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Peripheral Facial Paralysis and Hearing Loss as Expression of Neurosyphilis

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Abstract

Syphilis is an infectious venereal disease caused by the spirochete *Treponema pallidum*. Neurosyphilis is a clinical form that can occur at any stage of the disease, presenting itself with signs and polymorphic neurological symptoms that may mimic multiple disorders of the central nervous system. Thus, it is included in the differential diagnosis of many diseases, justifying the need for description of different forms of otosyphilis. This study reports two patients showing a rare form of presentation, demonstrating an exclusive and simultaneous involvement of the VII and VIII cranial nerves and the membranous labyrinth, cochlea and semicircular canals, without other neurological manifestations.

Keywords: Neurosyphilis; *Treponema pallidum*

Abbrevations: BAEP: Brainstem Auditory Evoked Potential; VDRL: Venereal Disease Research Laboratory; MRI: Magnetic Resonance Imaging; FTAAbs: Fluorescent Treponemal antibody – Absorption; ELISA: Enzyme-Linked Immunosorbent Assay

Introduction

Syphilis is an infectious venereal disease caused by the spirochete *Treponema pallidum*. The major transmission route is through sexual contact, but can also be transmitted from mother to fetus during pregnancy or at birth, resulting in congenital syphilis [1]. Clinically, it is divided into primary, secondary or tertiary syphilis and each has its forms of clinical presentation. Neurosyphilis is a clinical form that can occur at any stage of the disease, presenting itself with signs and polymorphic neurological symptoms that may mimic multiple disorders of the central nervous system [2]. Thus, it is included in the differential diagnosis of many diseases, justifying the need for description of different forms of otosyphilis. This study reports two patients showing a rare form of presentation, demonstrating an exclusive and simultaneous involvement of the VII and VIII cranial nerves and the membranous labyrinth, cochlea and

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semicircular canals, without other neurological manifestations.

Case Reports

Case 1

44-year-old African-American male came to the hospital's ENT service with complaints of hearing loss, tinnitus and vertigo for about two months and House-Brackmann facial paralysis grade III on the left for eight days (Figure 1). He denied comorbidities or sexually transmitted diseases. In admission, it was performed an audiometry which showed left anacusis and absence of response on the brainstem auditory evoked potential (BAEP), on the left (Figure 2).

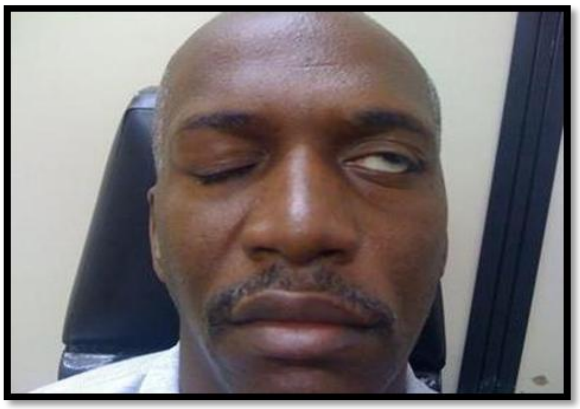

Figure 1: Patient with peripheral facial paralysis on the left.

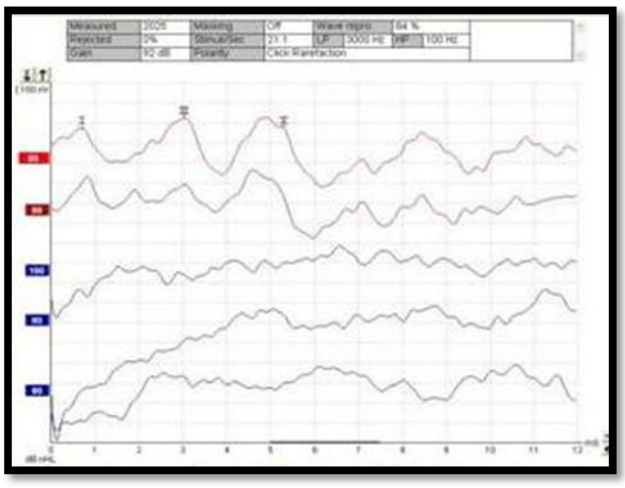

Figure 2: Brainstem Auditory Evoked Potential (BAEP), with no response on the left.

The only change in laboratory tests was a VDRL (Venereal Disease Research Laboratory) of peripheral blood 1:32. The brain MRI (magnetic resonance imaging) with emphasis on cerebellum angle showed a lesion with intense enhancement, after intravenous gadolinium injection, and thickening of the vestibulocochlear and facial nerves with impregnation of the membranous labyrinth of the cochlea and the semicircular canals to the left. A lumbar puncture was also asked, and showed an increase in cell at the expense of lymphocytes. The FTAAbs (fluorescent treponemal antibody - Absorption) and hemagglutination 1:16 were positive, reaching the diagnosis of neurosyphilis. All cultures and viral markers were negative.

The patient was admitted and submitted to therapy with crystalline penicillin, four million units every four hours for 14 days. A new MRI was performed after treatment and showed mild enhancement after the contrast injection involving the vestibulocochlear and facial nerves, showing an evolutionary improvement compared to the previous test. After treatment, the VDRL and FTAAbs in peripheral blood were negatives, and they have become non-reactive. There was a decrease of hemagglutination to 1:4, in the cerebrospinal fluid. The FTAAbs and ELISA remained reactants as expected. The patient was considered cured of neurosyphilis, but still presents anacusis and peripheral facial paralysis with slight improvement on the left. He continues the treatment as an outpatient in our service.

Case 2

34-year-old Caucasian male was admitted to the hospital's ER complaining of decrease in bilateral hearing acuity and facial paralysis on the right (Grade II House-Brackmann). The patient presented mild papular rash on thoracic, abdominal and palmar regions. The admission MRI was suggestive of neuritis of the facial and bilateral cochlear nerves (Figure 3), but the facial nerve left function was preserved and there weren´t other neurological complaints. VDRL were asked (1/1256 reagent) so as positive anti-HIV. In admission, an audiometry was performed and showed a profound bilateral sensorineural hearing loss.

It was also asked for a lumbar puncture which showed an increase in cell at the expense of lymphocytes, with positive VDRL at 1:32. The viruses serology and fungal research were negative, reaching the diagnosis of neurosyphilis.

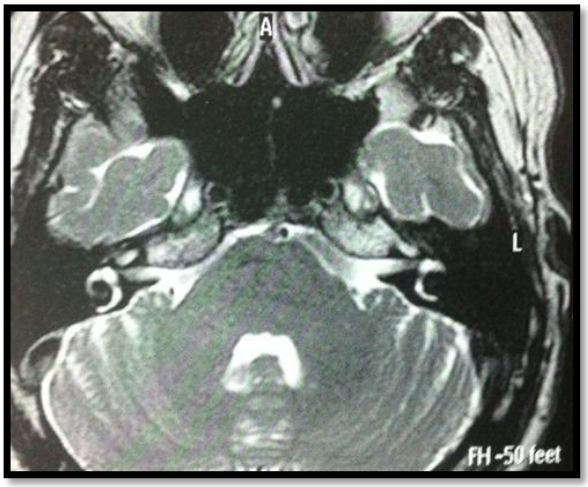

Figure 3: Thickening and impregnation with paramagnetic contrast of the cisternal portion and in the internal auditory canal of the facial nerves and vestibule-cochlear showing bilaterally inflammatory aspect (neuritis).

The patient had positive serology for HIV with CD4 T lymphocytes of 339 / mm 3 and viral load 82894CP / mL and 4.92 log. The patient was admitted and submitted to therapy with crystalline penicillin, four million units every four hours for 14 days. He remains in attendance in our service. The patient improved the facial paralysis and hearing, showing a regular BAEP threshold on the right and electrophysiological threshold near 60 dBnHL on the left (Figure 4).

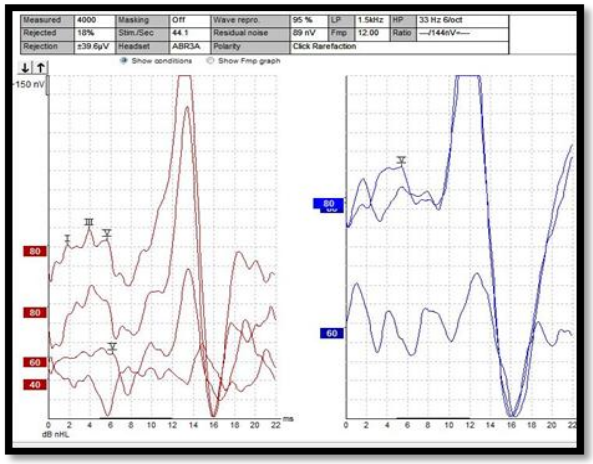

Figure 4: A regular BAEP threshold on the right and electrophysiological threshold near 60 dBnHL on the left.

Discussion

Neurosyphilis classification is extremely complex, not only because of taking multifaceted clinical conditions, but also because it is characterized by various histopathological aspects and the involvement of different anatomical areas. Roughly we can classify it in asymptomatic, meningeal, parenchymal and gummatous. However, this rigid classification does not correspond to clinical reality; in fact there are significant intersections of neurological disorders.

Thus, the most frequent neurosyphilis forms are the acute syphilitic meningitis, the meningovascular and the parenchymal. In acute meningeal form the incubation period is less than one year in most patients, and in 25% of cases, is the first manifestation of syphilis. The main neurological changes include cranial nerves' injury, particularly the II, VI, VII and VIII, and signs of intracranial hypertension. The sensorineural deafness, initially involving only high frequencies, occurs in 20% of cases. Acute syphilitic hydrocephalus's show clinical manifestations of intracranial hypertension and usually occur 3-7 months after the primary infection. Clinically, syphilitic meningitis usually manifests by headache, nausea, vomiting and stiff neck. The inflammatory process may also affect the ependyma and cerebral vessels, leading endarteritis, vascular occlusion and cerebral infarction, with consequent focal neurological signs such as aphasia and hemiplegia [3,4].

This study presents two rare cases of neurosyphilis with simultaneous impairment of the seventh and eighth cranial nerves. Yimtae et al. [5] reported 85 cases of otosyphilis in 16 years (1984-2000), the most common clinical manifestations were hearing loss (90.6% of patients), tinnitus (72.9% of patients) and dizziness (52.9% of patients). The paper did not report the presence of facial paralysis. Both forms of syphilis, acquired and congenital, can cause cochlea vestibular dysfunction [6], and sensorineural hearing loss is reported in late secondary and forms of syphilis [7,8]. The incidence of hearing loss in acquired syphilis is 17% in primary syphilis, 25% in late latent syphilis and 54% in tertiary syphilis [7-9]. There are few reports of facial paralysis in syphilis [10-12].

The diagnosis was possible due to increased gadolinium enhancement of contrast to MRI of the affected cranial nerves, the membranous labyrinth, the cochlea and the semicircular canals. Probably, this happens because of a discontinuation in the secondary brain barrier inflammation and the infection spread by perilymphatic and endolymphatic fluids. Thus, a wide range of imaging

findings can be seen in neurosyphilis. Many cases of clinical neurosyphilis have no imaging abnormalities. The highlight of the vestibular nerve and the labyrinth has been described in patients with syphilis labyrinthitis. However, the simultaneous engagement of the seventh and eighth pairs is rare [13-15]. The image combined with a high clinical suspicion and examinations with positive markers in cerebrospinal fluid and peripheral blood allowed the diagnosis and treatment of this rare form of Neurosyphilis.

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