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AJNR Am J Neuroradiol 2000, 21 (9) 1673-1675

<http://www.ajnr.org/content/21/9/1673>

This information is current as
of February 27, 2024.

Neurosyphilis as a Cause of Facial and Vestibulocochlear Nerve Dysfunction: MR Imaging Features

Michelle M. Smith and James C. Anderson

Summary: The prevalence of syphilis increased for several decades before the mid-1990s in the United States, particularly in the southern states. We report a case of neurosyphilis causing bilateral facial and vestibulocochlear nerve dysfunction in which the diagnosis was not initially suspected based on the patient's demographics and history. The MR imaging features helped to make the diagnosis in this case and to exclude other possible causes of multiple cranial nerve dysfunction in this patient. Hearing loss associated with neurosyphilis is one of the few treatable forms of progressive hearing loss, and it is essential that a diagnosis of neurosyphilis be made expeditiously.

Neurosyphilis is difficult to diagnose because of its variable time of onset and nonspecific clinical presentation. According to the Centers for Disease Control, in 1995, more than 68,000 cases of syphilis were detected in adults; the highest syphilis rates were reported in eight southern states (1). Subsequently, the prevalence of the disease declined; however, cases remain concentrated in the southern United States (2). Neurosyphilis may present as acute meningitis with cranial nerve dysfunction. We present the clinical and MR imaging findings in a case of neurosyphilis causing facial and vestibulocochlear nerve dysfunction. A high index of suspicion and knowledge of the imaging features are needed to make a diagnosis of neurosyphilis, and hearing loss associated with neurosyphilis can be treated (3, 4).

Case Report

A 51-year-old woman residing in Tennessee developed mild vertigo approximately 6 weeks before admission. The patient was able to conduct normal activities until the vertigo worsened 2 days before admission. She also noted progressive bilateral hearing loss, bilateral facial weakness, and severe headache. No history of sexually transmitted disease was elicited at the time of admission. A physical examination at the time of admission revealed left facial nerve paralysis and right facial weakness. Audiograms revealed severe-to-profound sensorineural hearing loss on the left and severe sensorineural hearing

loss on the right. The Distortion Product Otoacoustic Emissions test, which is a test of cochlear function, revealed bilateral cochlear dysfunction. Laboratory values on a lumbar puncture revealed 553 nucleated cells, 90% of which were lymphocytes and few of which were red cells. Glucose was 46 (normal range, 45-75), and protein was 236 (normal range, 15-45). Cryptococcal antigen, India ink capsule stain, serum rheumatoid, ANCA, S-PEP, thyroid-stimulating hormone, angiotensin-converting enzyme level, routine complete blood count, and comprehensive metabolic profiles were unremarkable. The Venereal Disease Research Laboratories (VDRL) CSF test and polymerase chain reaction for herpes, *Treponema*, and Lyme disease serologies were performed. The VDRL CSF result was 4+ positive. The remaining serologies were negative.

An MR imaging examination was performed on a 1.5-T MR unit. Transverse and coronal T1-weighted spin-echo images with a thickness of 3 mm were obtained before and after the administration of contrast medium. The T1-weighted imaging parameters were 450/15/4 (TR/TE/excitations). Axial T2-weighted turbo spin-echo images with a thickness of 5 mm and parameters of 5000/95/1 of the whole brain were also obtained.

Enhancement of the intracanalicular, labyrinthine, geniculate, and tympanic segments of both seventh nerves, as well as the cisternal segment of the right seventh and eighth cranial nerve complex, was present (Fig 1). Enhancement of the intracanalicular portions of the vestibulocochlear nerves, the cochleae, and the meninges lining the internal auditory canals was noted bilaterally (Fig 2). No abnormal enhancement or T2 signal was noted within the brain parenchyma, and no meningeal enhancement other than that within the internal auditory canals was present.

Penicillin G therapy, IV administered at a rate of 4 million units every 4 hours, was instituted. Within 1 week, the patient had improvement in hearing and facial nerve function. The patient was discharged to home and received 14 additional days of IV administered antibiotic therapy. Complete recovery of facial and vestibulocochlear nerve function was noted at the 1-month follow-up examination.

Discussion

Syphilis is a sexually transmitted disease caused by the spirochete bacterium *Treponema pallidum*. The prevalence of syphilis increased in the United States for several decades before the mid-1990s. The increase was secondary to the rise in unprotected homosexual activity and the increase in cases of AIDS. In 1995, the Centers for Disease Control reported that 84% of the counties in the United States with the highest syphilis rates were located in the southeast. Tennessee, Mississippi, and Maryland reported the highest rates, ranging from 13.5 to 15.4 cases per 100,000 (1). Higher rates of both syphilis and HIV infection are reported in African-

Received February 17, 2000; accepted after revision April 13.

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Presented at the 38th Annual Meeting of the American Society of Neuroradiology, Atlanta, GA, 2000.

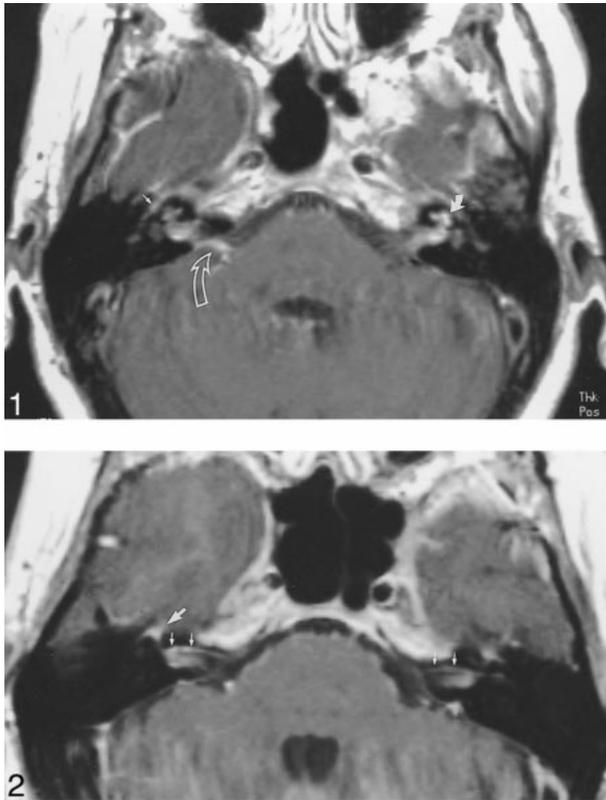


FIG 1. Axial T1-weighted MR image (450/15/4), obtained after the administration of contrast medium, shows enhancement within the cisternal segment of the vestibulocochlear nerve complex on the right (*curved open arrow*), within both internal auditory canals, within the left cochlea (*curved solid arrow*), and within the tympanic portion of the right facial nerve (*small arrow*). The enhancement within the internal auditory canals involves both the nerves within the CSF and the meninges lining the canal. Enhancement is also seen within the middle turn of the right cochlea.

FIG 2. Axial T1-weighted MR image (450/15/4), obtained after the administration of contrast medium from a position that is slightly superior to that of Figure 1, shows enhancement of the labyrinthine and geniculate portions of the right facial nerve (*large arrow*) and enhancement of the meninges lining both internal auditory canals (*small arrows*). Enhancement of the intracanalicular segments of the seventh and eighth cranial nerve complex is appreciated bilaterally.

Americans. The increased prevalence of syphilis in the southeastern United States is thought to reflect the larger African-American population existing in that region compared with the northern and western regions of the country. The rates of primary and secondary syphilis have declined in the United States since 1993; however, the disease remains geographically concentrated in the southern United States (2).

The infectious course of syphilis has three stages. The primary stage is characterized by the presence of a chancre, a round painless sore or ulceration at the inoculation site, and regional adenopathy. If untreated, within 2 to 4 weeks, hematologic dissemination and the secondary form of syphilis occur. Clinically, this manifests as a rash on the palms of the hands and soles of the feet.

CNS involvement can occur in the secondary stage and may produce aseptic meningitis. Progression to tertiary syphilis occurs after the secondary symptoms disappear. Tertiary syphilis usually does not present until 5 years or more after the primary infection. The bacterium damages organ systems, including the CNS, cardiovascular system, bones, joints, skin, and mucous membranes.

The CNS may become involved in any stage of the disease from weeks to years after initial infection. Neurosyphilis is seen primarily in the tertiary and occasionally in the secondary stages of the disease. Most patients are asymptomatic (3). The major forms of clinically symptomatic infection are acute syphilitic meningitis, meningovascular syphilis, and parenchymal neurosyphilis (4). The neurologic symptoms produced and the temporal relationship between the primary infection and the onset of symptoms vary between the forms of the disease, and mixed features are common.

The parenchymatous type of neurosyphilis is well known and results in general paresis secondary to widespread parenchymal damage and tabes dorsalis secondary to demyelination of the posterior columns, dorsal roots, and dorsal root ganglia. A more fulminant form of parenchymal disease, referred to as *necrotizing neurosyphilis*, has been reported to occur in patients with HIV (5). Meningovascular neurosyphilis is less frequent and may produce focal neurologic deficits or global CNS dysfunction and occurs as one of several subtypes. Meningovascular syphilis may also present as a subacute illness or stroke syndrome because of small vessel arteritis (4). The symptoms appear from months to a decade after the primary infection and may include seizures, hemiplegia, insomnia, personality changes, and dementia. Acute syphilitic meningitis typically presents with headache, meningeal irritation, and confusion. Cranial nerve palsies are also common, and patients become symptomatic within 2 years of primary infection. The symptoms and clinical presentation of the acute meningitic and meningovascular forms of neurosyphilis can be similar (6).

The symptoms and imaging findings in our patient were most consistent with those seen in acute syphilitic meningitis involving the basilar meninges. Meningeal irritation results in cranial neuropathies because the meningeal disease extends along the nerves at the base of the brain. Cranial nerves seven and eight are most frequently involved in syphilitic basilar meningitis (3). Involvement of the optic, oculomotor, and abducens nerves has been reported (7).

A wide range of imaging findings may be seen in neurosyphilis. Many cases of clinical neurosyphilis have no imaging abnormalities. In a 1989 series of HIV-positive Haitian men with neurosyphilis, one third had normal results of their imaging studies (5). In a 1997 study of 35 patients with documented neurosyphilis, 31% had no abnormalities detected on their CT scans and MR images (7).

Imaging findings in the parenchymal form of neurosyphilis may include atrophy, cerebral infarction, white matter lesions, and enhancing nodules referred to as *gummas*. Meningovascular syphilis may give rise to cerebral infarction, and the diagnosis should be considered in young patients presenting with stroke who are HIV-positive. The meningovascular and acute meningitic forms of neurosyphilis may produce a meningoneuritis, meningolabyrinthitis, or labyrinthitis (3). Enhancement of the leptomeninges and cranial nerves may be seen with contrast-enhanced MR imaging. Enhancement of the vestibulocochlear nerve and labyrinth has been described in patients with luetic labyrinthitis; however, facial nerve involvement in addition to eighth nerve involvement is rare. Enhancement of the meninges and membranous labyrinth is likely secondary to a breakdown of the blood-brain barrier secondary to inflammation with a concentration of contrast medium within the abnormal membranes (8). The enhancement of the cochlea in our patient, indicative of luetic labyrinthitis, may be secondary to a spread of infection via the endolymphatic or perilymphatic fluids. Hematogenous dissemination to the cochlea and spread of infection via the intracanalicular portion of the eighth cranial nerve to the cochlea are also modes of transmission to be considered.

Facial nerve dysfunction in association with vestibulocochlear dysfunction helped to localize the pathologic process in our patient to the cerebellopontine angle and/or temporal bone. The imaging features in our patient included enhancement of the seventh and eighth cranial nerves and cochlea bilaterally as well as the meninges lining the internal auditory canals. No parenchymal abnormalities within the brain were noted. Cranial nerve enhancement may be seen on MR images in a broad range of pathologic entities, including neoplastic, granulomatous, infectious, posttraumatic, and idiopathic disorders. The involvement of both the cranial nerves and the meninges, as well as the bilateral nature of the abnormalities, made primary neoplastic causes unlikely in our patient. Secondary neoplastic processes, such as leptomeningeal carcinomatosis and lymphoma, were considered; however, the patient had no history of malignancy. In addition, the meningeal enhancement was limited to the region of the temporal bone without other sites of leptomeningeal involvement. Head trauma, previous radiation, and chemotherapy may produce labyrinthitis; however, this patient had no such history. Granulomatous meningitis secondary to sarcoidosis may involve the leptomeninges and produce cranial neuropathies. Leptomeningeal sarcoidosis frequently involves the optic chiasm and infundibulum, and there was no such involvement in our case.

Infectious causes, including viruses such as herpes simplex virus, varicella, and herpes zoster, were considered less likely in this patient, although these viruses may cause facial paralysis, sensorineural hearing loss, and vertigo. No vesicles were identified

within the external auditory canal in this patient, and otalgia was not present, making herpes zoster infection (Ramsey-Hunt syndrome) unlikely. In addition, it would be unlikely that meningeal or internal auditory canal enhancement would be seen in association with Ramsey-Hunt syndrome. Cytomegalovirus infection or cerebritis may cause a similar appearance in patients with AIDS; however, this patient was HIV-negative. Bilateral facial nerve paralysis has been reported in association with Lyme disease (7); however, no history of tick bite was elicited and Lyme serology was negative in this case. Tuberculous meningitis and fungal infection may cause labyrinthitis; however, it is usually isolated to the middle and inner ear and internal auditory canal involvement is unusual. The positive result of the VDRL CSF test and rapid response to IV administered penicillin G therapy confirmed the diagnosis of neurosyphilis in this patient, despite that a history of sexually transmitted disease could not be elicited from the patient. Although the VDRL CSF examination has a high specificity of 94%, the reported sensitivity of the test for diagnosing neurosyphilis is only 27% (9).

Conclusion

The prevalence of syphilis has increased within the past several decades in the United States, particularly in the southeast. Neurosyphilis may occur in the secondary and tertiary forms of the infection. The acute meningitic and meningovascular forms of neurosyphilis may produce cranial nerve dysfunction. Involvement of the seventh and eighth cranial nerves results in symptoms of facial paralysis, sensorineural hearing loss, and vertigo. A high index of suspicion and knowledge of the MR imaging features is necessary to diagnose this highly treatable cause of progressive hearing loss and facial paralysis.

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