Int. Adv. Otol. 2012; 8:(2) 311-316

CASE REPORT

Cochlear Implantation in Cogan's Syndrome

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ABSTRACT: Cogan's syndrome is a rare multi-systemic disease which is thought to be autoimmune in origin. It targets various organs, but all patients will suffer some degree of audiovestibular involvement at some point through the disease process, with many sustaining a profound sensorineural hearing loss.

This article describes three cases of bilateral profound sensorineural hearing loss due to Cogan's syndrome treated at the Manchester Cochlear Implantation Centre with a focus on surgical and audiological outcome from cochlear implantation. The symptomatology, aetiology, diagnosis, treatment and prognosis of Cogan's syndrome are also discussed with evidence from the world literature to highlight the key points of this uncommon syndrome.

Submitted : 27 September 2011 *Accepted : 20 February 2012*

Introduction

Cogan's syndrome was described eponymously in 1945 by the American ophthalmologist Dr. David Glendenning Cogan[1], despite the first case having been described by Morgan and Baumgartner in 1934. The syndrome consists of non-syphilitic interstitial keratitis of the eye, vertigo, tinnitus and profound deafness.

This syndrome is relatively rare, with less than 200 cases having been reported in the literature in the 66 years since its first description. It predominantly affects young adults in their 3rd or 4th decade with no clear gender predilection, but has been described in children as young as 3 years old[2] and adults up to 70 years of age[3].

Symptoms are variable but can be classified as ocular, audiovestibular and systemic. Ocular symptoms can precede or follow otological symptoms whilst systemic symptoms usually occur later in the disease process. Ocular and audiovestibular symptoms can occur simultaneously in 25% of cases with the majority (85%) of cases occurring within 2 years of each other[3]. The disease tends to have a relapsing and remitting course, with up to 62% of patients having multiple flare-ups[3].

The Manchester Cochlear Implant Programme was established in 1988 and has implanted over 1100 adults and children with profound hearing loss due to a variety of aetiologies. We present a series of patients who underwent cochlear implantation (CI) to rehabilitate severe to profound hearing loss, with emphasis on mode of presentation, response to initial treatment and audiological outcome following CI.

Case Reports

Case 1:

A 45 year old lady presented to the ENT department in 1999 with a 5 year history of progressive deterioration of hearing in both ears, and on audiometry had a profound sensorineural hearing loss in the right ear and a 'dead' left ear. She had initially had episodes of rotatory vertigo which had settled by the time of presentation but she was left with a persistent imbalance. She also had intrusive tinnitus in the left ear. In addition to the audiovestibular symptoms, she had bilateral iritis and uveitis and a sensorimotor neuropathy of the lower limbs secondary to vasculitis, and hence a clinical diagnosis of Cogan's syndrome had been made. She was on prednisolone 15mg daily and methotrexate 20mg weekly to control the systemic

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symptoms and steroid eye drops for her ocular symptoms.

At her first audiological assessment, aided speech discrimination testing confirmed that her open set discrimination was too good to warrant implantation (Institute of Hearing Research (IHR) sentences – 54% keywords correct). Bithermal caloric testing showed no residual vestibular function. MR scanning confirmed normal cochleas although there was high signal change in the white matter around the posterior horns of the lateral ventricles. A subsequent cortical evoked response audiometry (CERA) test showed normal function of the central pathways.

Over the following 4 years, regular audiological follow up showed a gradual progression of the hearing loss to a point when her IHR sentence speech discrimination had deteriorated to 17%. In 2003 she underwent cochlear implantation with a Nucleus Contour implant. The operation was uneventful with a full insertion of the electrode array into a patent scala tympani.

Audiological speech discrimination testing postoperatively showed City University of New York (CUNY) sentence discrimination improved to 36% at 1 week and 56% at 3 months. However, despite the usual intensive audiological rehabilitation programme, her hearing did not continue to improve. Her best audiological outcome indicated AB word recognition of 23% (words) and 46% (phonemes), Bamford-Kowal-Bench (BKB) sentence recognition 23% and CUNY sentence recognition 25%. Further testing 4 years after implantation showed that the best CUNY sentence recognition score was achievable with lip-reading and digital hearing aid (95%), compared to lip-reading, hearing aid and implant (87%) and was only 28% with implant alone. It was eventually established that she had never been compliant with cochlear implant usage, having been using the implant for 1-2 hours per day only. As such, this patient has become a non-user of the implant.

Case 2:

A 75 year old lady presented to the rheumatology department in with evidence of vasculitis and iritis and a mild sensorineural hearing loss in 2002. She was diagnosed with Cogan's syndrome and was started on prednisolone 25mg daily and regular steroid eye drops. Within months of the diagnosis she experienced sudden onset of bilateral profound sensorineural hearing loss. Audiological assessment 2 years later indicated that she was a candidate for cochlear implantation (hearing thresholds >90dB through all frequencies; CUNY sentence recognition of 3%). MR scanning confirmed normal cochleas and she was therefore consented for a left cochlear implant. In 2005 she underwent CI with a left Nucleus Contour (soft tip) implant inserted into a patent scala tympani. Her postoperative recovery was uncomplicated.

Audiological testing showed an improvement in BKB sentences to 64% and 81% at 3 months and 9 months post-operatively, respectively. Similarly, CUNY sentence speech discrimination scores improved to 89% at 3 months and to 94% at 9 months. After 4 years of follow up, she was using the implant most of the time and was able to understand telephone conversation with an unfamiliar voice. At that time, her Arthur Boothroyd (AB) word recognition was 27% (words) and 47% (phonemes), BKB sentence recognition was 65% and CUNY sentence recognition was 86%.

Case 3:

A 26 year old lady presented to her local ENT department in June 1994 with a sudden onset of acute labyrinthine failure and profound hearing loss affecting her right ear only. Within 5 weeks she suffered the same symptoms in her left ear resulting in total bilateral vestibulocochlear failure. Despite vasodilators and systemic steroids there was no recovery of cochlear function noted. Within 2 months, she experienced episodes of bilateral scleritis which led to the clinical diagnosis of Cogan's syndrome. Her symptoms of systemic vasculitis were controlled with a further course of prednisolone and her ESR returned to normal, but her hearing loss remained unchanged.

Audiology showed no response from either ear and caloric testing showed no response to either hot or cold stimuli. MR scanning confirmed normal cochleas and in 1995 she proceeded to implantation with a right Clarion cochlear implant. At surgery areas of ossification were noted in the basal turn of the cochlea but a complete insertion of the electrode array was

achieved. She had no postoperative complications. Audiological testing postoperatively showed a rapid and sustained improvement in word and sentence recognition with, in 2005, CUNY sentence recognition of 93%, BKB recognition of 96% and AB word recognition of 40% (words) and 66% (phonemes).

Discussion

Audiovestibular involvement commonly presents as sudden onset of symptoms suggestive of Meniere's disease: nausea, vomiting, rotatory vertigo, fluctuating hearing loss and intermittent tinnitus. Up to 50% of patients present with a sudden onset sensorineural hearing loss[3]. All patients will experience some degree of hearing loss in both ears at some point in the disease process. Vestibular system involvement can cause nystagmus, oscillopsia and ataxia. Caloric testing may show evidence of absent or weakened vestibular response in 90% of cases, of which 75% involves bilateral vestibular systems[3]. Initially the hearing loss may be unilateral but usually progresses to bilateral hearing impairment. Untreated, hearing loss becomes profound in weeks to months whilst vestibular symptoms usually improve or resolve completely over a few years.

Typically, the ocular symptom seen in classical Cogan's syndrome is interstitial keratitis[2, 3]. Other ocular manifestations include scleritis, episcleritis, iritis, conjunctivitis, retinal haemorrhage or vasculitis, choroiditis, papillitis, orbital pseudotumour and tendonitis. Ocular inflammation tends to occur in intermittent episodes, alternating with periods of quiescence; hence repeated examinations may be required to demonstrate evidence of ocular involvement.

Systemic symptoms can be non-specific (fever, fatigue, headache, arthralgia, myalgia and weight loss) or secondary to vasculitis involving large- and medium-sized vessels. Significant morbidity due to vasculitis occurs in up to 72%[4] with multiorgan involvement. Cardiological involvement resulting in aortitis and aortic insufficiency[3, 5] may prove fatal in up to 10% of cases[7].

The aetiology of Cogan's syndrome has not been fully ascertained. It is thought to be autoimmune in nature although it is unknown whether this is mounted through a cellular or humoral process[6]. This theory is based on findings such as lymphocytes and plasma cell infiltration in the cochlea and cornea, and presence of organ specific antibodies. Possible targets for the antibodies are Cogan's peptide (homologous to autoantigen Connexin 26 on inner ear epithelium) and SSa/Ro, laminin, ladinin, kinesin and calcineurin[4]. Corticosteroids and immunosuppressants have a beneficial effect on symptoms further supporting an autoimmune aetiology. It is postulated that the autoimmune condition is initiated by a hypersensitivity reaction to Chlamydia pneumonia, other Chlamydia species or Borrelia, as up to 40% of patients have evidence of an upper respiratory tract infection immediately preceding their symptoms, with increased antibody titres to Chlamydia pneumonia[4, 7].

There is no single diagnostic test for Cogan's syndrome. In addition to clinical signs and symptoms, various non-specific findings are often present such as anaemia, thrombocytopenia, eosinophilia and raised ESR[8]. Radiological investigations are used to exclude other abnormalities and as supporting evidence of Cogan's syndrome. Both high resolution computed tomography and magnetic resonance imaging are utilised to assess the bony anatomy and the more frequent finding of soft tissue obliteration[9] of the cochlea respectively[10-12].

Temporal bone pathology in these patients has shown a range of findings including endolymphatic hydrops, atrophy of the organ of Corti, infiltration of the spiral ligament, osteoneogenesis of the round window, spiral ganglion cell degeneration, stria vascularis degeneration, demyelination of the eighth cranial nerve and vasculitis of the internal auditory artery[10, 13-14]. Repeated inflammatory episodes can cause gradual fibrosis and ossification[15], as early as 8 weeks from onset of symptoms[12].

The initial treatment of choice is high dose steroids (1-2mg/kg/day prednisolone), tapering to a maintenance dose after 2-4 weeks or stopped completely within 3-4 months[2]. Prompt treatment of Cogan's syndrome is crucial for a good response[16, 17], as the hearing loss may be reversible if treatment is started early e.g. within 2 weeks of the start of symptoms[18]. Other studies have found that it is not uncommon for patients who initially respond to steroids to then experience

deterioration of hearing despite continuation of appropriate treatment[3]. The response of hearing loss to systemic steroids is variable and can be complete, partial or no response. Immunosuppression is required in patients whose hearing loss does not respond to systemic steroids and in patients with systemic vasculitis. Cyclophosphamide, azathioprine, methotrexate, cyclosporine and entanercept have all been used[2,3]. Other treatments used include intravenous immunoglobulins and ethambutol[3].

The prognosis of ocular symptoms is usually excellent, responding promptly and completely to topical steroids, with few cases of permanent significant visual impairment[2]. Audiological prognosis, by contrast, is poor. Even when steroids are given promptly, 50-85% of patients develop profound deafness bilaterally[2,3, 19,20] and 70-95% have a moderate to severe loss at 3-5 years after diagnosis[2,9].

Patients with Cogan's syndrome are ideal candidates for cochlear implants, as they are post-lingually deaf, have often used hearing aids during the progressive sensorineural hearing loss and the duration of profound deafness is often short[8]. In the literature to date, there are 9 case series describing outcomes for cochlear implantation in a total of 30 patients with Cogan's syndrome, the results of which are summarised in Table 1. It is difficult to directly compare data from each case series due to the variety of outcome measures used and the differing duration of follow up. Many of the studies do not indicate the pre-operative values for the outcome measures used; one could presume that these scores were sufficient to fulfil cochlear implantation criteria, but calculating an absolute measure of improvement is often not possible.

Most studies have shown that cochlear implantation increases speech perception[4], word discrimination and sentence recognition in patients with Cogan's syndrome[2,3,19,21,22]. Indeed, the majority of case series in the literature show an excellent outcome with cochlear implantation in patients with Cogan's syndrome, as confirmed with our second and third cases whose sentence recognition improved to 94% and 93% respectively. However, some published studies report a wide range in their outcome measure[9,12,20,21] which demonstrates that patients with Cogan's syndrome do not universally have good audiological outcomes with a cochlear implant. The results from our first case (sentence recognition improving from 17% preoperatively to 25% at 12 months) concur with these findings.

Our patient went on to become a non-user of her implant due to the lack of improvement of speech perception. This could be attributed to her reluctance to use the implant on a regular or prolonged basis, but despite intensive audiological input this did not improve her compliance. It is unlikely that the 9 year duration of deafness before implantation in this patient is responsible for the failure of audiological rehabilitation as other studies[9,21] have shown periods of profound deafness of up to 15-19 years with no apparent negative effect on audiological outcome.

Cochlear implantation can be a technically challenging operation if cochlear obliteration has already occurred. The presence of cochlear obliteration should be identified preoperatively to allow appropriate planning and counselling of the patient but it is not an absolute contraindication to implantation[12,22], as we found in our third case. In our case series there were no postoperative complications but Cote et al have reported a complication of flap ischaemia, skin atrophy and pressure sores in their case report. Such complications could be due to vasculitis[20] or possibly the immune suppression from prolonged use of steroids or systemic immunosuppressants[23]. Pasanisi et al reported 2 patients who had an acute exacerbation of ocular symptoms in the immediate post-operative period, thought to be due to the stress of the surgery[22].

Conclusion

Patients with Cogan's syndrome typically present with a combination of audiovestibular and ocular symptoms and they are at risk of developing significant systemic symptoms. The diagnosis is largely based on clinical findings and the initial treatment of choice is high dose systemic steroids. In the presence of irreversible profound sensorineural hearing loss they are good candidates for cochlear implantation, although they are subject to variation in audiological outcome as with any other indication for implantation. The presence of cochlear osteoneogenesis or obliteration should be

Table 1. Cochlear implantation outcomes in Cogan's syndrome

Study	Number of patients	Age range (years)	Duration of deafness (months)	Outcome measure use	Pre-doperative score	Post-operative score	Operative complications
Cote[20], 1993	1	41	6	CID sentence recognition	0/200	110/200	Flap necrosis
				Iowa sentence recognition	0/88	35/88	
				Open set word recognition	0/50	15/50	
				Open set phoneme recognition	0/150	57/150	
Minet[9], 1997	4	32-56	12-180	Hearing threshold	>100dB	40-60dB (mean)	None
				Founiers phonemes	NS	42.5-100%	
				Comberscures phrases	NS	95.5-100%	
Cinamon[21], 1997	3	36-53	18-228	Open set 2-syllable word recognition		0-100%	None
				Open set single syllable word recognition		0-50%	
				Open set sentence recognition		0-96%	
Pasanisi[22], 2003	5	27-52	6-36	Open set word discrimination	0-17%	79-98%	None
				Sentence recognition	0-30%	85-100%	(Acute increased ocular symptoms in 2 patients)
Aschendorff[12], 2004	6	31.5 (mean)	1-132	Word recognition	NS	75-100%	None
				Monosyllable recognition	NS	25-80%	
Gluth[3], 2006	8	38 (mean)	NS	Sentence recognition	NS	78-100%	None
Vishwakarma[4], 2007	1	58	25	Speech reception threshold	NS	35dB	None
				Optimal speech discrimination score	NS	60%	
Im[15], 2008	1	25	6	Open set word discrimination	NS	91%	None
				Sentence recognition	NS	96%	
Kawamura[8], 2010	1	55	26	Monosyllable recognition	NS	80%	None
				Word recognition	NS	78%	
				Sentence recognition	NS	79%	

NS: Not stated

identified prior to proceeding to implantation. There appears to be no significantly increased risk of operative complications in Cogan's syndrome compared to implantation for other aetiologies, but close follow-up in the post-operative period allows for early management of any vasculitic complications.

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