Ophthalmic Images

Presumed Autoimmune Pigmented Paravenous Retinochoroidal Atrophy in a Female Patient Aged 69 Years

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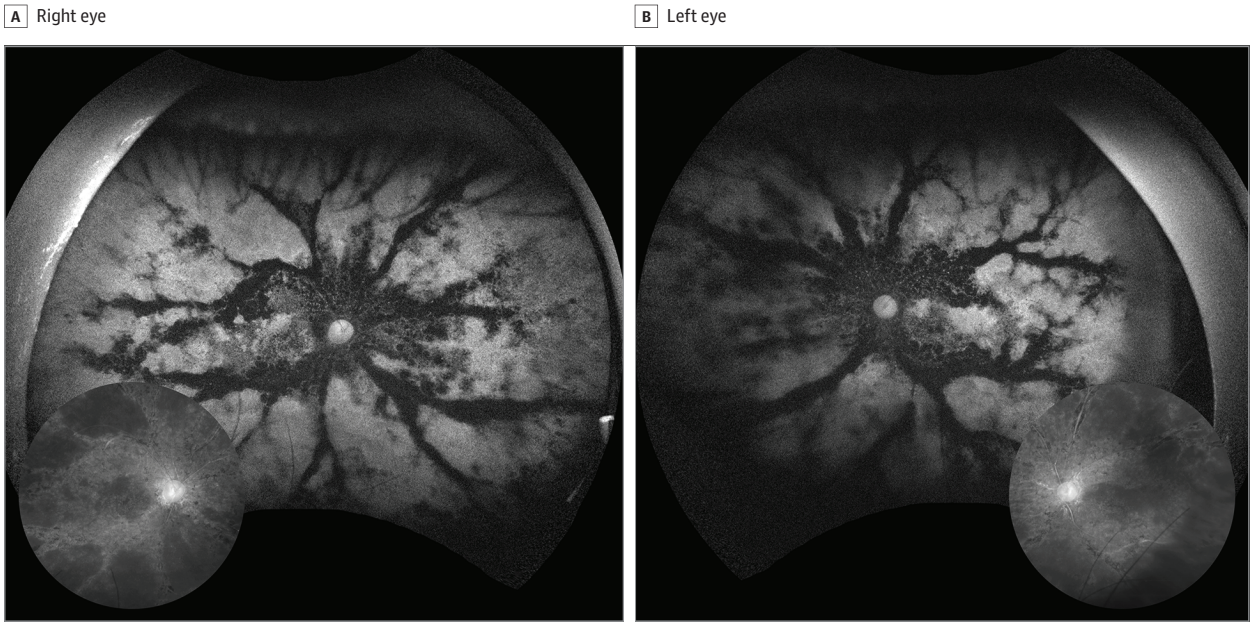

Figure. Presumed autoimmune pigmented paravenous retinochoroidal atrophy. Ultra-widefield fundus autofluorescence of the right (A) and left (B) eyes demonstrating bilateral and symmetrical hypoautofluorescence along the paravenous regions, with areas of isoautofluorescence between them. Patchy clumps of decreased fundus autofluorescence signal were identified in the macula, indicating macular atrophy. Macular-focused images are displayed in insets.

A 69-year-old White female with rheumatoid arthritis and scleroderma presented with long-standing and slowly progressive vision loss and nyctalopia. Family history was negative. At examination, visual acuity was hand motions bilaterally. Results of anterior-segment biomicroscopy were unremarkable. Ophthalmoscopy revealed bilateral optic disc pallor, vessel thinning, and retinochoroidal atrophy along the retinal vessels. Pigment clumping was evident in the atrophic areas. Macular atrophy was present bilaterally.

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On ultra-widefield fundus autofluorescence (FAF), patchy clumps of decreased FAF signal were identified in the atrophic areas (**Figure**). No clinically relevant variants were found on panel genetic testing. Pigmented paravenous retinochoroidal atrophy (PPRCA) is a rare disease of unknown etiology, although dysgenetic, degenerative, hereditary, and inflammatory causes have been suggested. It is usually slowly progressive, and visual acuity remains good unless the macula is involved.[1,2] Given the personal history of autoimmune disease and a negative genetic test result, a diagnosis of presumed autoimmune PPRCA was suggested.

ARTICLE INFORMATION

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