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Management of Idiopathic Sudden Sensorineural Hearing Loss: Experience in Newly Developing Qatar

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Abstract: Idiopathic sudden sensorineural hearing loss is a medical emergency that requires urgent diagnosis and treatment, and the adaptation of a proper protocol for management is a priority. In most cases, such treatment is rather controversial and depends on a variety of factors. The aim of this study was to determine and identify as early as possible those factors that play the important role in the prognosis of the condition, to describe the experience, and to suggest a treatment protocol that can be adopted in a tertiary hospital, such as Hamad General Hospital. Our study was retrospective and descriptive. It was conducted in the ear, nose, and throat outpatient clinics at Hamad General Hospital and the ear, nose, and throat wards at Rumailah Hospital. We enrolled a total of 21 patients with idiopathic sudden sensorineural hearing loss. The treatment protocol that was adopted—consisting of high-dose steroid therapy, full-dose antiviral drug (acyclovir), and a histamine analog, betahistine—resulted in hearing improvement in 57.4% of cases. Then the possible good and bad prognostic factors were discussed. The results of our study revealed that the steroid therapy protocol practiced in Qatar resulted in hearing improvement in patients with idiopathic sudden sensorineural hearing loss. Good prognostic factors include early diagnosis, marked reduction of symptoms, and improved shape of the audiometric curve.

Key Words: sensorineural hearing loss; sudden hearing loss; treatment protocol

Idiopathic sudden sensorineural hearing loss (ISSNHL) [1] is a well-known clinical condition characterized by a sudden occurrence in one or both ears of sensorineural hearing loss of more than 30 dB in at least three contiguous audiometric frequencies over a time course of no more than 72 hours. Its etiology is rather obscure and may involve a vascular, viral, or autoimmune cause [2]. Because of its various etiological possibilities and its prognostic characteristics, urgent attention and treatment are required, and the condition may be considered as an emergency. However, because of controversies regarding etiology and the need for urgent treatment, various diagnosis and treatment protocols were advocated by different centers. Recently, increasing interest is centered on the means of diagnosis and treatment, leading to the emergence of quite a variety of protocols adopted by different treatment centers.

Hearing impairment is a common problem worldwide and even more problematic in developing countries. A report prepared by the Department of Otolaryngology [3] at University of Texas Medical Branch documented approximately 15,000 reported cases of ISSNHL per year worldwide, with 4,000 of those cases occurring in the United States. One in every 10,000–15,000 people experience this condition, with the highest incidence occurring between 50 and 60 years of age. The lowest incidence is between ages 20 and 30 years. Of the patients suffering from ISSNHL, 2% have bilateral ISSNHL. In most series, the incidence was nearly equal in both men and women.

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Table 1. Distribution of Characteristics of Study Subjects (N = 21) with Idiopathic Sudden Sensorineural Hearing Loss

Variables	Number (%)
Gender	
Male	17 (80.9)
Female	4 (19.1)
Age group (years)	
20–29	1 (4.8)
30–39	4 (19.4)
40–49	8 (39.1)
50–59	6 (28.6)
60–69	2 (9.5)
Frequency of associated symptoms	
Tinnitus	16 (76.2)
Dizziness	13 (61.9)
Feeling of pressure	3 (14.3)
Ear ache	1 (4.8)
Headache	1 (4.8)
Ataxia	1 (4.8)
Duration	
<1 day	4 (19.0)
<2 weeks	7 (33.3)
<3 weeks	3 (14.3)
2–4 weeks	3 (14.3)
>4 weeks	4 (19.0)
Severity of hearing loss	
Mild	6 (28.6)
Moderate	5 (23.8)
Severe	3 (14.3)
Very severe	2 (9.5)
Profound	3 (14.3)
No response	2 (9.5)

Table 2. Investigation Results of Study Subjects (N = 21) with Idiopathic Sudden Sensorineural Hearing Loss

	ISSNHL Patients	
Variable	Normal No. (%)	Abnormal No. (%)
Audiological investigations		
Pure-tone audiometry	0 (0.0)	21 (100.0)
Tympanometry	21 (100)	0 (0.0)
Acoustic reflex	8 (38.1)	13 (61.9)
Speech audiometry	3 (14.3)	3 (14.3)
ABR	9 (42.8)	0 (0.0)
Laboratory investigations		
CBC	21 (100.0)	0 (0.0)
ESR	21 (100.0)	0 (0.0)
Blood film	21 (100.0)	0 (0.0)
Coagulation screening	21 (100.0)	0 (0.0)
Fasting blood glucose	15 (71.4)	6 (28.6)
VDRL	7 (33.3)	0 (0.0)
IM test	7 (33.3)	7 (33.3)
Immunoglobulin G	19 (90.5)	2 (9.5)
Immunoglobulin A	20 (95.2)	1 (4.8)
Immunoglobulin E	20 (95.2)	1 (4.8)
Radiological investigations		
Chest radiography	21 (100.0)	0 (0.0)
Computed tomography scan	69 (28.6)	0 (0.0)
Magnetic resonance imaging	17 (81.0)	4 (19.0)

ABR = auditory brainstem response; CBC = complete blood cell count; ESR = erythrocyte sedimentation rate; ISSNHL = idiopathic sudden sensorineural hearing loss; IM = infectious mononucleosis.

Sensorineural hearing loss [4–9] (or nerve-related deafness) involves damage to the ear caused by aging, prenatal and birth-related problems, viral and bacterial infections, heredity, trauma, exposure to loud noise, fluid backup, or tumor in the inner ear. Almost all ISSNHL can be effectively treated with hearing aids. The prevalence of hearing loss increases with age; most hearing losses develop over a period of 25–30 years.

ISSNHL may be a symptom of cochlear injury. It is characterized by sudden onset and, within a few hours, it reaches its maximum peak. It may be accompanied by vertigo and tinnitus. Many hypotheses have been advanced to explain its etiology: viral inflammation, vascular diseases, allergic reaction, rupture of intralabyrinthine membranes, and autoimmune diseases. The decrease in hearing may be unilateral or bilateral. Many studies have been carried out to assess the histopathology of the labyrinth in cases of ISSNHL and to provide a better understanding of the etiopathogenesis [10]. Finally, most causes of ISSNHL remain unknown; thus the term *idiopathic* is applied to these cases.

To date, no published study has addressed the incidence of ISSNHL in Qatar. Therefore, we have a pressing need to study the causes of ISSNHL and suggest a treatment protocol for ISSNHL. The aim of this study was to identify patients with ISSNHL as early as possible, to describe their experience, and to suggest a treatment protocol that can be adopted in a tertiary hospital, such as Hamad General Hospital.

PATIENTS AND METHODS

This retrospective study was conducted from January 2002 to December 2003 to identify patients with ISSNHL. We diagnosed ISSNHL in 21 patients (aged 24–67 years) who were fully evaluated for their main complaint; associated symptoms; disorder duration; and family history of similar illness and of such conditions as multiple sclerosis, upper respiratory tract infection, and other viral infections. Reports on clinical, ear, nose, and throat and neurological examinations were registered. Audiological evaluation, including pure-tone audiometry, tympanometry, speech audiometry, and auditory brainstem-evoked response testing results were included. We also recorded other results—of full blood count coagulation screening; blood biochemistry;

blood serology (including VDRL testing and immunoglobulin study); radiologic and magnetic resonance imaging; and follow-up hearing tests; and we tracked the treatment protocols adopted.

We collected data on the 21 patients with ISSNHL from their medical records and analyzed them for age range and median, associated symptoms, clinical findings, duration of the complaint, pure-tone audiometric curve, shape of audiogram, severity of hearing loss at speech frequencies (average HTL [hearing threshold level] for 0.5, 1, and 2 kHz), HTL at low frequency (0.25 kHz), and HTL at high frequencies (4 and 8 kHz).

The degree of improvement was graded as follows:

1. Grade 1: No change in HTL (comprises seven patients)
2. Grade 2: Mild improvement; mild but noticeable clinical recovery and improvement in HTL of up to 15 dB for speech frequencies (comprises two patients)
3. Grade 3: Moderate improvement; clearly noticeable clinical reduction of symptoms and improvement of HTL by more than 20 dB without return of audiometric curve to normal (comprises two patients)
4. Grade 4: Return of HTL to normal (averaged speech frequencies comparable to the unaffected side) with or without complete improvement of symptoms (comprises eight patients)
5. Grade 5: No results recorded (comprises two patients)

RESULTS

Table 1 shows the distribution of characteristics of the studied patients with ISSNHL. Of the studied subjects, 81% were male and 19% were female (median age, 43 years; range, 24–67 years). With respect to age, we observed that the prevalence of ISSNHL was 38.1% in the age group 40–49 years, followed by 28.6% in the age group 50–59 years. All patients except one had unilateral ear involvement and revealed almost equal frequency of involvement for both left and right ears. Frequently identified associated symptoms were tinnitus (76.2%), dizziness (61.9%), and feeling of pressure (14.28%). Earache, headache, and ataxia each represented 4.76%. In respect to the duration of illness, 33.3% of the studied patients had the complaint for 2 weeks, and 19% had it for more than 4 weeks.

Table 2 shows the investigation results of the study subjects. Pure-tone audiometry results were abnormal in all patients. Tympanometry showed a type-A curve in all patients, and acoustic reflex results were negative in the affected ear in 13 patients. Only three patients had

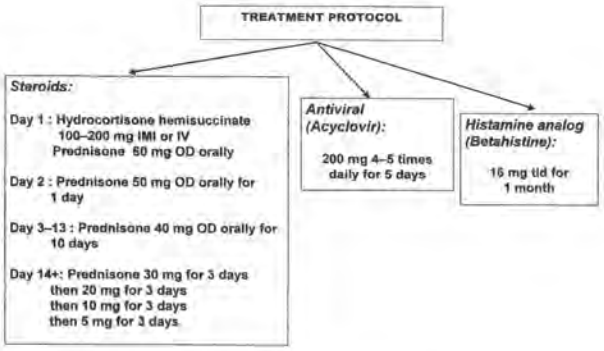

Figure 1. Treatment protocol.

speech audiometry and, for auditory brainstem response, results in nine patients were normal except for decreased HTL. With respect to laboratory investigations, results of complete blood cell count, erythrocyte sedimentation rate, blood film for abnormal cells, and coagulation screening were normal in all patients. A total of 28.6% of the patients were diabetic. Serological investigation results were negative in all tested patients.

Radiology and imaging investigations revealed that all patients had normal chest x-ray results; six patients had normal computed tomography scans; and, of all patients, four had abnormal cranial magnetic resonance imaging (MRI) results: one patient with small lacunar infarcts in the temporal and parietal regions, two patients with small pontine infarcts, and one patient with obliteration of the left common carotid artery (diagnosed as Takayasu's disease).

Figure 1 shows the treatment protocol adopted for patients. Twelve patients (57.4%) showed variable degrees of improvement; of these, eight (37.6%) had full improvement (grade 1) with return of the audiometric curve to normal hearing. Two patients (9.5%) showed mild improvement (grade 2), and another two patients (9.5%) exhibited reasonable improvement (grade 3). Six patients of the grade 4 group had flat, basin-shaped

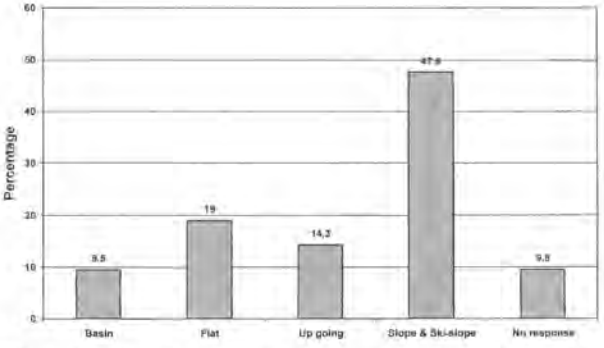

Figure 2. Shape of audiometric curve in patients with idiopathic sudden sensorineural hearing loss.

or up-turning audiometric curves (Fig. 2). In ten of the twelve patients who showed improvement, the duration of onset was less than 14 days and was less than 7 days in six of eight patients in grade 4.

We found no age relation or significant side predilection, and the duration of improvement was variable, extending from less than 1 week to up to 6 weeks. Of the seven patients (33.3%) who showed no improvement in their hearing both audiologically and clinically (grade 1), six patients had dizzy spells when first seen, four had audiometric HTLs of 65 dB or no response at 8 kHz, five had high-frequency sloping audiometric curves, and all showed no improvement at 6 weeks' duration.

The prognosis of tinnitus involved full recovery in all but one of the grade 4 patients; the unrecovered patient had at 8 kHz a dip that did not improve. Incidentally that patient developed recurrence of hearing loss 12 week later but in the opposite ear, which was also treated with steroid therapy and showed remarkable improvement. Tinnitus decreased but did not disappear in patients in grades 2 and 3.

All but three patients who developed dizziness (those with very severe hearing loss) had fully recovered in a few days; the unrecovered group had dizzy spells that lasted more than a few days. We have no follow-up notes on the rest of their symptoms. Long-term follow-up revealed recurrence of hearing loss in four patients: One had established autoimmune, fluctuating-type ISSNHL; the second developed recurrent hearing loss in the contralateral side in 12 weeks' time and improved with a second dose of steroid therapy; and the third and fourth had low-frequency loss unilaterally and improved with betahistine and diuretic therapy for 1–4 weeks. Vestibular testing was not done.

DISCUSSION

ISSNHL is a symptom of cochlear injury and is characterized by sudden onset; within a few hours, it reaches its maximum peak. ISSNHL is the most intractable of hearing loss types, because its fundamental cause is hair cell death. Spectacular advances have recently been made in our understanding of hair cell physiology and molecular biology, offering new hope of alleviating ISSNHL [10].

Although called *sudden*, hearing loss seems unlikely to be abrupt but rather probably evolves over a few hours [6]. It occurs most frequently in the age 30–60 group and affects the genders equally. Also, we observed in our study that most patients who had ISSNHL ranged in age from 40 to 60 years, although the highest rate was found among men.

Research conducted by the Institute of Audiology in Turin documented that 5,925 cases were diagnosed over a 5-year period (1990–1995) [7]. The researchers concluded that sudden idiopathic hearing loss, often attributed to general vascular causes, was one of the most frequent reasons for monolateral hearing loss. Monolateral sensorineural hearing loss, which is not seriously disabling, represents a handicap from the social point of view.

Sudden hearing loss can affect different people very differently. ISSNHL is usually unilateral and is often accompanied by tinnitus, vertigo, or both. The degree of hearing loss may vary from mild to severe and may involve different parts of the hearing frequency range. Roughly one-third of people with ISSNHL awaken in the morning with a hearing loss [5]. Our study revealed that the most frequently identified associated symptoms were tinnitus (76.2%) and dizziness (61.9%). The incidence of tinnitus compares well with that in the literature [11,12]; however, tinnitus did not assist well in determining which patients would suffer hearing loss. However, a high incidence of associated dizziness may be responsible for poor recovery incidence in our series, so it can be considered to indicate a poor prognosis [13–15].

Many causes can trigger sudden hearing loss [6–10] but, despite extensive evaluation, the majority of cases elude definitive diagnosis and, therefore, remain idiopathic. Reports estimate that the etiology of ISSNHL is diagnosed in only 10% of the cases. Suggested causes of ISSNHL include viral infections, immunologic vascular compromise, and intracochlear membrane breaks. We observed in our study that some of the causes of sudden or fluctuating sensorineural hearing loss were meningitis, multiple sclerosis, or Friedreich's ataxia. Also, our study showed that the good prognostic signs were early diagnosis; low-severity hearing impairment; and flat, basin-shaped, and up-turning audiometric curves, whereas poor prognostic signs were late presentation of patients, late diagnosis, history of very severe to profound or total hearing loss, high-frequency hearing loss (> 60 dB), and high-frequency sloping curve. In all the studied cases, patients had an intact tympanic membrane and revealed no other ear, nose, and throat abnormality.

CONCLUSION

Our study revealed that the treatment protocol involving steroid therapy in Qatar resulted in hearing improvement in 57.4% of patients with ISSNHL. Furthermore, we concluded that results can be improved if the disorder is diagnosed early in patients using proper protocols and if early treatment is undertaken.

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