Sector retinitis pigmentosa

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Background: Retinitis pigmentosa (RP) is one of the most common hereditary retinal dystrophies and causes of visual impairment affecting all age groups. The reported incidence varies, but is considered to be between 1 in 3,000 to 1 in 7,000. Sector retinitis pigmentosa is an atypical form of RP that is characterized by regionalized areas of bone spicule pigmentation, usually in the inferior quadrants of the retina.

Case Report: A 57-year-old Hispanic man with a history of previously diagnosed retinitis pigmentosa came to the clinic with a longstanding symptom of decreased vision at night. Bone spicule pigmentation was found in the nasal and inferior quadrants in each eye. He demonstrated superior and temporal visual-field loss corresponding to the areas of the affected retina. Clinical measurements of visual-field loss, best-corrected visual acuity, and ophthalmoscopic appearance have remained stable during the five years the patient has been followed.

Discussion: Sector retinitis pigmentosa is an atypical form of RP that is characterized by bilateral pigmentary retinopathy, usually isolated to the inferior quadrants. The remainder of the retina appears clinically normal, although studies have found functional abnormalities in these areas as well. Sector RP is generally considered a stationary to slowly progressive disease, with subnormal electro-retinogram findings and visual-field defects corresponding to the involved retinal sectors.

Conclusion: Management of RP is very difficult because there are no proven methods of treatment. Studies have shown 15,000 IU of vitamin A palmitate per day may slow the progression, though this result is controversial. Low vision rehabilitation, long wavelength pass filters, and pedigree counseling remain the mainstay of management.

Key Words: Atypical retinitis pigmentosa, electro-retinogram, retinitis pigmentosa, sector retinitis pigmentosa, visual-field defects

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Retinitis pigmentosa (RP) is a term used for a collection of progressive hereditary disorders that diffusely affect photoreceptor and pigment epithelial function.[1] It is one of the most common hereditary retinal dystrophies and causes of visual impairments in all age groups.[2] The incidence of RP varies, depending on the study cited, but is considered to be 1 in 3,000 to 1 in 7,000.[1,3,4] An estimated six newborns out of 1,000,000 in the United States are diagnosed with RP per year.[4] It generally does not respect any racial boundaries or age group, and is slightly more prevalent among males.[5] Sector retinitis pigmentosa is an atypical variant of this condition, in which only isolated areas of the fundus show pigmentary changes. It is characterized by regionalized areas of bone spicule pigmentation usually found in the inferior quadrants of the retina, subnormal electro-retinograms, visual-field defects, and slow to no progression. Dark adaptation, fluorescein angiography, and electrofunctional testing have revealed impaired function, not only in the affected retina, but in the normal-appearing retina, indicating that even those areas that may appear normal on ophthalmoscopic examination are also affected.[6-9]

Case Report

A 57-year-old Hispanic man came to the clinic with a symptom of a broken right lens in his glasses and a need for a new spectacle prescription. He reported being diagnosed with retinitis pigmentosa at age 44. He stated pigmentary changes were found in his eyes and electrodiagnostic testing was performed. The results from the testing were unavailable. He also stated he had difficulty with night vision since he was in the military—around the age of twenty—as well as acquired hearing loss, for which he was

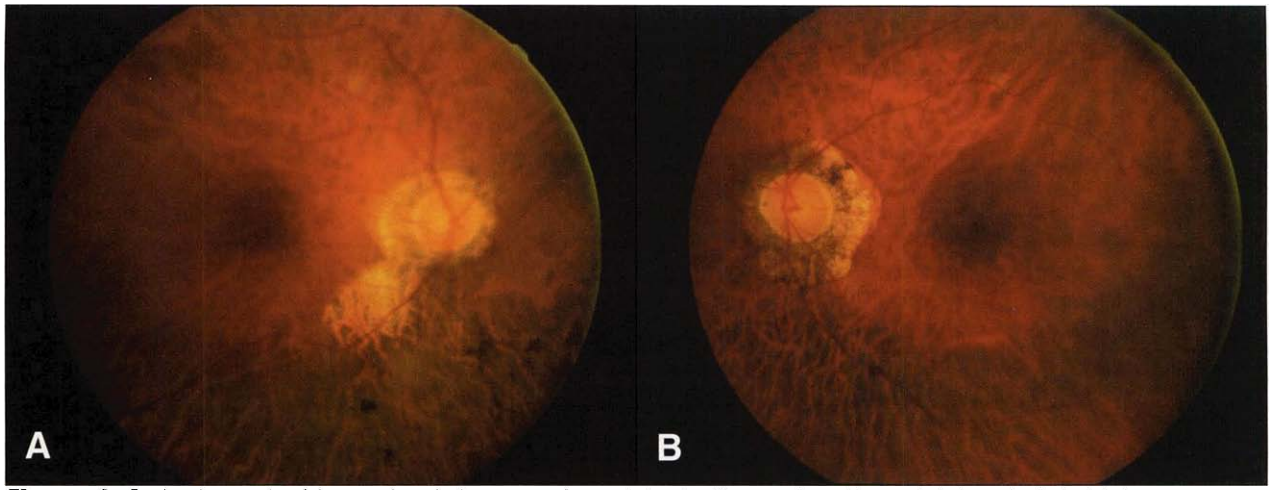

Figure 1 Fundus photographs of the posterior pole demonstrate ***A***, extended peripapillary atrophy and bone spicule formation in the inferior nasal aspect of the retina O.D. and ***B***, peripapillary atrophy with no pigmentary changes found O.S.

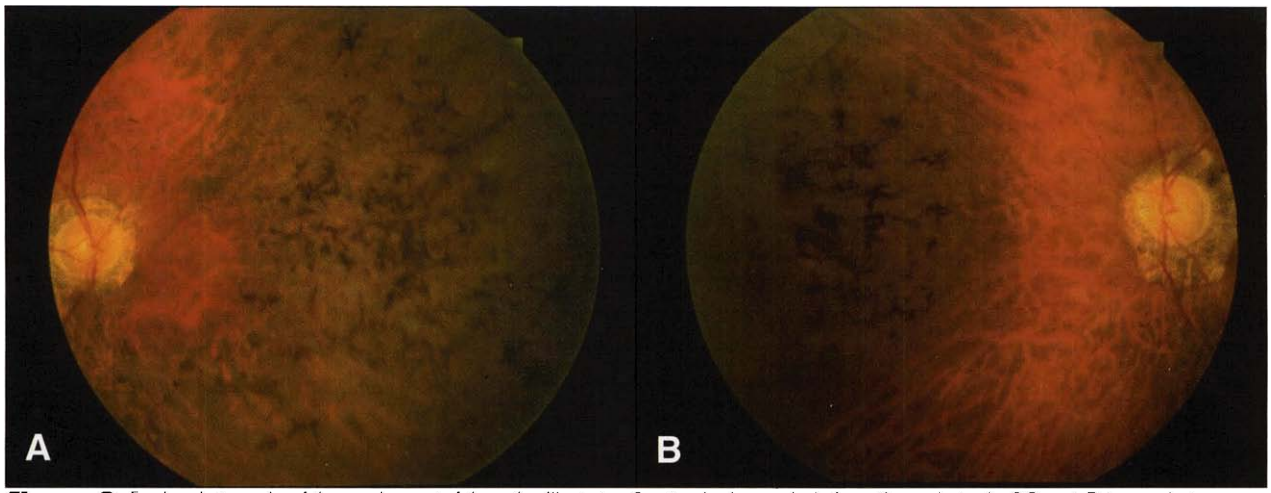

Figure 2 Fundus photographs of the nasal aspect of the retina illustrates ***A***, extensive bone spicule formation and atrophy O.D. and ***B***, bone spicule formation O.S.

fit with binaural hearing aids at the age of 54. Medical history was remarkable for hypertension, gout, and an irregular heartbeat. His current medications included probenicid, verapamil, and aspirin. The patient denied head trauma, syphilitic or rubella infection, or family history of RP. Family ocular history was positive for glaucoma, diagnosed in his mother.

Entrance testing was unremarkable, with best-corrected visual acuities of 20/20 OU. His refractive error was -4.75 -0.75 × 095 O.D. and -3.00 -0.75 × 105 O.S., with a +2.00 add OU. Slit-lamp biomicroscopy was remarkable for corneal arcus and posterior vitreous detachments OU. Goldmann applanation tonometry yielded intraocular pressures of 19 mmHg OU. Dilated fundus examination revealed bone spicule pigmentary changes in the inferior nasal and nasal quadrants OU and extensive peripapillary atrophy O.D. > O.S. (*see* Figures 1, *A* and *B*, and 2, *A* and *B*). The cup-to-disk ratio was 0.45 round OU, with mild pallor. Goldmann visual-field testing demonstrated superior temporal and temporal field defects, as well as paracentral to midequatorial scotomas O.D. Testing O.S. revealed a temporal visual-field defect much more pronounced with a small target (I-4e) and an enlarged blindspot (*see* Figure 3, *A* and *B*). The patient was diagnosed with early sector RP, and was counseled regarding the nature of his disease. New spectacles were ordered and a follow-up visit was scheduled for one year.

One year later the patient returned with a symptom of decreased vision O.D. for the previous 3 to 4 months. Medical history was unchanged, except for a breast lipoma that had been found and was scheduled for removal. Best-corrected