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Clinical Record

Side selection for cochlear implantation in a case of Cogan's syndrome

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Abstract

Cogan's syndrome is a rare clinical entity that is characterised by non-syphilitic interstitial keratitis and audiovestibular symptoms. The cause of Cogan's syndrome is considered to be autoimmune disease, which is supported by the resolution of hearing loss after steroid treatment, and the association with other autoimmune diseases. The sensorineural hearing loss of Cogan's syndrome is progressive over a few months, and sudden, bilateral deafness often occurs, which may be an indication for cochlear implantation. This paper presents the case of a young woman suffering from Cogan's syndrome and sudden, bilateral deafness. With reference to this case, we describe problems regarding cochlear implantation for Cogan's syndrome and radiological findings aiding selection of cochlear implantation side.

Key words: Cogan's Syndrome; Cochlear Implants

Introduction

Cogan's syndrome was first described by David Cogan in 1945. It is characterised by non-syphilitic interstitial keratitis, Ménière-like episodes of vestibular symptoms, and bilateral, sensorineural hearing loss.[1] The most common cause of Cogan's syndrome is considered to be autoimmune, which is supported by resolution of hearing loss after steroid treatment and an association with other autoimmune diseases, such as rheumatoid arthritis.[2] Cogan's syndrome can also be triggered by vaccinations and infections in some cases. This can be explained by the fact that Cogan's syndrome is mediated by a hypersensitivity response to one or more infectious agents that are associated with vasculitis.[3]

Classification of Cogan's syndrome is based on the ocular findings. The typical form is defined by non-syphilitic interstitial keratitis, and is associated with aortitis and aortic insufficiency in 12 per cent of cases. The atypical form may involve every ocular structure, leading to chronic or recurrent conjunctivitis, episcleritis, uveitis, optic disc oedema, or retinal vasculitis. It is associated with rheumatological syndromes in approximately 20 per cent of cases and has a more unfavourable prognosis than the typical form.[4,5] Systemic manifestations occur in approximately 70 per cent of cases and are basically attributable to systemic vasculitis. Sudden hearing loss and systemic features, except for aortic insufficiency, are more frequent in the atypical form. Adult cases are of the typical form in 70 per cent and of the atypical form in 30 per cent, although typical and atypical Cogan's syndrome closely resemble each other.[2,5]

One report found that bilateral hearing loss affected 43.5 per cent of patients within three months of the onset of initial symptoms. Audiovestibular dysfunction requires systemic corticosteroids and cytotoxic drugs, such as methotrexate and cyclophosphamide.[5] Cogan's syndrome patients usually progress to postlingual deafness, and cochlear implantation should be considered in order to restore hearing. Although cochlear ossification and other conditions that are associated with autoimmune disease should be borne in mind when considering cochlear implantation for Cogan's syndrome, excellent post-operative audiological results can be expected. We present the considerations for operative side selection and the considerable surgical issues, with reference to a case of Cogan's syndrome undergoing cochlear implantation.

Case reports

A 25-year-old woman with a known history of Sjögren's syndrome, which included interstitial keratitis, presented with initial complaints of profound, bilateral sensorineural hearing loss and vertigo.

An ophthalmological examination revealed interstitial keratitis in the form of patchy, deep, subepithelial corneal infiltrates (Figure 1).

Laboratory tests for anti-nuclear autoantibody and rheumatoid factor were positive, and serological tests for syphilis were negative. The patient was diagnosed with Cogan's syndrome.

The sudden sensorineural hearing loss was treated initially with oral steroids and methotrexate. However, the patient progressed to total bilateral deafness within one month.

A high resolution, temporal bone computed tomography (CT) scan showed general thickening of the membranous labyrinth on the right side and a soft tissue density in the right cochlear basal turn (Figure 2). This CT image may have represented calcific

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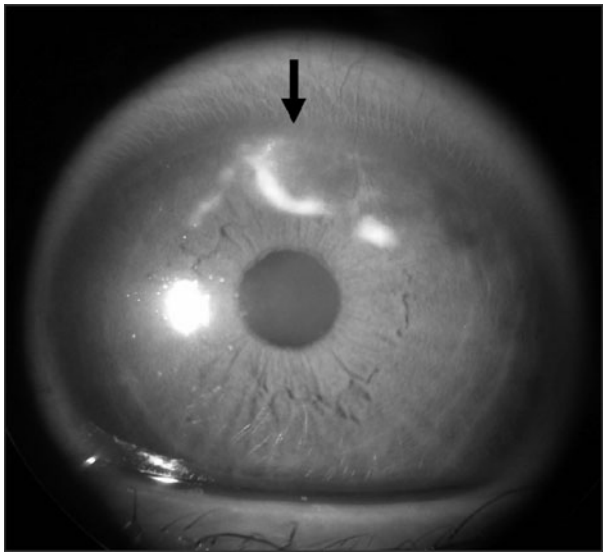

FIG. 1

Interstitial keratitis found on ophthalmological examination, indicated by the presence of patchy, deep, subepithelial corneal infiltrates (arrow).

obliteration and/or fibrosis in the right cochlea. A T2-weighted magnetic resonance imaging (MRI) scan demonstrated obliteration of the right cochlea. A contrast-enhanced, T1-weighted MRI scan demonstrated high signal intensity in the right membranous labyrinth, which indicated contrast leakage through the damaged labyrinthine membrane into the cochlear turn space (Figure 3). The radiological studies revealed that the findings in the right cochlea might be the result of an acute, active stage of Cogan's syndrome, since the cochlea was filled with granulation or fibrosis and there was partial ossification of the membranous labyrinth.

The patient underwent cochlear implantation with a Combi 40+ device (Med-El, Innsbruck, Austria) in her left ear, six months after the initial diagnosis. The Combi 40+ is safe for MRI studies at 1.5 Tesla without surgical removal of the internal magnet (Figure 4). An inferiorly based, inverted 'J' flap was used. Twelve pairs of electrodes of the standard electrode array type were inserted into the scala tympani through a standard cochleostomy. Ossification and fibrosis were not found, and full electrode insertion was easily achieved.

In the post-operative period, there were no complications during a one-year follow up, and no flap-related problems occurred. Our patient achieved excellent speech perception abilities, reaching mean scores of 91 per cent on open-set word tests and 96 per cent on everyday sentence recognition tests after one year of device use. However, she suffered from cutaneous skin lesions, and used steroids and methotrexate intermittently for symptom relief. In the future, monitoring for skin atrophy and flap ischaemia will be necessary.

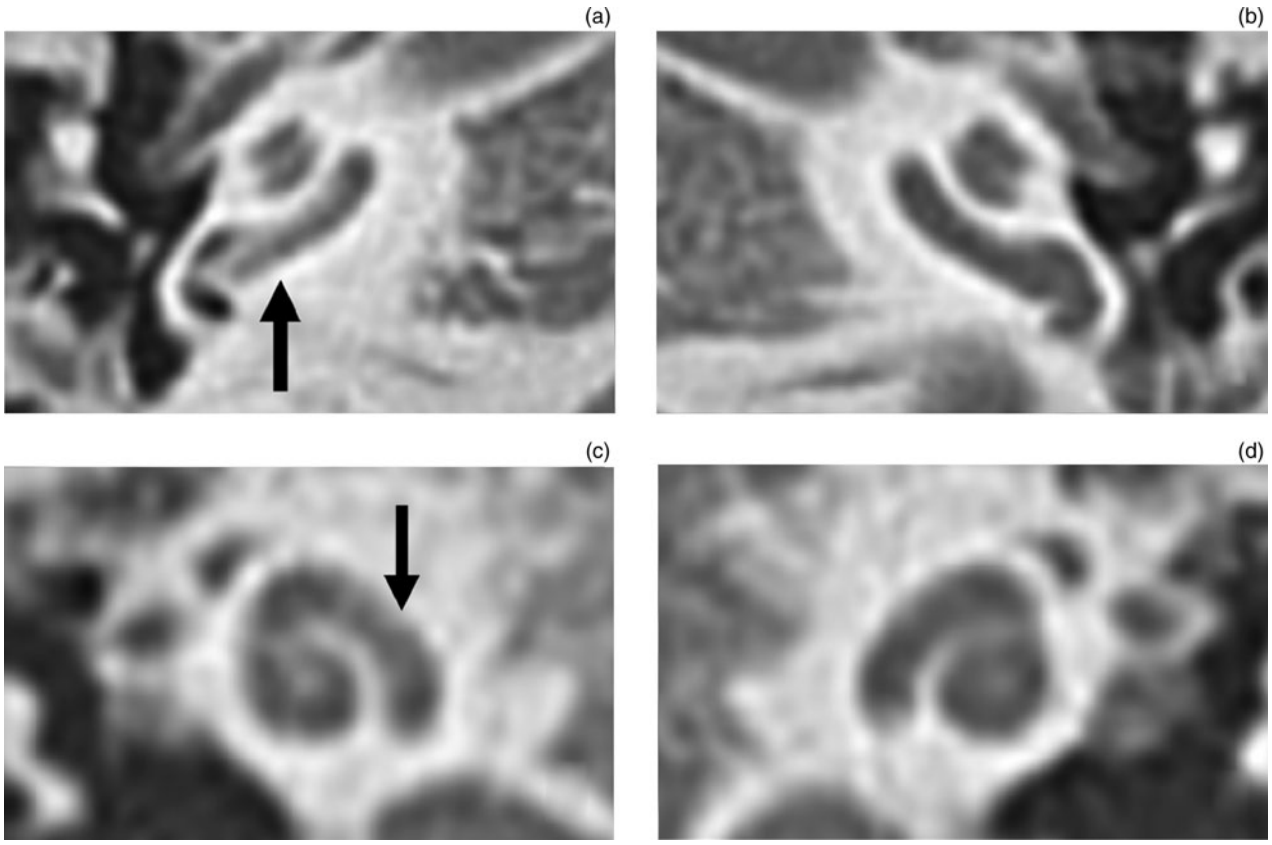

FIG. 2

High resolution, temporal bone computed tomography images: (a) right axial; (b) left axial; (c) right coronal; (d) left coronal. Images show a general thickening of the membranous labyrinth on the right side and a soft tissue density in the right cochlear basal turn, which may represent calcific obliteration and/or fibrosis in the right cochlea (arrow).

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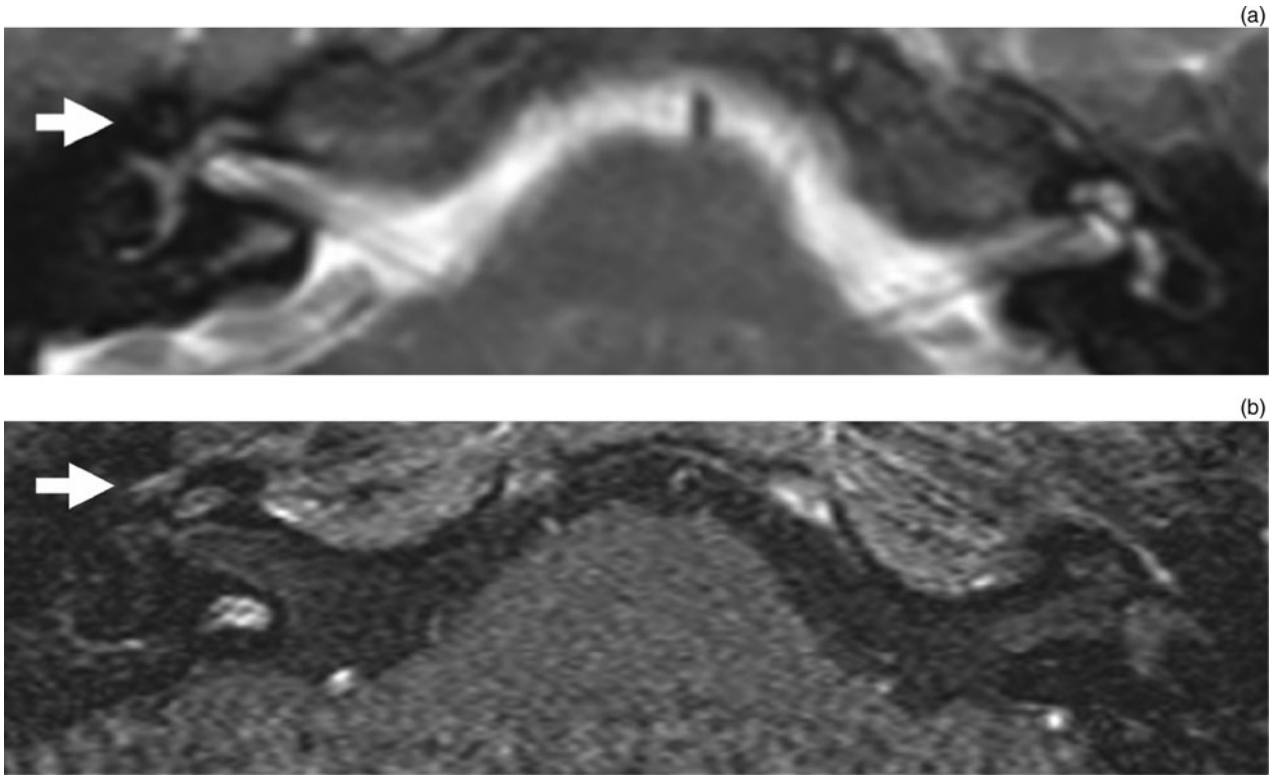

FIG. 3

(a) T2-weighted magnetic resonance imaging (MRI) scan demonstrating obliteration of the right cochlea. (b) Contrast-enhanced T1-weighted MRI demonstrating high signal intensity in the right membranous labyrinth, which indicates an acute, active stage of Cogan's syndrome (arrow). (The left cochlea had a normal appearance.)

- **Cogan's syndrome is characterised by non-syphilitic interstitial keratitis, Ménière-like episodes of vestibular symptoms, and bilateral sensorineural hearing loss**
- **This paper presents a young woman suffering from Cogan's syndrome and sudden, bilateral deafness. In Cogan's syndrome, careful radiological evaluation can classify the cochlear disease state into normal, active and ossification**
- **In Cogan's syndrome, cochlear implantation side selection depends on audiological results and radiological findings. If there is no audiological difference between the two sides, the surgeon should choose the side with a normal or active disease state rather than the side with an ossification state**

Discussion

This case demonstrates the problems encountered with cochlear ossification and the radiological findings that determine operative side selection. A histological study of the findings in Cogan's syndrome revealed acute labyrinthitis, with inner-ear tissue atrophy and diffuse fibrosis, such as that seen in other autoimmune diseases.[2] Pathological analysis of specimens from a patient with acute Cogan's syndrome revealed infiltrating lymphocytes and plasma cells in the cochlea and a generalised thickening of the membranous labyrinth.[6] In Cogan's syndrome, calcific obliteration and soft tissue obliteration of the intralabyrinthine fluid spaces can be observed radiologically. Surgeons should carefully evaluate thickening of the membranous labyrinth, using temporal bone CT, and should prepare for possible ossification and/or fibrosis in the cochlea.

Cochlear hypointensity on pre-contrast, T2-weighted images may represent an obliterative state or active cochlear disease. Hyperintensity inside the membranous labyrinth on the pre-contrast, T1-weighted images and enhancement on the contrast-enhanced, T1-weighted images probably represent an active disease state in Cogan's syndrome.[7] In contrast, the absence of high signal lesions and enhancement in pre-contrast or contrast-enhanced T1-weighted images indicate that there is no inflammation and/or ossification of the cochlea.

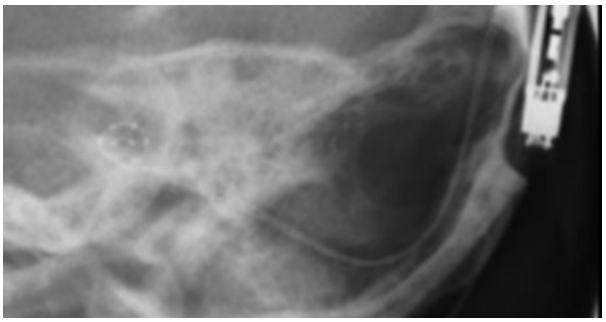

FIG. 4

A Combi 40+ device was implanted in the left ear.

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The presence of inner-ear enhancement did not predict the ossification of the cochlea, but it did indicate active cochlear inflammation and/or granulation. Although repeated inflammation can cause gradual ossification, the active disease state of Cogan's syndrome does not represent a contraindication to cochlear implantation, compared with ossific obliteration. Careful review of pre-operative radiological findings can enable appropriate selection of a suitable side for cochlear implantation.

Previous reports of cochlear implantation in Cogan's syndrome indicated that scala tympani ossification occurred in three out of a total of 12 cases, and that successful insertion into the scala vestibuli was possible in obliterated cases.[8–10] This can be explained by the fact that fibrosis of the cochlea may occur more frequently than does calcific obliteration, and that the area of greatest obliteration was the scala tympani of the basal turn.[11] Partial labyrinthine ossification has been reported in as many as 80 per cent of patients with profound, post-meningitic deafness.[11] However, cochlear ossification of deaf patients in Cogan's syndrome may occur less frequently, compared with patients with post-meningitic deafness, because autoimmune labyrinthitis is less severe than meningitic labyrinthitis.

In Cogan's syndrome, cochlear implantation side selection depends on the audiological results and radiological findings. If there is no difference between the two sides regarding audiological results, the surgeon should choose the side with a normal or active disease state rather than the side with an ossification state.

A special electrode, such as a compressed or split electrode, should be prepared pre-operatively for an obliterated cochlea. Also, a cochlear implant device that is appropriate for MRI scanning should be considered, due to the frequent MRI scanning required in autoimmune disease patients. Early intravenous antibiotics are needed because patients with Cogan's syndrome are more prone to infections due to their long-term steroid use. Inverted 'J' flap incisions, which provide a wide blood supply from the posterior auricular and occipital arteries, are recommended in order to prevent flap ischaemia. Long-term mastoid compressive dressings should be avoided in order to prevent skin ischaemia and pressure sores. Post-operatively, the surgeon should watch for skin atrophy, due to frequent use of steroids in autoimmune diseases, and flap ischaemia, caused by vasculitis.

Although these challenging problems should be considered in the management of patients suffering from Cogan's syndrome, the potential gain in auditory and speech abilities is significant. Cogan's syndrome is also associated with visual loss, which impedes adequate language communication. For these reasons, cochlear implantation is an excellent treatment modality for the profound hearing loss found in Cogan's syndrome.

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