Extensive Macular Atrophy with Pseudodrusen (EMAP) in a middle-aged patient - AN INTERESTING CASE

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PURPOSE:

To describe a case of Extensive Macular Atrophy with Pseudodrusen (EMAP) appearance in female middle-aged patient.

METHODS:

Case report with multimodal examination.

RESULTS:

A 56-year-old female patient, Caucasian, married, from São Paulo, retired for 4 years, who came to our service with a history of progressive low visual acuity for 5 years.
Five years ago (early 2018) she started having low visual acuity at work due to computer use associated with nyctalopia.

PERSONAL HISTORY:

- Systemic arterial hypertension in use of Metoprolol Succinate 50mg/day.
- Rheumatic Fever 7-year-old treated with Benzathine Penicillin 1,200,000 IU every 15 days for 1 year then monthly for 7 years.

OPHTHALMOLOGICAL HISTORY:

- Myopia (-3.00)
- Radial keratotomy 22 years ago
- Bilateral facectomy in 2018

VISUAL ACUITY:

- OD: +1.25 -2.00 100 (v = 0.8q)
- OS-1.00 -1.00 90 (v= 0.6q)

BIOMICROSCOPY:

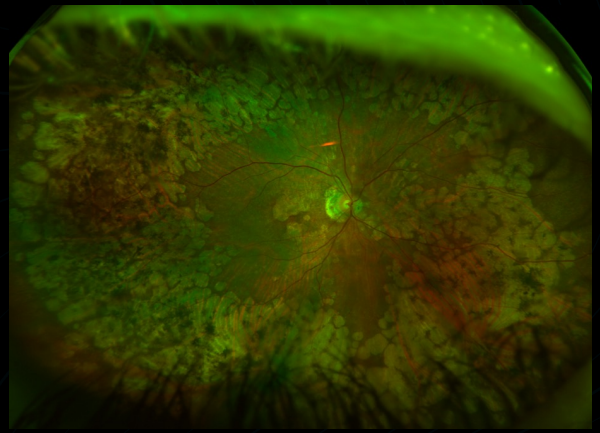
- Clear conjunctiva, transparent cornea with RKs scars, Pseudophakic, IOL centered, anterior chamber formed, no anterior chamber reaction in both eyes.

FUNDOSCOPY:

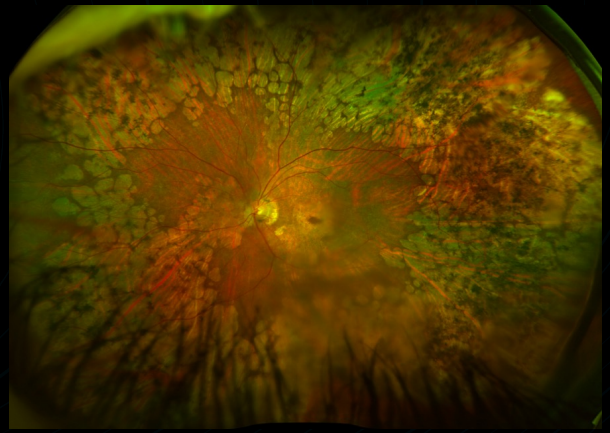
- Image 1 and 2.

AUTOFLUORESCENCY (AF):

- Image 3 and 4.



(Image 1)



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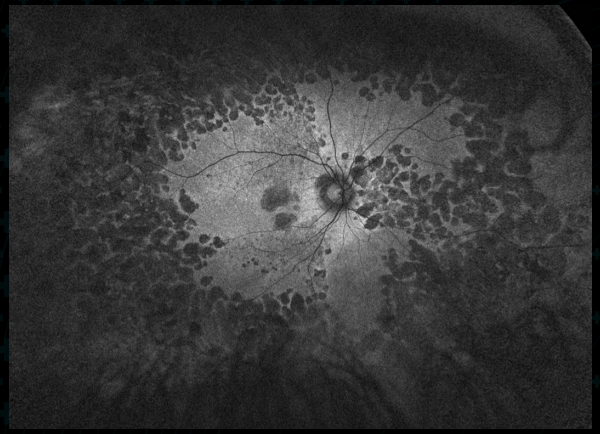
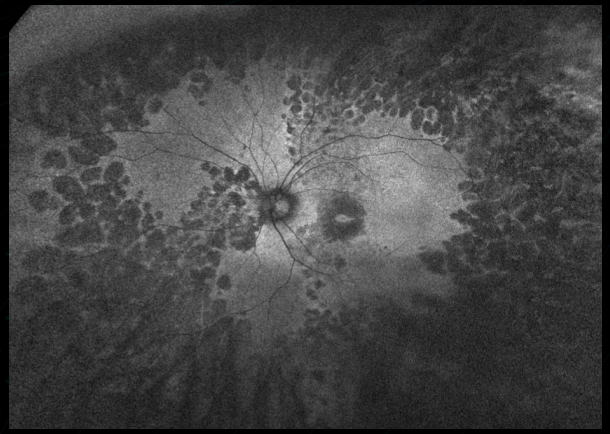


(Image 3)



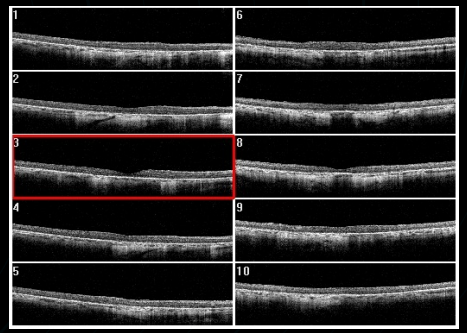
(Image 4)

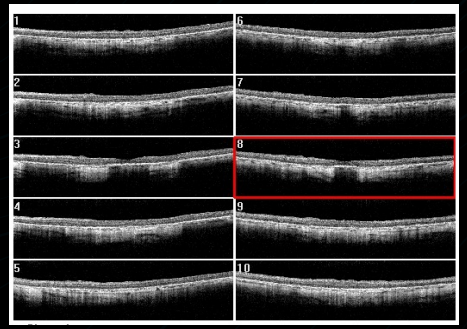
RETINOGRAPHY:

- In the wide-angle retinography of both eyes, it's possible to observe a normal optic disc, C/D 0.3, beta peridisc atrophy, macular region with areas of atrophy and pigment mobilization, vessels with normal calibers and distributions, atrophic lesions in the retina nasal, middle and extreme periphery compatible with "*Paving Stones*" grouped 360• and applied retina.

AUTOFLUORESCENCE (AF):

- Areas of hypoautoflorescence in the macular region, middle and extreme periphery corresponding with complete atrophic lesions of the retinal pigment epithelium in both eyes.





OPTICAL COHERENCE TOMOGRAPHY (OCT):

OD: Areas of atrophy of the outer retina, photoreceptors and perimacular EPR in Bull's Eye pattern.

OS: Presence of ERM, thinning of the retinal pattern, important areas of atrophy in the outer retina, photoreceptor layer and perimacular RPE.

MULTIFOCAL ELECTRORETINOGRAM:

OD: important reduction in central retinal function (0• to 25• of retinal eccentricity).

OS: important reduction in central retinal function (0• to 25• of retinal eccentricity).

VISUAL FIELD 30:2:

OD: VFI 19%, MD -24.51 dB (p <0,5) and PSD (7.97 dB).

OS: VFI 22%, MD -23.20 dB (p <0,5) and PSD (9.63 dB).

FULL FIELD ELECTRORETINOGRAM:

OD: Undetectable rods system response and presence of 7% cones system response.

OS: Undetectable rods system response and presence of 5% cones system response.

COMPLETE GENETIC PANEL FOR VITREORETINAL DYSTROPHIES AND ORNITHINE DOSAGE:

- Negative genetic test.
- Ornithine within the normal range.

DISCUSSION AND CONCLUSION:

EMAP (Extensive Macular Atrophy with Pseudoatrophy) is a new entity, for the first time described under that name in 2009 and has many obstacles for diagnostic elucidation and correct management. This disease usually appears in middle-aged patients (mean age 50 years), associated with night blindness, rapid progression of atrophy with a vertical pattern, association with grouped paving stones, bilateral, absence of AMD risk factors (advanced age, smoking, sun exposure and family history), in addition to a negative genetic pattern.

In this case, specifically, the patient had a hypothesis of AMD rising, being treated with Lutein and Zeaxantine replacement, in addition to being submitted to intravitreal injections of Anti-VEGF (Aflibercept) in both eyes and bilateral cataract surgery. All methods to improve visual acuity.
What did not happen.

Recently, new studies and case series have demonstrated a probable association of EMAP with patients who used Benzathine Penicillin for long periods of time for treatment or prophylaxis of Rheumatic Fever. Their real association has not yet been established.

EMAP is still a diagnosis of exclusion, and it is mandatory to exclude other diagnoses such as early AMD, Retinitis pigmentosa in its different spectra, Late Onset Retinal Dystrophy (LORD), Girate Atrophy.

Correctly managing these patients means first starting with an early diagnostic elucidation and guidance regarding lifestyle adequacy for patients with low vision until, with the advent of science, an effective treatment is discovered that aims to reduce or delay their atrophy pattern.

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