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Case Report

# Medulloblastoma Manifesting as Sudden Sensorineural Hearing Loss

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We present a rare case of medulloblastoma which presented with unilateral sudden sensorineural hearing loss as an initial symptom. A 19-year-old man was admitted to our hospital with a chief complaint of dizziness and facial numbness on the right side. His illness had begun two years previously with sudden hearing loss on the right side, for which he had been treated as an idiopathic sudden hearing loss. Magnetic resonance imaging demonstrated abnormal signals located mainly in the right middle cerebellar peduncle. We performed partial resection of the tumor by suboccipital craniotomy. The histopathological diagnosis was medulloblastoma. Intrinsic brain tumor is an extremely rare cause of sudden sensorineural hearing loss and is therefore easily overlooked as was in the present case. The present case highlights not only the need to evaluate patients with sudden sensorineural hearing loss by magnetic resonance imaging but also the importance of paying attention to intrinsic lesions involving the brainstem. Although this condition like the presented case might be rare, intrinsic brain tumor should be considered as a potential cause of sudden sensorineural hearing loss, as it may be easily missed leading to a delay in appropriate treatment.

**Key Words :** Medulloblastoma · Sudden hearing loss · Sensorineural hearing loss · Intrinsic tumor · Brain tumor.

## INTRODUCTION

Sudden sensorineural hearing loss is not a rare condition, and thus may occasionally be encountered by clinicians in daily practice. The incidence of sudden hearing loss has been reported to range from 5 to 20 per 100,000 subjects per year, but the precise incidence is estimated to be higher[14]. The etiology of sudden hearing loss is diverse[4], and patients with sudden sensorineural hearing loss may show magnetic resonance (MR) imaging abnormalities corresponding to the clinical picture such as labyrinthine hemorrhage, cochlear inflammation, multiple sclerosis, and neoplastic disease[1,16]. Although several previous studies recommended evaluating patients with sudden sensorineural hearing loss by MR imaging to rule out underlying organic disease[1,12,16], few clinicians seem to be aware that even a small intrinsic lesion has the potential to produce sudden sensorineural hearing loss.

We describe a case of medulloblastoma located in the middle cerebellar peduncle that manifested as unilateral sudden sensorineural hearing loss followed by cranial nerves pareses and cerebellar dysfunction.

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## CASE REPORT

A 19-year-old man presented with a one-year history of dizziness and facial numbness on the right side. His symptoms had begun two years previously with sudden hearing loss on the right side, for which he had consulted an otorhinolaryngologist. The otorhinolaryngologist examined the patient with MR imaging (Fig. 1) and patient was treated based on a diagnosis of idiopathic sudden hearing loss, but his hearing disturbance did not improve. Details of the treatment were not available. Subsequently, he started suffering from the aforementioned symptoms and consulted our department.

On admission, neurological examination revealed decreased facial sensation, absent corneal reflex, slight peripheral-type facial weakness, hearing disturbance on the right side, and horizontal and alternate lateral gaze nystagmus. His motor strength was normal, but limb ataxia was observed on the right side. Pure tone audiometry showed right side sensorineural hearing loss up to 102.5 dB in all frequency bands. MR imaging demonstrated abnormal signals located mainly in the right middle cerebellar peduncle shown as iso to hyperintensity on T1-weighted images and hyperintensity on T2-weighted images. This lesion was faintly enhanced by contrast medium (Fig. 2). A retrospective review of the MR imaging results obtained at the previous

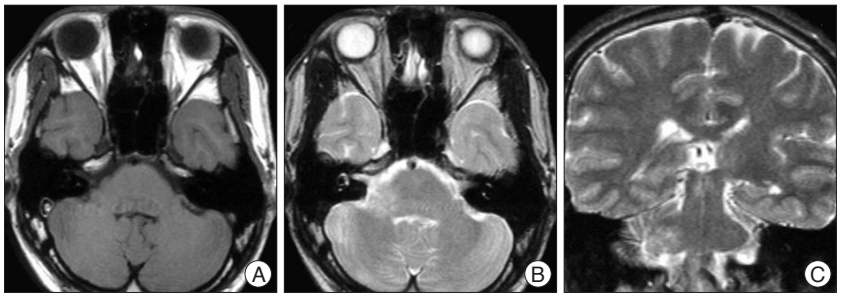

**Fig. 1.** T1-weighted (A) and T2-weighted (B and C) magnetic resonance images checked at the previous hospital approximately 34 months before the patients consulted our department. An undemarcated intrinsic lesion localized mainly in the right middle cerebellar peduncle is shown as hyperintensity on T2-weighted images, but is not clear on the non-contrast T1-weighted image.

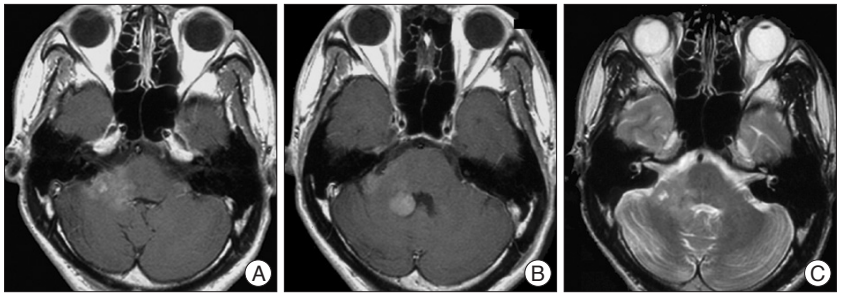

**Fig. 2.** T1-weighted with contrast medium (A and B) and T2-weighted (C) magnetic resonance images on admission show the lesion mainly located in the right cerebellar peduncle. The lesion is slightly enhanced by contrast medium.

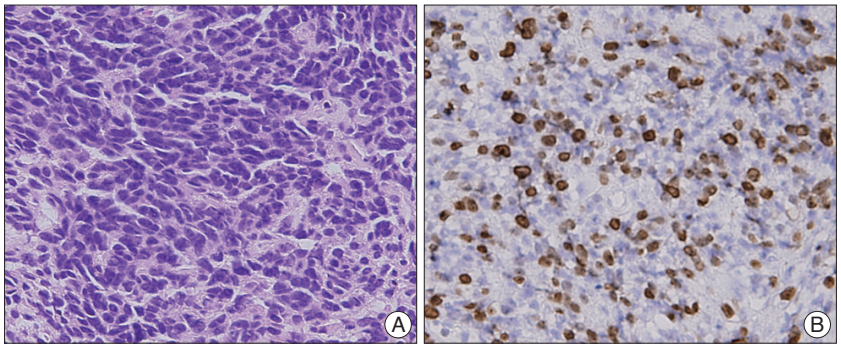

**Fig. 3.** Histopathological finding is compatible with medulloblastoma (H&E stain, ×400) (A). MIB-1 staining for Ki-67 antigen with a value of 36% (B).

hospital suggested that the lesion had been present at the first presentation of sudden hearing loss (Fig. 1), and this small intrinsic lesion was considered to be responsible for the hearing loss in this patient. T1-weighted images with contrast medium were not performed at the previous hospital.

The patient underwent suboccipital craniotomy for partial resection of the lesion in the fourth ventricle. Intraoperatively, the lesion appeared to be invasive into the adjacent brainstem, but the exophytic part of the lesion was localized in the fourth ventricle. The eighth cranial nerve of the cisternal segment was not observed in the operative field. Histopathological diagnosis was medulloblastoma, and MIB-1 staining for Ki-67 antigen was 36% (Fig. 3).

Thereafter, the patient received radiation therapy at a dose of 23.4 Gy for the whole brain, an extended local dose of 27 Gy, and 24 Gy for the whole spinal axis followed by three courses of chemotherapy with ifosfamide, cisplatin, and etoposide. After these adjuvant therapies, there has been no evidence of tumor recurrence on MR imaging for seven years, and the patient is now well except for slight limb ataxia. However, persistent deafness on the right side is still observed.

## DISCUSSION

Medulloblastoma is classified as a brain tumor of grade IV according to the World Health Organization classification, and accounts for 1.1% of all primary central nervous system tumors in all ages and up to 11.9% of those in adolescents under the age of 15 years[3]. The most common presenting features of medulloblastoma are usually associated with increased intracranial pressure, such as papilledema (75%) and headache (65%) or are associated with cerebellar dysfunction, such as truncal (62%) and limb (35%) ataxia[9]. This report describes a case of medulloblastoma in a 19-year-old man initially presenting with unilateral sudden sensorineural hearing loss. Unilateral sudden sensorineural hearing loss can be recognised as an initial symptom of brain tumor, but is most often associated with vestibular schwannomas[2,5,8,11-13,18]. Sudden sensorineural hearing loss caused by medulloblastoma is extremely rare and, to our knowledge, there have been only three previous reports of medulloblastoma manifesting as sudden sensorineural hearing loss[6,7,9].

Sudden hearing loss has been defined as 30 dB or more sensorineural hearing loss over at least three contiguous audiometric frequencies occurring within three days or less[17]. The etiology of sudden sensorineural hearing loss is variable and often difficult to identify. As a consequence, many cases are labelled as "idiopathic". However, clinicians must always be aware that idiopathic sudden sensorineural hearing loss still remains an exclusion diagnosis after the multiple possible cases are eliminated. Several pathological conditions are known to cause sudden sensorineural hearing loss[4], but intrinsic brain tumors seem to have attracted little attention, and are consequently overlooked as seen in our case. Although intrinsic brain tumors manifesting as sensorineural hearing loss have been reported previously, these cases were also misdiagnosed on first presen-

tation of hearing loss[6,9,15].

From an anatomical viewpoint, intrinsic lesions are unlikely to cause unilateral sensorineural hearing loss, because the auditory neural pathway distinctively has bilateral neural innervations[9,15] after the cochlear nucleus in the brainstem receives the afferent neural fibers through the eighth cranial nerve. That is, only dysfunction of the fibers of the eighth cranial nerve to the cochlear nucleus or the unilateral cochlear nucleus itself can theoretically result in unilateral sensorineural hearing loss. In the present case, the cisternal segment of the eighth cranial nerve appears to be intact according to MR imaging obtained at the initial onset of the hearing disturbance. On the basis of this finding, we considered the unilateral sensorineural hearing loss in this patient to have been caused by impairment of the cochlear nucleus or the afferent neural fibers of the ipsilateral eighth cranial nerve within the brainstem due to tumor invasion or peritumoral edema.

## CONCLUSION

The present case calls attention not only to lesions involving the cochlea, internal auditory meatus, or cerebellopontine angle, but also to lesions involving the brainstem itself in examining patients with sudden sensorineural hearing loss, as a small intrinsic lesion can be easily missed. Clinicians should be aware of this unique presentation of intrinsic brain tumors and of the possibility of patients having an organic cause involving the brainstem for sudden sensorineural hearing loss, as misdiagnoses may lead to a delay in treatment.

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