Conductive hearing loss is the result of any process that attenuates the sound energy reaching the cochlea. The differential diagnosis for conductive hearing loss is broad; however, a thorough patient history in conjunction with the physical examination, audiologic testing, and imaging may narrow the differential diagnosis. The most common cause of progressive conductive hearing loss in the absence of chronic otitis media is otosclerosis, yet other entities causing ossicular chain fixation, ossicular discontinuity, and third window phenomena should be considered [1].

In this report, we present the unusual case of a patient who underwent middle ear exploration for presumed otosclerosis, but who was ultimately found to have a temporal bone encephalocele as the etiology of conductive hearing loss. Preoperative evaluation did not reveal any specific signs or symptoms strongly suggestive of this diagnosis, with an aerated middle ear and inconclusive imaging studies. Temporal bone encephaloceles can present with conductive hearing loss, though usually in the context of persistent middle ear effusion or otitis media. However, as this case demonstrates, they can also be the cause of conductive hearing loss in the presence of a normal otoscopic exam.

A 50 year old healthy female was referred for evaluation of unilateral hearing loss. The patient weighed 250 lbs with body-mass index of 41. Hearing loss began 6 months prior with associated aural fullness. She denied tinnitus, vertigo, or dizziness, sound or pressure-induced dizziness, otorrea, otalgia, history of ear disease, or trauma. She also denied a family history of hearing problems.

On examination, the right ear tympanic membrane was intact and the middle ear aerated. Tuning fork examination was consistent with a significant conductive hearing loss in the right ear. The remaining otoneurological examination and the general head and neck examination were unremarkable.

Audiologic testing revealed a right mixed hearing loss with an air-bone gap at all frequencies (most pronounced at 2000 to 4000 Hz) and type A tympanograms bilaterally (Figure 1). Right ipsilateral acoustic reflexes were absent. Left contralateral reflexes were present at 500 Hz, 1000 Hz, 2000 Hz, and absent at 4000 Hz (Figure 2). A high resolution CT scan of the temporal bone was obtained to evaluate for the possibility of cochlear otosclerosis (given the mixed hearing loss) as well as a third window disorder (given the presence of acoustic reflexes). CT imaging was essentially normal, with no evidence of cochlear otosclerosis or a third window disorder. The tegmen appeared thin in places but was not clearly dehiscent. Scent opacification was noted in the mastoid.

The patient elected to have a right middle ear exploration to address the conductive hearing loss. Intraoperatively, no specific ossicular abnormality was identified. Scent middle ear fluid was present and the possibility of an encephalocele with associated CSF (cerebrospinal fluid) otorrea was entertained, though an encephalocele was not directly visualized. A small amount of fluid was collected and sent for beta-2 transferrin (though this test was ultimately negative).

Given the intraoperative findings, a magnetic resonance imaging (MRI) of the brain was ordered postoperatively to evaluate for possible encephalocele (Figure 3). Possible encephalocele was identified, and there appeared to be more extensive opacification of the mastoid compared to the preoperative CT scan. A repeat CT scan of the temporal bone was obtained, again suggesting increased opacification of the mastoid, as well as an area of tegmen dehiscence (Figure 4). As such, the patient underwent a combined transmastoid and middle cranial fossa repair, with intraoperative confirmation of the encephalocele and CSF leak. Postoperatively, the patient had an uneventful recovery, with postoperative audiogram demonstrating resolution of conductive hearing loss.

There are multiple potential diagnoses for an adult patient presenting with progressive conductive hearing loss. In addition to a thorough history and examination, it may also be important to obtain audiometric and radiologic evaluation. In this report, the patient had no history of ear disease or trauma, an unremarkable otoscopic examination who presented with worsening conductive hearing loss because of an encephalocele impeding normal ossicular movement. Now temporal bone encephalocele must also be considered when evaluating a patient with conductive hearing loss and partially intact acoustic reflexes.