Case report: We present the case of a 65-year-old man who was seen in the casualty department complaining of bilateral visual loss. His past medical history was unremarkable except for chronic lymphocytic leukaemia diagnosed two years previously: he was no longer receiving treatment for this disorder. His visual acuity was 20/200 in the right eye and 20/63 in the left eye and he had a bilateral papilledema. Computed tomography and magnetic resonance imaging of the brain and orbit were performed and showed an infiltrative mass in the orbit with extension to the ethmoid bone and both optic nerves. Biopsy of the mass was compatible with chronic lymphocytic leukaemia.

Discussion: Leukaemic infiltration of the optic nerve and central nervous system is a rare complication of chronic lymphocytic leukaemia, although it is common in the acute forms of this disease. Optic nerve infiltration is the most important ophthalmologic manifestation of leukaemia because it threatens vision and it is treatable (Arch Soc Esp Oftalmol 2007; 82: 303-306).

Key words: Chronic lymphocytic leukaemia, optic nerve, leukaemic infiltration, papilledema.

Caso clínico: Varón de 65 que consulta por pérdida de agudeza visual bilateral. Había sido diagnosticado de leucemia linfocítica crónica hacía 2 años y se encontraba sin tratamiento. Su agudeza visual era de 20/200 en el ojo derecho y 20/63 en el izquierdo y presentaba edema bilateral de papila. Se realizaron tomografía axial computerizada y resonancia magnética orbital y cerebral encontrándose una masa orbitaria con extensión al etmoides e infiltración de ambos nervios ópticos cuya biopsia reveló leucemia linfocítica crónica.

Discusión: La infiltración leucémica del nervio óptico y del sistema nervioso central es extremadamente rara en la leucemia linfocítica crónica al contrario de lo que sucede en las formas agudas. La infiltración del nervio óptico es la manifestación oftalmológica leucémica que implica mayor trascendencia clínica y terapéutica por amenazar seriamente la función visual y ser tratable.

Palabras clave: Leucemia linfocítica crónica, leucemia linfática crónica, nervio óptico, infiltración leucémica, papiledema.
INTRODUCTION

Leukemic infiltration of the optic nerve (ON) is particularly significant since it is a treatable cause for the irreversible loss of vision (1-4); this is the reason why it is highly advisable to perform in all cases of leukemia an ophthalmic examination at the time of diagnosis (2).

There are several types of leukemia and not all are equally capable of affecting the optic nerve.

Thus, chronic lymphocytic leukemia (also known as chronic lymphatic leukemia) rarely misses the ON, so that there are very few published cases on the issue (1,5). The present article illustrates the case of a patient suffering from chronic lymphocytic leukemia and bilateral infiltration of the ON.

CLINICAL CASE

A 65-year-old male checks in the ophthalmology emergency unit reporting a sudden bilateral loss of visual acuity.

He had a history of chronic lymphocytic leukemia diagnosed two years earlier. At the time of the ophthalmologic episode, he was not under treatment and was being followed up at the hospital’s hematology unit.

He exhibited visual acuity in long-distance vision and its regular correction of 20/200 had not improved with the stenopeic glasses on the right eye (RE), while the stenopeic glasses on the left eye (LE) had improved from 20/80 to 20/63.

Exploration of pupillary reflexes revealed a discreet pupillary defect relative to the RE.

Biomicroscopic exploration only revealed a pseudophakia with intraocular lens in the posterior chamber in both eyes.

Eye fundus exploration revealed a large papillary swelling with total loss of papillary limits in AO. Furthermore, the RE presented a large peripapillary hemorrhage concentric to the papilla, approximately 3 disk diameters and temporarily extending up to the macula (figs. 1 and 2).

An emergency cranial and orbital computed axial tomography (CAT) revealed an infiltrating mass covering both orbital vortexes and extending to the upper orbital fissures and cavities; the injury covered up to the preptontine cistern, involving the basilar artery and facing the ethmoid (fig. 3).
A brain and orbital magnetic resonance confirmed the CAT findings, revealing as well the infiltration of the optic nerve on both orbital vortexes (fig. 4).

Finally, the systemic exploration was completed with a cervical-thoracic-abdominal CAT that did not reveal any changes with respect to the patient’s previous condition.

Jointly with otolaryngology unit, a biopsy of the ethmoidal infiltration was performed since it was the most accessible of the affected areas. The hystopathologic study was compatible with the chronic lymphocytic leukemia.

From an ophthalmologic perspective, the diagnosis was a secondary papillary edema with retrobulbar infiltration of the optic nerve.

Faced with the anatomical condition of leukemic infiltration and its severe visual consequences, chemotherapy was immediately prescribed with fludarabine and cyclophosphamide, as well as cranial radiotherapy and the administration of systemic corticoids.

Evolution was good, observing 4 months after treatment a regression of the tumoral mass in both imaging tests; additionally, exploration of the eye fundus showed a resolution of the papillary infiltration, papillas being clearly marked with a slight pallor (figs. 5 and 6). Visual acuity at this point improved up to 20/50 in the right eye and 20/32 in the left eye.

**DISCUSSION**

Infiltration of the ON is an extremely rare complication in chronic lymphocytic leukemia, although there are previous cases described in the literature (1,5). When this or another type of leukemia affects the ON, two types of involvement may be distinguished, one prelaminary, the other retrolaminary. In those cases where the prelaminary portion of the optic nerve is involved, a tumor may
be observed directly emerging along the papilla. In those cases where affectation is retrolaminary, the image of the eye fundus is that of a papillary edema (3,5). In this case, infiltration of the optic nerve in the orbital vortexes generated the already described papilledema image.

It is precisely those cases involving retrolaminary affectations that raise most differential diagnostic issues with respect to the papilledema (4); in those cases, the unilaterality of the process leads to leukemic affectation. Bilateral leukemic infiltration of the ON is an atypical condition (4,5) and may lead to confusion in terms of diagnosis in the sense of an intracranial expansion process related to the papilledema (4,5). In this case, the previous diagnosis of chronic lymphocytic leukemia allowed physicians to suspect leukemic affectation and imaging tests may determine that the cause of the papillary swelling was a retrolaminary infiltration of the optic nerve. The differential diagnosis of the swelling of the optic nerve’s head involves, in addition to the papilledema caused by intracranial hypertension, the previous neuritis, anterior ischemic optic neuropathy, the optic nerve drusen, the pseudopapilledema and Leber’s disease.

The infiltration of the optic nerve is the ophthalmologic leukemic manifestation of greatest relevance in clinical and therapeutic terms since it seriously compromises vision and is treatable (2-5). Based on the above, a full ophthalmologic examination should be part of any exploration for any patient diagnosed with leukemia or leukemic relapse (3). One must keep in mind that vision may be preserved during the initial stages of leukemic affectation of the ON when ophthalmologic signs of affectation are already present (4-5).

When faced with an ophthalmologic exploration compatible with papilledema, it is extremely important to consider the differential diagnosis with a bilateral leukemic infiltration of the ON. In such cases, complementary tests such as hemograms, bone marrow tap, spinal tap and imaging tests may guide physicians in one direction or the other, and help them implement the appropriate treatment in each case (4).

**BIBLIOGRAFÍA**