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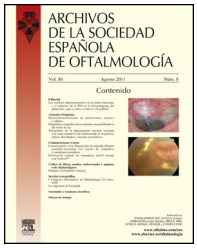

Short communication

# Unilateral optic disk edema with central retinal artery and vein occlusions as the presenting signs of relapse in acute lymphoblastic leukemia☆,☆☆

*R. Salazar Méndez*[a,*], *M. Fonollá Gil*[b]

[a] *Servicio de Oftalmología, Hospital Comarcal Jarrio, Coaña, Asturias, Spain*
[b] *Servicio de Oftalmología, Hospital Universitario Central de Asturias, Oviedo, Asturias, Spain*

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ABSTRACT

*Clinical case:* A 39-year-old man with Philadelphia chromosome-positive acute lymphoblastic leukemia (LAL Ph+) developed progressive vision loss to no light perception in his right eye. He had optic disk edema and later developed central artery and vein occlusions. Panphotocoagulation and radiotherapy of the whole brain were performed in several fractions. Unfortunately the patient died of hematological relapse 4 months later.
*Discussion:* Optic nerve infiltration may appear as an isolated sign of a leukemia relapse, even before a hematological relapse occurs. Leukemic optic neuropathy is a critical sign, not only for vision, but also for life, and radiotherapy should be immediately performed before irreversible optic nerve damage occurs.



## Edema de papila y obstrucción de arteria y vena central de la retina como manifestación inicial de una recaída leucémica

*Palabras clave:*
Leucemia linfoblástica aguda
Nervio óptico
Obstrucción arteria central retina
Obstrucción vena central retina
Radioterapia

RESUMEN

*Caso clínico:* Varón con amaurosis en ojo derecho y diagnóstico previo de leucemia linfoblástica aguda con cromosoma Philadelphia positivo (LAL Ph+). Presenta edema sectorial del disco óptico y, posteriormente, obstrucción de arteria y vena central de la retina. Se realiza panfotocoagulación retiniana y tratamiento radioterápico holocraneal. A los 4 meses sufre recaída hematológica, siendo finalmente exitus.
*Discusión:* La afectación del nervio óptico puede presentarse como único hallazgo en una recurrencia de una LAL, precediendo incluso a la recaída hematológica. Constituye por tanto

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☆☆ This clinical case was presented as a board communication during the 88th National Congress of the Sociedad Española de Oftalmología [Spanish Society of Ophthalmology], from 26 to 29 September 2012, Barcelona, Spain.

* *Corresponding author.*
E-mail address: raquelsalazarmendez@gmail.com (R. Salazar Méndez).

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una urgencia no solo visual sino, sobre todo, vital, que precisa de un tratamiento intensivo muchas veces complementado con radioterapia.



## Introduction

The incidence of ophthalmological manifestations in patients with leukaemia has increased notably in recent years due to the longer patient survival resulting from the effectiveness of the available therapies.[1–3] Despite longer remission periods, a relapse after a full remission continues to represent an unfavourable prognostic factor, especially in cases where the central nervous system (CNS) is involved.[4]

The optic nerve (ON) is affected in 13–18% of all patients with leukaemia. This may occur with any cytological variety and at any stage of the disease, even as the only finding of an extramedullary relapse, and even several months before a haematological relapse.[4] Visual loss due to leukaemia infiltration of the ON constitutes an ophthalmological emergency and requires the urgent use of radiotherapy before irreversible neuronal damage occurs.[3]

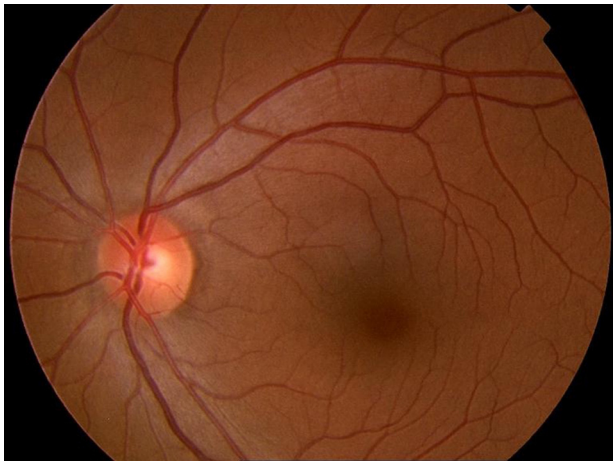

**Fig. 1 – Retinography of left eye.**

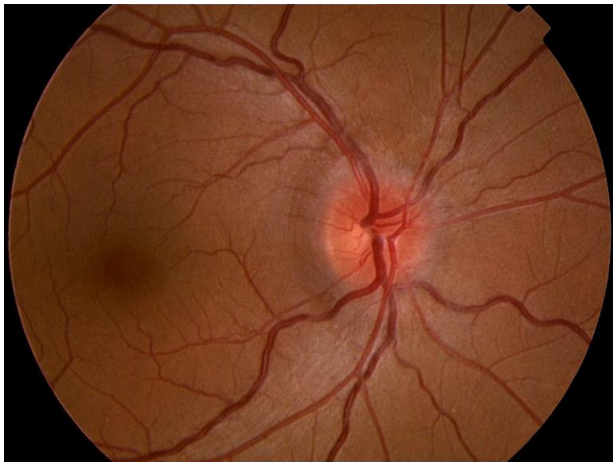

**Fig. 2 – Retinography of right eye. Note the blurring on the nasal edge of the optic disk.**

## Clinical case

A 39-year-old male patient with a one-week course episode of loss of vision in the right eye (RE) and supraorbital pain. His medical history is notable for a diagnosis of Philadelphia chromosome-positive acute lymphoblastic leukaemia (Ph+ ALL) five years before. Chemotherapy was administered as per Pethema protocol and he later received an umbilical cord blood allotransplantation; he suffered two relapses following complete remission.

On the physical examination, the RE does not have light perception and the visual acuity in his left eye (LE) is 1.0. The intraocular pressure is normal and there is an afferent pupillary defect in the RE. The LE fundus does not show abnormalities (Fig. 1), whereas the RE displays some raised blurring of the optic nerve's nasal edge (Fig. 2).

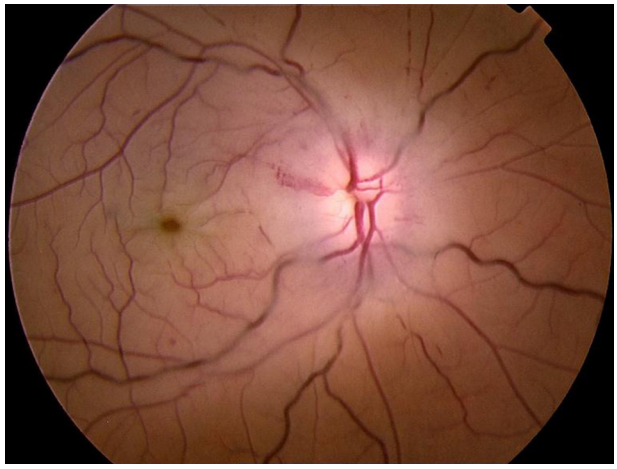

**Fig. 3 – Funduscopic image at four days post-admission with diffuse retinal oedema, disruption of the blood column and cherry-red stain.**

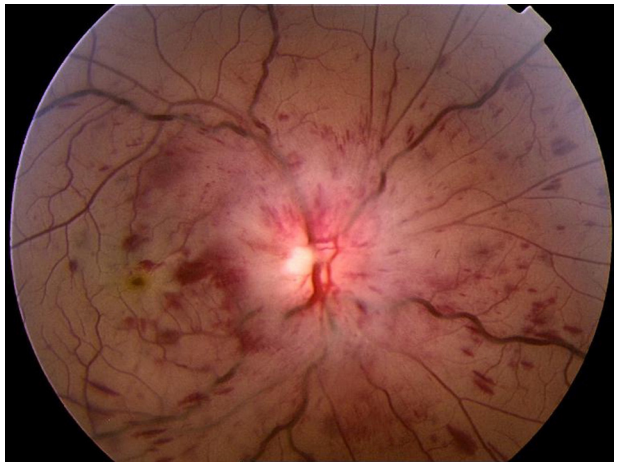

**Fig. 4 – Funduscopic image at eight days post-admission, with blot haemorrhages appearing along the venous tracts.**