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EDITORIAL

EMAP in focus: aspects of extensive macular atrophy with pseudodrusen-like appearance in Brazil

EMAP em foco: aspectos da *extensive macular atrophy with pseudodrusen* no Brasil

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Extensive macular atrophy with pseudodrusen-like appearance (EMAP) was first described by Hamel et al. in 2009 and has been diagnosed with increasing frequency in Brazil. However, it remains an underdiagnosed disease, mainly because of its phenotypic similarity to age-related macular degeneration (AMD), especially in older patients[1].

The classic features of EMAP include multilobulated atrophy of the retinal pigment epithelium (RPE), often distributed along the vertical axis; a predilection for the superior perifoveal region; and relative foveal preservation in the early stages[1-3]. Other findings include pseudodrusen-like subretinal deposits in the posterior pole, areas of peripheral pavingstone degeneration, and symptom onset at a younger age compared to classic AMD, usually between the fourth and sixth decades of life.

More recent studies have significantly broadened the spectrum of the disease in terms of structures, describing separation of the RPE from Bruch's membrane, progressive choroidal thinning, rupture of Bruch's membrane, and choroidal neovascularization[2,4]. In terms of function, electrophysiological tests show diffuse involvement of the outer and inner retina, along with changes in full-field electroretinography and a reduction in the photopic negative response, which suggests the involvement of photoreceptors and bipolar and ganglion cells[5,6].

Patients with EMAP present different clinical characteristics from those with AMD and those with hereditary retinal dystrophies. The typical presentation includes progressive and insidious visual loss, often accompanied by difficulty adjusting to the dark, dyschromatopsia, and paracentral scotomas. Central visual acuity can remain preserved for years; however, once foveal involvement is established, visual progression tends to be rapid and clinically significant.

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In Brazil, EMAP was first documented in 2018 in a report that described the clinical progression of a patient, including that of the involved structures, who was followed up for eight years. This study contributed to the recognition of the entity in the national context[7]. Subsequently, Brazilian studies expanded on this observation by highlighting relevant epidemiological specificities, including the high frequency of previous history of rheumatic fever in childhood[8,9].

Rheumatic fever is a late-onset autoimmune inflammatory disease triggered by oropharyngeal infection with group A Streptococcus pyogenes, which is still responsible for significant cardiovascular morbidity and mortality in developing countries[10,11]. National estimates indicate that millions of individuals are at risk of developing rheumatic fever after streptococcal tonsillitis, which remains an important public health problem in Brazil[10].

A frequent history of rheumatic fever associated with prolonged use of benzathine penicillin G as secondary prophylaxis has been detected in Brazilian cohorts with APEM[8,9]. Although this association does not establish a causal relationship, it raises the hypothesis of an underlying chronic immune-mediated mechanism. To date, there is no evidence of retinal toxicity directly related to prolonged use of benzathine penicillin, and its mention is relevant as an indirect marker of exposure to rheumatic fever.

Some conceptual similarities exist between rheumatic fever and autoimmune retinopathy, including the presence of elevated inflammatory mediators, a chronic subclinical course with episodic exacerbations, the absence of specific diagnostic markers, and difficulty in objectively assessing progression and treatment response[4,11,12]. In this context, we hypothesize that, in genetically susceptible individuals, EMAP may be a late manifestation of a chronic immunological cross-reaction after previous exposure to group A streptococcus[5]. This hypothesis is supported by the previous description of systemic inflammatory changes in patients with APEM, such as lymphocytosis, eosinophilia, elevated erythrocyte sedimentation rate, and increased C3, which indicate an underlying systemic inflammatory or allergic state[2].

Advances in multimodal imaging, including panoramic retinography, fundus autofluorescence, and high-resolution optical coherence tomography, allow a more accurate diagnosis and better structural monitoring. Electrophysiological tests are widely used in retinal diseases, and their role in the diagnosis and follow-up of autoimmune retinopathies is well established. The latter show electrophysiological patterns that vary based on the paraneoplastic association, the presence of melanoma, the non-paraneoplastic form, and the profile of anti-retinal autoantibodies[13-15]. Although the literature on these entities is limited due to their rarity and diagnostic complexity, EMAP shares several clinical characteristics with autoimmune retinopathy, such as bilaterality, painless progressive visual loss, nyctalopia, dyschromatopsia, photophobia, and central and peripheral scotomas, which are often associated with systemic autoimmune diseases.

Currently, there is no consensus on the pathophysiology, risk factors, or disease-modifying treatment of EMAP. In the absence of specific treatments, the main goals of its clinical management should be early diagnosis, rigorous structural and functional monitoring, treatment of associated complications such as choroidal neovascularization, and visual rehabilitation.

The recognition of EMAP as a distinct entity, potentially influenced by systemic immunological factors prevalent in developing countries, prompts the need for prospective studies, genetic and immunological analyses, and international multicenter collaboration. In this context, Brazil is a unique and important setting for the advancement of knowledge about this disease.

REFERENCES

1. Hamel CP, Meunier I, Arndt C, ben Salah S, Lopez S, Bazalgette C, et al. Extensive macular atrophy with pseudodrusen-like appearance: a new clinical entity. Am J Ophthalmol. 2009;147(4):609–620.
2. Douillard A, Picot MC, Delcourt C, Lacroux A, Zanlonghi X, Puech B, et al. Clinical characteristics and risk factors of extensive macular atrophy with pseudodrusen: The EMAP Case-Control National Clinical Trial. Ophthalmology. 2016;123(9):1865–1873.
3. Antropoli A, Bianco L, Condroyer C, Antonio A, Navarro J, Dagostinoz D, et al. Extensive Macular Atrophy with Pseudodrusen-like appearance: Progression kinetics and late-stage findings. Ophthalmology. 2024;131(10):1175-1184.
4. Antropoli A, Bianco L, Romano F, Trinco A, Arrigo A, Benadji A, et al. Extensive macular atrophy with pseudodrusen-like appearance (EMAP) clinical characteristics and diagnostic criteria, and insights from allied inherited retinal diseases and age-related macular degeneration. Prog Retin Eye Res. 2025 Jan:104:101320.
5. Watanabe SES, Quercia AZF, Sacai PY. Electrophysiological findings in extensive macular atrophy with pseudodrusen. Doc Ophthalmol. 2023;147(2):121-130.
6. Watanabe SES, Quercia AZF, Assis GPSM, Borges ML, Sacai PY. Ganglion cell function loss in extensive macular atrophy with pseudodrusen. Int Ophthalmol. 2025;45(1):338.

7. Moreira-Neto CA, Moreira Júnior CA. Extensive macular atrophy with pseudodrusen-like appearance resembling atrophic age-related macular degeneration: a fully documented eight-year clinical history. e-Oftalmo. 2015;1(3):1-7.
8. Moreira-Neto CA, Andujar RAS, Chao JCT, Vasconcelos H, Alves FEE, Rodrigues GD, et al. Rheumatic fever and long-term use of benzathine penicillin as possible risk factors for extensive macular atrophy with pseudodrusen in a Brazilian cohort. Int J Retina Vitreous. 2024;10(1):75
9. Audi LO, Carvalho RAP, Casella AM, Maniero LAH, Zett C, Messias AMV, et. al. Visual outcomes and clinical features of extensive macular atrophy with pseudodrusen. Retina. 2025;45(10): 1842-1853.
10. Leão SC, Lima MRM, Nascimento HM, Octacilio-Silva S, Rodrigues TMA. IL-10 and ET-1 as biomarkers of rheumatic valve disease. Rev Bras Cir Cardiovasc. 2014;29(1):25-30.
11. Oliveira SG, Marossi LM, Spaziani AO, Frota RS, Filho LSG, Monteiro STF, et al. Epidemiologia da doença reumática crônica cardíaca no Brasil nos anos de 2014 a 2018. Braz J Health Rev. 2020;3(1):857-872.
12. Grange L, Dalal M, Nussenblatt RB, Sen HN. Autoimmune retinopathy. Am J Ophthalmol. 2014;157(2):266-272.e1.
13. Azevedo PM, Pereira RR, Guilherme L. Understanding rheumatic fever. Rheumatol Int. 2012;32(5):1113-20.
14. Braithwaite T, Vugler A, Tufail A. Autoimmune retinopathy. Ophthalmologica. 2012;228(3):131-42.
15. Canamary AM Jr, Takahashi WY, Sallum JMF. Autoimmune retinopathy: a review. Int J Retina Vitreous. 2018 Jan 3:4:1.

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