Case Report

Bilateral Subdural Hematomas and Hearing Disturbances Caused by Spontaneous Intracranial Hypotension

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Spontaneous Intracranial Hypotension (SIH) is an uncommon headache syndrome. Patients classically present with orthostatic headache, tinnitus, and diplopia. The authors reported a 43 year-old man who presented with orthostatic headache, tinnitus, and hearing impairment for 3 months. Physical examination was unremarkable except for auditory impairment. The audiogram revealed minimal low-frequency neurosensori hearing loss suggesting a cochlear lesion. Computed tomography of the brain revealed bilateral thin chronic subdural hematomas. He underwent burr-hole surgery. Headache and auditory symptoms persisted and reevaluation of this syndrome was performed. MRI of the brain showed diffuse smooth enhanced dura mater, low lying position of midbrain, pons, medullar and cerebellar tonsil, as well as enlarged pituitary gland compatible with low CSF pressure syndrome. MRI of the whole spine could not demonstrate the site of CSF leakage. The patient was much improved after conservative treatments with hydration and bed rest. One year after treatment, he had no headache and only mild tinnitus was reported.

Keywords: *Spontaneous intracranial hypotension, Bilateral subdural hematomas, Tinnitus, Hearing loss, Audiogram*

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Spontaneous Intracranial Hypotension (SIH) is an uncommon headache syndrome caused by leakage of cerebrospinal fluid (CSF) through tiny dural defects usually at the level of the spine[1-4]. Although the cause of dural defects is rarely identified, it was hypothesized to be related with minor trauma or fragile dura mater[5-7], caused by preexisting connective tissue diseases[8,9]. Patients classically present with orthostatic headache, tinnitus, and diplopia. Some patients also have hearing impairment, blurring of vision and facial numbness or facial weakness[3]. Other rarer reported clinical presentations include non-orthostatic headache, dysgeusia[10], hyperprolactinemia[11], ataxia[12,1], parkinsonism[12], frontotemporal dementia[13], subdural hematoma[1,4,14-16], or even stupor and coma[17]. The authors reported a case of SIH who presented with subdural hematoma and hearing impairment. Detailed history taking and appropriate neuroimaging uncovered SIH as the cause of the problems.

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Case Report

A 43-year-old Thai male engineer had a 3-month history of orthostatic headache, tinnitus, and subsequently developed hearing impairment. The orthostatic headache began 3 months before admission and was characterized by symmetrically occipital pain radiating to temporal and orbital areas. It usually lasted for 30 minutes and was relieved in supine position and worsening during sitting up or coughing and sneezing. Headache progressed and became severe causing him to give up his work during the attacks. Three weeks after the onset of headache, he deve loped tinnitus, following by impaired hearing acuity bilaterally. His past history was unremarkable, except a 3-year history of noise exposure, due to his working environment in an industrial plant. He consulted an ENT specialist and an audiogram was

performed, which revealed minimal low-frequencies (250 Hertz and 500 Hertz) sensori-neural hearing loss with the hearing threshold of 25 decibel (dB) on the right and bilateral high-frequency (4kHertz and 8kHertz) sensorineural hearing loss. Computed tomography (CT) of the brain revealed bilateral thin SDH (Fig. 1). The routine biochemistry and coagulogram were unremarkable. He underwent burr-hole surgery at the left frontal region with removal of 40 ml of subdural clotted blood. The headache was slightly improved, but the hearing problem persisted. Neurological evaluation after surgery including vital sign and direct ophthalmoscope revealed no abnormalities. MRI of the brain with gadolinium enhancement showed diffuse smooth enhanced dura mater (Fig. 2), downward displacement of midbrain, pons, medullar and cerebellar tonsils as well as enlarged pituitary gland (Fig. 3), compatible with low CSF pressure syndrome.

SIH causing SDH was diagnosed. MRI of the whole spine with gadolinium enhancement revealed no evidence of dural defects. Radionuclide cisternography was not performed. He was managed conservatively with intravenous hydration and bed rest. His headache was gradually resolved. Three months later, tinnitus became less severe while hearing ability was much improved. The follow-up audiogram at 3 months showed normal hearing ability with hearing threshold level of 10 decibel at low frequencies (250 Hertz and 500 Hertz).In addition, hearing thresholds at high frequencies (4kHertz and 8kHertz) had also improved. Upon 1-year follow-up, he reported only mild tinnitus and audiogram showed no significant difference from the previous audiogram at 3 months. The follow-up audiograms revealed nearly complete recovery of the hearing impairment (Fig. 4-6). No symptoms of recurrent SIH were observed during one-year follow-up.

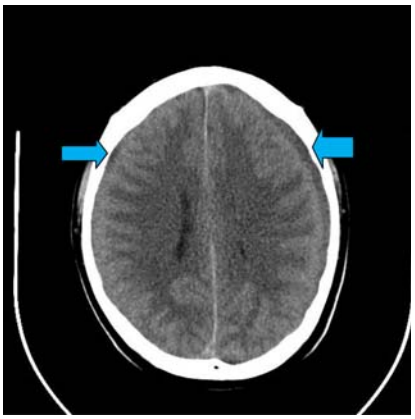

Fig. 1 Non contrast CT brain showed bilateral chronic subdural hematoma

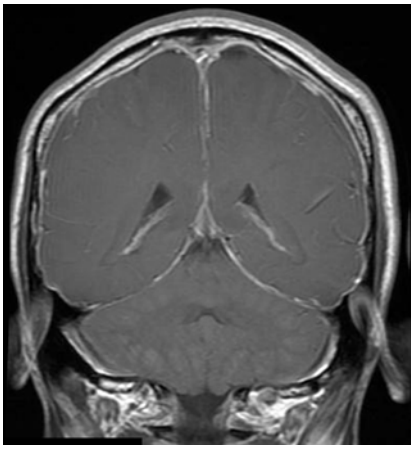

Fig. 2 Gadolinium T1WI MRI coronal view showed diffuse enhancement of the pachymeninges

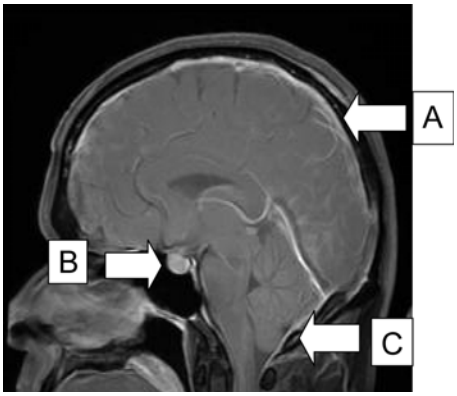

Fig. 3 Gadolinium T1WI MRI sagittal view showed [A] enhancement of the pachymeninges, [B] enlargement of pituitary gland, [C] downward displacement of cerebellar tonsil and brainstem

Discussion

SIH is an uncommon headache syndrome that affects women two times more frequently than men with an estimated annual incidence of 5 per 100,000[3]. Because of its rarity, it is commonly misdiagnosed. The initial diagnosis was incorrect in 17 of 18 patients and the diagnostic delay times ranged from 4 days to 13 years[10].

SIH is caused by CSF leakages, leading to decreased CSF pressure[5-7] and downward displacement of the brain [2,18,16]. This pathophysiological changes further cause compensatory dilatation of the cerebral

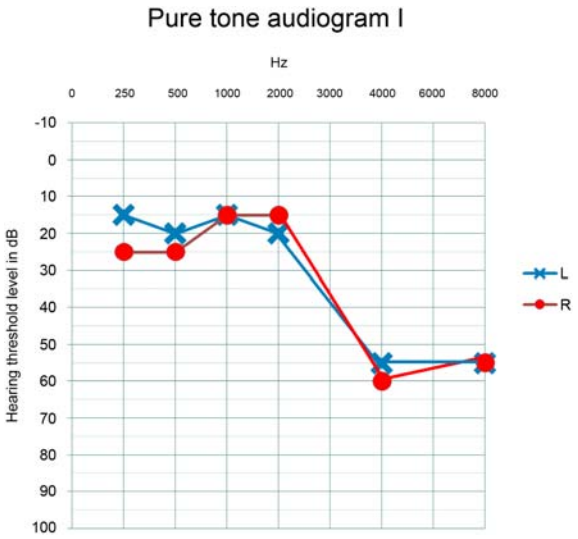

Fig. 4 Audiogram preoperative evaluation revealed minimal low-frequencies (250 Hertz and 500 Hertz) sensori-neural hearing loss with the hearing threshold of 25 decibel (dB) on the right and bilateral high-frequency (4kHertz and 8kHertz) sensori-neural hearing loss

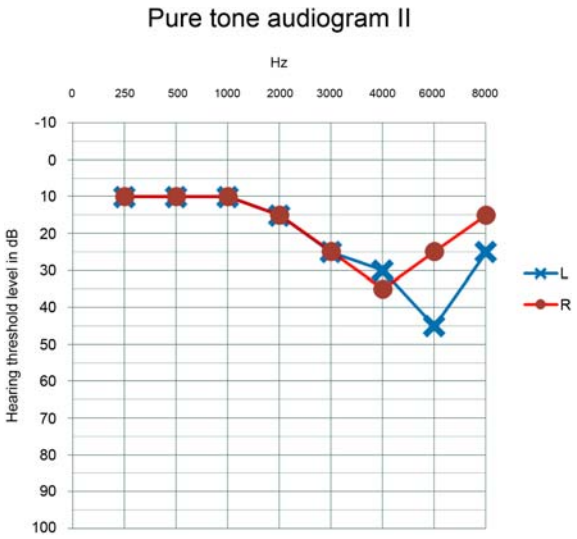

Fig. 5 Audiogram 3 months after operation revealed normal hearing acuity with the hearing threshold level of 10 decibel at low-frequencies (250 Hertz and 500 Hertz). Bilateral high-frequencies (4 kHertz and 6 kHertz) sensori-neural hearing loss and 8 kHertz frequency sensori-neural hearing loss on the Lt ear

venous system[19] and congestion of the pituitary gland[20]. Downward displacement of the brain[2,18,16] causes traction on the dura mater, cranial nerves, and other brain parenchyma structures, which can explain the clinical syndrome of SIH such as orthostatic headache, cranial neuropathies, dementia, and stupor.

SDH has also been observed in patients with SIH[1,4,16]. The mechanism is hypothesized probably due to rupture of bridging veins in the subdural space[21], which may be easily broken when the dura mater is being retracted[21,22]. The terms SDH, subdural hygroma and subdural fluid collection, related to SIH, have been changeably used in most of the literature[3,1,23]. Schievink reported subdural fluid collections in half of 40 cases with SIH[24]. Among these, 20 patients had subdural fluid collection, eight patients had SDH with significant mass effect. Three out of eight patients required evacuation, while in the other five patients SDHs resolved within 1 to 3 months without surgical drainage. SDH seen in SIH is mostly thin, bilateral, located at the cerebral convexities, and usually required no drainage. Several authors suggested conservative treatment and treatment of the underlying spinal CSF leakages. However, patients with large and life-threatening SDH may require emergency intervention[3]. The presented patient was initially

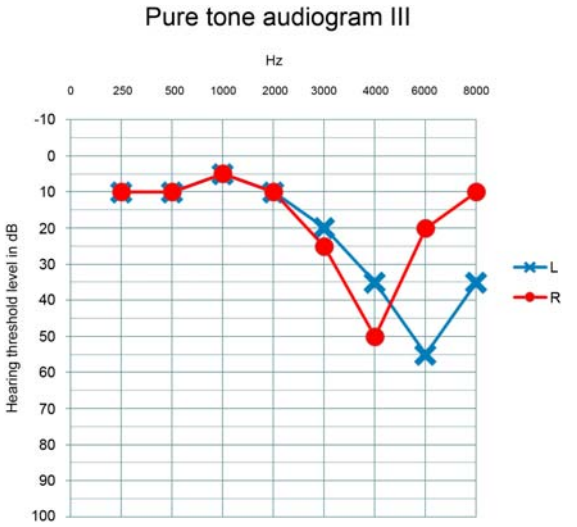

Fig. 6 Audiogram 1 year after follow-up revealed normal hearing acuity with the hearing threshold level of 10 decibel at low-frequencies (250 Hertz and 500 Hertz). Bilateral High-frequency (4 kHertz) sensori-neural hearing loss both ears and high-frequencies (4kHertz and 6kHertz) sensori-neural hearing loss on the Lt ear. In comparison, hearing thresholds at low-frequencies (250 Hertz and 500 Hertz) were improved from 25 dB to 10 dB and high-frequencies (4kHertz and 8kHertz) sensori-neural hearing loss were also improved from the preoperative audiogram

diagnosed as SDH with unknown cause and underwent possible unnecessary surgical intervention. Orthostatic headache, tinnitus, and hearing impairment are important diagnostic clues to uncover etiology of SDH due to SIH. Typical MRI findings were the final clues to the diagnosis of SIH in the presented patient.

Auditory syndrome in SIH is common, however, hearing impairment is unusual, and audiograms are rarely reported in the literature. The mechanism of auditory symptoms is also an interesting issue. The proposed mechanism is the effect of low CSF pressure causing secondary perilymph hypotension and subsequently, secondary endolymphatic disturbances[25]. This mechanism was supported in two previously reported cases. Audiographic findings in these cases revealed low-frequency hearing loss with the pattern typically seen in Meniere's disease, a well-known endolymphatic disturbance[26-28]. Mild unilateral or bilateral sensori-neural hearing loss could be seen in the other more common causes of low CSF pressure such as post ventriculo-peritoneal shunt or spinal puncture[27]. The studies in these cases showed mild and reversible low-frequency hearing loss. In the presented case, the audiogram was studied three times during the course of the illness. The audiographic studies showed a pattern of minimal right low-frequency hearing loss, which was reversible after 3 months of conservative treatment, suggesting the process of secondary endolymphatic disturbances. Bilateral high-frequency hearing loss was also demonstrated and this was improved after the treatment. However, high-frequency hearing loss, especially at 4kHertz, typically found in noise-induced hearing loss still persisted in the presented case. The cause of this residual auditory impairment may be due to chronic noise exposure in his industrial plant.

Demonstration of spinal CSF leakage is important, investigations may not always be able to detect site of CSF leakages. In the presented case, MRI of the whole spine with gadolinium enhancement revealed no definite sign of CSF leakage[1-3,23]. Radionuclide cisternography may be performed for further detection of the site of CSF leakage[2,3]. Since the presented patient was improved clinically after conservative treatment with bed rest and hydration, the radionuclide cisternography was not done.

Treatments of SIH consist of bed rest, hydration, epidural blood patch, and surgery to close the site of CSF leakage. Recurrence of SIH may occur, especially in cases with persistent site of CSF leakage[14]. The presented patient responded to conservative treatment. He had no further complications or other signs of recurrence of the disease during 1-year follow-up.

Conclusion

The authors reported a case of SIH who presented with bilateral thin SDHs of unknown cause. History of orthostatic headache and MRI with gadolinium enhancement were crucial clues to diagnose SIH. Hearing loss in the presented case was confirmed by audiograms. To the authors' knowledge, the presented case is among a few previous reported cases of SIH, which had unilateral low-frequency hearing loss, confirmed by audiograms.

Acknowledgement

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ภาวะเลือดออกใต้เยื่อหุ้มสมองชั้นดูราและระดับการได้ยินเสียงผิดปกติที่มีสาเหตุจากภาวะความดันในช่องกะโหลกศีรษะต่ำ

ดวงพล ศรีมณี, ณัฐ พสุธารชาติ, กัมมันต์ พันธุมจินดา

ภาวะความดันในช่องกะโหลกศีรษะต่ำ (spontaneous intracranial hypotension) เป็นกลุ่มอาการปวดศีรษะที่พบไม่บ่อย มักมีอาการและอาการแสดง คืออาการปวดศีรษะเมื่อเปลี่ยนท่า มีเสียงในหู และเห็นภาพซ้อน รายงานผู้ป่วยชายไทยคู่อายุ 43 ปี ปวดศีรษะ 3 เดือนสัมพันธ์กับการยืนร่วมกับอาการเสียงในหู และระดับการได้ยินที่ผิดปกติ การตรวจระดับการได้ยิน พบการสูญเสียการได้ยินระดับความถี่ต่ำเล็กน้อย ลักษณะความผิดปกติที่โคเคลียร์ เอกซเรย์คอมพิวเตอร์พบ เลือดเก่าออกเป็นชั้นบางในกะโหลกศีรษะใต้ชั้นดูราทั้ง 2 ข้าง ผู้ป่วยได้รับการผ่าตัดเจาะรูกะโหลก อาการปวดศีรษะและระดับการได้ยินที่ผิดปกติยังคงอยู่ จึงมีการตรวจทบทวนผู้ป่วยอีกครั้ง การตรวจเอกซเรย์สนามแม่เหล็กไฟฟ้าศีรษะพบสารรังสีกระจายทั่วศีรษะ ตำแหน่งของก้านสมอง ส่วนก้านสมอง และสมองน้อยส่วนทอนซิลเคลื่อนต่ำลง ต่อมพิทูอิทารี่โตขึ้น เข้าได้กับภาวะความดันในกะโหลกศีรษะต่ำ การตรวจเอกซเรย์สนามแม่เหล็กไฟฟ้า ส่วนไขสันหลังทุกระดับไม่พบรอยรั่วของน้ำไขสันหลัง ผู้ป่วยอาการดีขึ้นหลังได้รับการรักษาแบบประคับประคองด้วยการนอนพักร่วมกับให้สารเหลวทางหลอดเลือดดำ 1 ปี หลังเข้ารับการรักษาอาการปวดศีรษะหายสนิทและเสียงในหู ลดลงเกือบเป็นปกติ