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Cochlear Implantation of a Patient with Definitive Neurosarcoidosis

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

Keywords

neurosarcoidosis, systemic sarcoid, retrocochlear lesion, cochlear implant, bilateral sensorineural hearing loss, internal auditory canal enhancement

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Noncaseating epithelioid granulomas are found in the central nervous system in 5% to 26% of patients diagnosed with systemic sarcoidosis. The most common presentation of central nervous system sarcoidosis (neurosarcoidosis) is cranial neuropathy, followed by meningeal disease, including aseptic meningitis and mass lesions.[1]

Neurosarcoidosis has a predilection for basal meninges that surround the cranial nerves with infiltrative, perivascular granulomas. In the case of cranial nerve involvement, eighth nerve symptoms occur in up to 20% of patients and are associated with other cranial nerve neuropathies or overt systemic disease.[1,2] The incidence of sensorineural hearing loss (SNHL) is only 5% to 9% among those diagnosed with neurosarcoidosis.[2] The likely mechanism of injury is vasculitis that leads to transient ischemia and neural damage.[3] Approximately 70% of patients will recover at least some hearing, either spontaneously or with corticosteroid therapy.[2] Progression to profound hearing loss is exceedingly rare. Consequently, little is known about cochlear implantation as a rehabilitation option in this group of patients.

Case Report

A 54-year old man presented to the otology clinic with sudden left moderate SNHL (speech reception threshold [SRT], 50 dB; speech discrimination score [SDS], 60%) and intermittent dizziness. He was treated with oral and intratympanic corticosteroids without improvement. Four months later, the hearing on the left declined to severe and unaidable (SRT, 80 dB; SDS, 18%); that on the right progressed from normal (SRT, 5 dB; SDS, 100%) to mild (SRT, 25 dB; SDS, 86%). Magnetic resonance imaging (MRI) demonstrated bilateral enhancing lesions of the internal auditory canal (**Figure 1**, left). Eighteen months after presentation, the hearing on the left fluctuated but did not recover, and the hearing on the right deteriorated to severe and unaidable (SRT, 70 dB; SDS, 8%). Subsequent MRI demonstrated the internal auditory canal lesions fluctuating in size and not correlating to the degree of SNHL. During this time, the patient was seen by multiple specialists, and the workup yielded negative results, including serum angiotensin-converting enzyme (ACE) levels, Lyme titers, and antinuclear cytoplasmic antibodies. His chest radiograph finding was negative for sarcoidosis. In addition to corticosteroid therapy, he was treated with etanercept and valacyclovir. Despite medical treatment, his hearing did not recover, and cochlear implantation was performed on the left, followed by the right 2 months later. Postoperative consonant-nucleus-consonant word scores were >88%.

Six months after cochlear implantation, the patient developed neurologic symptoms (jaw chatter, oscillopsia, cognitive dysfunction) discovered to be complex partial seizures, culminating in a grand mal seizure. MRI revealed a new enhancing left hippocampal lesion. Lumbar puncture was negative for inflammatory or infectious processes, including antineuronal antibodies. Cerebrospinal fluid ACE level was normal. Despite treatment with acyclovir for presumed viral encephalitis, the hippocampal lesion continued to grow (**Figure 1**, right). An image-guided

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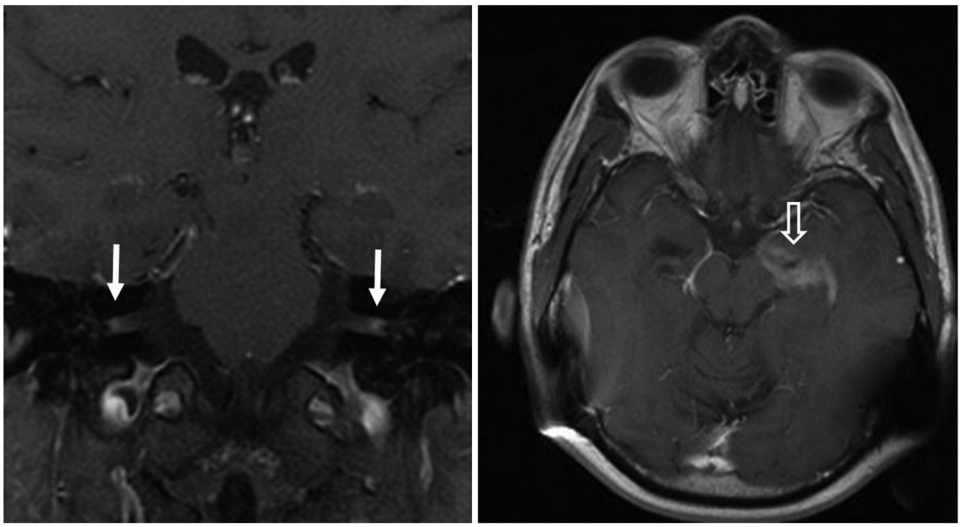

Figure 1. Magnetic resonance imaging: left, coronal T1 gadolinium sequence shows enhancing lesions in the distal internal auditory canals (arrows); right, axial T1 gadolinium sequence shows an enhancing lesion of the left hippocampus (open arrow).

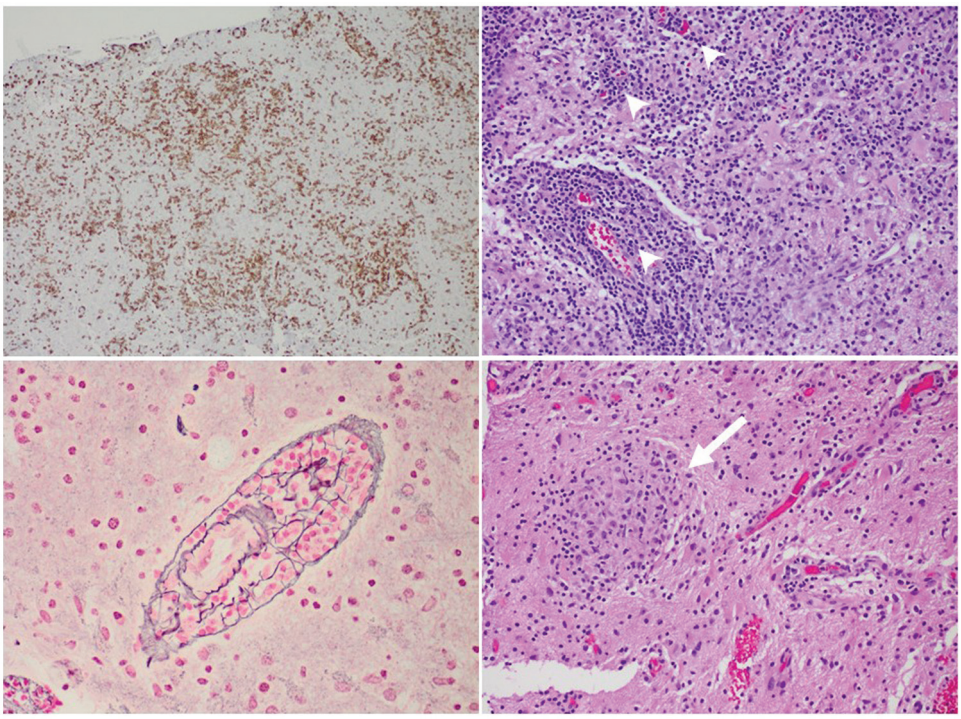

Figure 2. Top left: CD3+ stain demonstrating a T-cell lymphocytic infiltrate. Hematoxylin and eosin stains show a noncaseating granuloma (lower right, arrow) and perivascular infiltrates (upper right, arrowheads). Bottom left: reticulin stain highlighting reticulin surrounding and permeating the histiocytic cluster.

biopsy was obtained through the temporal gyrus approach. The abnormal tissue appeared friable and slightly hemorrhagic.

Pathologic examination revealed nonnecrotizing granulomas with a perivascular lymphocytic infiltrate within the brain parenchyma, most consistent with neurosarcoidosis (**Figure 2**). Nationally recognized advanced diagnostic laboratories analyzed the specimens, excluded a lymphoproliferative process, and did not identify a causative infectious agent. Two years later, in addition to seizures, the patient continues to suffer further neurologic manifestations of sarcoid, including dysphonia, dysphagia, dysarthria, worsening ambulation, and some cognitive decline despite immunosuppressive therapy.

Discussion

The typical clinical course of neurosarcoid-associated SNHL is an asymmetric sudden or rapidly progressive mild

to moderate loss, fluctuating and involving both ears.[2] In rare cases of profound loss, patients may demonstrate radiologic abnormalities, such as inflammatory lesions along the cochlear nerve, labyrinthine enhancements, and even cochlear ossification.[4] While definitive diagnosis of neurosarcoid depends on a central nervous system biopsy, for most patients the diagnosis of probable neurosarcoidosis is obtained by a combination of radiologic findings, abnormal cerebrospinal fluid profile, biopsy of extraneural sites, chest radiography, and serum ACE levels.[5] With SNHL, most patients experience concurrent neurologic symptoms, and half the patients will have associated cranial nerve neuropathies.[1-3] Successful cochlear implantations have been reported in only a handful of cases with probable neurosarcoidosis. Our case report is the first to describe implantation in definitive neurosarcoid confirming that cochlear implants remain a viable option and should be considered for patients with unaidable, medically unresponsive disease despite retrocochlear or intralabyrinthine lesions.[1,4]

Authors' Note

The presentation of this case was conducted in accordance with the NYU School of Medicine Institutional Review Board policies, available at https://med.nyu.edu/research/research-resources/clinical-research/sites/default/files/nyu-som-irb-policies-and-procedures-for-human-subjects-research-protection.pdf.

Author Contributions

Maja Svrakic, concept and design, interpretation of data, drafting, final approval, accountability for all aspects of the work; **John G. Golfinos**, acquisition of data, critical revision, final approval, accountability for all aspects of the work; **David Zagzag**, acquisition of data, interpretation of data, critical revision, final approval, accountability for all aspects of the work; **J. Thomas Roland Jr**, concept and design, acquisition of data, critical revision, final approval, accountability for all aspects of the work.

Disclosures

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