


# A 15-Year-Old Boy With Facial Palsy and Progressive Hearing Loss

Global Pediatric Health  
Volume 3: 1–3  
© The Author(s) 2016  
Reprints and permissions:  
sagepub.com/journalsPermissions.nav  
DOI: 10.1177/2333794X16658575  
gph.sagepub.com  


Paola America Huaynate-Cuadrado, MD<sup>1</sup>,  
Janet M. Poulik, MD<sup>1</sup>, and Jocelyn Yu Ang, MD<sup>1</sup>

Received June 1, 2016. Accepted for publication June 14, 2016.

## Case Report

A 15-year-old previously healthy African American male developed peripheral facial palsy over the course of 10 days. His primary care physician prescribed steroids and acyclovir to which he initially showed improvement, which lasted for about a month. After the facial palsy was resolved, he started complaining of unilateral, intermittent headaches and progressive right-sided hearing loss that worsened over a month associated to dizziness. He was taken to the emergency department.

## Hospital Course

On his initial admission, a brain magnetic resonance imaging (MRI) was performed, which showed right labyrinthitis and bilateral neuritis of cranial nerves, dural enhancement consistent with meningitis, and partial opacification of the mastoid air cells with right side predominance (Figure 1A-C). A spinal tap was performed and revealed no evidence of bacterial or viral meningitis.

The patient was admitted to the Infectious Disease service for a workup. His hearing loss had progressed to the left side as well and was observed to be mostly sensorineural. Blood and cerebrospinal fluid (CSF) cultures for bacterial, fungal, mycobacterial, and viral microorganisms were performed; however, no growth was shown. Serology was reactive for Epstein-Barr virus and varicella zoster virus, showing a resolved infection and previous immunization, respectively. Mycoplasma IgM also was positive. He completed 5 days of azithromycin and 7 days of acyclovir followed by 7 days of valacyclovir. A rheumatologic workup was performed showing the following: antinuclear antibody, weakly positive, with a 1:80 titer for both homogeneous and speckled pattern; anti-dsDNA antibody, negative; rheumatoid factor, 2.1 IU/mL (reference range = 0.0-6.0 IU/mL); extractable nuclear antigen screen, negative; angiotensin-converting enzyme (ACE), 106 Units/L (reference range = 18-101 Units/L). A high-resolution thorax computed tomography was normal. Full sequencing of the NOD2 gene was negative for the mutation associated with Blau's syndrome. The patient was

started on systemic steroids and was discharged. He continued to follow with ENT and was not seen by other subspecialties.

Over the course of 2 months, ENT continued with the steroids, but the hearing loss and the facial palsy worsened when a taper was attempted. Audiograms from this period showed profound bilateral sensorineural hearing loss, and because of the clinical progression the child was admitted to the Infectious Disease service. CSF and blood studies were repeated, and for the first time, fluid from the right and left middle ear was sent for serology and culture. All the cultures remained sterile. Polymerase chain reaction (PCR) for herpes simplex virus (HSV) 1 and 2 from the right middle ear came back positive, for which intravenous acyclovir was given with a total duration of 21 days. The laboratory would later inform that the sensitivity and specificity of PCR for HSV had not been determined for middle ear fluid. Intravenous steroids were administered as well as 2 doses of intravenous immunoglobulin therapy. In addition, 2 doses of intratympanic steroid were administered on the right side by a neurotologist consultant. Repeat audiograms showed no improvement from the previous audiograms. ACE level was 47 Units/L (reference range = 18-101 Units/L). After completion of the 21-day course of acyclovir, a repeat MRI showed unchanged inflammation of the seventh and eighth cranial nerves, continued bilateral labyrinthitis, and partial opacification of the right mastoid air cells without meningeal enhancement (Figure 2).

The patient was discharged on a steroid taper course and continued with the intratympanic steroid injections as an outpatient with no improvement on his hearing loss. Over a course of 3 months after his discharge, his facial palsy was noted to have improved almost completely, and he was scheduled for a right cochlear implant. During the

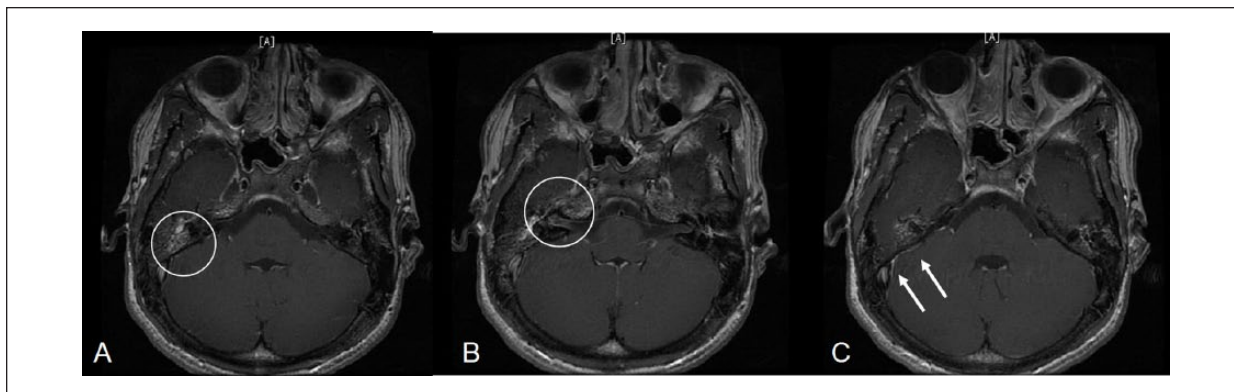
<sup>1</sup>Children's Hospital of Michigan, Detroit, MI, USA

### Corresponding Author:

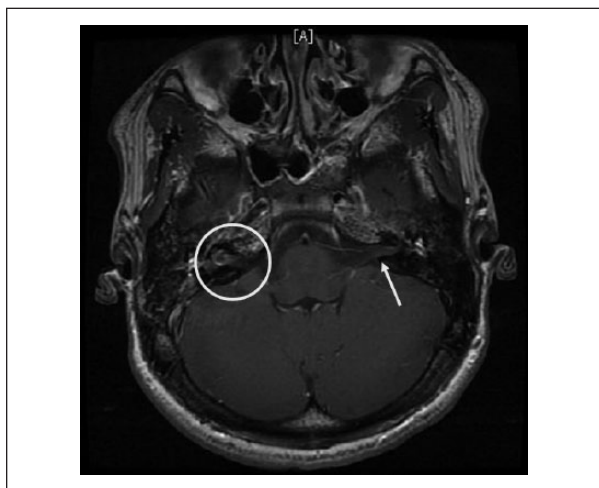
Paola America Huaynate-Cuadrado, MD, Department of Pediatrics, Children's Hospital of Michigan, 3901 Beaubien St, Detroit, MI 48201, USA.

Email: phuaynat@dmc.org



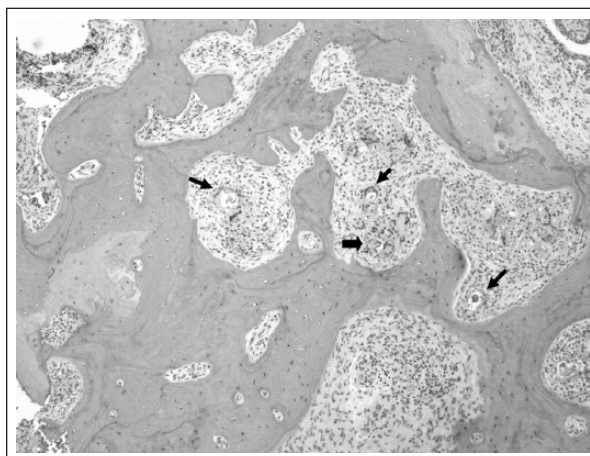


**Figure 1.** Brain MRI, axial view. Right cochlear enhancement (A, white circle). Enhancement of right acoustic nerve (B, white circle). Right-sided meningeal enhancement (C, white arrows).



**Figure 2.** Brain MRI, axial view. Persistent enhancement of both acoustic nerves without obvious meningeal enhancement (white circle and arrows).

surgery, it was noticed that his right mastoid contained sclerotic tissue, suggestive of inflammation versus infection, and the implant was aborted. A biopsy of the tissue was submitted and was described as consisting of  $0.3 \times 0.2 \times 0.2$  cm of soft tissue and a  $0.7 \times 0.5 \times 0.3$  cm portion of bone. Microscopically, sections of the soft tissue consisted of respiratory mucosa with a heavy chronic inflammatory infiltrate consisting of lymphocytes, plasma cells, scattered neutrophils, and multinucleated giant cells containing nonpolarizable inclusions. Also present were poorly formed nonnecrotizing epithelioid granulomas. Similar findings were seen between the bony trabeculae. No fungal or acid-fast microorganisms were identified by GMS or Fite acid-fast stains. The von Kossa stain for calcium was negative. The features were compatible with sarcoidosis if infections were excluded clinically (Figure 3). The patient received a cochlear implant on the right



**Figure 3.** Bone with poorly formed granulomas and multinucleated giant cells with nonpolarizing inclusions Schaumann bodies (arrows) and poorly formed granuloma (block arrow). Hematoxylin-eosin, magnification  $4\times$ .

side after 2 weeks. Biopsies procured during cochlear implantation microscopically showed respiratory mucosa with chronic inflammation consisting of lymphocytes and plasma cells, dense fibrosis consistent with scar, and fragments of viable reactive bone. No granulomatous inflammation was identified. The patient continued to follow with Rheumatology for the treatment of sarcoidosis for the next months.

## Final Diagnosis

Neurosarcoidosis.

## Discussion

Neurosarcoidosis is a diagnosis of exclusion, representing 5% to 15% of cases of adult sarcoidosis.<sup>1,2</sup> It is

even more rare as initial presentation in the pediatric population. It should remain in the differential of unexplained sudden-onset sensorineural hearing loss,<sup>3,4</sup> which is an otolaryngological emergency.

Sarcoidosis in children shows 2 different patterns: an early-onset type marked by rash, uveitis, and arthritis, and a second type, later in childhood, with involvement of several organs, mostly affecting the lungs with 60% of patients showing bilateral hilar or paratracheal lymphadenopathy on chest X-ray upon initial presentation.<sup>5</sup> While about 80% to 90% of cases of neurosarcoidosis involve the cranial nerves, the presentation of sudden hearing loss has been reported in about 30 pediatric cases in the reviewed literature.<sup>6,7</sup>

This presentation can be very insidious; however, the clinician should have a low threshold for suspicion for an autoimmune disease so that appropriate immunosuppressive therapy can be initiated early in the course of disease. As in systemic disease, corticosteroid therapy can decrease the inflammation and thus reverse the hearing loss.<sup>1</sup> In our case, the final diagnosis was reached with a mastoid tissue biopsy. The gold standard is biopsy of the affected cranial nerve but this is seldom feasible.

Viral infections are also often implicated in sudden sensorineural hearing loss.<sup>8</sup> In our patient, a positive result of the PCR for HSV was obtained in the right middle ear but not in CSF or the left middle ear. Case reports of HSV infection causing hearing loss usually involve a positive result in CSF and/or positive cultures,<sup>9</sup> which in this case resulted in concurrent treatment with acyclovir, since the final diagnosis was not available until the pathology was obtained.

## Conclusion

Pediatric neurosarcoidosis presenting as cranial nerve involvement without pulmonary compromise is an extremely rare occurrence that requires a high level of clinical suspicion. The diagnosis was delayed due to the marginal elevation of the ACE value, which later normalized after steroid courses.

We are of the impression that, in this case, the positive HSV result was of little or no clinical relevance since the pathology demonstrated abnormalities associated to sarcoidosis.

## Acknowledgments

We would like to thank Swati Mody, MD, a pediatric radiologist at Children's Hospital of Michigan, for reviewing the magnetic resonance images.

## Author Contributions

PAHC contributed to conception and design; contributed to acquisition; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

JP contributed to acquisition; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

JYA contributed to conception and design; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

## Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

## Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

## References

1. Loor RG, van Tongeren J, Derks W. Multiple cranial nerve dysfunction caused by neurosarcoidosis. *Am J Otolaryngol*. 2012;33:484-486.
2. Nozaki K, Judson MA. Neurosarcoidosis: clinical manifestations, diagnosis and treatment. *Presse Med*. 2012;41:e331-e348.
3. Cama E, Santarelli R, Muzzi E, et al. Sudden hearing loss in sarcoidosis: otoneurological study and neuroradiological correlates *Acta Otorhinolaryngol Ital*. 2011;31:235-238.
4. Schreiber BE, Agrup C, Haskard DO, Luxon LM. Sudden sensorineural hearing loss. *Lancet*. 2010;375:1203-1211.
5. Fretzayas A, Moustaki M, Vougiouka O. The puzzling clinical spectrum and course of juvenile sarcoidosis. *World J Pediatr*. 2011;7:103-110.
6. Baumann RJ, Robertson WC Jr. Neurosarcooid presents differently in children than in adults. *Pediatrics*. 2003;112:e480-e486.
7. Rheault MN, Manivel JC, Levine SC, Sinaiko AR. Sarcoidosis presenting with hearing loss and granulomatous interstitial nephritis in an adolescent. *Pediatr Nephrol*. 2006;21:1323-1326.
8. Chau JK, Lin JR, Atashband S, Irvine RA, Westerberg BD. Systematic review of the evidence for the etiology of adult sudden sensorineural hearing loss. *Laryngoscope*. 2010;120:1011-1021.
9. Rabinstein A, Jerry J, Saraf-Lavi E, Sklar E, Bradley WG. Sudden sensorineural hearing loss associated with herpes simplex virus type 1 infection. *Neurology*. 2001;56:571-572.