CASE REPORT

Bilateral Optic Nerve Involvement Combined with Unilateral Facial Palsy in a Patient with Acute Myeloid Leukaemia: A Case Report

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ABSTRACT

Central nervous system (CNS) involvement, including optic nerve involvement, in a patient with acute myeloid leukaemia (AML) is an extremely rare condition. We report a case of bilateral optic nerve involvement combined with unilateral facial palsy in a patient with AML who achieved complete remission following allogeneic peripheral blood stem cell transplantation as a young patient. After further evaluation, the patient was diagnosed with a recurrence of AML with CNS involvement. The presentation of multiple types of CNS involvement in AML may be suspicious evidence of AML recurrence.

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Introduction

Acute myeloid leukaemia (AML) is the most common acute leukaemia in adults. In younger patients (< 55 years), the clinical outcomes are generally better.¹

Ocular involvement in leukaemia has long been reported, and several ocular tissues may be affected. Signs of ocular involvement of the central nervous system (CNS) in leukaemia include optic nerve infiltration, cranial nerve palsies, and papilledema.²

After complete remission, optic nerve involvement associated with cranial nerve palsy is extremely rare. We report an unusual case of bilateral optic nerve involvement associated with unilateral facial palsy in a patient with AML believed to be in complete remission.

Case report

A 36-year-old woman presented unilateral facial palsy with progressively worsening blurred vision in her left eye for 10 days. She had a medical history of AML, was treated with a chemotherapy agent, and was in complete remission 2 years prior to presentation. At the initial visit, she had been observed and treated with cyclosporine in the

oncologic department, since allogeneic peripheral blood stem cell transplantation (allo-PBSCT) had been performed.

On ophthalmic examination, best-corrected visual acuity was 20/20 in her right eye and 20/70 in her left eye. The pupillary reflex did not show an apparent pupillary defect in either eye. On slit-lamp examination, there was no remarkable abnormality in the cornea or anterior segment. There was no pathologic finding in the measurement of intraocular pressure, and no limitation in binocular movement in any field of gaze. A funduscopy examination showed bilateral diffuse papillary swelling with blurred margins (Figure 1). A visual field test (Carl Zeiss Meditec, Dublin, CA, USA) using the 30–2 program showed asymmetrical bilateral visual field defects (Figure 2). Brain and orbit magnetic resonance imaging showed diffuse sulcus hypersensitivity on fluid-attenuated inversion recovery imaging with enhancement (Figure 3). A complete blood count showed a leukocyte count of 65,000 per cubic millimetre (64.3% neutrophils), a haemoglobin level of 12.1 g/dL, and a platelet count of 93,000 per cubic millimetre.

After 1 week, she complained of progressively worsening blurred vision, extending to her right eye. On funduscopy examination, bilateral papillary swelling remained, and was more prominent in the

Figure 1. (A) A fundusoscopic examination shows bilateral papillary swelling with a blurred margin at initial visit. (B) One week later, progressive papillary swelling and multiple flame-shaped papillary haemorrhages in the right fundus photography. (C) One month later, after intrathecal chemotherapy, bilateral reductions in papillary swelling and haemorrhage.

right eye with multiple flame-shaped haemorrhages (Figure 1). The visual field defects using 24–2 program in both eyes had also progressed since the last visit (Figure 2). The patient was referred to the oncologic department for further evaluation. She was admitted, and cerebrospinal fluid (CSF) tapping was performed for a neurologic evaluation. Lumbar puncture revealed an opening pressure of 27 cm of water. CSF analysis showed 1600 white blood cells per microliter (90% lymphocytes) with malignant cells in cytology test (Figure 4). She was diagnosed with a recurrence of AML with CNS involvement. Intrathecal chemotherapy with methotrexate and cytarabine (Ara-C) was performed for the treatment of AML.

The patient visited the clinic 1 month later with improved visual acuity of 20/20 in both eyes. Her unilateral facial palsy was completely resolved. On fundusoscopic examination, findings of papillary swelling and haemorrhage remained, but were reduced compared with the previous visit (Figure 1). A repeat visual field test using the 30–2 program showed improvement in both eyes (Figure 2).

Discussion

The prevalence of ocular involvement in leukaemia was reported to range from 9% to 90%.^{3,4} In leukemic patients with ocular involvement, optic nerve infiltration occurred variably, and ranged

Figure 2. (A) A visual field test shows asymmetrical bilateral visual field defects at initial visit. (B) One week later, progressed bilateral visual field defects. (C) One month later, after intrathecal chemotherapy, bilateral improvement of the visual field defects.

Figure 3. Brain and orbit magnetic resonance images. (A) Diffuse sulcus hypersensitivity on an axial fluid-attenuated inversion recovery image. (B) No definite enhancement of optic nerve on axial and coronal T2-weighted fat-suppressed images.

Figure 4. Monomorphic appearing atypical cells in cerebrospinal fluid analysis (May-Grünwald-Giemsa stain, $\times 600$).

from 5% to 13%.⁵ However, optic nerve infiltration in AML is a relatively uncommon condition.

Karesh et al. reported three (5.3%) cases with optic nerve involvement among 56 patients with AML,⁶ and Reddy et al. reported only one (0.9%) case with optic nerve involvement among 113 patients with AML.⁷

In this case, unilateral facial palsy and bilateral visual loss with optic nerve infiltration presented simultaneously following complete remission in a patient with AML. Based on the CSF analysis and other blood tests, the patient was diagnosed with a recurrence of AML with CNS involvement. Additionally, the increased intracranial pressure (27 mm of water) might cause the bilateral papillary swelling concurrently with CNS involvement. After chemotherapy, her facial palsy and visual acuity were improved and stable.

CNS involvement is a well-recognized complication of acute lymphocytic leukaemia (ALL). However,

the exact prevalence of CNS involvement in AML is unknown, but is considered uncommon and reported to range from 2% to 4%.⁸ Russo et al.⁹ demonstrated that the presence of specific ocular lesions was associated with a higher frequency of bone marrow relapses and CNS involvement, leading to a lower survival rate in ALL and AML. The exact pathophysiology of CNS involvement in AML is unknown. We speculated that spreading of malignant cells into CNS even after complete remission might be occurred. Thus, clinical outcomes in AML with CNS involvement are poor, and the optimal treatment is yet to be established because of the rarity of this condition.¹⁰ The CNS involvement in AML might occur rarely.

Therefore, clinicians should be aware of the possible ocular manifestations associated with multiple neurologic symptoms, such as peripheral nerve palsy, and possible causes in patients with AML, even if they have achieved complete remission. Moreover, other causes of vision loss in AML should be excluded, such as infection, inflammation, radiation-related artefacts, and side effects of chemotherapeutic agents.

Declaration of interest

The authors declare that there are no conflicts of interest. The authors alone are responsible for the writing and content of the article.

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