INTRODUCTION

Sweet’s syndrome or acute febrile neutrophilic dermatosis is a skin disease characterized by the sudden onset of fever, leukocytosis, and tender, erythematous, well-demarcated papules and plaques which show dense infiltrates by neutrophil granulocytes on histologic exam. The pathogenesis of this condition is not completely defined, but the response to corticosteroids points towards an autoimmune etiology. There have been two prior reports of progressive, bilateral, sensorineural hearing loss (SNHL) in the background of Sweet’s syndrome. It is speculated that an autoimmune process within the inner ear is responsible for this manifestation. Prior descriptions of this phenomenon demonstrate a significant improvement in auditory outcomes and quality of life following cochlear implantation in these profoundly deafened patients. We describe a patient with Sweet’s syndrome presenting with sudden, progressive, bilateral SNHL. As in prior studies, our patient also underwent cochlear implantation. Our outcomes were limited however, by Sweet’s skin flares interfering with surgical wound healing along with a failure of the Nucleus-5 cochlear implant upon initial stimulation.

CASE REPORT

A 49-year-old woman with neutrophilic dermatosis controlled with oral steroids diagnosed two years prior presented to the emergency department with a one day history of vomiting, tinnitus, vertigo, bilaterally decreased hearing, and blurred vision. Evaluation and imaging ruled out stroke or any other ischemic etiology. The patient denied any recent travel or sick contacts. Except for a transient leukocytosis, basic labs including complete blood count, comprehensive metabolic panel, and coagulation profiles were otherwise within normal limits. Infectious serum workup for syphilis, human immunodeficiency virus, hepatitis B and hepatitis C were all within normal limits. Autoimmune screening with c-ANCA, p-ANCA, rheumatoid factor, and ANA were negative as well. Analysis of cerebrospinal fluid (CSF) was normal for cryptococcal antigen, protein electrophoresis, and protein/glucose/cell counts. Viral culture of CSF initially grew herpes simplex virus, but confirmatory polymerase chain reaction failed to grow any viral DNA. Audiometry revealed worsening hearing loss, culminating in a profound bilateral sensorineural loss by day of hospitalization (Figure 1). Screening head, temporal bone, and neck computed tomography scanning, magnetic resonance imaging (MRI), and magnetic resonance angiography were negative for collection, hemorrhage, mass, or ischemia. A non-specific right periatrial white matter focus on T2/FLAIR imaging (Figure 2) was observed on MRI. The patient was administered a tapering course of high-dose oral prednisone upon admission to the hospital and three doses of intra-tymanic dexamethasone 4 days later with no improvement in hearing.

An adequate trial of hearing aids provided no benefit, and she subsequently underwent a left-sided cochlear implantation. Proper placement of implant and electrodes was confirmed with radiography. Despite having no inflammatory skin lesions at the time of implantation, the surgical site developed a Sweet’s flare with drainage that responded to antibiotics. Following self-administration of an oatmeal bath, a collection formed: which resolved with surgical drainage and prolonged antibiotics. At the time of initial stimulation, the Nucleus-5 device was only able to establish thresholds at electrode 16. Due to the initial implant failure and complications with the surgical site, the device was removed with plans to place a new implant on the contralateral side with stringent precautions on wound care.

REFERENCES