Commentary: Posterior segment manifestations of rickettsial disease

Post-febrile retinitis can be associated with a number of infective diseases like dengue, chikungunya, rickettsia, typhoid, malaria, West Nile virus, bartonellosis, cat scratch disease, Rift Valley fever, and herpes viridae family.[1] Diagnosis of rickettsiosis is particularly difficult due to lack of uniform diagnostic criteria and absence of easily available low-cost confirmatory laboratory tests. Fever, headache, malaise and skin rash in an endemic area should be investigated with high index of suspicion to diagnose rickettsiosis.[2,3]

In Indian subcontinent, three groups of rickettsiosis are commonly encountered a) Typhus group (scrub and murine typhus), b) Spotted fever group (tick typhus) and c) Q fever group.[4] Rickettsia has the propensity to damage endothelial cells of small blood vessels, causing microvascular leakage, edema and tissue necrosis.[5] Retinal involvement can either be due to invasion by microorganism or due to immune complex formation. Retinal and vascular involvement is common due to presence of glutamic acid in the retinal tissue and angiotrophic nature of the organism.[6]

The ophthalmic manifestations of rickettsiosis are varied with a predilection for the posterior segment. Ocular involvement may be subclinical and asymptomatic in the early stage or may develop even after a month following fever. White retinal lesions or focal retinitis are often the most frequent presentation. They can vary from 0.5 disc diameter (DD) to 3 DD in size, can be unifocal or multifocal and usually located along the vessels either at the posterior pole or at the periphery. Other posterior segment features include vitritis, vasculitis, retinal hemorrhages, serous retinal detachment, cystoid macular edema, vascular occlusion and neuroretinitis.[2,7] Miscellaneous ocular features described in the literature include ocular flutter, conjunctival congestion, conjunctivitis, keratitis and anterior granulomatous uveitis.[3]

In the current study on ocular manifestations of rickettsiosis predominantly involving paediatric cohort from South India, the authors have highlighted few of the uncommon manifestations in the form of cranial nerve palsies, optic neuritis and papilledema. Other common posterior segment findings noted in this study were retinitis (25.9%), vasculitis (22.2%) and macular edema (14.8%). Another interesting observation is that the ocular features were evident in almost all patients with central nervous system (CNS) involvement. Bilaterality, pediatric age and CNS involvement had been associated with poor visual outcome in this study.[8]

In a study involving 30 patients of Mediterranean spotted fever, 33.3% patients had angiographic abnormalities despite clinically normal fundus. While the smaller white retinal lesions were isofluorescent or hypofluorescent without

staining at late phase of angiogram, the larger lesions exhibited staining. Other angiographic features included vascular and disc leakage. Multiple hypofluorescent dots at choroidal level were also noted in some clinically inapparent cases.[7,9]

Leakage of dye from the site of active inflammation in late phases of conventional angiography may obscure the underlying details. Optical coherence tomography angiography (OCTA) may be helpful in such scenario. Isolated reports on OCTA manifestations of rickettsial retinitis include hypointense dark areas in the superficial and deep capillary plexus (more in latter), disruption of the foveal avascular zone with pruning of vessels at macula, and projection artifacts with signal void areas in the choriocapillary slab. Following treatment, gradual vascular remodeling with reduction in non-perfusion area was noted in one case whereas the other case showed persistent hypointense area.[9,10]

OCT features in active stage of the disease are inner retinal layer hyperreflectivity with retinal thickening and back shadowing. Associated features may include subretinal fluid, intra-retinal cystic spaces, hyperreflective dots, vitreous cells, thickened posterior hyaloid and disc edema. Predominant inner layer involvement is more in favor of rickettsiosis as compared to other causes of retinitis (toxoplasmosis, cytomegalovirus and varicella zoster virus) which cause full-thickness involvement. Inner retinal layer atrophy with disruption of ellipsoid zone may be noted following resolution.[9]

Although early treatment is beneficial, delay due to asymptomatic presentation and presence of innocuous lesion is frequently encountered. Oral antibiotic therapy with doxycycline (100 mg twice a day) for 7–10 days is the drug of choice for rickettsiosis although prolonged therapy may be required in some patients.[3,11] Alternatives include chloramphenicol, other tetracyclines, and azithromycin. Most of the cases are treated with concurrent systemic steroids. Systemic steroids may be indicated in retinitis lesions involving the macula with exudative detachment or extensive retinitis, optic nerve involvement, vascular occlusion, severe vitritis and macular edema. Isolated retinitis has been reported to resolve without steroids also.[11] Most of the lesions resolve completely within 3-10 weeks without any sequelae. But there are reported cases that develop choroidal neovascular membrane, retinal pigment epithelium atrophy, and disc pallor upon resolution with resultant visual decline.[1,2]

Physicians must be aware of the extended spectrum of ocular manifestations in rickettsiosis and should exercise high index of suspicion and seek detailed ophthalmic care promptly to avoid long-term visual morbidity.

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