

# Neurosensory Detachment in Acute Myeloid Leukemia

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## ABSTRACT

Neurosensory retinal detachment is a rare ocular finding in acute myeloid leukemia. We describe a case of a 25 year-old female with history of acute myeloid leukemia under chemotherapy that was referred to the Ophthalmology Department due to blurred vision and metamorphopsia in the left eye for the last two weeks. On examination, there was an oval, high, paracentral lesion, superior to the macula with adjacent small hemorrhage. She performed macular spectral domain optic coherence tomography that showed macular edema with neurosensory detachment and pigment epithelial detachment adjacent. Fluorescein angiography revealed a multifocal diffusion with hyperfluorescence and indocyanine green hypofluorescent spots. The chemotherapy was continued and, one month after, progressive visual improvement was identified. A systemic disease with atypical retinal findings should alert to diagnose extramedullary disease and a multidisciplinary approach is necessary.

**Keywords:** Retinal detachment, acute myeloid leukemia, chemotherapy, multidisciplinary approach

## RESUMO

O descolamento da retina neurosensorial é um achado raro em doentes com diagnóstico de leucemia mielóide aguda. Descrevemos um caso clínico de uma doente com 25 anos de idade, antecedentes pessoais de leucemia mielóide aguda sob quimioterapia, referenciada ao Departamento de Oftalmologia por visão turva e metamorfopsias no olho esquerdo com duas semanas de evolução. Ao exame oftalmológico, no olho esquerdo observou-se uma lesão elevada, paracentral, oval, superior à macula com uma pequena hemorragia adjacente. Foram realizados exames complementares de diagnóstico, dos quais se destacam: tomografia de coerência óptica macular com evidência de um descolamento neurosensorial e do epitélio pigmentar da retina (EPR), angiografia fluoresceínica que mostrou uma hiperfluorescência com difusão multifocal e angiografia com verde indocianina que demonstrou uma área hipofluorescente. A doente manteve a quimioterapia e, após um mês, o descolamento neurosensorial desapareceu, o descolamento do EPR melhorou e a acuidade visual foi de 20/20. Este caso evidencia uma apresentação ocular rara de uma doença sistémica e demonstra a importância de uma abordagem multidisciplinar.

**Palavras-chave:** descolamento neurosensorial da retina, descolamento EPR, leucemia mielóide aguda, quimioterapia, abordagem multidisciplinar

## INTRODUCTION

Leukemia can affect all ocular structures. Ocular involvement has been reported more commonly in patients with acute leukemia (than in chronic forms) and the clinical features affect mainly the posterior pole.<sup>1</sup> Ocular involvement may be caused by a myriad of entities: direct infiltration by neoplastic cells, affection by hematological abnormalities (thrombocytopenia, anemia, hyperviscosity states) or secondary to leukemia treatment. Typical retinal findings can range from intraretinal hemorrhages, cotton wool spots, white-centered hemorrhages, vessel occlusion to opportunistic infections.<sup>1</sup> Choroidal involvement is much more uncommon than retinal consequences. The majority patients with choroidal infiltration are asymptomatic, however, in some cases, symptoms can result from neurosensory or retinal pigment epithelial detachments.<sup>2</sup>

## CASE REPORT

A 25-year old woman presented with blurred vision and metamorphopsia in the left eye. The patient had a medical history of acute myeloid leukemia undergoing chemotherapy (FLAG IDA: fludarabine, cytarabine, idarubicin, granulocyte colony-stimulating factor) and active fungal septicemia (*Fusarium* spp) secondary to immunosuppression treated with voriconazole and amphotericin. Under observation, best corrected visual acuity (Snellen scale) was 20/20 and 20/200 in right and left eye, respectively, intraocular pressure was normal and the anterior segment was unremarkable. Fundus examination revealed multiple small intraretinal hemorrhages in the right eye and an oval, high, paracentral lesion superior to the fovea with a small hemorrhage adjacent in the left eye.

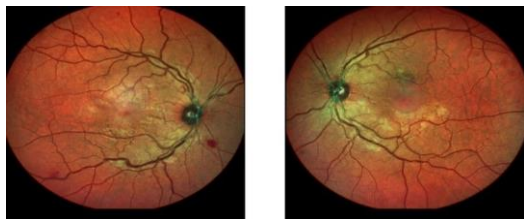


Figure 1 – Multicolor fundus photography

Amsler grid showed a paracentral inferior quadrant distortion in the left eye. Macular spectral domain optic coherence tomography was normal in the right eye and

presented a foveal neurosensory detachment, a disruption of the ellipsoid zone/external limiting membrane, a pigment epithelial detachment and a disorganization of internal layers in the left eye.

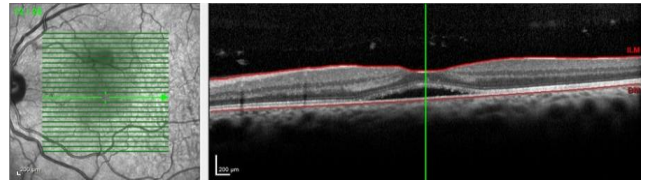


Figure 2 – Macular spectral domain optic coherence tomography: subretinal fluid (left eye)

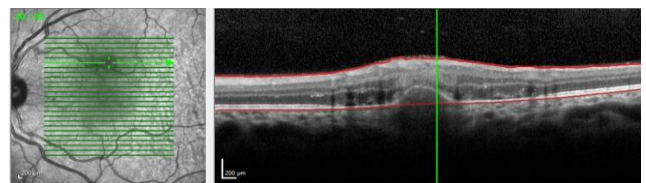


Figure 3 – Macular spectral domain optic coherence tomography: disruption of the ellipsoid zone/external limiting membrane, a pigment epithelial detachment and a disorganization of internal layers (left eye)

Fluorescein angiography disclosed a hyperfluorescent multifocal diffusion that increases during the exam and indocyanine green hypofluorescent spots.



Figure 4 – Fluorescein angiography and indocyanine green (left eye)

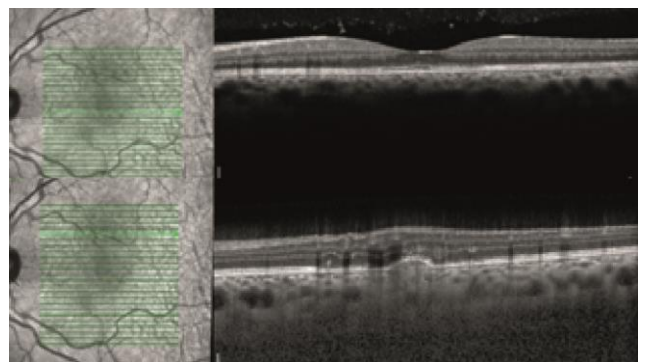


Figure 5 – Macular spectral domain optic coherence tomography macular: A - neurosensory detachment resolution; B - pigment epithelial detachment (left eye)

## DISCUSSION

Acute myeloid leukemia may present signs of choroidal infiltration without having leukemic retinopathy. Choroidal involvement is frequently affected in histopathologic studies but clinical manifestations are rarely found. In our case, the patient had mainly loss of vision due to subretinal fluid at the fovea which is unusual observed.<sup>2-5</sup> When performed imaging tests, a pigment epithelial detachment was observed too. A leukemic choroidal cells infiltration or a fungal choroidal affection might be responsible for this serous and pigment epithelial detachments. After one month of systemic chemotherapy and antifungal treatment, neurosensory detachment disappeared, pigment epithelial detachment was diminished and visual acuity was recovered which suggest the role of leukemic cells in the detachment area or fungal choroidal affection. The mechanism behind detachments in leukemia are still poorly understood. Some authors propose decrease blood flow in the choriocapillaris, disruption of outer blood-retinal barrier, alterations to local oncotic and/or hydrostatic forces. Thus, choroidal hypoxia lead to accumulation of subretinal fluid by retinal pigment epithelium damage. Chemotherapy is recommended as the first line treatment to solve intraocular affection and no topical treatment is indicated.<sup>6,7</sup>

The main differential diagnosis of serous and epithelial pigment detachments in this case was central serous choroidopathy. However, OCT didn't reveal a thickened choroid, fluorescein angiography didn't show early hyperfluorescent spot that gradually enlarges or indocyanine green angiography hyperfluorescent areas. Inflammatory causes were promptly excluded due to the absence of anterior or posterior inflammation.

A prompt recognition of ocular manifestations as a sign of extramedullary disease is essential. Atypical retinal manifestations should alert the hematologist and ophthalmologist that this can be a sign of choroidal involvement.<sup>8</sup>

## CONCLUSIONS

Serous plus pigment epithelial detachment of the retina is seldom reported. The visual function can be fully restored through undertaking appropriate treatment. An early identification of ocular manifestations can help to diagnose extramedullary disease.

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No conflicts of interest to disclose.

The authors declare that there is no conflict of interest.