Bilateral Multifocal Choroiditis and Optic Neuropathy in a Patient With AIDS: A Diagnostic Dilemma

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By AIDS Reader [1]

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Here we describe the case of a young man with HIV infection who presented with bilateral multifocal choroiditis, optic neuropathy, and altered sensorium. In the literature, most cases of multifocal choroiditis in patients with AIDS were diagnosed at autopsy, underscoring the life-threatening nature of disseminated opportunistic infections in persons with HIV/AIDS. [1-3] Laboratory investigation, imaging studies, and therapeutic response to systemic amphotericin B therapy in our patient indicated disseminated Cryptococcus neoformans infection.

CASE SUMMARY
A 30-year-old HIV-positive man presented to the emergency department after experiencing 3 episodes of generalized tonic-clonic seizures the previous day. He had a history of fever, multiple episodes of headache, vomiting for 2 days, and blurred vision for about 2 weeks. HIV infection had been diagnosed 3 months earlier by a general physician. The patient was not taking any medication.

On systemic examination the patient was disoriented and was making incomprehensible sounds. He exhibited eye opening and withdrawal (flexion) motor response to painful stimuli. His Glasgow coma scale score was 8. There was no sign of any focal neurological deficits or nuchal rigidity.

Multiple umbilicated nodules were noted in the periorbital area and face. Bilateral pupillary reflexes (direct and consensual) were absent. On funduscopic examination, both optic discs appeared pale, but disc margins were distinct. There were also bilateral multiple patchy areas of retinal (retinal pigment epithelial) hypopigmentation with irregular margins of different sizes as well as multiple deep yellow-white choroidal lesions with an indistinct border within patchy retinal hypopigmentation. There were also areas of intraretinal hemorrhages (Figure 1). These features suggested bilateral multifocal choroiditis and optic neuropathy. There was no sign of anterior chamber inflammation or vitreitis.

Figure 1. Fundus photography shows bilateral multiple patchy areas of retinal (retinal pigment epithelial) hypopigmentation with irregular margins of different sizes and multiple deep yellow-white choroidal lesions with indistinct borders within patchy retinal hypopigmentation. There are also areas of intraretinal hemorrhages. These features suggested bilateral multifocal choroiditis.
A lumbar puncture was performed. The opening pressure was normal. His cerebrospinal fluid (CSF) was analyzed and included the following: total and differential leukocyte counts; protein, glucose, chloride, globulin, and adenosine deaminase levels; cytology; Gram, India ink, and Ziehl-Neelsen staining; bacterial and fungal culture; latex agglutination test for Cryptococcus polysaccharide antigen; and polymerase chain reaction assay for Mycobacterium tuberculosis. Results of the CSF analysis were normal except for the India ink smear of centrifuged CSF, which revealed encapsulated yeast cells with budding suggestive of C neoformans infection (Figure 2). The latex agglutination test was positive for Cryptococcus polysaccharide antigen.

A battery of additional laboratory investigations was subsequently conducted and included a complete blood cell count; examination of a peripheral blood smear; determination of the erythrocyte sedimentation rate and fasting blood glucose and serum electrolyte levels; blood culture; serological tests for Treponema pallidum and Toxoplasma (IgG titers); CD4+ cell count; urinalysis and urine culture; stool analysis; chest radiography; ultrasonography of the abdomen; and contrast-enhanced CT of the chest, head, and orbit. Findings from most of the investigations were unremarkable. However, scans of the chest showed a mild ground-glass opacity in the subpleural region of right lower lobe and scans of the brain showed mild cerebral atrophy. The CD4+ cell count was 71/µL.

MRI brain scans revealed multiple parenchymal lesions in the basal ganglia, thalamus, midbrain, and cerebellum. A cystic lesion was present in the left caudate nucleus, as evidenced by hypointensity on T1-weighted images and hyperintensity on T2-weighted images with no contrast enhancement, which was suggestive of cryptococcoma (Figures 3 and 4). Abnormal meningeal enhancement was not seen, and there was no edema. Mild mass effect was seen with compression of the frontal horn of the left lateral ventricle. Both optic nerves were normal. A cutaneous biopsy specimen of periorbital nodular lesions revealed molluscum contagiosum.
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Figure 3. Axial T1-weighted (A) and T2-weighted (B) images show a lesion in the left caudate nucleus, which is hypointense on T1-weighted imaging (arrow) and hyperintense on T2-weighted imaging. There is no edema. A mild mass effect is seen with compression of the frontal horn of the left lateral ventricle. Additional punctate lesions are seen in right caudate nucleus and bilateral lentiform nuclei on T2-weighted images. The lack of hypointensity and absence of a rim on T2-weighted images suggest that tuberculoma or lymphoma is unlikely. The signal intensity of the larger lesion is different from that of cerebrospinal fluid (CSF)—suggesting a cryptococcoma. Dilated Virchow-Robin spaces, which are seen in cerebral cryptococcosis, would show signal intensity similar to that of CSF on all sequences.

Figure 4. Postgadolinium-enhanced image shows no contrast enhancement of the lesions. No abnormal meningeal enhancement is seen.

With the presumptive diagnosis of bilateral cryptococcal choroiditis and optic neuropathy with disseminated cryptococcal infection, intravenous therapy with liposomal amphotericin B (0.7 mg/kg/d) was initiated for 2 weeks. This was followed by fluconazole (400 mg/d) and a twice-daily antiretroviral regimen of lamivudine 150 mg, stavudine 30 mg, and nevirapine 200 mg. The CSF and urine samples taken at admission were positive for *C neoformans* after cultured for 2 weeks.

Within 2 weeks of treatment, the patient was symptomatically better, with an improved sensorium; the choroiditis lesions showed some resolution with residual pigmentary changes. However, the
perception of light in both eyes was absent secondary to optic atrophy. On fundus examination 2 months later, only multiple areas of hyperpigmentation with pale optic discs were noted, suggestive of resolution. Follow-up cultures of CSF, urine, and serum showed that the fungemia had resolved. The molluscum contagiosum was also absent on follow-up.

**DISCUSSION**

The choroidal circulation has one of the highest rates of blood flow in the body. In persons with AIDS, multifocal choroiditis is usually secondary to endogenous infectious emboli in the choriocapillaris and reflects systemic dissemination of opportunistic infection. Early diagnosis and specific treatment are imperative and may be lifesaving. The causative agents may be *Pneumocystis jiroveci* (formerly *carinii*), *M tuberculosis*, *C neoformans*, *Histoplasma capsulatum*, *Candida albicans*, *Aspergillus fumigatus*, *Toxoplasma gondii*, *Mycobacterium avium-intracellulare*, and *T pallidum*.

HIV-associated primary lymphoma has also been reported as a cause of multifocal choroiditis. Usually, cryptococcal lesions appear as yellow-white multiple choroidal lesions with indistinct borders with or without intraretinal hemorrhage. A confirmed diagnosis may be possible only by vitreous or choroidal biopsy. Optic neuropathy in an HIV-infected patient may be the result of cryptococcal meningitis, tubercular meningitis, neurosyphilis, cytomegalovirus meningitis and encephalitis, CNS toxoplasmosis, herpesvirus infection, or HIV infection itself. The characteristic appearance of multifocal choroiditis, CSF analysis, and MRI brain scans suggestive of cryptococcoma guided us to consider a presumptive diagnosis of disseminated cryptococcal infection in this patient. The therapeutic response to systemic amphotericin B and fluconazole confirmed our diagnosis.

*C neoformans* is the most common life-threatening fungal pathogen that infects persons with HIV/AIDS. The primary site of infection is the lung, but the infectious process may manifest as meningitis and as a secondary neuro-ophthalmic complication. Intraocular invasion by *Cryptococcus* presenting as multifocal choroiditis or as endophthalmitis is an uncommon complication. Optic neuropathy in cryptococcal meningitis may be caused by direct optic nerve involvement or by chiasmal involvement; it may occur secondary to increased intracranial pressure or perineuritic adhesive arachnoiditis or, rarely, as a result of compression of a portion of visual sensory system from an adjacent cryptococcoma. In our patient, MRI scans did not reveal ventriculomegaly or meningeal enhancement and revealed only a mild mass effect. Optic neuropathy in this patient may have been the result of direct invasion of the optic nerve by *C neoformans*.

Most published reports show that multifocal choroiditis in persons with AIDS was diagnosed only at autopsy. This underscores the high mortality associated with disseminated opportunistic infection in this setting. Our case report highlights the importance of early diagnosis of multifocal choroiditis by an ophthalmologist. As this case demonstrates, good coordination between the primary care physician and the ophthalmologist can allow a correct and timely diagnosis of a disseminated and potentially fatal disease.

No potential conflict of interest relevant to this article was reported by the authors.

**References:**

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