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Idiopathic (benign) intracranial hypertension-induced sudden hearing loss: A rare case

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The authors stated that the written consent was obtained from the patient presented with images in the study.

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Abstract

Sudden hearing loss (SHL) is a clinical condition characterized by the acute onset of sensorineural hearing loss. Although many etiological factors have been reported, it is primarily idiopathic. Idiopathic Intracranial Hypertension (IIH) is a syndrome that presents signs of increased intracranial pressure in the absence of any intracranial lesion, meningeal inflammation, or venous obstruction. In our case, a patient diagnosed with SHL presented with hearing loss and visual field constriction. An investigation into etiological factors identified IIH. Although rare, IIH can cause SHL. It is crucial to consider this possibility during the diagnostic process and carefully plan the treatment based on the potential etiology.

Keywords: sudden hearing loss, pseudotumor cerebri, ıdiopathic, benign ıntracranial hypertension, ıntracranial causes of sudden hearing loss, sensorineural hearing loss

Introduction

Idiopathic intracranial hypertension (IIH) is a syndrome that presents with symptoms and signs of increased intracranial pressure in the absence of intracranial mass lesions, meningeal inflammation, or venous obstruction [1,2]. The incidence of IIH is 0.9 per 100,000, with a higher prevalence amongst obese women of childbearing age. Sudden hearing loss (SHL) is characterized as a sensorineural hearing loss of a minimum of 30 dB at three consecutive frequencies occurring within 72 h. This condition requires prompt diagnosis and treatment [3,4].

In this case report, informed consent was obtained from the patient, with their information kept confidential.

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Case presentation

A 56-year-old female patient presented at an outpatient clinic with complaints of hearing loss, tinnitus, and headaches in both ears that had begun 3 days prior. The patient had no history of systemic disease, trauma, or acute infections, except for hyperlipidemia and diabetes mellitus.

The otoscopic examination yielded normal results. The audiological evaluation revealed severe sensorineural hearing loss, with thresholds of 85/68 dB in the right ear and 86/69 dB in the left. The patient was diagnosed with sudden hearing loss (SHL) and began treatment with 1 mg/kg intravenous prednisolone. Informed consent was obtained. The patient continued her oral medications, which included metformin, gliclazide, and pitavastatin, for the management of diabetes mellitus and hyperlipidemia.

The patient was referred to neurology due to headaches. A mini-mental state examination was conducted, which came back normal and the patient was found to be alert and cooperative. Fundoscopy revealed visual field constriction and enlarged blind spots, thus leading to a referral to ophthalmology. An examination by the ophthalmologist showed visual field constriction in the left eye and swelling of the optic nerve. Thyroid function tests, B12, folate, sedimentation, and homocysteine levels were found to be within the normal range.

A radiological examination, which included contrast-enhanced temporal, cranial, and orbital MRI, was performed. No intracranial mass lesion or retrocochlear pathology was detected in the cranial and temporal contrast-enhanced MRI. The sagittal sections of the orbital MRI revealed increased tortuosity of the bilateral optic nerves. Cranial MR venography and Visual Evoked Potential (VEP) were conducted to confirm the increased intracranial pressure. The MR venography showed increased intracranial pressure, while VEP reported normal bilateral P100 latencies and amplitudes. These findings led to the conclusion that the sudden hearing loss and visual impairment in the right eye were caused by the increased intracranial pressure. The patient was subsequently transferred to the neurology department for further investigation and treatment of IIH. A lumbar puncture was carried out to reduce intracranial pressure and collect cerebrospinal fluid (CSF) for Anti-myelin Oligodendrocyte Glycoprotein IgG (MOG-IgG) testing. However, the patient continued to experience headaches, visual disturbances, and hearing loss, necessitating an adjustment in medical treatment to diazoxide 250 mg and topiramate 25 mg. The patient was then discharged.

A behind-the-ear hearing aid was prescribed for severe bilateral sensorineural hearing loss. However, even after three months of use, the patient-derived no benefit from the hearing aid, necessitating a cochlear implant procedure at our clinic.

Discussion

SHL is defined as sensorineural hearing loss of at least 30 dB across at least three consecutive frequencies, occurring within three days or less [4,5]. SHL affects both genders equally, primarily between the ages of 30–60, and accounts for 1% of all sensorineural hearing losses. The annual incidence of SHL is estimated to be between 5 and 20 cases per 100,000 people [6]. However, the actual incidence may be higher due to the low hospital admission rate associated with patients demonstrating a spontaneous recovery tendency. SHL is typically unilateral, affecting one ear in 90% of cases. In an estimated 80–90% of SHL cases, no identifiable cause can be found, and these cases are termed “idiopathic sudden hearing loss.” Recognizable causes of SHL include infections, neoplasms, vascular diseases, trauma, labyrinthine membrane ruptures, pharmacological toxicity, immunological disorders, and, rarely, neurological diseases [7].

Corticosteroids are the most commonly used drugs in the treatment of idiopathic SHL. Prednisolone is typically started with an oral dose of 1 mg/kg/day and gradually reduced [8]. If a cause is identified for SHL, treatment should be planned accordingly.

IIH is typically a self-limiting clinical condition. Although the etiology remains unknown, various mechanisms have been proposed. Reports suggest that the increase in intracranial pressure is a result of an escalation in cerebral blood flow, cerebral blood volume, and CSF production. This leads to compromised cerebral microcirculation and the development of intracellular or extracellular edema [9,10].

IIH is common in obese women of childbearing age, with a prevalence of 0.9 per 100,000 in the general population. The female/male ratio varies between 4.3/1 and 8/1. Female gender and obesity are major risk factors for IIH [9,10]. IIH has also been reported during pregnancy; menstrual disorders; antibiotic and contraceptive use; iron deficiency anemia; Behçet’s disease; systemic lupus erythematosus; protein C and S deficiency; antiphospholipid antibody syndrome; Addison’s disease; hypoparathyroidism; endocrine disorders (such as obesity); hypervitaminosis A (due to excessive liver consumption);the use of isotretinoin, synthetic growth hormone, and tetracycline; and steroid withdrawal [10].

Clinically, IIH manifests with symptoms such as headaches, visual loss, pulsatile tinnitus, and diplopia. Additional minor symptoms may include neck pain, paresthesia, arthralgia, and ataxia.

It is estimated that approximately 70% of IIH patients experience visual disturbances. Serial visual field testing holds significant importance in monitoring optic nerve function. Optic nerve compression resulting from increased CSF pressure can lead to visual disturbances [11]. Papilledema represents the most critical finding. Other than sixth cranial nerve palsy, neurological examination findings are typically normal.

The vestibulocochlear nerve, similar to the optic nerve, is also susceptible to compression, which explains the hearing and balance symptoms displayed by patients with IIH [11].

Unilateral tinnitus is one of the earliest symptoms reported by 70% of patients with IIH. Therefore, it is important to include IIH in the differential diagnosis for patients presenting with tinnitus. In IIH, tinnitus is believed to result from pressure exerted by brain structures on the venous sinuses, disrupting laminar blood flow and converting it into turbulent flow. This is perceived as pulsatile tinnitus in one or both ears [12].

A significant amount of literature has demonstrated that low-frequency hearing loss is more prevalent in patients with IIH. It is believed that the cause of hearing loss in IIH is due to the transmission of elevated intracranial pressure to the perilymph via the cochlear aqueduct. Rarely, IIH patients may also present with

symptoms typical of endolymphatic hydrops such as vertigo, hearing loss, tinnitus, and aural fullness. The cause of these symptoms correlates to the increase in fluid pressure (both endolymph and perilymph) in the inner ear, proportional to the increase in intracranial pressure. Notably, these symptoms tend to improve after the normalization of CSF pressure [13].

To accurately diagnose IIH, it is necessary to consider the patient's history, including medication use, carry out ophthalmological and neurological examinations, measure CSF pressure, utilize neuroimaging to confirm the presence of normal or small ventricles and establish that no mass lesion is causing increased intracranial pressure. Radiologically, normal or small ventricles, as well as normal CSF composition, are characteristic features of this condition.

The principal aim of medical treatment is to decrease intracranial pressure. Carbonic anhydrase inhibitors (CAIs), often employed for this purpose, reduce CSF pressure by lessening CSF secretion from the choroid plexus. This process requires carbonic anhydrase for CO_2 hydration in active CSF secretion. CAIs act by blocking the dehydration of carbonic acid into water and CO_2. There is not an ideal dosage, and it is adjusted according to the patient's symptoms and observations. Typically, treatment commences with a dosage of 0.5–1 g/day, which is progressively increased until the patient's symptoms improve and the dosage is tolerated [14].

Topiramate, a partial CAI, has gradually found use in the off-label treatment of IIH, either as an adjunct or an alternative to CAIs, owing to its headache-reducing and weight-loss effects. Alongside CAIs, furosemide may be incorporated in IIH treatment to leverage its diuretic properties. Furosemide (20–100 mg/day) can also diminish CSF secretion from the choroid plexus [14].

The role of corticosteroids in the treatment of IIH is controversial. While they are sometimes used in IIH treatment, both their usage and subsequent withdrawal can potentially exacerbate the disease. Surgical treatments are considered for IIH cases that are unresponsive to medical treatment. Options for surgical intervention in IIH include lumboperitoneal and ventriculoperitoneal shunt procedures, optic nerve sheath fenestration (ONSF), bariatric surgery, and venous sinus stenting [14].

The pathogenesis of IIH remains unclear. Reasons why the disease follows a benign course in some patients while displaying rapid progression in others are still unknown. At present, no definitive treatment for IIH exists. Despite surgical treatments being considered when medical interventions fail, the high rate of complications necessitates caution when considering these options. Current research is exploring the efficacy of treating venous sinus disease and endovascular stenting in the context of IIH.

Conclusion

In conclusion, the etiology of SHL, tinnitus, and accompanying left eye visual field reduction in this case was identified as IIH. Treatment was planned accordingly. The CSF pressure was reduced by lumbar puncture, and severe bilateral hearing loss was managed with cochlear implantation. The hearing thresholds improved from 85/68 dB to 86/69 dB in audiological testing. When dealing with cases of SHL, IIH should be taken into consideration. Treatment should be planned according to the identified etiology, as demonstrated in this case.

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