our patient, whose consultation was prompted by progressive blurring of her central vision in the left, had vision of counting fingers with RPE atrophy and scars noted on both fundus.

The macular pattern is a variation of the classic type but the typical chorioretinal serpentine lesions in the macula are map-like and are not continuous with the optic nerve. It has a higher risk of poorer visual outcome due to its proximity to the fovea, and is usually late or under-diagnosed as age-related macular degeneration, toxoplasmosis, or macular dystrophy.

Ampiginous or atypical choroidopathy usually occurs in the periphery in isolation or in a multifocal pattern but with less foveal involvement.[1]

Fluorescein angiography (FA) had characteristic features of serpiginous choroidopathy but were not pathognomonic. The active lesions (borders) showed blocked fluorescence due to choroidal nonperfusion and late diffuse staining and leakage (Figures 2B-D, 3B-D). Inactive lesions show early hypofluorescence and late progressive hyperfluorescence at the margins due to staining of the underlying sclera and fibrosis.[1] These angiographic findings maybe similar to acute posterior multifocal placoid pigment epitheliopathy (APMPPE)

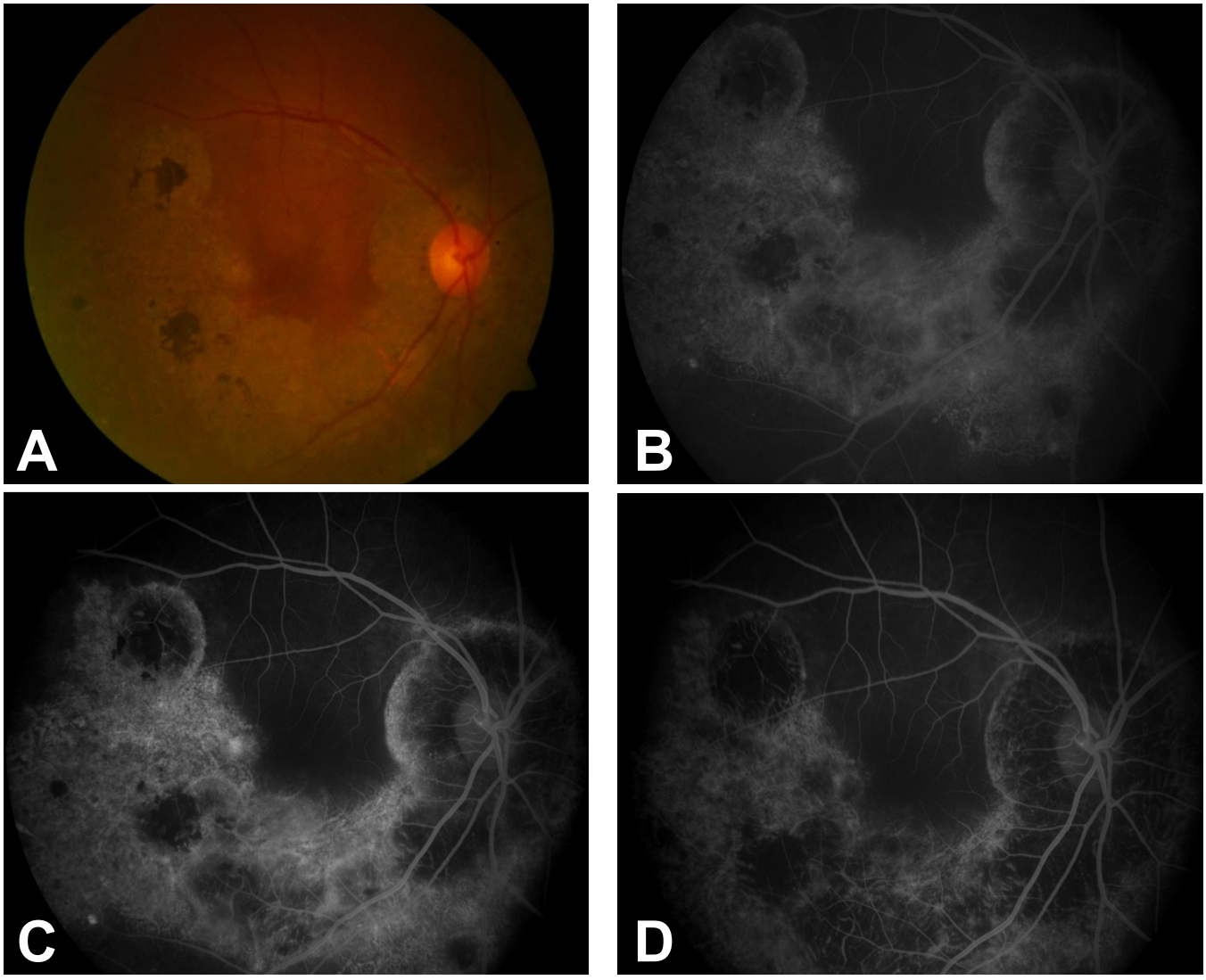

Figure 2. Fundus photo of the right eye at the time of consultation (A); midphase fluorescein angiogram (FA) at time of consultation (B); midphase FA at 3 months follow-up (C); midphase FA at 6 months (D).

and choroidal ischemia which all need to be excluded in the differential diagnosis.

Serpiginous choroidopathy can be assessed with various diagnostic tools. Because it is a disease of the choriocapillaris and RPE, indocyanine green angiography (ICG) can also be used. Early hypofluorescence of the lesions extending beyond the area of clinically observable lesions is present. Some reports noted that occult lesions became apparent on ICG (Saenz, 2003).[2] Optical coherence tomography imaging shows hyper-reflectivity of the outer retina with disintegration of the RPE and choriocapillaris producing the waterfall effect.[3] Visual-field testing demonstrates scotoma corresponding to the geographic lesions which become less dense as active lesions resolve over months. Electrooculogram and electroretinogram usually show normal wave forms except in extensive and late disease.[1]

The pathogenesis of serpiginous choroidopathy remains unknown but various studies have mainly proposed an autoimmune pathology. The inflammatory nature of serpiginous choroidopathy is supported by the presence of vitritis, anterior uveitis, and phlebitis in some patients; histopathological finding of lymphocytic infiltrates in the choroid and vessel walls in one report; and association of HLA B27 and decreased C3 in three reports. Other less supported proposed etiologies included infectious, vascular and degenerative.[1]

The natural history of serpiginous choroidopathy is multiple recurrences and progressive scarring which may eventually involved the fovea. Without treatment, the active lesions typically resolve over a few months with gradual extension of the primary lesions. However,

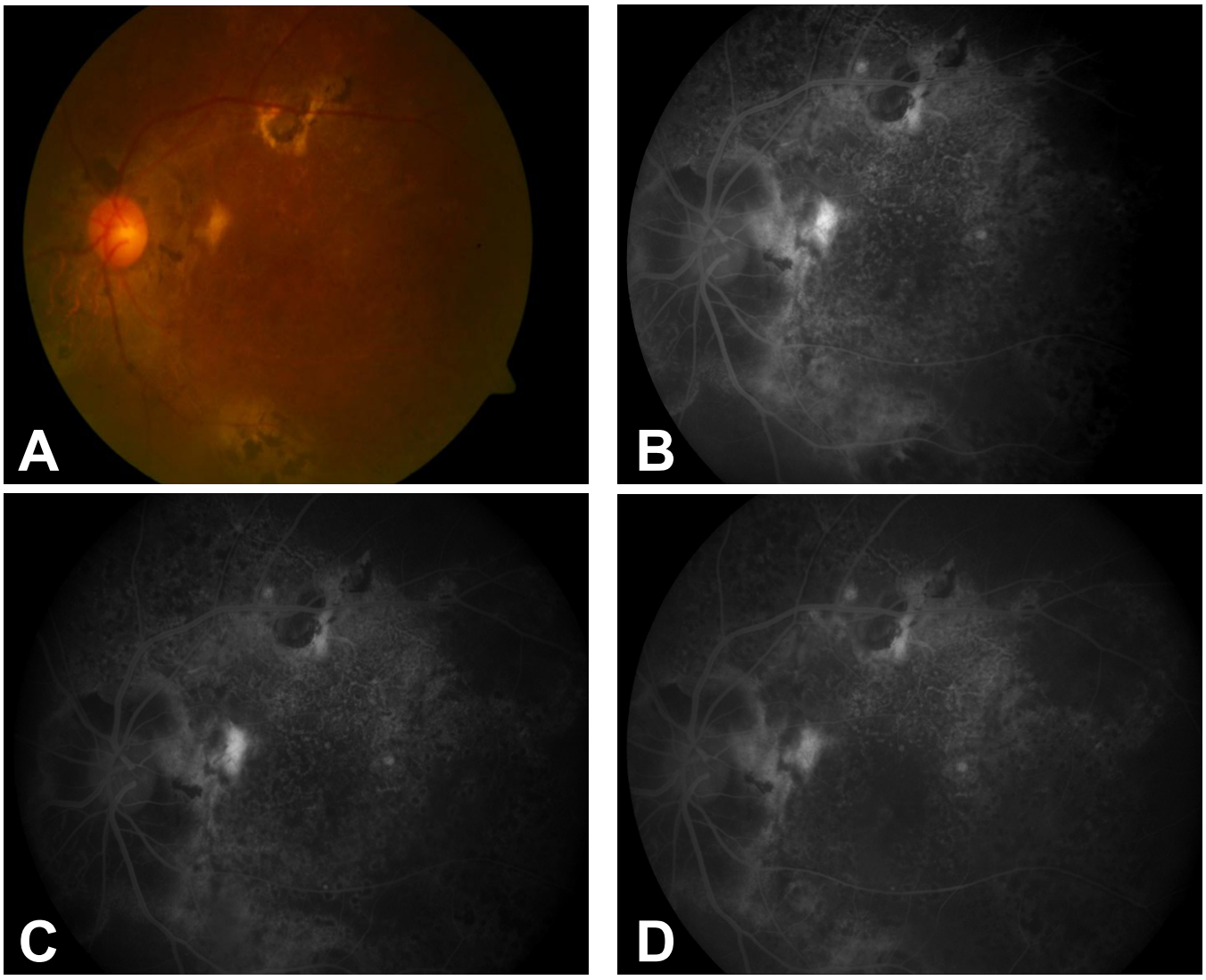

Figure 3. Fundus photo of the left eye at the time of consultation (A); midphase fluorescein angiogram (FA) at time of consultation (B); midphase FA at 3 months follow-up (C); midphase FA at 6 months (D).

active lesions may also remain active for as long as nine months.

The most common and visually significant ocular complication is choroidal neovascularization (CNV), present in 13 to 35% of patients with serpiginous choroidopathy. It is a direct response to intraocular inflammation altering balance between pro and anti-vascular-endothelial-growth factor observed as poorly defined subretinal lesions with early hyperfluoresence.[4] Other sequelae include branch retinal vein occlusion, periphlebitis, pigment epithelium detachment, serous retinal detachment, cystoid macular edema, optic disc neovascularization, subretinal fibrosis, and anterior uveitis.[1] The goals of treatment are rapid control of active lesions during recurrences and prevention of further recurrences, progression of the disease, and complications.

Immunosuppressive therapy is the treatment of choice based on available evidence. In complicated cases, laser photocoagulation is the management for CNV. In two retrospective studies, cyclosporine, azathioprine, and mycophenolate mofetil in one review and chlorambucil or cyclophosphamide in another, the authors concluded that immunosuppressive treatment ultimately inhibited proliferation of lymphocytes and long-term use for at least 6 months, ensured inflammatory quiescence, prolonged remission, and preserved vision.[5-6] No randomized clinical trial on the treatment of serpiginous choroidopathy has been performed, as it is a rare condition.[1]

Serial fundus photography and FA are utilized to document non-progression and to demonstrate the success of any therapeutic approach. Visual acuity is not an objective measurement of non-progression. Even if central visual acuity is preserved, the ensuing scotoma caused by atrophic parafoveal lesions can be debilitating and be a nidus for future CNV.[1]

CONCLUSION

This is a case of serpiginous choroidopathy with inactive pattern. It is a rare, vision-threatening disease in the presented case, producing unilateral decrease in central vision, scotoma, and RPE atrophy in a serpentine pattern originating from the disc with macular involvement in one eye. Fluorescein angiogram aids in the diagnosis and monitoring of inflammatory activity. Goals of management include monitoring, prevention of recurrences and progression, and rapid control of sequelae with potential use of immunosuppressive therapy.

References

1. Lim WK, Buggage RR, Nussenblatt RB. Serpiginous choroidopathy: major review. *Surv Ophthalmol* 2005; 50: 231-244.
2. Garcia-Saenz MC, Gili MP, Banuelos BJ, et al. DiazIndocyanine green angiography in chorioretinal inflammatory diseases. *Arch Soc Esp Oftalmologia* 2003; 78: 675-683.
3. Gallagher MJ, Yilmaz T, Cervantes-Castaneda RA, Foster CS. The characteristic features of optical coherence tomography in posterior uveitis. *Br J Ophthalmol* 2007; 91: 1680-1685.
4. Lee DK, Suhler EB, Augustin W, Buggage RR. Serpiginious choroidopathy presenting as choroidal neovascularization. *Br J Ophthalmol* 2003; 87: 1184-1197.
5. Akpek EK, Jabs DA, Tessler HH, et al. Successful treatment of serpiginous choroiditis with alkylating agents. *Ophthalmology* 2002; 109: 1506-1513.
6. Akpek EK, Baltatzis S, Yang J, Foster CS. Long-term immunosuppresive treatment of serpiginous choroiditis. *Ocular Immunology & Inflammation* 2001; 9: 153-167.