Arteritic anterior ischemic optic neuropathy associated with chronic myelomonocytic leukemia (CMML): a case report

Abstract

Ocular involvement in patients with chronic myelomonocytic leukemia (CMML) is rarely reported, owing to the severe illness faced by many of these patients limits the feasibility of ocular examinations. Nevertheless, post-mortem examination of eyes from patients without ophthalmological symptoms and diagnosis of CMML has revealed ocular infiltration of atypical cells. We presented a 72-year-old man with two days of left-sided blurring of vision and an episode of transient visual obscuration associated to periorbital pain. To our knowledge, this is the second case in peer-reviewed literature reporting the association between arteritic anterior ischemic optic neuropathy (AAION) and chronic myelomonocytic leukemia. A possible explanation is the fact that small vessel vasculitis, of unclear pathogenesis so far, complicates approximately 10% of CMML and presents as cutaneous vasculitis and lupus-like syndromes. Therefore, this may be in relation with the vasculitis of the short posterior ciliary arteries, which happens in cases of AAION.

Key words: chronic myelomonocytic leukemia; optic neuropathy; giant cells arteritis

Introduction

Arteritic anterior ischemic optic neuropathy (AAION), also known as giant cell arteritis, typically presents in elderly patients with acute severe visual loss (visual acuity less than 20/200) and is occasionally preceded by transient visual loss similar to that associated to carotid artery disease. Reduction of visual acuity is associated with headaches and, more specifically, scalp tenderness and jaw claudication.

AAION is caused by an ischemia of the posterior ciliary arteries and/or the ophthalmic artery due to a granulomatous vasculitis of the vessel walls. Therapy is immediate intervention with systemic steroids, especially to protect against vision loss in the fellow eye. 1

Chronic myelomonocytic leukemia (CMML) is a disease formerly classified solely as a type of myelodysplastic syndrome (MDS) but reclassified in 1999 as a mixed MDS/myeloproliferative disorder.

MDS is a term that refers to a heterogeneous group of clonal bone marrow disorders associated with changes in marrow cellularity accompanied by dysmyelopoiesis and peripheral blood cytopenias. Many case reports exist in the literature demonstrating ocular involvement in patients with MDS, such as corneal ulcer, iridocyclitis, vitreous hemorrhage, retinal hemorrhage, and nerve-fiber layer infarcts. 2 In 2009, a case of non-arteritic anterior ischemic optic neuropathy in a patient with a MDS associated with chronic anemia was described for the first time. 3

In contrast to other MDS, ocular involvement in patients with CMML is rarely reported, owing to the fact that the severe, progressive illness faced by many of these patients limits the feasibility of ocular examinations.
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**Case report**

A 72-year-old man presented with two days of left-sided blurring of vision associated to periorbitary pain. Four days ago he had suffered a transient visual obscuration in his left eye (LE). His medical history included hypertension, hypercholesterolemia, diabetes mellitus, chronic renal insufficiency and a recent diagnosis of chronic myelomonocytic leukemia (CMML).

Best-corrected visual acuity (BCVA) (decimal notation) was 1.0 in right eye (RE) and light perception in LE. Anterior segment examination was unremarkable in both eyes. A relative afferent pupillary defect was present in the LE. Fundus examination of RE was normal (Figure 1), and fundus examination of LE revealed optic disc swelling, which corresponded to optic disc filling delay and leakage in late phases on fluorescein angiography (Figure 2A and 2B). Perimetry was normal in RE and it showed generalized depression in LE. Palpation of temporal arteries showed mild enlargement of left temporal artery. The erythrocyte sedimentation rate (ESR) was 120 mm (1st hour) and C-reactive protein was 7.8. A full blood count showed anemia and monocytosis. Complementary examinations, including infectious serology, lumbar puncture, and imaging studies (magnetic resonance imaging –MRI- and computed tomography –CT-) yielded normal findings. Leukemic optic nerve infiltration was excluded based on the absence of abnormalities in MRI and CT.

A presumptive diagnosis of anterior ischemic optic neuropathy was made. Hypertension, hypercholesterolemia, diabetes mellitus and anemia suggested non-arteritic ischemic optic neuropathy. However, the ESR pointed more towards an arteritic form of ischemic optic neuropathy (AAION).

Therefore, treatment with endovenous bolus of methylprednisolone, followed by slow-tapering of oral steroids, was started. One week later, temporal artery biopsy revealed the presence of giant cells, which confirmed the diagnosis. One month later, BCVA in LE remained stable, and vision in the fellow eye was preserved. Fundus examination of LE showed partial resolution of optic disc edema, although slight pallor of the disc was noted (Figure 3).

**Figure 1.** LE. Fundus examination of RE was normal.

**Figure 2.** A: Fundoscopy appearance of left eye at first examination. Note the optic disc swelling.

**Figure 2.** B: Fluorescein angiography of left eye showing hyper fluorescence in intermediate phases.
Figure 3. Fundus color photograph at last examination. Partial disappearance of optic disc edema can be appreciated.

Discussion
As previously mentioned, ocular involvement in patients with CMML is rarely reported, owing in part to the fact that the severe and progressive illness limits the feasibility of ocular examinations.

Nevertheless, post-mortem examination of the eyes of patients without ophthalmological symptoms and diagnosis of CMML, has revealed infiltration of atypical cells within the vasculature of the choroid bilaterally. This fact can arise the suspect that eye involvement in patients with CMML may be underdiagnosed.

Acute suprachoroidal hemorrhage with acute angle closure glaucoma, keratitis, keratouveitis and exudative retinal detachment are some of ocular manifestations described in patients affected by CMML.

However, optic neuropathy in the context of CMML has only been described once in the literature. Specifically, the first case of optic neuropathy associated to CMML was described in 2012.

Conclusion
To our knowledge, this is the first case in the peer-reviewed literature reporting the association between AAION and chronic myelomonocytic leukemia.

A possible explanation for this occurrence is the fact that small vessel vasculitis, of unclear pathogenesis so far, complicates approximately 10% of CMML and presents as cutaneous vasculitis and lupus-like syndromes. Therefore, this may be in relation with the vasculitis of the short posterior ciliary arteries which happens in cases of AAION.

Further research aiming to determine the likelihood of developing AAION in association with CMML is warranted.

REFERENCES