

# CASE REPORT OF AN EXTENSIVE MACULAR ATROPHY WITH PSEUDODRUSEN

Arthur Baldim Terra, MD; Isabela Soares Bôa Morte, MD; Jéssica Pimentel Lino, MD;  
Luísa Machado dos Santos Rocha, MD; Lina Porto Hermeto, MD; Liz de Oliveira Moura  
Sales, MD

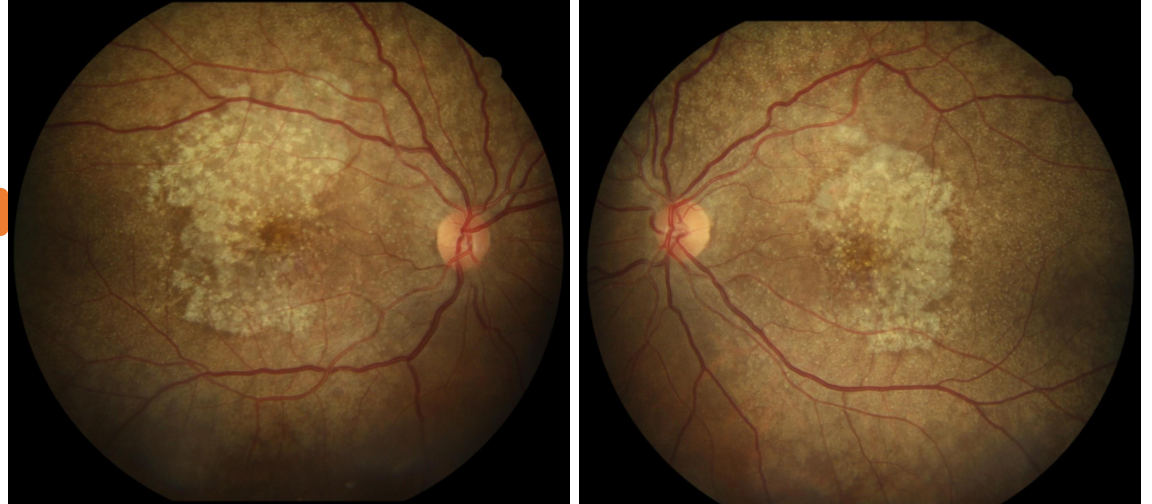
## PURPOSE

Report a case of an extensive macular atrophy with pseudodrusen (EMAP) and discuss its challenging follow up.

## METHODS

Case report. Clinical histories, examination and multimodal imaging findings were analyzed.

## CASE REPORT



A 54 year-old checks herself in the Ophthalmology Department at her local hospital reporting photophobia, a burning sensation, nyctalopia, and low visual acuity on both eyes, with a 2-year evolution. She did not present any changes upon biomicroscopy examination. The corrected visual acuity was 20/30 in the right eye and 20/60 in the left eye. After performing a retina mapping, it was found physiological optic disk and vitreous, vertical macular atrophy area extending to the upper and lower arcades and difuse pseudodrusen throughout the medium periphery in both eyes.

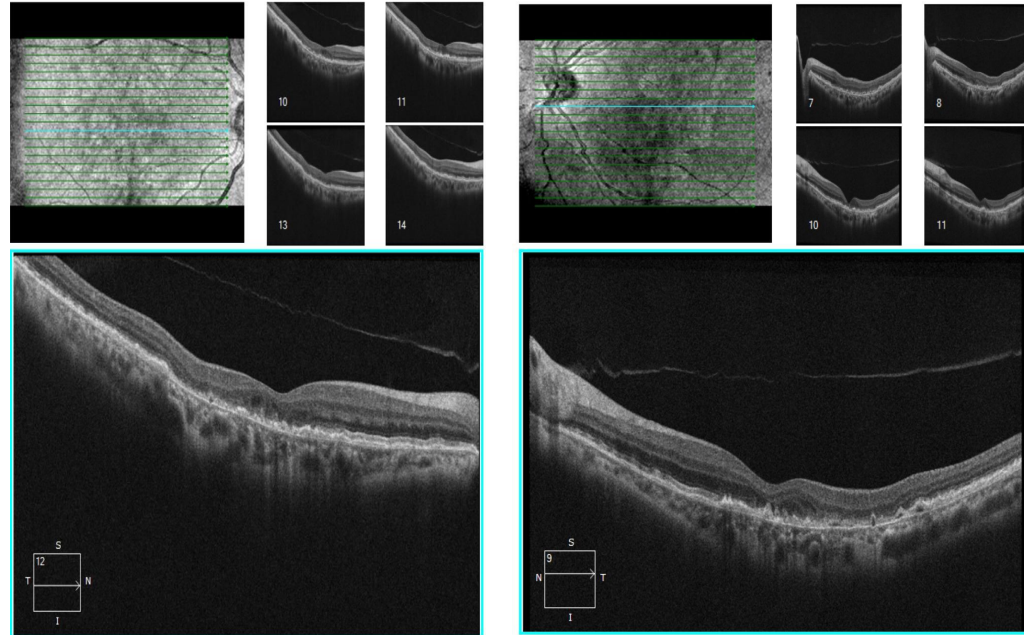
# CASE REPORT OF AN EXTENSIVE MACULAR ATROPHY WITH PSEUDODRUSEN

**Arthur Baldim Terra, MD; Isabela Soares Bôa Morte, MD; Jéssica Pimentel Lino, MD; Luísa Machado dos Santos Rocha, MD; Lina Porto Hermeto, MD; Liz de Oliveira Moura Sales, MD**

## CASE REPORT

Optical coherence tomography (OCT), retinography and autofluoresceinography were performed to investigate the etiology of macular atrophy. OCT showed areas of atrophy in external retina in foveal region symmetrically in both eyes and diffuse pseudodrusen in the medium periphery.

The autofluoresceinography showed bilateral symmetrical areas of hypoautofluorescence affecting the macula and extending to the arcades consistent with atrophy. The changes in the exams performed suggested the possibility of EMAP. Due to the lack of treatments for this condition, it was decided to follow up at the retina department and maintain the patient under observation for any potential clinical progression.

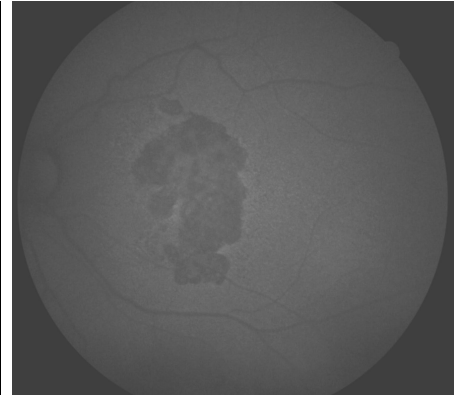
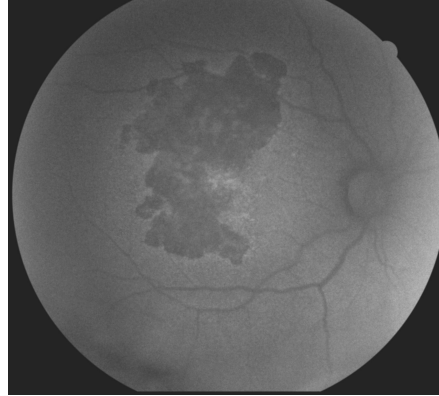


# CASE REPORT OF AN EXTENSIVE MACULAR ATROPHY WITH PSEUDODRUSEN

**Arthur Baldim Terra, MD; Isabela Soares Bôa Morte, MD; Jéssica Pimentel Lino, MD; Luísa Machado dos Santos Rocha, MD; Lina Porto Hermeto, MD; Liz de Oliveira Moura Sales, MD**

## DISCUSSION

Extensive macular atrophy with pseudodrusen (EMAP) is a rare type of macular degeneration first described in 2009. It is a rapidly progressing bilateral macular degeneration with an onset at a relatively early age, typically between ages 40 and 55, with the presentation of night blindness being a frequent sign. EMAP causes macular degeneration with early involvement of the fovea which results in a longitudinal atrophy that crosses the vascular arcades. Pseudodrusen can extend throughout the retina and form a cobblestone degeneration in the peripheral region. There is still uncertainty about the correlation of EMAP and choroidal neovascularization, but it is known that one of its major differential diagnoses is the dry age-related macular degeneration (AMD).

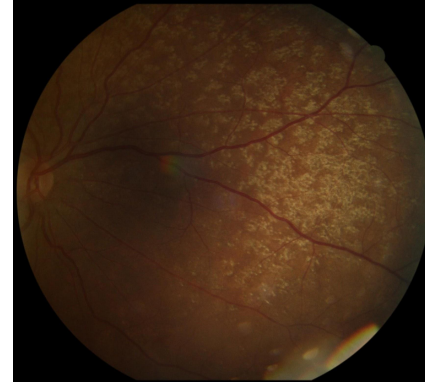
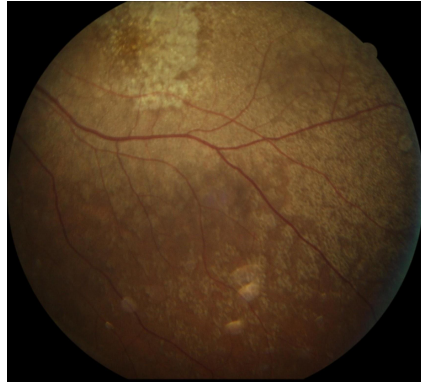
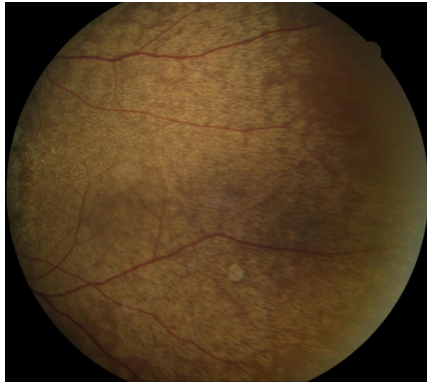


# CASE REPORT OF AN EXTENSIVE MACULAR ATROPHY WITH PSEUDODRUSEN

**Arthur Baldim Terra, MD; Isabela Soares Bôa Morte, MD; Jéssica Pimentel Lino, MD; Luísa Machado dos Santos Rocha, MD; Lina Porto Hermeto, MD; Liz de Oliveira Moura Sales, MD**

## DISCUSSION

This condition is relatively new, so well-defined diagnostic criteria has not been developed yet.. Initially, it was described as having the following clinical characteristics: (1) younger onset (40-55 years) compared to dry age-related macular degeneration (AMD); (2) bilateral symmetrical extensive macular degeneration spreading vertically; (3) absence of foveal preservation; (4) pseudodrusen throughout the retina; (5) peripheral cobblestone degeneration; (6) absence of choroidal neovascularization; (7) lack of family history; (8) night blindness and/or photophobia. In addition, a higher incidence of EMAP in women has been reported.





# CASE REPORT OF AN EXTENSIVE MACULAR ATROPHY WITH PSEUDODRUSEN

**Arthur Baldim Terra, MD; Isabela Soares Bôa Morte, MD; Jéssica Pimentel Lino, MD; Luísa Machado dos Santos Rocha, MD; Lina Porto Hermeto, MD; Liz de Oliveira Moura Sales, MD**

## DISCUSSION

Extensive macular atrophy with pseudodrusen is associated with family history of AMD, glaucoma, female gender, diets high in processed food, abnormal plasma levels of C3, CH50, eosinophils and lymphocytes, and people who have had contact with chemical and toxic products (such as fungicides) for long periods. On the other hand, a protective factor would be the Mediterranean diet, rich in omega-3 and virgin olive oils, which are natural non-steroidal antioxidants.

Many recent clinical trials have evaluated the efficacy of complement inhibitors in reducing the progression of dry AMD. However, ongoing clinical trials do not distinguish between various subtypes of macular degeneration, potentially excluding individuals with conditions that could benefit from such interventions (e.g. EMAP). Therefore, due to the fact that EMAP is a relatively new condition with limited studies, its patients suffer from the lack of treatment options to contain the disease.

## REFERENCES

1. Douillard, Aymeric et al. "Clinical Characteristics and Risk Factors of Extensive Macular Atrophy with Pseudodrusen: The EMAP Case-Control National Clinical Trial." *Ophthalmology* vol. 123,9 (2016): 1865-73.
2. Douillard, Aymeric et al. "Dietary, environmental, and genetic risk factors of Extensive Macular Atrophy with Pseudodrusen, a severe bilateral macular atrophy of middle-aged patients." *Scientific reports* vol. 8,1 6840. 1 May. 2018.
3. Sato, Shigeru et al. "Extensive Macular Atrophy with Pseudodrusen in a Japanese Patient Evaluated by Wide-Field OCTA." *Case reports in ophthalmology* vol. 13,3 847-854. 16 Nov. 2022.
4. Hamel, Christian P et al. "Extensive macular atrophy with pseudodrusen-like appearance: a new clinical entity." *American journal of ophthalmology* vol. 147,4 (2009): 609-20.