

## Isolated otosyphilis; a case report

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### Abstract

Syphilis is a systemic disease and its presentations can be altered with wide use of antibiotics and HIV co-infection, particularly neurosyphilis. Involvement of eighth cranial nerve is recognized as a part of meningeal involvement as well as an isolated phenomenon. Isolated otosyphilis is potentially reversible with early antibiotic treatment. Clinician should suspect otosyphilis in the presence of vestibular cochlear involvement in patients with syphilis.

The definite diagnosis of isolated otosyphilis is challenging and it should be made on the basis of a typical clinical presentation and positive serological test results for syphilis. Over diagnosis is justifiable as this is a potentially reversible cause of hearing loss with early treatment.

**Key words:** Syphilis, Neurosyphilis, Otosyphilis, Sri Lanka, case report

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### Full article

#### Introduction

Syphilis is a systemic disease and its presentations can be altered with wide use of antibiotics and in HIV co-infection; this is particularly true in neurosyphilis. Involvement of eighth cranial nerve is recognized as a part of meningeal involvement as well as an isolated phenomenon. Isolated otosyphilis is potentially reversible with early antibiotic treatment. Clinician should suspect otosyphilis in the presence of vestibular cochlear involvement in patients with syphilis.

#### Case Report

Mr. A, 30-year-old male was found to have both HIV and hepatitis C infections. Examination was unremarkable at the STD clinic, Colombo. His baseline CD4 count was 464 cells/ $\mu$ l and had chronic Hepatitis C

infection with genotype 3a. Serum VDRL and TPPA were negative. He was assessed by a gastroenterologist and decided to start treatment for hepatitis C infection.

Six months later he presented to the clinic with a non-itchy skin rash and low grade fever of 5-day duration. During this six-month period, he admitted having unprotected sexual exposures with male partners. Examination revealed macular papular rash involving both palms and soles and generalized lymphadenopathy with normal central nervous system (CNS) examination. A diagnosis of secondary syphilis was made with a VDRL titre of 1:64 and a positive TPPA test. He was treated with single dose of benzathine penicillin 2.4 million units intramuscularly.

Initially he had adequate clinical and serological response to treatment but suddenly complained of some auditory and vestibular symptoms like dizziness, vertigo, tinnitus, and left sided hearing impairment after three months of benzathine penicillin injection. Examination of the CNS revealed no abnormality other than left sided hearing impairment. The CT and MRI scan of brain were normal and specialist ENT opinion was obtained. Audiogram showed moderate mixed hearing loss of left ear and minimal sensorineural hearing loss in right ear. The diagnosis of Meniere's disease was made and he was treated with vestibular sedatives.

However, a rash suggestive of secondary syphilis appeared again within few days. Investigations revealed that his VDRL titre was 1:128 which was four fold higher than the last follow up VDRL titre. Considering the hearing loss in the presence of clinical and laboratory evidence of syphilis, lumbar puncture was performed. Examination of cerebrospinal fluid (CSF) revealed following; WBC - nil, RBC - nil, protein - 20 mg/dl, sugar - 4.3 mmol /dl (RBS - 6.6 mmol/dl), CSF-VDRL - Weakly reactive, CSF-TPPA - positive, culture - sterile and acid-fast bacilli (AFB) - negative. He was treated with intravenous aqueous crystalline penicillin 4 million units 4 hourly for 14 days under steroid cover in the first 3 days. Vertigo was improved completely following treatment and hearing loss too, improved to some degree.

### Discussion

Syphilis is a systemic disease caused by *Treponema pallidum* subsp. *pallidum* and is still common in most countries. Central nervous system involvement can occur during any stage of syphilis, and CSF abnormalities are common with CNS involvement (1).

As the incidence of neurosyphilis has continued to decline in the antibiotic era, atypical presentations have been noted. Further, importantly clinical presentation can be altered by concomitant presence of HIV infection (1). Atypical and rare forms of presentations need to

be considered by Venereologists especially in this era.

Neurosyphilis often detected in persons whose HIV infection has not yet progressed to an advanced stage as AIDS (1). Ootosyphilis is a rare cause of sensorineural hearing loss and dizziness, and is important for clinicians to consider early because the hearing loss is potentially reversible with early diagnosis and treatment. Many experts believe Venereologists need to be more vigilant on this issue.

Cochlear vestibular symptoms of neurosyphilis can result via two main mechanisms based on published literature. First, as a part of acute syphilitic meningitis with abnormal CSF findings. In this situation, hearing loss is usually accompanied by other neurological deficits and findings on cerebrospinal fluid examination will usually be abnormal. Second, and more commonly, without any other features of coexisting neurosyphilis and CSF parameters are usually found to be normal in this situation. Cochlear vestibular symptoms are thought to result from direct damage to the vestibule-cochlear apparatus (2).

Sudden onset of vestibular cochlear symptoms, rapid rise in VDRL titre and nearly normal CSF findings (weakly reactive CSF-VDRL would have been due to a subjective reading in the absence of cells) were more in favour of the diagnosis of isolated otosyphilis in this patient. Definitive diagnosis of isolated otosyphilis is difficult as it needs inner ear fluid evaluation (3). This patient had normal MRI scan and audiogram confirmed the asymmetrical neurological involvement with evidence of sensorineural defect.

The definitive diagnosis of isolated otosyphilis is challenging and clinical diagnosis should be made on the basis of a typical clinical presentation and positive serological test results for syphilis. Obviously this approach leads to over diagnosis of cases but useful as otosyphilis is a potentially reversible cause of hearing loss (2).

The optimal treatment for otosyphilis is not established and the published data is limited. However, all standard guidelines recommend intravenous crystalline penicillin for 10 - 14 days for patients with secondary syphilis and neurological involvement as for neurosyphilis. Very few studies showed effectiveness of doxycycline as an alternative. This patient was treated with IV crystalline penicillin for 14 days with pre-treatment adjuvant steroids.

Patient showed marked improvement following the treatment and his hearing improved greatly while symptoms of vertigo and unsteadiness disappeared completely. Unfortunately, we could not perform follow up audiogram which is the objective way of confirming the recovery as the patient went back to his home country.

There were no previous reported cases of otosyphilis among HIV infected patients in Sri

Lanka. According to the available evidence, early treatment carries better prognosis and in general, improvement of cochlear symptoms following treatment is much evident than hearing improvement. Fine clinical alertness is very important in dealing with patients with syphilis and cochlear vestibular symptoms.

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