1-1-2014

Acute cranial neuropathies heralding neurosyphilis in human immunodeficiency virus-infected patient.

Saeed Alqahtani
George Washington University

American Journal of Case Reports

ISSN 1941-5923
© Am J Case Rep, 2014; 15: 411-415
DOI: 10.12659/AJCR.892292

Received: 2014.08.20
Accepted: 2014.09.06
Published: 2014.09.25

Acute Cranial Neuropathies Heralding Neurosyphilis in a Human Immunodeficiency Virus-Infected Patient

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF Saeed Alqahtani

Department of Neurology, School of Medicine and Health Sciences, George Washington University, Washington, DC, U.S.A.

Corresponding Author: Saeed Alqahtani, e-mail: sqanea@gwu.edu
Conflict of interest: None declared

Patient: Male, 31
Final Diagnosis: Neurosyphilis
Symptoms: Diplopia •facial droop • facial nerve palsy • headache
Medication: —
Clinical Procedure: —
Specialty: Infectious Diseases

Objective: Unusual clinical course

Background: Symptomatic early neurosyphilis with isolated acute multiple cranial nerves palsy as initial manifestation of HIV infection is very rare. It is caused by direct invasion of the central nervous system by the spirochete *Treponema pallidum*.

Case Report: A 31-year-old African-American homosexual man presented with bilateral hearing loss, constant vertigo, intermittent horizontal diplopia, and bilateral facial droop, which was associated with occipital headache without fever. Neurological examination revealed bilateral vestibulocochlear and facial nerve palsy. On brain magnetic resonance imaging (MRI) before and after administration of gadolinium, he was found to have extensive isolated basilar meningeal enhancement involving the midbrain, pons along the seven and eight nerves complex bilaterally, consistent with basal meningoencephalitis.

Conclusions: Neurosyphilis can present as initial manifestation of HIV infection with early involvement of basal meninges and cranial nerves. It is important to understand that neurosyphilis is still a significant disease with complex neurological presentation. Early diagnosis and treatment of neurosyphilis is crucial due to potential persistent disabilities that can be easily treated or even prevented.

MeSH Keywords: Cranial Nerve Diseases • HIV • Magnetic Resonance Imaging • Meningoencephalitis • Neurosyphilis

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/892292

Background

Neurosyphilis is very challenging to diagnose due to its variable and complex presentation. This case illustrates a rare manifestation of isolated cranial neuropathies due to neurosyphilis revealing undiagnosed human immunodeficiency virus (HIV) infection in a young male.

Case Report

A 31-year-old African-American homosexual healthy man who presented to the emergency room with 2 weeks history of acute bilateral hearing loss and constant vertigo associated with intermittent horizontal binocular diplopia. He also had progressive difficulty closing both eyes, with increased tearing. His symptoms were associated with continuous occipital headache and neck stiffness without any other neurological complaints or fever. The review of systems was unremarkable and there had been no previous similar presentation. Neurological examination revealed a drowsy patient but awake and following commands, with obvious bilateral facial droop and positive Bell's phenomenon. He had significant decrease in hearing, mainly in the left ear, with abnormal Weber test. The remaining of cranial nerves and neurological examination were intact except for limited neck flexion due to pain and stiffness. On admission, brain magnetic resonance imaging (MRI) before and after administration of gadolinium was obtained and showed extensive basal meningeal enhancement surrounding the midbrain and pons (Figure 1A, 1B). There was associated increased T2 and fluid-attenuated inversion recovery (FLAIR) signal intensity in the brain stem parenchyma (Figure 2A, 2B). There was also subtle enhancement of the cisternal segments of the facial and vestibulocochlear nerves complex, mainly over the left side (Figure 3). The image finding confirmed the diagnosis of basal meningoencephalitis. Cerebrospinal fluid (CSF) analysis demonstrated a lymphocytic pleocytosis, with a white blood cell count of 298/μL, a glucose level of 22 mg/dL, and significant elevation of protein level of 595 mg/dL. Laboratory test results were positive for human immunodeficiency virus (HIV) antibody, which was confirmed with positive HIV polymerase chain reaction (PCR) test result and elevated viral load. The serum CD4 count was low (237 cells/μL), serum rapid plasma reagin (RPR) level was very high (1:64), *Treponema pallidum* particle agglutination assay (TPPA) was positive, the Venereal Disease Research Laboratories (VDRL)-CSF dilution titer result was positive and elevated at 1:16. The other laboratory test results, including the flow cytometry of the CSF and cytology, QuantiFERON test for tuberculosis, CSF India ink, and CSF gram stain and culture, were all negative. The patient was started on intravenous penicillin G at dose of 4 million units every 4 hours for 21 days. The patient had

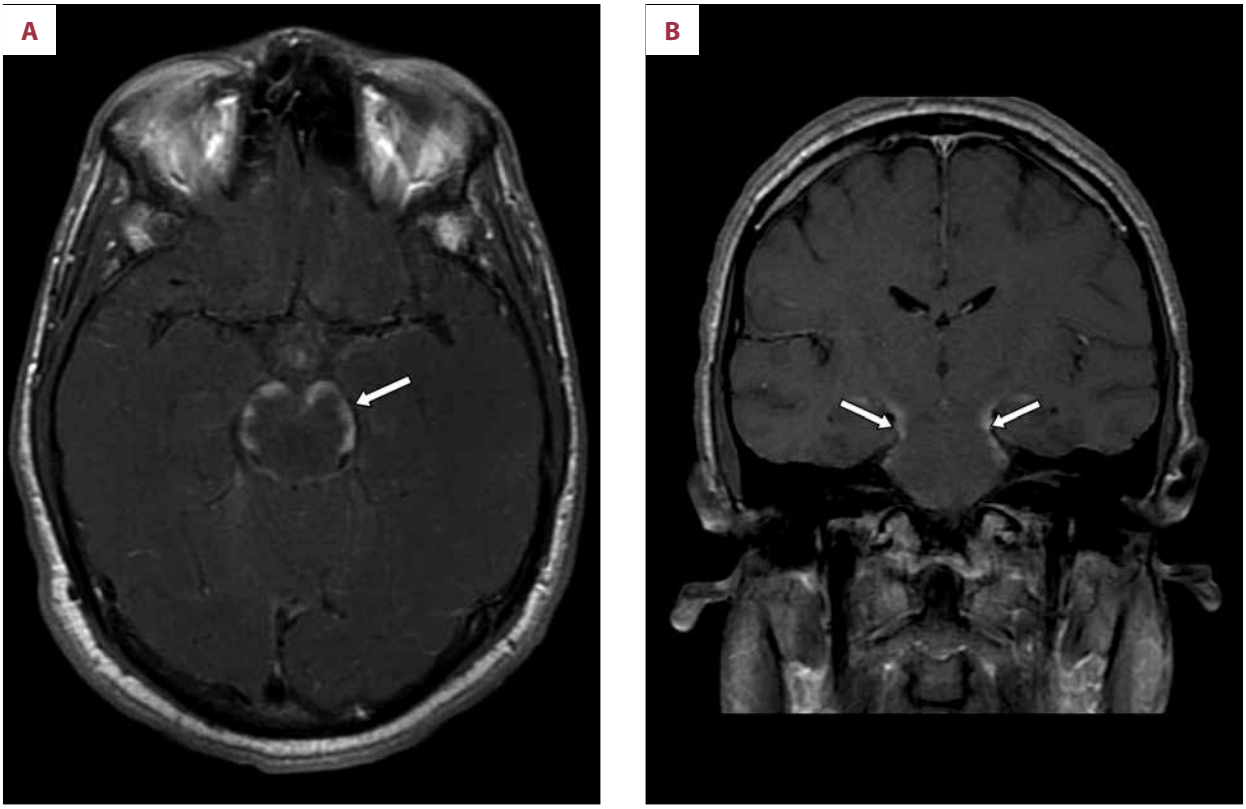
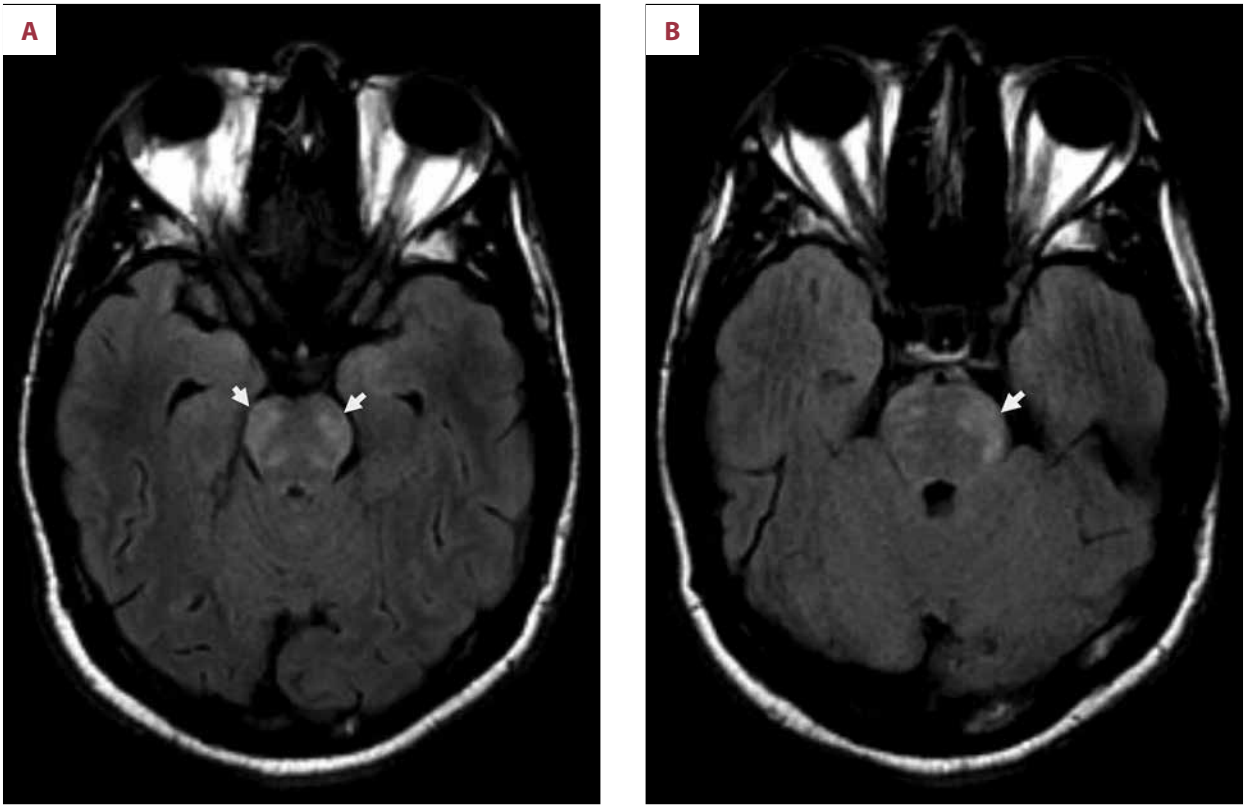


Figure 1. (**A, B**) Axial and coronal postgadolinium T1-weighted images, respectively, showing diffuse nodular as well as smooth basal meningeal enhancement around the midbrain and pons (arrows).



Figures 2. (**A, B**) Axial brain image showing increased T2 and FLAIR signal intensity in the brain stem parenchyma (mid-brain and pons, respectively) due to syphilitic meningitis (short arrows).

improvement gradually over couple days, with complete clinical recovery observed at 3 months. Antiretroviral therapy was initiated later in the infectious disease clinic after the patient was discharged. Repeated brain MRI after 2 months showed resolution of the initial abnormal image findings (Figure 4A, 4B).

Discussion

Symptomatic early neurosyphilis is a rare serious condition notable for its complex array of presentations, which is caused by the spirochete *Treponema pallidum* [1]. Neurosyphilis is almost unheard of in the United States in the era of penicillin treatment for syphilis, but reemerged in the 1980s among persons with HIV infection. In the developed world, the rate of syphilis declined rapidly with the widespread use of penicillin. In 2000, the incidence of syphilis reached its lowest point since the 1940s, at 2.1 cases per 100 000 population. The rates of syphilis have been increasing in the United States and other

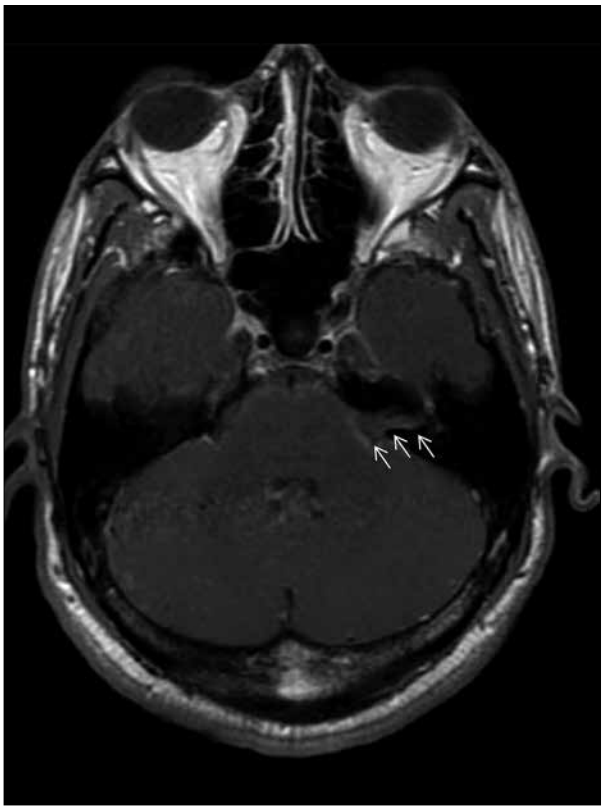
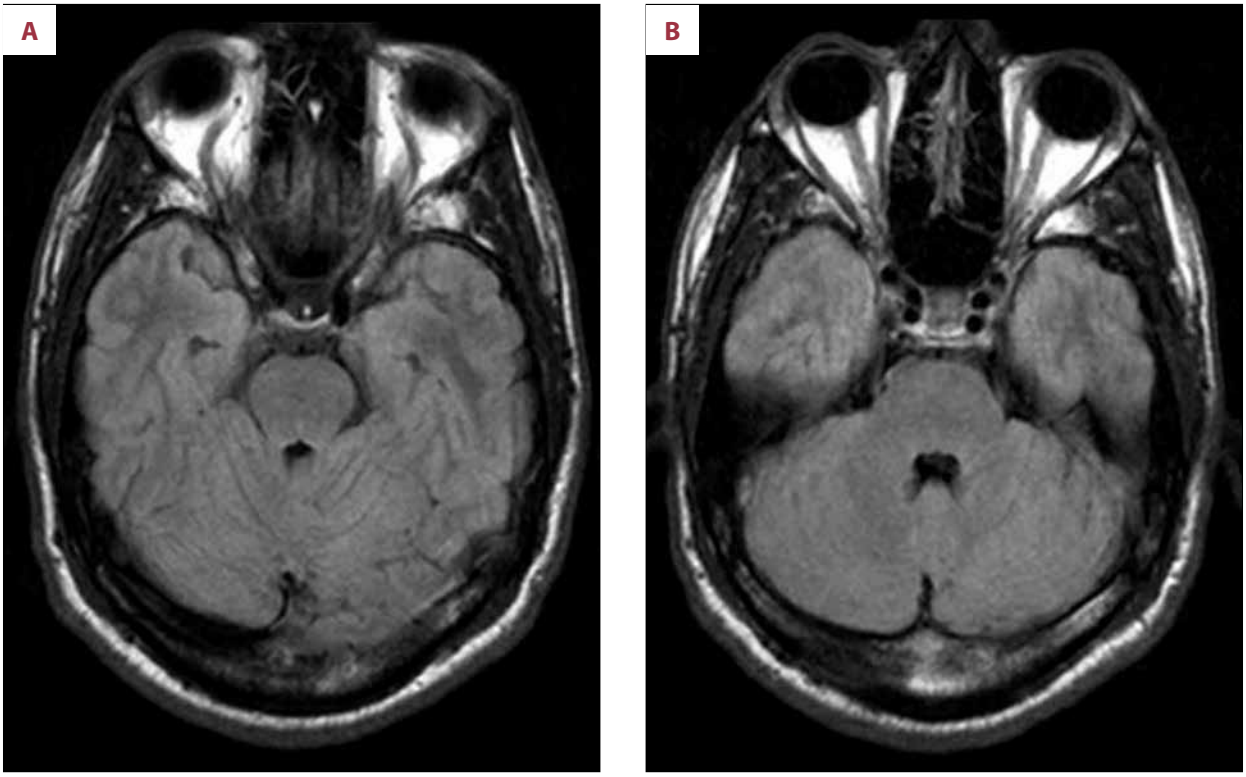


Figure 3. Axial postgadolinium T1-weighted image showing subtle enhancement of the cisternal segments of the facial and vestibulocochlear nerves complex in the left side (thin arrows). Such features are consistent with basal meningoencephalitis.



Figures 4. (**A, B**) Axial brain image showing complete resolution of previously increased T2 and FLAIR signal intensity in the brain stem parenchyma (mid-brain and pons, respectively) 2 months after initial presentation and after anti-syphilitic therapy.

Western countries, primarily among homosexual men. Although the incidence of syphilis in the United States declined by 89.7% during the 1990s and 2000s, the rate increased annually since then [2]. Traditionally, neurosyphilis is considered a form of late or tertiary syphilis, but CNS invasion by *Treponema pallidum* can occur at any time after the initial infection and might actually occur more often than was previously thought [3]; delayed recognition and treatment may result in irreversible sequelae [4]. Patients with concomitant HIV and syphilis often present differently than syphilis patients without HIV. They may present with more a fulminant picture, overlapping disease stages, multiple chancres, atypical skin rashes, and a more rapid progression to neurosyphilis [3]. Of note, syphilis has also been shown to negatively affect the immune status and viral load of patients with HIV [5]. Early neurosyphilis among HIV patients results in acute or sub-acute meningitis, cranial nerves palsy, and inflammatory vasculitis, leading to a cerebral infarction [6] or even intracranial hemorhhage [7]. HIV-infected patients with symptomatic early neurosyphilis are known to have variable clinical presentation; in a case series, approximately 75% of the patients had visual disturbances, ocular manifestation, and headache; cranial nerve palsy occurred in about 6%, 12% had acute meningitis, 4% developed acute stroke, and almost half of the patients were asymptomatic, and HIV infections were undiagnosed in nearly one-quarter of patients [6]. Herein, the image findings and patient presentation is consistent with syphilitic meningitis involving the basilar meninges resulting in basal meningoencephalitis and acute multiple cranial neuropathies as initial HIV manifestation. Such complex presentation or unexplained neurological symptoms in a young healthy patient should always raise the suspicion of neurosyphilis related to immune incompetence states such as undiagnosed HIV infection. All patients with suspected HIV infection or syphilis presenting with neurological symptoms should have brain MRI with contrast and undergo a lumber puncture with CSF analysis.

Conclusions

It is important to understand that neurosyphilis is still a significant disease, with complex neurological presentation. It is highly recommended to screen for neurosyphilis in patients with cranial nerves palsy or other unexplained neurological findings, with special consideration in HIV patients. Brain MRI with and without contrast and CSF studies are essential tools and very helpful in planning management. Early diagnosis and treatment of neurosyphilis is crucial due to potential persistent disabilities that can be easily treated or prevented.

Conflict of Interest Disclosures

None reported.

References:

1. Kamath M, Rizvi M, O'Nions J et al: A complex neurological presentation of syphilis. BMJ Case Rep, 2013; 2013. pii: bcr2013200309
2. Sadeghani K, Kallini JR, Khachemoune A: Neurosyphilis in a man with human immunodeficiency virus. J Clin Aesthet Dermatol, 2014; 7(8): 35–40
3. Kurtz SD, Rollin F: Ocular syphilis in a patient with HIV. JAAPA, 2014; 27(4): 32–35
4. Cassilde AL, Barnaud G, Baccar S et al: Sudden-onset bilateral deafness revealing early neurosyphilis. Eur Ann Otorhinolaryngol Head Neck Dis, 2014; pii: S1879-7296(14)00061-1
5. Jarzebowski W, Caumes E, Dupin N et al: Effect of early syphilis infection on plasma viral load and CD4 cell count in human immunodeficiency virus-infected men: results from the FHDH-ANRS CO4 cohort. Arch Intern Med, 2012; 172(16): 1237–43
6. Symptomatic Early Neurosyphilis Among HIV-Positive Men Who Have Sex with Men, Four Cities, United States, January 2002-June 2004, Centers for Disease Control and Prevention, Atlanta, GA. Available from: URL: *http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5625a1.htm*. Accessed July 1,2014
7. Koh M, Kashiwazaki D, Yamatani K et al: Intracranial hemorrhage caused by neurosyphilis: a case report. No Shinkei Geka, 2014; 42(8): 711–16