Case Report

Pediatric otosyphilis—An unusual cause of conductive hearing loss

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ABSTRACT

We present the case of a 5 year old female with a unilateral conductive hearing loss which had a relapsing and remitting course over a 3 year period. An initial noncontrast CT temporal bone study was unremarkable and a diagnosis of otitis media was made in the first instance. However, a second CT temporal bone study performed 3 years later demonstrated bilateral demineralisation of the ossicles and abnormal lucency affecting both the otic capsules. A diagnosis of otosyphilis was proposed on the basis of the imaging features and a prior medical history of previously treated congenital syphilis. With the benefit of hindsight, early per cochlear lucency was identified on the initial CT temporal bone study. There has been a steady rise of syphilis cases since the millennium with resurgence in many high income countries. Otosyphilis has a highly variable clinical presentation and there is limited data to establish the pattern of hearing loss in pediatric patients with a background of congenital otosyphilis. Temporal bone and otic capsule demineralisation carries a broad differential diagnosis including osteogenesis imperfecta, otosclerosis, Paget’s disease and radiation related changes. Otosyphilis is a rare but potentially treatable cause of deafness and a high index of suspicion is required to make the diagnosis. In conjunction with a positive syphilis serology, a noncontrast temporal bone CT can aid the diagnosis and expedite the treatment.

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Introduction

The WHO have estimated that there are 11 million new syphilis cases per year globally in adults aged 15-49 yrs. [1]. The 2 routes of transmission are through sexual contact or transplacental vertical transmission during pregnancy. The rate of primary and secondary syphilis has increased steadily since the millennium [2], and presents a significant health problem in low income countries. There has been resurgence in high income countries, particularly affecting certain at risk groups such as MSMs [2,3]. The growing number of infected women of reproductive age has led to resurgence of congenital syphilis cases; with many high-risk women not being identified and treated during pregnancy [4].

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Syphilis infection is characterized by active infection, interspersed with periods of latency. Left untreated, syphilis can progress through 4 disease stages, which are outlined in Table 1.

Both acquired and congenital syphilis are known to cause cochleovestibular dysfunction [5]. Sensorineural hearing loss is frequently described in secondary and tertiary syphilis [6,7].

Otosyphilis has a variable clinical presentation and a high index of suspicion is required to make the diagnosis. The Centre for Disease Control recommends a treatment regimen of IV penicillin in order to prevent further disease progression and the development of complications as a result of multisystemic involvement [8].

Late adult presentation of hearing loss, decades after the primary syphilis infection has been documented, however pediatric cases are rare. We report a case of a 5-year-old female with an unusual presentation of unilateral conductive hearing loss, which had a relapsing and remitting presentation over several years. The diagnosis of otosyphilis was proposed by radiological findings on a noncontrast temporal bone CT.

### Table 1 – The stages of syphilis infection.

| Stage | Description |
|-------|-------------|
| Primary | A painless chancre develops at the site of inoculation following a 2-6 week incubation period [9]. The affected sites are usually the anal, genital or oral mucosa, this is frequently accompanied by local lymphadenopathy. After several weeks, there is spontaneous healing of the chancre due to macrophage directed phagocytosis of the treponemes [9,10]. However, some of the treponemes escape and systemically disseminate to multiple organs, which results in the development of chronic disease. |
| Secondary | This stage follows the multiplication of the disseminated treponemes. This can occur at the same time as, or 6 months following, the appearance of the chancre. It is characterized by systemic symptoms such as headache, malaise, pyrexia, condylomata lata, and a generalized rash affecting the palms or soles [11]. This stage can last between several weeks and months. |
| Latent | Bacteria is present in body but without associated signs or symptoms. Can occur years to decades after the primary infection. This stage is rare today due to early treatment with curative antibiotics [9,12]. The treponemes can invade any organ system or tissue [11] and provoke inflammation due to a delayed hypersensitivity response. This can result in cardiovascular syphilis, neurosyphilis or gummatous syphilis. The latter is characterised by the formation of granulomatous lesions (gumma) which are detected predominantly within the skin and bone [11]. |
| Tertiary | |
to be more effective in reaching treponemocidal levels in the cerebrospinal fluid (CSF) [13–16]. Several studies have additionally supplemented this with a corticosteroid for otoysphilis [16,17]. Bradshaw et al. undertook a small retrospective study using 7 confirmed adult otoysphilis cases. There was a subjective hearing improvement in 67% of those receiving treatment 1 month after the onset of hearing loss [18]. Gleich et al. conducted a prospective cohort study involving 18 adult patients with cochleovestibular dysfunction and positive syphilis serology, treated with the IV antibiotics and steroids. The median age was 53 and symptoms were present between 2 weeks and 20 years. 31% achieved an improved speech recognition threshold by 15 dB [5]. It was found that patients with hearing loss greater than 5 years did not respond [5] and generally a younger age was associated with a better prognosis [5]. However there is a paucity of data evaluating treatment outcomes in pediatric patients specifically.

A systematic review by Chau et al. identified a lack of longitudinal audiometric follow-up data for patients previously either fully or partially treated for congenital syphilitic infection [19]. Only one study reported audiological data for a cohort of infants with a serologically confirmed diagnosis of congenital syphilis [20]. Although none of the 75 neonates had evidence of hearing impairment at birth, no audiological follow up was performed to assess the delayed onset of hearing loss in children treated for congenital syphilis.

Hearing loss secondary to congenital syphilis is typically sensorineural and bilateral in nature and can present suddenly [21]. The current case was atypical in that the patient had presented with conductive hearing loss with a fluctuating and variable course over several years.

The most common cause of conductive hearing loss in the pediatric population is otitis media, and our patient developed this which acted as a complicating factor.

Salomone et al. discuss a case of delayed diagnosis of juvenile otosclerosis after the conductive hearing loss in a young child was found to be refractory to grommet insertion [22].

In the present case, the diagnosis of otoysphilis was proposed by the radiological findings of the temporal bone CT. The patient was otherwise systemically asymptomatic without other stigmata of a syphilis infection. The clinical and audiological findings were nonspecific. Interestingly, bilateral radiological findings were demonstrated despite generally unilateral symptoms and puretone audiogram abnormalities; the reason for this is unclear. Kivekas et al. (2014) describe a case of a 75-year-old female who developed a bilateral sensorineural hearing loss. A temporal bone CT study demonstrated bilateral demineralisation of the otic capsules without associated ossicular chain involvement [23]. The development of conductive hearing loss, as seen in our case, is considered secondary to the development of osteitic lesions of the ossicles which results in fixation of the incus or malleus to the lateral wall of the atic [21,24–26].

Histopathological changes within the temporal bones are identical for congenital and acquired syphilis [21,27]. In milder cases, connective tissue infiltration is observed which leads to an inflammatory fibrosis [27]. However, in more severe cases, an obliterator endarteritis and mononuclear infiltration

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**Fig. 1 – Axial and coronal reformats from a temporal bone CT study performed 3 years earlier. There is subtle pericochlear lucency noted bilaterally (blue arrows) (Color version of figure is available online.)**
The radiographic findings reflect the underlying histopathological process with an irregular permissive lucency within the osseous labyrinth [29]. The same process can extend to involve the ossicular chain resulting in lucencies within the malleus and incus [29]. The differential diagnosis for the radiological findings of bilateral otic capsule demineralisation includes osteogenesis imperfecta, otosclerosis, and Paget’s disease [30]. Radiation related change is another increasingly recognised cause of temporal bone demineralisation [31].

Osteogenesis imperfecta is a genetic disorder of collagen synthesis. The clinical phenotype includes blue sclera, abnormal dentition and ligamentous laxity in addition to hearing loss [32]. The imaging findings are similar to those of syphilis [30] and otosclerosis [33] but are often more extensive. There is proliferation of thickened demineralised bone surrounding the osseous labyrinth [32] with extension into the labyrinth [32]. There may be an associated conductive hearing loss component due to fixation of the stapes footplate or atrophic changes to the long process of the incus [32].

Otosclerosis or otospongiosis is a disorder with a complex genetic and environmental aetiology that results in abnormal bone remodelling, resorption, new bone deposition and vascular proliferation [34]. Symptoms commonly affect patients in their 20s-40s. There is female gender predominance and most cases are bilateral [35].

The 2 main subtypes are fenestral and cochlear. With fenestral disease, the fisula ante fenestram is the region most commonly involved. This is an important differentiator from otosclerosis which spares the fisula ante fenestram [36]. The pathology may also extend to involve the otic capsule, round window niche and footplate of the stapes [34] which frequently results in progressive conductive hearing loss. The early stages of the disease are characterized by demineralisation anteromedial to the oval window which reflects underlying spongiotic bone resorption. Eventually this can progress to proliferation of demineralised bone which results in narrowing of the oval window, fixation of the stapes footplate and possible obliteration of the round window niche [33].

The ossicular chain can be involved with the formation of a web of

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Fig. 2 – Axial and coronal reformats from a temporal bone CT that demonstrate abnormal and widespread pericochlear lucency affecting the otic capsule bilaterally (blue arrows). There is marked demineralisation of the ossicular chains that was not evident on the prior CT study (white arrows on the current study and compare to Fig. 1) (Color version of figure is available online.)
bone between the incus or malleus and the wall of the attic [37].

Cochlear otosclerosis is less common than, but will always co-exist with the fenestral subtype [35].

It is characterised by demineralised spongiotic vascular bone around the cochlear capsule which can additionally extend to surround the vestibule, semicircular canals and internal auditory canal.

On CT, this corresponds to a ‘double ring’ of demineralisation around the cochlear [35], in addition to findings of fenestral otosclerosis.

Paget’s disease is characterised by excessive bone turnover due to osteoclastic resorption followed by osteoclastic repair [38]. Normal bone is replaced by abnormal lamellar bone which is soft and porous, deforming under stress and therefore susceptible to fracture. When the temporal bone is involved, the most common symptom is bilateral sensorineural hearing loss, and possibly also tinnitus and headache [38]. Conductive hearing loss component may also be present due to fixation of the stapes footplate [38]. During the early stage of the disease, well-defined lytic lesions are demonstrated in the skull. Demineralisation begins at the petrous apices of the temporal bones (due to greater marrow deposition) which progresses inferiorly and laterally [38]. Bone resorption starts at the peripheral aspect of the otic capsule and extends centrally [38]. The intermediate and later stages of Paget’s disease involve bony sclerosis and expansion, and are therefore more readily distinguishable from otosclerosis.

The temporal bone is frequently included within the radiation field during the treatment of head and neck cancer. Clinical symptoms can present 3 months to 40 years from the date of the initial treatment [31]. The pathophysiology involves an obliterator endarteritis and periarteritis which causes avascular necrosis [39]. Histologically, there is death of osteocytes, increased osteolysis and fibrosis in the region of the dead bone with loss of marrow [40]. On CT, mild cases of osteoradionecrosis involve bony erosion around the external auditory canal and mastoid opacification [31].

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Summary

Syphilis is a rare but increasingly prevalent cause of hearing loss. Otoysphilis has a highly variable clinical presentation with limited available long term data to establish the pattern of hearing loss in pediatric patients with a background of congenital otosphilis. In conjunction with positive serology, non-contrast temporal bone CT imaging can aid diagnosis and expedite treatment.

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