AN ATYPICAL CASE OF RETINITIS PIGMENTOSA, MANAUS-AM

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PURPOSE

Case report of na atypical case of Retinitis Pigmentosa (RP)

METHODS

Female, 59, white, diabetic, with a previously treated breast cancer, referring a gradual decrease in visual acuity (VA) and visual field (VF) over two years.

RESULTS

VA two years ago was 20/60 in the right eye and 20/80 on the left eye. Fundoscopy on both eyes showed a healthy optic disc with no signs of glaucoma, general vessel and retinal pigmented epithelium (RPE) thinning and an absence of the characteristic foveal luster. Spicule-like pigmented lesions were isolated in the extreme peripheral region. VA and VF tests, however, often fluctuated, and while in some light conditions the patient presents with the same vision as before, but with tubular vision, she is completely unable to roam by herself and often appears to show a hand motions-like VA.

Due to the atypical onset of complaints and drastic VF worsening progression, Retinitis Pigmentosa (RP) was not considered at first. While she did have spicule-like lesions in the periphery, the posterior pole didn't match a patient presenting tubular vision that often emulated a hand motions VA. Ischemic Optic Neuropathy was considered, but neurological evaluation, magnetic resonance imaging and electroencephalogram were all normal. However, macular Optical Coherence Tomography showed a large-scale absence of the photoreceptor line, notably in the foveal region, tying it back to the retinal dystrophy diagnosis.

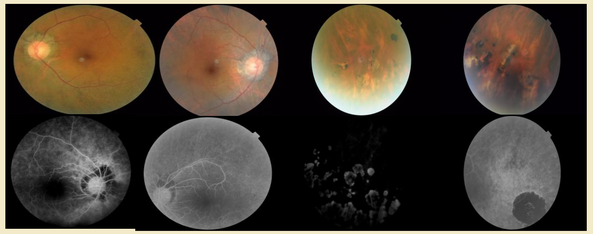

Figure 1 – Fluorescent Angiography (FAG) in both eyes showed changes in peridiscal hypofluorescencia in early stages, hypofluorescent of 3 ODD by blockade in the terminal branch of LTA suggesting scarial process and multiple peripheral lesions with hypofluorescent center and hyperfluorescent edges smaller than 1 ODD in LE, increased FAZ in RE and preserved size in LE, was not visualized areas of contrast leakage

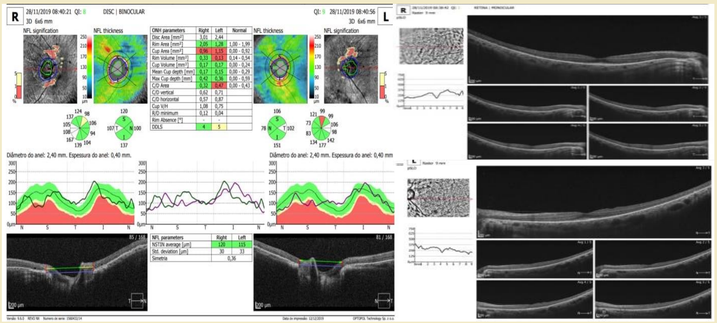

Figure 2- Optical coherence tomography (OCT) evidencing high lesion on optical disc, with high reflectivity and shading layer, absence of photoreceptor line.

CONCLUSIONS

RP is a heterogeneous group of hereditary diseases, with progressive and generalized loss of photoreceptors and RPE, along with visible pigment deposits called bone spicules, which gradually cause nocturnal blindness and a progressive decrease in VF and VA. Diagnosis criteria include bilaterality, loss of peripheral vision and progressive deterioration of the photoreceptors function. The classic triad is arteriolar thinning, bone spicules and disc pallor. This report aims to present an atypical presentation of RP, with a late-onset of VF worsening and general disrelation between fundus examination and disease progression.

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