We present a man with bilateral progressive hearing loss.

**Imaging Findings:**

A 44 year old male patient presented with bilateral hearing loss. According to the patient, the hearing loss occurred progressively, starting 3 years earlier after a mild head injury. Otomicroscopical examination showed intact tympanic membranes and no middle ear effusion. Pure tone audiometry showed a bilateral, severe to profound, mixed conductive and sensorineural hearing loss.

High-resolution computed tomography of the temporal bones was performed (Fig 1-3), showing bilateral and symmetric bulbous dilatation of the internal auditory canal, and bilateral absence of the cochlear bony modiolus. There was a bilateral dilatation of the labyrinthine section of the facial nerve, the canal of the superior vestibular nerve, and the singular nerve canal. These abnormalities were more pronounced on the left side. The middle ear cavities and ossicles appeared normal. The semicircular canals and vestibules also had a normal appearance. In view of the clinical history and the imaging findings, the diagnosis of X-linked congenital hearing loss was established.
Discussion:

X-linked congenital hearing loss is a congenital disease causing perilymphatic hydrops due to a fistulous connection between the internal auditory canal and the cochlea. The genetic abnormality is situated on the X-chromosome. The disease manifests itself in young men as profound mixed or sensorineural hearing loss, often progressive and possibly associated with vestibular dysfunction. Female carriers are asymptomatic or may have a much less severe form of this condition.

The underlying developmental anomaly is an absence of the bony partition between the fundus of the internal auditory canal and the basal turn of the cochlea, allowing a direct communication between the subarachnoid space and the perilymphatic space. The intracranial pressure is transmitted into the perilymphatic space, and the raised perilymphatic pressure, exerted on the cochlear duct and the stapes footplate, results in a mixed pattern of sensorineural and conductive hearing loss. Widening of the other nerve canals at the fundus of the internal auditory canal is also explained by the deficiency of bone in that region.

The differential diagnosis is otosclerosis and other developmental disorders of the inner ear, such as large vestibular aqueduct syndrome. Differentiating X-linked deafness by CT, in subjects presenting with progressive sensorineural or mixed hearing loss from otosclerosis is important. Because of the communication between the subarachnoid space in the internal auditory canal and the perilymphatic space in the cochlea, attempting to correct the conductive hearing impairment by stapedotomy (as is done in otosclerosis) carries the risk of a profuse egress of clear cerebrospinal fluid (gusher) when manipulating the stapes, leading to permanent deafness.

**Differential Diagnosis List:** X-linked congenital deafness.

**Final Diagnosis:** X-linked congenital deafness.

**References:**


**Figure 1 a**

**Description:** The singular nerve canal (white arrowhead) is widened. The cochlear duct is widened (black arrow), and the cochlear modiolus is not visible. **Origin:**
Figure 2

Description: Wide cochlear duct and absence of cochlear modiolus (black arrow). Origin:
Description: Dilatation of the labyrinthine facial nerve (white arrow) and superior vestibular nerve canal (black arrow). Origin: