

Acute Bilateral Sensorineural Hearing Loss Associated with Myelodysplastic Syndrome

Daniel R. Bunker, MD*; Peter D. Gorevic, MD; John O. Mascarenhas, MD; Eric Smouha, MD

*Daniel R. Bunker, MD

Department of Medicine, Division of Rheumatology, Icahn School of Medicine at Mount Sinai, New York, NY 10029, USA

Tel: 212-241-8740, Fax: 212-987-5584; Email: daniel.bunker@mountsinai.org

Abstract

Importance: Acute bilateral sensorineural hearing loss can be associated with hematologic malignancies. We present the second reported case of bilateral sudden sensorineural hearing loss in the setting of myelodysplastic syndrome.

Observations: A patient with recently diagnosed myelodysplastic syndrome (MDS) presented with acute bilateral sensorineural hearing loss. Labs and radiographic studies were unrevealing for an underlying cause. He was treated with high dose steroids without significant clinical improvement.

Conclusion: Acute bilateral sensorineural hearing loss is a well described phenomenon in the setting of leukemia but can also be seen in myelodysplastic syndrome. Given the well-recognized association of MDS with autoimmune disease, an autoimmune cause of our patient's hearing loss must be considered. Unfortunately our patient did not response to corticosteroids; more research is needed to determine the pathogenesis of sudden sensorineural hearing loss in patients with hematologic malignancies.

Keywords

sensorineural hearing loss; myelodysplastic syndrome

Introduction

Sudden sensorineural hearing loss is an otologic emergency. Most cases are unilateral and idiopathic [1]. The occurrence of acute bilateral sensorineural hearing loss is rare and when present often indicates serious systemic pathology such as infection, malignancy, or toxic insult [2]. Sudden sensorineural hearing loss is well described in leukemic and myeloproliferative conditions, however we can find only one such case in the literature occurring in the setting of myelodysplastic syndrome (MDS)[3]. Here, we present a case of a 52 year old man with MDS who developed acute bilateral sensorineural hearing loss.

Case Presentation

A 52 year old man with recently diagnosed MDS was transferred to our hospital because of acute bilateral hearing loss. A month before presentation he had developed fatigue, weakness, and shortness of breath. Labs showed new anemia and thrombocytopenia, and a bone marrow biopsy found ineffective hematopoiesis and multi-lineage dysplasia, including hypolobated megakaryocytes, consistent with MDS. Cytogenetic analysis revealed a complex monosomal karyotype in 75% of the evaluable cells. Soon

afterwards, he developed what he described as a “clogged” sensation in his left ear. Over the next two days, he developed severe left sided hearing loss and moderate right sided hearing loss. Additionally, he endorsed bilateral tinnitus as well as disequilibrium without vertigo. He denied otalgia and otorrhea, or any preceding rhinorrhea, sinus congestion, or sore throat.

On exam, both external auditory canals were normal with intact tympanic membranes. There was no evidence of auricular inflammation, no vasculitic skin rashes, and no synovitis. His cognitive function was intact and other than decreased hearing full neurologic evaluation was unremarkable. Anti-neutrophil cytoplasmic antibody titers and antiphospholipid serologies were normal. A serum paraneoplastic antibody panel (anti-Hu, anti-Yo, amphiphysian, CRMP-5 IgG, striational, P/Q type calcium channel, N-type calcium channel, Ach Receptor, AChR ganglionic neuronal, neuronal K⁺ channel) was normal, and CSF studies showed no elevated protein and no evidence of malignancy. CT of the head and temporal bones showed no abnormalities of the external, middle, or internal auditory canals (MRI could not be done because of cervical hardware placed after a previous motor vehicle accident). Audiogram revealed bilateral sensorineural hearing loss, profound on the left and moderate on the right (figure 1). The patient was given 1gm of methylprednisolone intravenously for 3 days, and transitioned to prednisone 60mg daily followed by a two week taper. His hearing function stabilized but did not improve (figure 2). Additional immunomodulators like intravenous gammaglobulin (IVIG) were considered but it was felt that his hearing might improve with treatment of his underlying malignancy. He has been treated with a hypomethylating agent for over six cycles for his underlying MDS, with a hematologic and cytogenetic response, however his hearing loss remains unchanged. He is currently being evaluated for cochlear implants.

Discussion

Acute bilateral sensorineural hearing loss is a well-described phenomenon in the setting of leukemia, both acute [4] and chronic [5]; rarely it can be the presenting manifestation of the disease [4-6]. The pathogenesis of this phenomenon in leukemia has traditionally been attributed to inner ear hemorrhage, infection, or leukemic infiltration [7]; most recently hyperviscosity [8] has been invoked as a potential cause in patients with severe leukocytosis. Previous studies have found increase signal intensity on T1-weighted MRI with abnormal enhancement of the cochlea [6]. Pathologic evaluations in patients with acute lymphocytic leukemia and otologic symptoms most commonly have shown hemorrhage and leukemic infiltration [9]. In general, the prognosis for recovery of hearing in these settings is poor.

MDS is a heterogeneous group of diseases characterized by dysregulated hematopoiesis leading to variable cytopenias and risk of transformation to acute myeloid leukemia. Unlike acute leukemia, MDS is characterized by the relative lower frequency of circulating blasts. Bilateral sudden sensorineural hearing loss has been described previously in the setting of MDS [3], but is extremely rare and the pathology is uncertain. Hemorrhage due to thrombocytopenia is one potential mechanism, but leukemic infiltration and hyperviscosity are unlikely causes given the absence of high numbers of circulating blasts.

Interestingly, MDS has been associated with various autoimmune manifestations in approximately 10-20% of patients; cutaneous vasculitis and a seronegative polyarthritis are most

common, but systemic vasculitic syndromes have also been reported [10]. Given our patient's underlying MDS as well simultaneous bilateral symptoms, it is possible that his hearing loss was not due to any of the previously mentioned mechanisms, but rather to autoimmune mediated inner ear disease (AIED). Though a number of inner ear antigens have been proposed as the primary target for this disease, the true pathophysiology is still unclear; obtaining pathologic tissue from the temporal bone is virtually impossible in a live patient, and there are still no reliable serologic tests for this disease [11]. While our patient's lack of response to high dose corticosteroids does not support an autoimmune cause, AIED is not ruled out either. For example AIED in the setting of malignancy, termed paraneoplastic cochleovestibulopathy [12], is not uniformly responsive to immunosuppression. In the absence of a serum marker or histopathologic analysis, we cannot be certain of the etiology of the hearing loss.

Conclusion

We present a case of acute bilateral sensorineural hearing loss occurring in the setting of MDS. To our knowledge, this is the second report of an association between these diseases. As MDS is known to be associated with autoimmune phenomena, we hypothesize that the patient's sudden bilateral sensorineural hearing loss was autoimmune in nature. However, the patient did not respond to high dose corticosteroids. More research is needed to determine the pathogenesis of sudden sensorineural hearing loss in patients with hematologic malignancies.

Figures

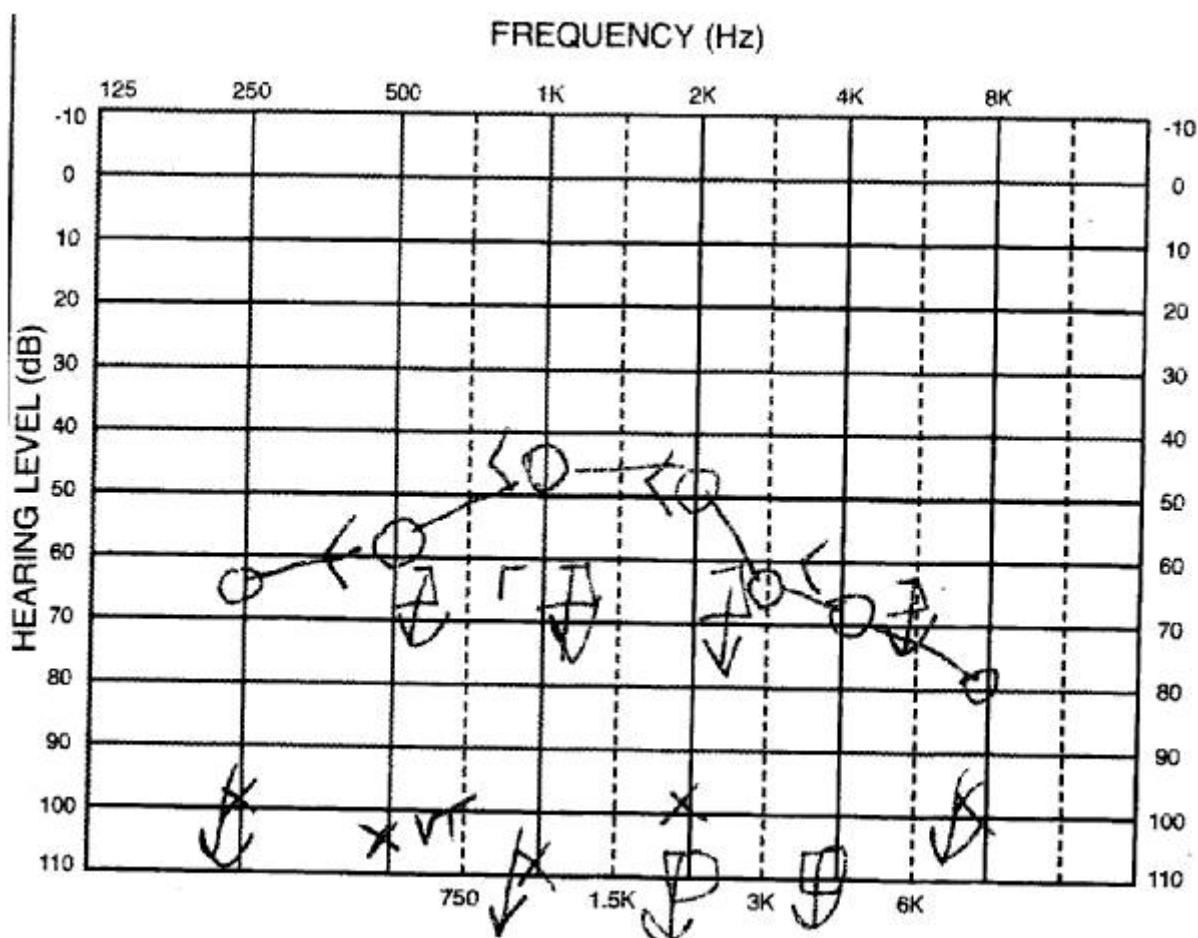


Figure 1: Audiometry Report On Presentation

Patient's audiogram showing moderate sensorineural hearing loss in the right ear (circles) and severe sensorineural hearing loss in the left ear (x). The frequency in hertz of the test tones is shown on the horizontal axis. Loudness in decibels is measured on the vertical axis. An ear with normal hearing has thresholds 25 dB or less at each test frequency.

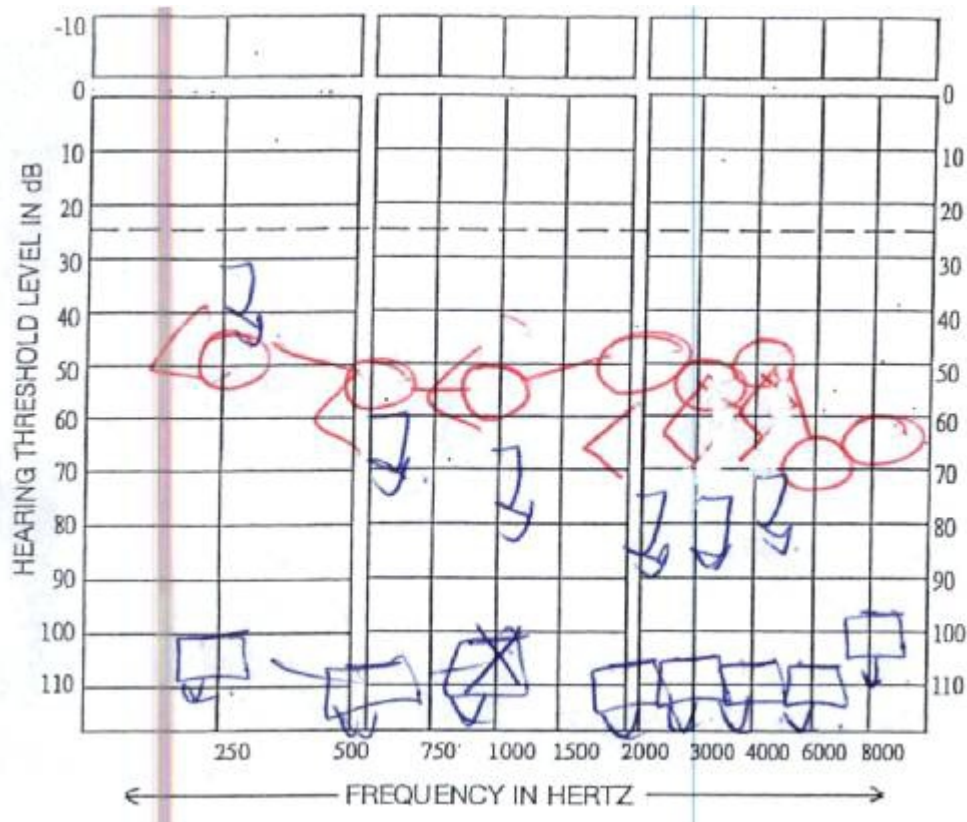


Figure 2: Audiometry Report after Three Weeks of High Dose Corticosteroids

References

1. Rauch, S.D., Clinical practice. Idiopathic sudden sensorineural hearing loss. *N Engl J Med*, 2008. 359(8): p. 833-40.
2. Sara, S.A., B.M. Teh, and P. Friedland, Bilateral sudden sensorineural hearing loss: review. *J Laryngol Otol*, 2014. 128 Suppl 1: p. S8-15.
3. Lee, E.J. and Y.J. Yoon, Bilateral sudden sensorineural hearing loss as an initial presentation of myelodysplastic syndrome. *Am J Otolaryngol*, 2012. 33(6): p. 782-3.
4. Harada, T, S. Namiki, and I. Kawabata, Acute profound sensorineural hearing loss as the initial manifestation of acute leukemia--report of a case. *Auris Nasus Larynx*, 2000. 27(4): p. 359-62.
5. Genden, E.M. and R.S. Bahadori, Bilateral sensorineural hearing loss as a first symptom of chronic myelogenous leukemia. *Otolaryngol Head Neck Surg*, 1995. 113(4): p. 499-501.
6. Hsu, Y.C., C.Y. Su, and R.F. Hsu, Unilateral sudden hearing loss as a presenting manifestation of chronic myeloid leukemia: case report. *Otolaryngol Head Neck Surg*, 2004. 130(2): p. 271-3.
7. Druss, J.G., Aural manifestations of leukemia. *Arch Otolaryngol*, 1945. 42: p. 267-74.
8. Chae, S.W., et al., Sudden hearing loss in chronic myelogenous leukaemia implicating the hyperviscosity syndrome. *J Laryngol Otol*, 2002. 116(4): p. 291-3.
9. Terao, K., et al., Pathologic correlations of otologic symptoms in acute lymphocytic leukemia. *Am J Otolaryngol*, 2011. 32(1): p. 13-8.

10. Giannouli, S. and M. Voulgarelis, A comprehensive review of myelodysplastic syndrome patients with autoimmune diseases. *Expert Rev Clin Immunol*, 2014. 10(12): p. 1679-88.
11. Baruah, P., Cochlin in autoimmune inner ear disease: is the search for an inner ear autoantigen over? *Auris Nasus Larynx*, 2014. 41(6): p. 499-501.
12. Greene, J.J., et al., Paraneoplastic syndrome: a masquerade of autoimmune inner ear disease. *Otol Neurotol*, 2015. 36(1): p. e3-10.

Manuscript Information: Received: May 20, 2016; Accepted: July 05, 2016; Published: July 07, 2016

Authors Information: Daniel R. Bunker, MD^{1*}; Peter D. Gorevic, MD¹; John O. Mascarenhas, MD²; Eric Smouha, MD³

¹Department of Medicine, Division of Rheumatology, Icahn School of Medicine at Mount Sinai, New York, NY 10029, USA

²Department of Medicine, Division of Hematology and Medical Oncology, Icahn School of Medicine at Mount Sinai, New York, NY 10029, USA

³Department of Otolaryngology, Icahn School of Medicine at Mount Sinai, New York, NY 10029, USA

Citation: Bunker DR, Gorevic PD, Mascarenhas JO, smouha E. Acute bilateral sensorineural hearing loss associated with myelodysplastic syndrome. *Open J Clin Med Case Rep*. 2016; 1137

Copy right statement: Content published in the journal follows Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>). © **Bunker DR 2016**

Journal: Open Journal of Clinical and Medical Case Reports is an international, open access, peer reviewed Journal focusing exclusively on case reports covering all areas of clinical & medical sciences.

Visit the journal website at www.jclinmedcasereports.com

For reprints & other information, contact editorial office at info@jclinmedcasereports.com