Ocular manifestations of the antineutrophil cytoplasmic antibody and antiphospholipid syndromes

Sir,

I read with interest the article by Kumar Saurabh and colleagues on the patient profile of retinal vasculitis from Eastern India,[1] and would like to comment on the importance of a more comprehensive laboratory work-up to ensure that the vasculitides syndromes associated with significant morbidity and mortality are not missed. The antineutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAV) and antiphospholipid antibody syndrome (APS) both can initially present with only ocular manifestations such as uveitis and work-up to exclude these vasculitides is more important in younger patients without known systemic risk factors.[2,3]

The AAVs include Wegener’s granulomatosis (cytoplasmic staining pattern seen on immunofluorescence of ethanol-fixed human neutrophils, cANCA, with specificity for proteinase 3 [PR3]) while microscopic polyangiitis and Churg–Strauss syndrome (usually have a perinuclear pattern on immunofluorescence, pANCA, with specificity for myeloperoxidase [MPO]) can present with various ophthalmic manifestations. But in a subset of patients, these findings may be the earliest indicators of systemic disease (up to 50% in children).[2,4] Orbital and anterior segment findings are most common, whereas posterior segment complications such as retinal vasculitis and optic neuropathy occur much less frequently. However, a study on patients with pANCA-associated vasculitis reported that ocular surface (scleritis and peripheral keratitis) and posterior segment manifestations (central or branch retinal vein occlusion, optic neuropathy, acute posterior multifocal placoid pigment epitheliopathy) were the important eye presentations.[5] However, a significant proportion of patients will have systemic symptoms, but may not initially present, and the finding of abnormal chest X-ray or serum creatinine or glomerulonephritis should warrant an urgent laboratory request to exclude an ANCA-associated vasculitis.

The autoimmune disease APS is characterized by the consistent presence of antiphospholipid antibodies (or anticyclic lupus anticoagulant) at 12 weeks, arterial or venous thromboses and repetitive foetal loss. Diluted Russel viper venom time (dRVTT) is often used as one of the phospholipid-dependent tests that shows a prolonged APTT which gets corrected only with increased phospholipid concentrations (suggesting the presence of lupus anticoagulant). This can be present without ds-DNA antibodies with a nonspecific positive antinuclear antibody (ANA) only, and hence the addition of lupus anticoagulant in the diagnostic criteria of systemic lupus erythematosus. Ocular involvement in APS occurs in 8-88% of patients, of which 75% with ocular presentation were women with mean age at 40 years.[6] Ocular findings in APS include anterior uveitis, unilateral or bilateral blurring of vision, transient scotoma, unilateral amaurosis fugax (bilateral suggests CNS ischemia), visual field defects and various retinal pathologies (hemorrhages, vasculitis, retinopathy) discussed by Utz VM and Tang J in their review.[7]

Clinicians should therefore have a high index of suspicion of AAV and APS, in especially younger patients, which would be the first step in avoiding the inevitable complications of arterial inflammation and visual loss. Emphasis to lower additional risks of thrombosis such as treating cardiovascular risk factors (hypertension, diabetes, hyperlipidemia, tobacco use or obesity), during immobilization or malignancy, treating microalbuminuria, screening for inherited thrombophilias, and counseling regarding oral contraceptive use remain equally important.

Sujoy Khan
Department of Allergy and Immunology,
Apollo Gleneagles Hospital, Kolkata, India
Correspondence to: Dr. Sujoy Khan,
Department of Allergy and Immunology, Apollo Gleneagles Hospital, 58 Canal Circular Road, Kolkata - 54, India.
E-mail: sujoykhan@gmail.com

References