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Acute exsudative polymorphous vitelliform maculopathy: a case report

Maculopatia viteliforme polimorfa exudativa aguda: um relato de caso

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Abstract

Acute exudative polymorphous vitelliform maculopathy is an extremely rare retinal disorder, that has been considered as a form of paraneoplasic retinopathy, found in patients with a underlying primary tumor. Symptoms of acute exudative polymorphous vitelliform maculopathy include preceding headache followed by acute onset of vision loss. The fundus of a patient with this condition typically demonstrates bilateral, subretinal white-yellow deposits in the macular region. The report of a rare disease which has a strong association with underlying neoplasia is extremely relevant whereas it helps better comprehend its genuine history, possible complicacy and prognosis.

Keywords: *Macula lutea/pathology; Retina/pathology; Neoplasms/diagnosis; Low vision; Case reports*

Resumo

A maculopatia viteliforme polimorfa exsudativa aguda é um distúrbio retiniano extremamente raro, que tem sido considerado como uma forma de retinopatia paraneoplásica, encontrada em pacientes com um tumor primário subjacente. Os sintomas de maculopatia viteliforme polimorfa exsudativa aguda incluem dor de cabeça precedente seguida de perda aguda da visão. O fundo de olho de um paciente com essa condição demonstra geralmente depósitos bilaterais, branco-amarelados na região macular. O relato de uma doença rara e que tem uma forte associação com neoplasia oculta é de extrema relevância, pois ajuda a conhecer melhor a sua historia natural, possíveis complicações e prognóstico.

Descritores: Macula lutea/patologia; Retina/patologia; Neoplasias/diagnóstico; Baixa visão; Relatos de casos

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Introduction

Acute exudative polymorphous viteliform maculopathy (AEPVM) is a rare retinal disease, which was first described in 1988 by Gass et al.[1] So far, about 20 cases were described in the world literature[2]. The first cases reported were of two white men who had a sudden onset of headache and visual loss associated to various creamy-white sub-retinian injuries and serous retinal detachment in macular areas of both eyes[1].

The etiology of the disease has not been clarified yet. However, it is considered that the changes are resulting from a paraneoplastic retinopathy found in patients with some hidden primary neoplasm.

The present study aims to report a case of acute exudative polymorphous viteliform maculopathy in order to facilitate the identification and better understanding of the disease.

Case report

A 19-year-old patient complaining of sudden low visual acuity in the right eye for about 20 days, with no personal and family history of disease, as well as previous ophthalmologic changes. The patient reported frequent headaches and denied prodromes of viral diseases. Visual acuity with correction was of counting fingers in the right eye, and 20/20 in left eye one. The anterior segment did not have changes. The aplanation tonometry was 12 mmHg in the right eye and 13 mmHg in the left eye.

The fundus exam showed a regmatogenic retinal detachment in the right eye, with rupture involving the inferior temporal region associated to a yellowish and round subfoveal lesion. In the macular region of the left eye, yellowish sub-retinal lesions with a viteliform appearance were seen, similar to those of the right eye, but in greater quantity (Figure 1).

In the autofluorescence examination (Figure 2), a hyper-autofluorescence of the regions corresponding to the sub-retinal accumulation of yellowish material was observed in both eyes, bordered by areas of hypo-autofluorescence.

The fluorescein angiography (Figure 3) showed marked hyperfluorescence in the central areas of the maculae in both eyes, corresponding to serous retinal detachment and exudative deposit. No areas of contrast leakage were observed.

Optical coherence tomography (OCT) of the right eye revealed intra retinal cysts associated to retinal detachment. The OCT of the left eye (Figure 4) showed serous detachment of the neuroepithelium, with subretinal rounded deposits.

The retinal detachment found in the right eye was treated by posterior vitrectomy via pars plana, using perfluorocarbon, endolaser and octafluoropropane C3F8 gas. About 2 months after surgery, the visual acuity in this eye was 20/100, and the retina was completely glued.

Discussion

The article reports a case of AEPVM in a 19-year-old young man, without previous comorbidities. The findings of the case coincide with the alterations already described for the disease, of small and numerous bilateral yellowish lesions affecting the retinal pigment epithelium (RPE), causing serous retinal detachment[3-5]The low visual acuity identified in the right eye of the patient was related to a regmatogenic retinal detachment, which had no relation to the presently diagnosed AEPVM.

The pathophysiology of the disease is still the subject of speculation. It is presumed that the individual who develops AEPVM has a primary malignancy that has not yet been diagnosed due to the absence of signs and symptoms. The most common primary neoplasia reported in patients with AEPVM is cutaneous or choroidal melanoma. However, the condition has been documented in patients with other types of neoplasms,

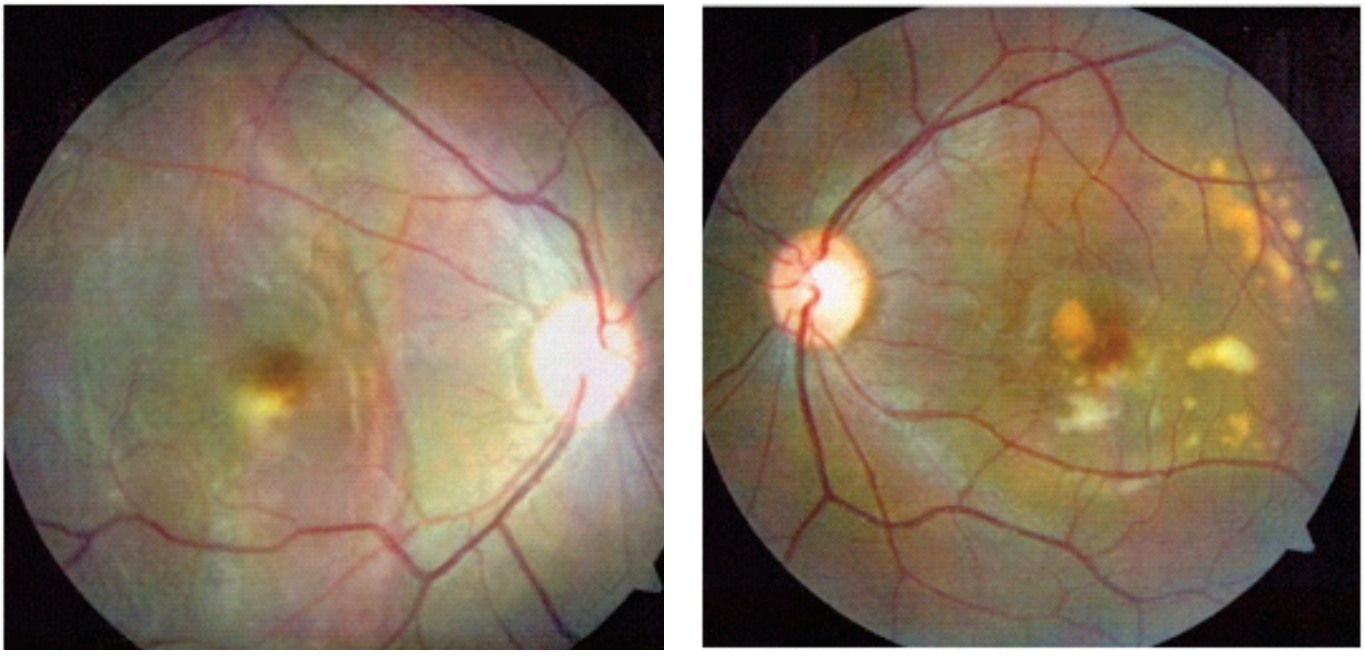

Figure 1. Retinographies: right eye shows extensive regmatogenic retinal detachment and a yellowish subfoveal rounded lesion. In the left eye, there are multiple yellowish lesions in the macular region.

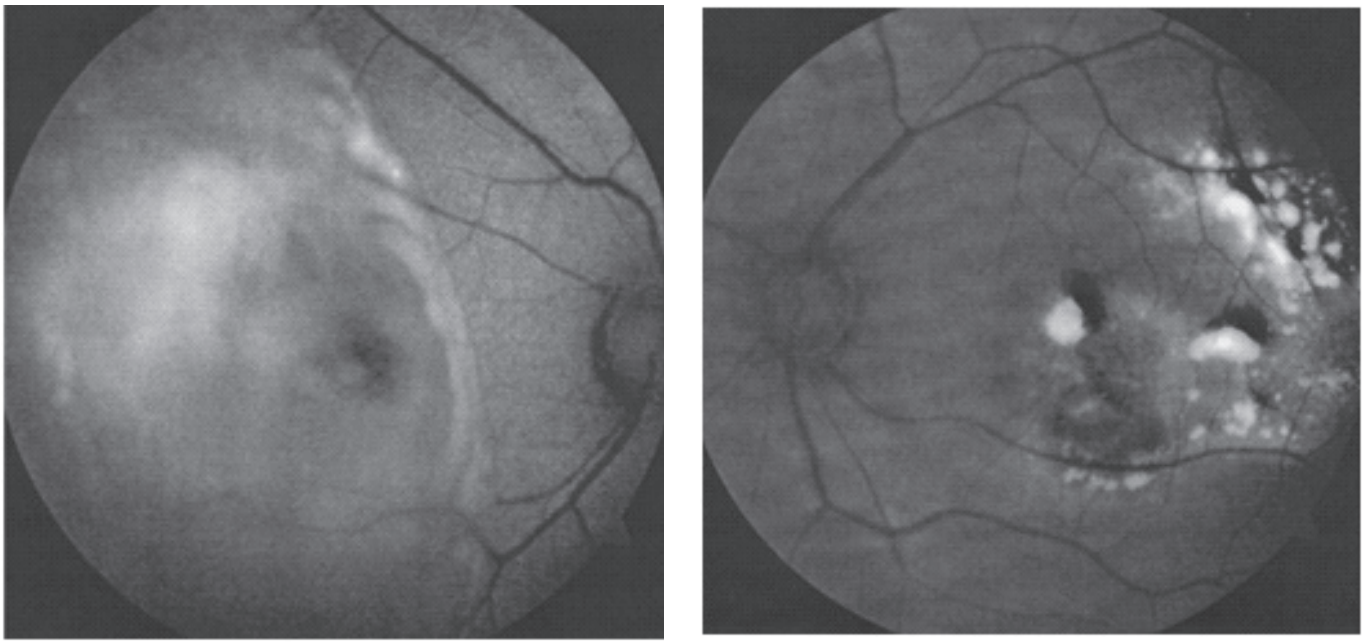

Figure 2. Autofluorescence: hyper-autofluorescence in the areas corresponding to the subfoveal yellowish lesions, bordered by areas of hypo-autofluorescence, in both eyes

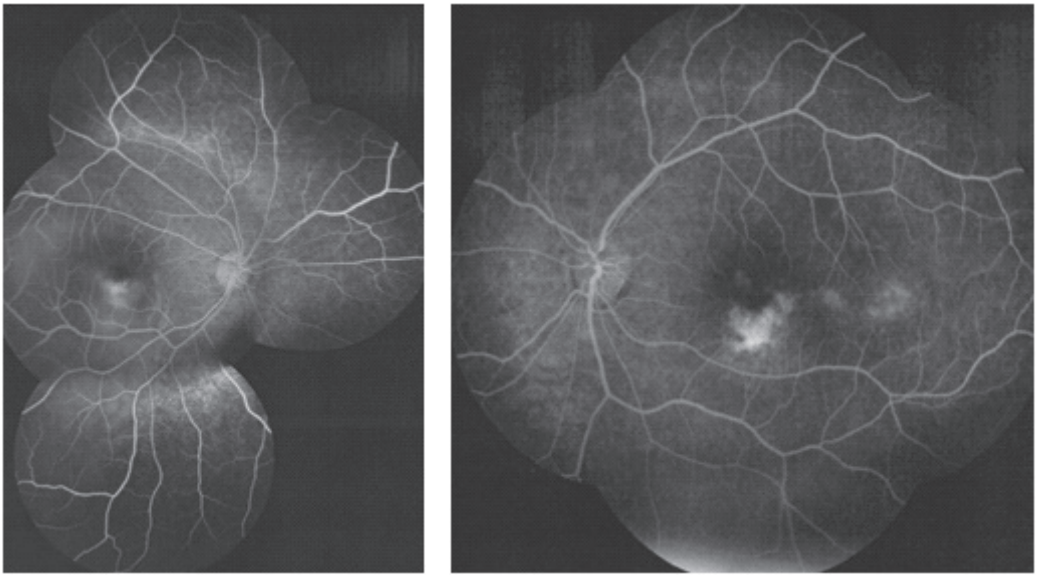

Figure 3. Angiofluoresceinography: subfoveal hyperfluorescent areas in both eyes

such as carcinomas[6].

There is considerable suspicion that RPE is the main structure affected in this condition, and its dysfunction could explain the overload of lipofucsin seen in autofluorescence[7]. The hypothesis is that a cross-reaction occurs between the autoantibodies produced against primary neoplasia and antigens of the RPE cells, and that the yellowish pigment is an EPR product and/or layer of damaged photoreceptors[8].

Autoimmune etiology was considered due to the detection of anti-peroxiredoxin 3 autoantibodies (PRDX3) in the serum of a patient during the acute phase of the disease (9). Symptoms of upper airway infection similar to influenza and headache have been reported prior to the manifestations of the ocular symptoms, and some studies suggest that there may be some viral trigger for the development of AEPVM[6]. Experiments in rats detected viral antigens and autoantibodies against RPE following coronavirus infection, and associated the findings to retinopathy [6].

The hypothesis of inflammatory pathology was also raised, since some patients presented a favorable response to the treatment with steroidal anti-inflammatory drugs[1]. However, treatment with corticosteroids is still controversial, since visual acuity tries to improve with or without treatment in a few weeks or months[6].

The natural history of the disease has not been fully elucidated yet, as the cases described are few, and some of them end up succumbing in a short time due to the aggressiveness of the primary disease, as occurred in a study in which 2 of the 5 patients being followed died in less than 5 months[9].

The most defended hypothesis is that AEPVM is a pa-

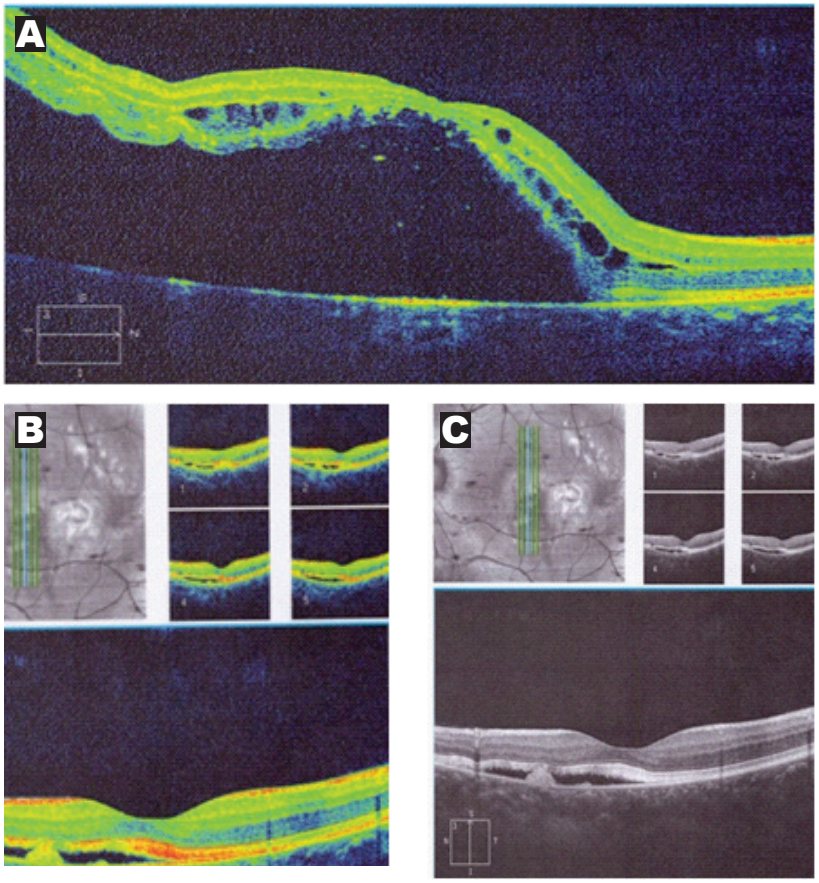

Figure 4. Optical Coherence Tomography (OCT): (A) Retinal detachment in the right eye associated to intra-retinal cysts. (B-C) Serous retinal detachment with deposits and accumulation of subretinal fluid in the left eye.

raneoplastic retinopathy and thus, in the case of AEPVM, it is mandatory to perform a screening for occult primary neoplasias. The mean time between onset of AEPVM lesions and the diagnosis of primary neoplasm is generally 42 months (10).

In the case described, tests were performed for cancer screening, but no findings were found for this disease. The young man had no lesion suggestive of malignancy, and also he was black, which is usually a protective factor for skin cancer. However, even if the probability is minimal, screening should be performed.

In summary, AEPVM is a rare disorder, with few cases described in the literature published, whose etiology and pathogenesis are not well known, but may represent the first manifestation of a neoplasm. Considering this, a thorough and extensive propaedeutics should be performed to search for any underlying cancer, and thus start as soon as possible the appropriate treatment.

References

1. Gass JD, Chuang EL, Granek H. Acute exudative polymorphous vitelliform maculopathy. Trans Am Ophthalmol Soc. 1988; 86:354-66.
2. Massaro et al. Acute exudative polymorphous vitelliform maculopathy. Eur J Ophthalmol. 2015; 25(5): e91-e94.
3. Chan CK, Gass FJ, Lin SG. Acute exudative polymorphous vitelliform maculopathy syndrome. Retina. 2003;23(4):453–62.
4. Spaide R. Autofluorescence from the outer retina and subretinal space: hypothesis and review. Retina. 2008;28(1):5–35.
5. Vaclavik V, Ooi KG, Bird AC Robson AG, Holder GE, Webster AR. Autofluorescence findings in acute exudative polymorphous vitelliform maculopathy. Arch Ophthalmol. 2007;125(2):274–7.
6. Modi, Kunjal K, Roth, Daniel B., Green, Stuart N. Acute Exudative Polymorphous Vitelliform Maculopathy in Young Man: a case report. Retinal Cases Brief Rep. 2014; 8(3):200-4.
7. Wiklund A, Peebo BB. Acute exudative polymorphous vitelliform maculopathy in young woman: presymptomatic findings and 21-month follow-up. Retinal Cases Brief Rep. 2013; 7(2):123–7.
8. Gass JD. Acute idiopathic exudative polymorphous vitelliform maculopathy. In: Gass JDM, ed. Stereoscopic Atlas of Macular Diseases: Diagnosis and Treatment. 5th ed. St Louis, MO: CV Mosby. 1997, Vol. 2, p.168–9.
9. Koreen L, He SX, Johnson MW, Hackel RE, Khan NW, Heckenlively JR. Anti-retinal pigment epithelium antibodies in acute exudative polymorphous vitelliform maculopathy: a new hypothesis about disease pathogenesis. Arch Ophthalmol. 2011;129(1):23-9.
10. Al-Dahmash SA, Shields CL, Bianciotto CG, et al. Acute exudative paraneoplastic polymorphous vitelliform maculopathy in five cases. Ophthalmic Surg Lasers Imaging. 2012; 43(5): 366-73.

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