

Commentary: Importance of ocular imaging in macular serpiginous choroiditis

We congratulate the authors for a well-written and informative paper on the clinical and imaging aspects of macular serpiginous choroiditis (MSC).^[1] Classically, serpiginous choroiditis (SC) is described as a white-dot syndrome of unknown etiology affecting the choriocapillaris and overlying retinal pigment epithelium.^[2,3] It is a rare, usually bilateral, and chronically recurring inflammatory disease beginning in the peripapillary area and spreading centrifugally to involve macula in a snake-like manner over months or years.^[4] However, in a few patients, the lesion arises at the macula. This finding is described as MSC.^[5]

Classically, SC is a noninfectious immune-mediated disease affecting choriocapillaris and sparing deep choroidal stroma.^[6] Infectious diseases such as tuberculosis (TB) also present with classic or macular serpiginous-like choroiditis (SLC).^[7] Failure to consider it as a differential diagnosis of intraocular inflammation can have catastrophic consequences as the immunosuppressive agents used to manage intraocular inflammation may be fatal for both vision and life in patients with active disease. Morphological variations of SLC lesions such as placoid and dendritic lesions have been described in the literature, of which placoid lesions are associated with choroidal thinning and poor visual outcome.^[8,9]

Clinically, MSC mimics many posterior pole pathologies such as APMPPPE, VKH disease, posterior scleritis, CSCR, acute idiopathic maculopathy, choroidal neovascular

membrane (CNVM), and retinal pigment epithelitis. Clinical appearance of macular lesions and their spread in a snake-like fashion helps in differentiating MSC from other macular pathologies with newer imaging techniques. Clinically, patients diagnosed as MSC are similar to classic SC as per age, gender, and laterality. In MSC, patients present early with vision loss and scotoma due to macular involvement.

Imaging in MSC is important to establish diagnosis and check disease activity.^[10] Active disease stage on fluorescein angiography (FA) shows early phase hypofluorescence of the lesion due to blocked choroidal fluorescence secondary to overlying edematous and inflamed RPE/choriocapillaris. In the mid-phase, lesions show hyperfluorescent borders due to leakage from choriocapillaris at the margins. Late-phase angiogram shows stippled hyperfluorescence with the entire lesion later becoming hyperfluorescent with irregular borders due to diffuse leakage from larger choroidal vessels. Healed lesions appear as marked hypofluorescent areas with sharply delineated margins in early phases due to extensive destruction of choriocapillaris. Later, phase films show hyperfluorescence due to diffuse staining of lesions as the dye diffuses into scarred areas from surrounding healthy choriocapillaris.

In active cases, indocyanine green angiography (ICGA) shows dye blockage in both early and late phases in the lesion area due to nonperfusion of the choriocapillaris and choroidal vessels as well as blocked fluorescence due to inflamed RPE and retina. The extent of the involved area delineated by ICGA seems to be larger than that on FA in active cases.

Fundus autofluorescence (FAF) has recently emerged as the investigation of choice due to its noninvasiveness and ability

to differentiate active and healed lesions in SC.^[11] FAF shows hypo FAF lesions due to melanin pigment accumulation, with hyper FAF margins showing disease activity. Healed lesions show complete hypo FAF with sharp irregular margins. All FAF lesions appear larger than those on FFA.

Optical coherence tomography (OCT) imaging of active lesions in MSC shows inflammation of RPE and outer retina, seen as hyperreflective, irregular bands suggestive of photoreceptoritis in outer retina with loss of ellipsoid zone and IS-Os junction. Healed lesions show retinal thinning and decreased subfoveal choroidal thickness in areas of chorioretinal scarring. OCT is useful for identifying and monitoring CNVM secondary to SC.

OCT angiography (OCTA) has emerged as a novel imaging technique to visualize retinal and choroidal microvasculature at optic disc and macula.^[12] It provides *in vivo* 3D vascular information by analyzing the movement of flowing RBCs, thereby enabling visualization and quantification of functional vessel networks within microcirculatory tissue beds in a noninvasive manner without the use of dye.^[13] In MSC, OCTA shows decreased vascularity in the choriocapillaris slab and choroidal region with relatively retained inner retinal vascularity. Following treatment, flow voids seen in the choriocapillaris layer reduce in area and are replaced with an irregular and nonuniform capillary-like network.

Visual field evaluation shows dense absolute and/or relative, mostly central scotoma in active MSC corresponding to size, shape, and location of the lesions. With treatment, the scotoma reduces in size and density.

Corticosteroids remain the mainstay of treatment. In severe vision-threatening lesions, pulse-dose intravenous methylprednisolone along with systemic corticosteroids are given. Immunosuppressive agents such as azathioprine, mycophenolate mofetil, and cyclosporine are also used for treating MSC. In infectious tuberculous macular SLC, systemic anti-TB drugs (four drugs for 2 months followed by two drugs for 7 months – standard regimen) are needed. Worsening following initiation of therapy may indicate paradoxical worsening or presence of multidrug-resistant TB.^[14,15] The treatment response to systemic steroids and immunosuppressive agents in MSC is similar to classic SC. However, due to the macular location of lesions in MSC, visual prognosis is poor without aggressive treatment. Recurrences are common after a period of latency and new lesions develop at the edges of healed ones. Recurrent episodes are managed with systemic corticosteroids and immunosuppressants as per required dosage with longer treatment duration and closer monitoring.

To conclude, multimodal imaging helps in diagnosing MSC, identifying the extent and activity of the disease, and diagnosing early recurrences with regular follow-up so that the disease can be controlled with appropriate and adequate therapy with minimal damage.

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