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Case report: 52-year-old woman with rare form of macular serpiginous choroiditis.

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Abstract

Purpose: To report a 52-years old female with macular involvement of Serpiginous choroidopathy (choroiditis).

Methods: Total clinical systemic examination, computed tomography, and optical coherence tomography was performed in this case.

Results: The eye condition was managed by oral corticosteroids, Mycophenolate Mofetil and Methotrexate. Despite aggressive four-year treatment the disease is still active, lesions increase in size and shape, and new foci.

Conclusions.SC is a chronic,bilateral, and recurrent disease with an unknown etiology.The applied treatment in this case turned out to be insufficient to stop the progression of the disease and there was no achievement of remission.The use of biologic drugs is questionable in this case.

Key words. Serpiginous choroiditis (SC),Serpiginous-like choroiditis (SLC), tuberculosis

Introdaction

Serpiginous choroiditis (SC) is a chronic, usually bilateral inflammation of the retinal pigment epithelium outer retina and choriocapillaris. The disease typically starts peripapillary and then gradually spreads, rarely affecting the macula.The main problem of this autoimmune disease is usual recurrence and the formation of new foci, which leads to retinal scarring and macular atrophy.[1]

The cause of SC is still unknown;however, it seems to be inflammatory or autoimmune. There are associations with an autoimmune response to the retinal S antigen and other infectious diseases such as tuberculosis and syphilis. [2]

Case report

A 52-year-old woman came to theclinic with complains of decreased, blurred vision and grey-white visual fields of one month duration in the left eye. Primary vision examination showed best corrected visual acuity (BCVA) of 1,0 in both eyeswith hypermetropic correction +4.00. Slit-lampexamination of the anterior segment and intraocular pressure were normal.

Humphrey's 120-point full field screening test demonstrated peripheral visual field defects in the righteye and central scotoma of 15 degrees in the left eye.**(Fig.1.)**

A fundoscopic examination of the right eye had one yellowish lesion peripapillary and macula was not involved. **(Fig. 2.)**In the left eye were found multiple subretinal yellowish-greyish plaques with localization around the optic nerve disc and macula. There was found large, elongated, yellowish atrophic chorioretinallesion with pigmentation in the upper border and paramacular localization.**(Fig. 4.)**Also,multiple yellowish lesions peripapillary were found. **(Fig. 5.)**

Fig.1. Visual field testing demonstrated peripheral visual field defects in the right eye and central scotoma in the left eye.

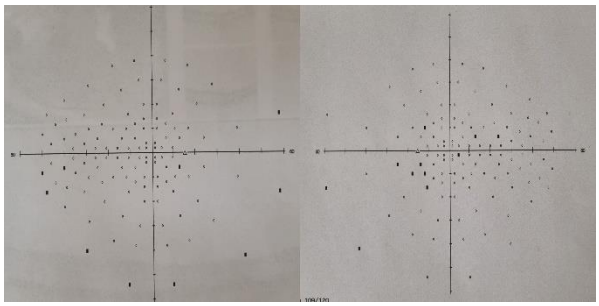


Fig.2. Fundus and FA photo of the right eye

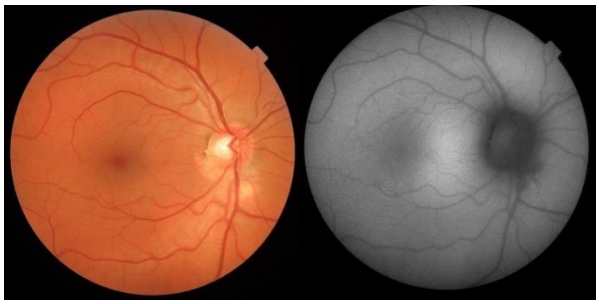


Fig.3. Color fundus photo of the left eye

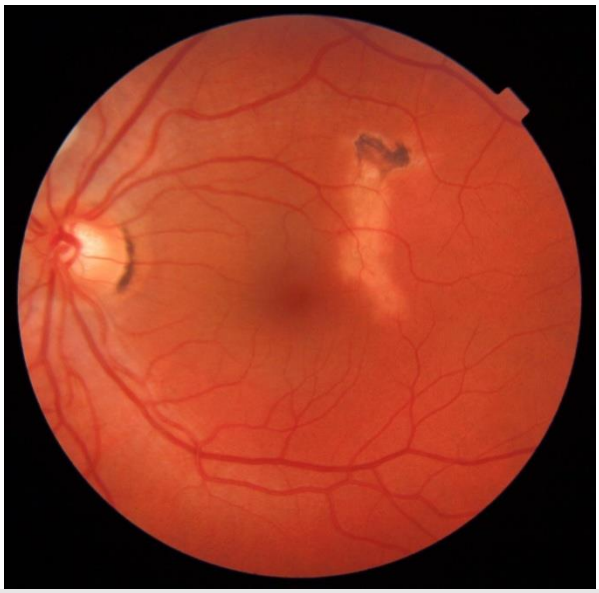


Fig.4. Color fundus photo of the left eye

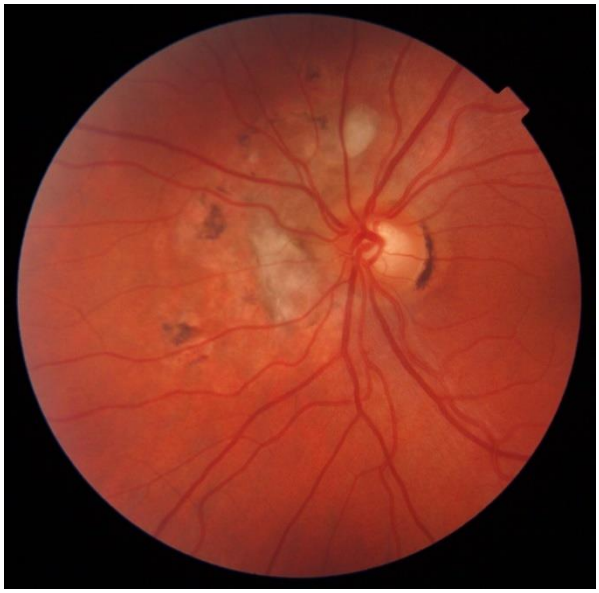
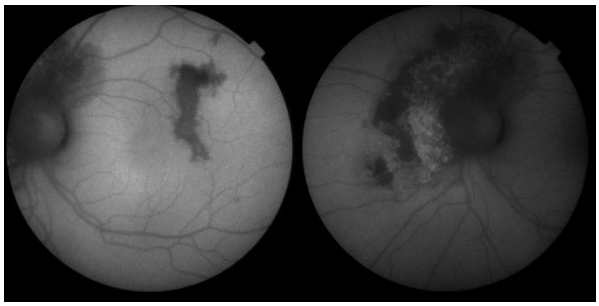


Fig.5. FA of the left eye

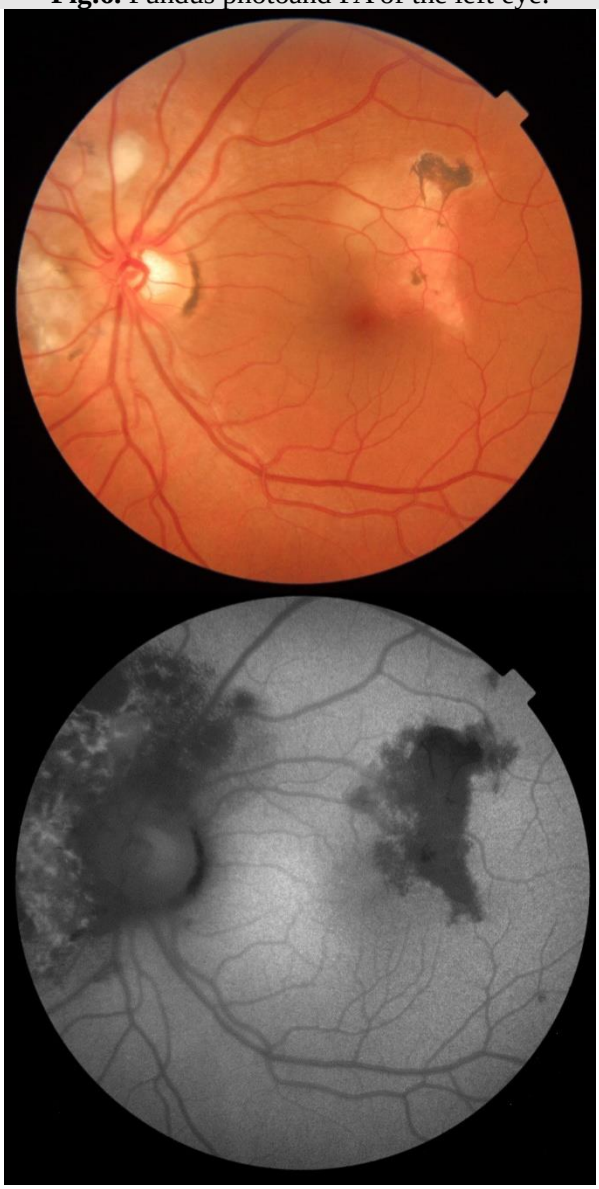


To rule out the diagnosis, a complete systemic study was done (complete blood count, erythrocytic sedimentation rate, blood sugar and coagulation). Immune profile tests (ANCA, ANA, HLA - B51, HLA-A29, HLA – B27, antistreptolysin, Rheumatoid factor, anti CCP) results were within normal limits. All infectious diseases were excluded due to negative results of chest computed tomography, QuantiFERON-TB Gold test and serum analysis (HBsAg, Anti-HCV, TPHA, Anti-HIV 1/2 and HIV 1 Ag, Toxocaracanis IgG, Toksoplasma gondii IgM/IgG and PCR for HSV, VZV, CMV). According to negative results, the etiology of SC in this case was idiopathic and autoimmune.

The eye condition was managed by oral corticosteroids, methotrexate, mycophenolate mofetil.In the period from 2019 to 2021 she was treated with oral corticosteroids (16 mg per day). The dosage of oral corticosteroids was reduced to a minimally effective dosage. During treatment there was seen positive dynamics, however, the lesions increased in size and new lesionsoccurred. In this regard, oral corticosteroid dosage was increased (32 mg per day) and immunosuppressive therapy asMycophenolate Mofetil (CellCept in dosage 2 g per day) was added. After a month of treatment, due to patients drug intolerance (nausea and vomiting, elevated ALAT and ALAT) Mycophenolate Mofetil was substituted for subcutaneous MTX (15 mg once a week) and folic acid.

Despite treatment with oral corticosteroids and immunosuppressive therapy, the disease rapidly spreads to the left eye. The new foci continue to appear, and the macular lesion increases in shape and gets active. **(Fig. 6.)**

Fig.6. Fundus photoand FA of the left eye.



The current treatmentis oral corticosteroids (8 mg per day) in combination with MTX (20 mg once a week) and folic acid. The disease is active and remission in not achieved during aggressive treatment.

Discussion

The etiology of SC is still unclear; however, most authors suggest autoimmune and infectious causes. [3]

Individualspredisposed to the HLA-B7, HLA-A2, HLA-B8, and HLA-Dw3 are more likely to have an autoimmune responsein case of SC. [4]Broekhuyse and colleagues found auto-reactivity of circulated lymphocytes to the retinal S antigen and opsin in case of SC and acute posterior multifocal placoid pigment epitheliopathy (APMPPE). [5] Outer retina is richly endowed with S antigen and causes autoimmune to this layer, which can lead to secondary involvement of RPE and choriocapillaris. [4]Other infectious causesincludeexposure to Mycobacterium tuberculosis, Treponema Pallidum and Herpes virus. [1., 4., 8]

Active lesions are slightly yellowish with poorly defined margins. [1] Characteristic features of the OCT are hyper-reflectivity and thickening of the outer retina, as well as increased reflectance of the

choroid. Healed lesions have sharp demarcation line, while atrophic lesions are hypofluorescent. A common feature is disruption of the photoreceptor inner and outer segment junction in both active and inactive lesions. [3]

Patients often complain about blurred vision and central or paracentral scotoma. Typically, new lesions occur within the previous chorioretinal scars, patients rarely have solitary separated active lesions. Areas of chorioretinal atrophy could more often be seen in the contralateral, symptom free eye, within the optic nerve disc. [2]

Many disorders may resemble SC,therefore it is important to exclude other diagnoses. **(Fig.7.)**Firstly, the diagnostic should be performed to the infectious causes of SC, because immunosuppressive drugs could cause exacerbations of disease. [2]Laboratory testing should be performed prior to start immunosuppressive treatment. [4]

Fig.7. Differential diagnosis of SC.

Infectious Diseases
- Syphilitic choroiditis
- Tuberculous choroiditis
- Herpes virus choroiditis
- Ocular histoplasmosis
- Ocular toxoplasmosis

Noninfectious Diseases
- Acute posterior multifocal placoid pigment epitheliopathy
- Ampiginous choroiditis
- Relentless placoid chorioretinitis
- Multifocal choroiditis
- Tumors (benign and malignant, ocular and metastatic)
- Posterior scleritis

Serpiginous-like choroiditis (SLC)is observed in patients with MT infection and could closely mimic SC.However, lesions are more pigmented in contrast to SLC. [6]The diagnosis of active tuberculosis should be ruled out with chest computed tomography and QuantiFERON test. [2., 6]

Other infection diseases that have been reported with serpiginous like appearance include Herpes infections and syphilis. [2]Syphilis could present as multifocal chorioretinitis. [7] In some cases, SC was found to be PCR positive for VZV. [4]

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is characterized with yellowish lesions mostly involving RPE. The disease has an acute onset, lesions usually disappear within 2 weeks and are undetectable on angiography examination. [1., 2., 3]

The most common complication in SC is choroidal neovascularization (CNV), which occurs within both active and healed lesions. [1., 3., 8]Subretinal fibrosis and preretinal neovascularization could also be presented in case of SC. [9]

In cases of a positive test result for syphilis and tuberculosis,etiological treatment should be performed immediately. In cases when infectious diseases are excluded as primary diagnosis, oral steroid are the first line treatment to stop the inflammation. [1., 3., 8]In macular involvement pulse intravenous methylprednisolone (1 g/day for 3 days) could be performed. [3]

Due to the recurrence of the diseaseand appearance of new foci,immunosuppressive treatment in combination with oral corticosteroids are effective to control disease activity and achieve long-term remission [3., 8] In more aggressive course of disease pulse cyclophosphamide therapy is effective to achieve long-term drug-free remissions.[9]

Biologic drugs could also be used in treatment of SC. Sobaci and colleagues used TNF alfa inhibitors to treat SC. [10] However, the role of biologic drugs remains unclear.Spanish case report presented with lethal outcomes; A 48-year-old patient with SC was treated with infliximab for 8 weeks. [11]

In this case, we were faced with an active disease progression during oral corticosteroid and immunosuppressive treatment. The use of biologic drugs is also questionable due to the lack of studies and long-term treatment outcomes.

Conclusion

SC is a chronic,bilateral, and recurrent disease ofunknown etiology. The disease affects the RPE, choriocapillaris and the outer retina. The mainclinical symptoms include blurred vision, recurrent floaters, and visual field defects. Syphilis, tuberculosis, and Herpes infection may mimic Serpiginous choroiditis and should be excluded prior to immunosuppressive treatment.

However, the role of long-term use of biologic drugs is still unclear, and due to fatal case, reports are questionable in this case.

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