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Bilateral sudden sensorineural hearing loss due to subcortical cerebral hemorrhage in a patient with Moyamoya disease: case report

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Abstract

Background Moyamoya disease (MMD) is a chronic cerebrovascular occlusive disease that can results in a variety of neurological deficits. We describe a very rare presentation of MMD in the form of bilateral sudden sensorineural hearing loss after a second attack of subcortical cerebral haemorrhage.

Case presentation A 44-year-old female presented with bilateral sudden sensorineural hearing loss (SSNHL) secondary to an acute intracerebral haemorrhage in the left temporoparietal region. She had a previous haemorrhagic event in the right temporoparietal region two years ago. The two haemorrhagic events have apparently led to interruption of the auditory pathways bilaterally, resulting in sudden complete deafness. Digital subtraction angiography was done, and it showed bilateral occlusion of the internal carotid arteries, with the abnormal anastomotic vascular network at the base of the brain characteristic for MMD.

Conclusions Subcortical hemorrhage due to MMD is a potential cause of bilateral SSNHL that can be diagnosed with early proper imaging. Bilateral damage of the auditory pathways is the likely cause of cortical deafness in this clinical scenario.

Keywords Moyamoya, Deafness, Sudden sensorineural hearing loss

Background

Moyamoya disease (MMD) is a rare chronic cerebrovascular occlusive disease first described by the Japanese neurosurgeons Takeuchi and Shimizu in 1957 [1]. The hallmark of this disease is bilateral severe stenosis or occlusion of the internal carotid arteries, associated with the development of abnormal anastomotic vascular network located at the base of the brain [2]. We demonstrate a very rare case of a patient with MMD who developed bilateral sudden sensorineural hearing loss (SSNHL) as a main presentation of a second hemorrhagic event. Moyamoya disease can lead to cortical deafness either via ischemic or hemorrhagic lesions that lead to damage of the subcortical white matter and disconnection between the brainstem and cortical auditory areas [3]. Ischemic lesions are more common in pediatric MMD patients, while hemorrhagic lesions are more common in elderly MMD patients [4].

Case summary

A right-handed 44-y-old female patient was referred to our center from another hospital, where she had been admitted eight days earlier under the chief complaint of acute headache, nausea, and sudden complete hearing loss. The initial CT scan done at the referring hospital showed left temporo-parietal acute intracerebral hematoma, measuring 30×27x17 mm in maximum dimensions, and extending into the left lateral ventricle, the third and the fourth ventricles, with mild hydrocephalic

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changes. A follow-up CT was done two days after admission and showed no significant changes, compared to the initial. The patient was referred to our center to investigate any underlying vascular disease. The patient's family members reported that she was hospitalized two years ago due to cerebrovascular accident that was associated with seizures and was treated with Levetiracetam since then. The patient is otherwise normal, her coagulation profile was normal, and she has no significant past medical history or a family history of similar conditions.

Upon admission to our center, the patient was fully conscious (GCS 15/15), she had no focal motor or sensory impairments. She had bilateral sensorineural hearing loss, she had normal speech, and we could communicate with her through written text. A follow-up CT was done, and it showed a resolving left temporo-parietal acute hematoma, surrounded by a hypodense rim of edema, extending only to the left lateral ventricle, with no hydrocephalic changes, (Fig. 1).

The patient was prepared for diagnostic cerebral angiography. The angiography was performed using a right trans-femoral approach. The angiography revealed bilateral complete occlusion of the middle cerebral arteries (MCA) and the anterior cerebral arteries (ACA) (Fig. 2) and severe attenuation of the left posterior cerebral artery (PCA), (Fig. 3). The characteristic anastomotic vascular network of MMD was evident on both sides (Fig. 2). The vertebrobasilar circulation appeared normal, however, the right posterior inferior cerebellar artery (PICA) showed smaller caliber, compared to the left PICA, (Fig. 3). Collateral supply was offered to the posterior portions of the right ACA and MCA territories via anastomoses with the parieto-occipital and splenial branches of right PCA and the marginal branch of right superior cerebellar artery (SCA), (Figs. 2, 3). A few anastomotic connections were found between the dural and osseous branches of external carotid arteries (ECA) and the cortical branches of MCA and ACA, (Fig. 4).

Surgical options to halt the progress of MMD were discussed with the patient and her family and they opted against surgery at this stage. The patient was repatriated to the primary referring hospital to complete the investigations and treatment of hearing loss. The patient had been lost to follow up at the primary referring hospital. One year later, we managed to contact one of the family members through a phone call, and we knew that the patient has started to regain part of the auditory function six months after her discharge from our service, however her hearing abilities are still far away from being normal.

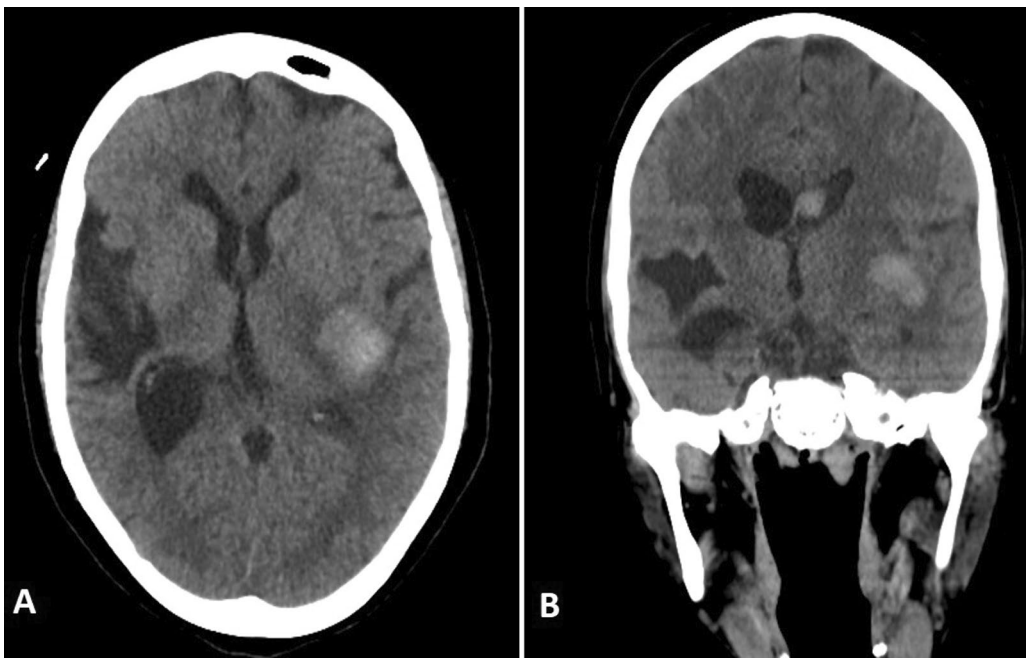

Fig. 1 CT scan upon admission to our center, eight days after the hemorrhagic event: **A** axial, **B** coronal. CT scan shows the left sided resolving acute temporoparietal hematoma and an area of encephalomalacia on the right temporoparietal region, corresponding to the old hemorrhagic event and causing dilatation of the adjacent temporal horn

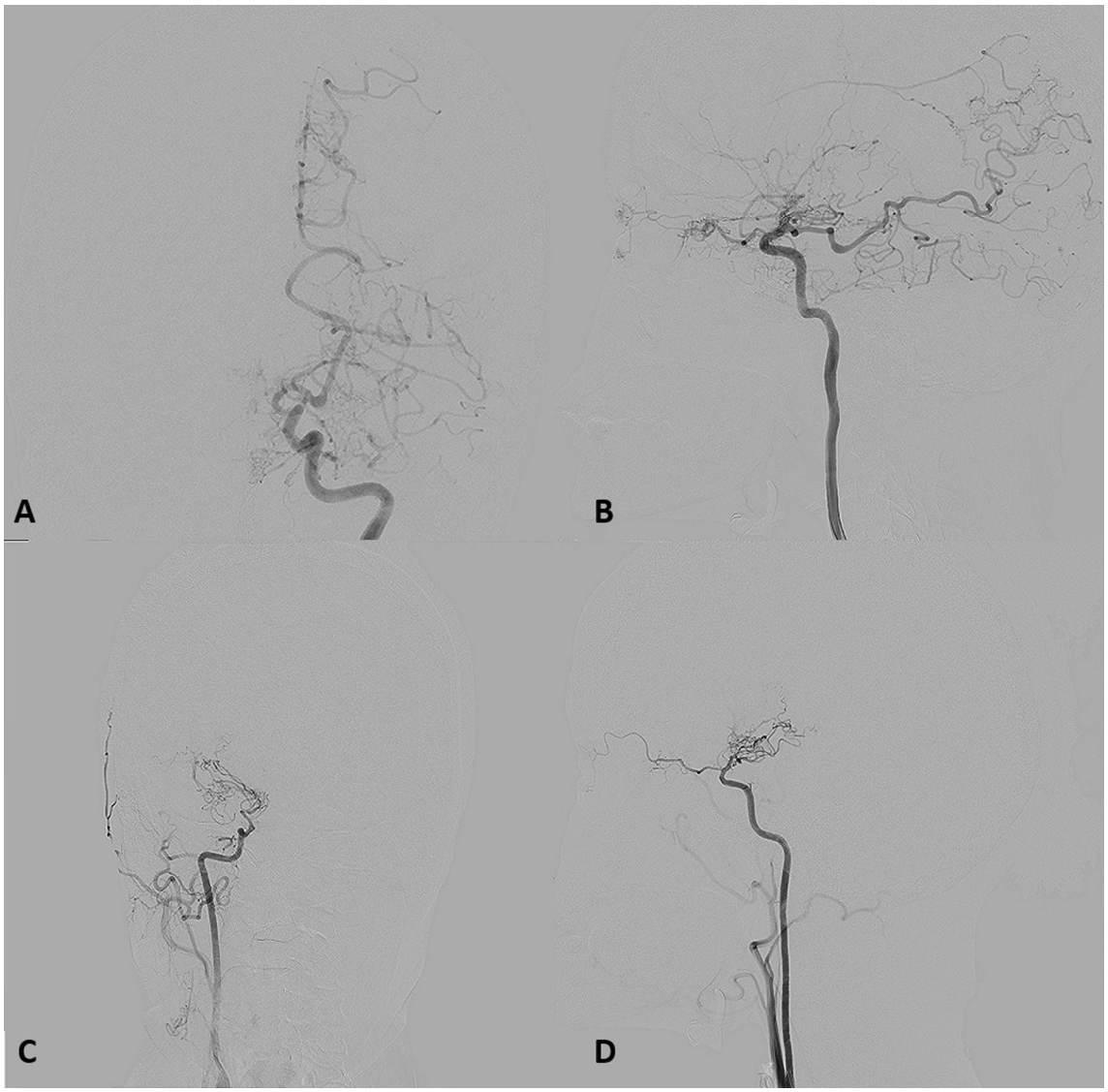

Fig. 2 Selective angiography for left ICA: AP **A** and lateral **B** views, right ICA: AP **C** and lateral **D** views. The angiograms show complete occlusion of the MCA and ACA bilaterally and the characteristic Moyamoya abnormal vessels at the base of the brain

Discussion

Bilateral SSNHL is a severe and rare form of deafness that results in loss of response to all types of sounds. Owing to the bilateral representation of auditory pathways, bilateral SSNHL is very rare [5]. To our knowledge, this is the second case report about bilateral SSNHL in an adult patient with MMD, after the first case published by Wakabayashi in 1999 [6]. Unilateral SSNHL in patients with MMD was described in two other case reports [7, 8]. Different clinical scenarios about the association between MMD and hearing loss have been described in two other case reports [9, 10].

Unfortunately, we couldn't find any audiometric studies for the patient as she was lost to follow up as previously mentioned. Conductive hearing loss was primarily excluded by performing Weber and Rinne tests. We see that bilateral damage of the auditory pathways by the new (left) and old (right) hemorrhagic insults is responsible for the sudden deafness in our patient. This etiology was previously demonstrated by Tanaka and colleagues who concluded that severity of cortical deafness is mainly affected by bilateral damage to the white matter tracts adjacent to the posterior half of the putamen [11]. The possibility of bilateral anterior inferior cerebellar artery

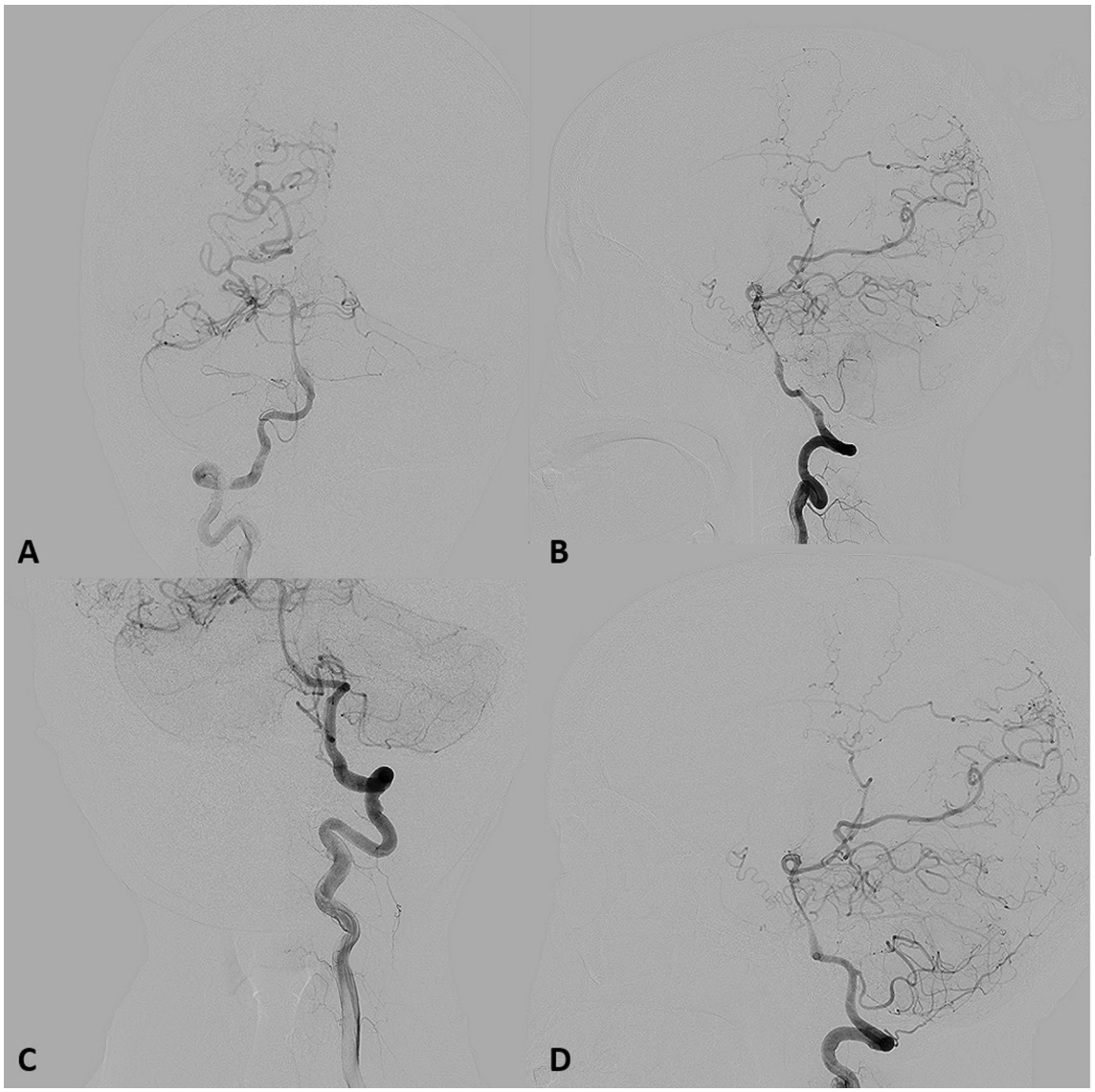

Fig. 3 Selective angiography for right VA: AP **A** and lateral **B** views, left VA: AP **C** and lateral **D** views. The angiograms show normal vertebrobasilar circulation, however the left PCA shows marked decrease in caliber, and the right PICA has smaller caliber, compared to the left PICA

(AICA) occlusion is neither supported by the clinical scenario, nor by the angiography.

In 2019, the updated clinical practice guidelines on sudden hearing loss strongly recommended that clinicians should not order routine head CT scans in the initial evaluation of a patient with presumptive SSNHL except in patients with focal neurological deficits or history of trauma. In these guidelines, the panel has recommended MRI brain and Auditory brainstem response (ABR) for evaluation of patients with SSNHL due to retrocochlear pathology at some point of their care without a specific time frame for that [12]. We believe that subcortical haemorrhages due to MMD can be overlooked based on these guidelines, and that's why we believe that this case report is important to delineate the linkage between SSNHL and this type of brain haemorrhage. We also believe that ordering brain CT or MRI early in the course of SSNHL is important because performing proper neurological examination in these patients may be difficult due to the poor communication.

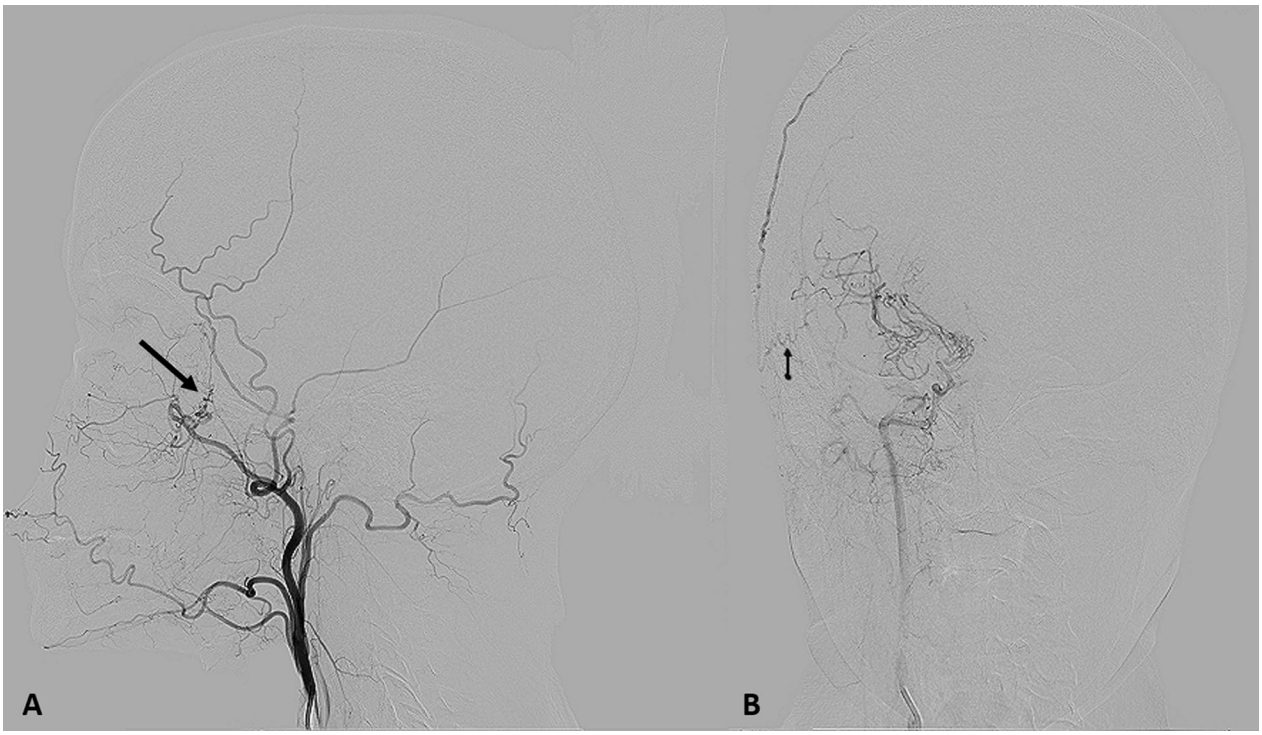

Fig. 4 Selective angiography for left ECA: lateral **A** and AP **B** views. The angiograms show anastomotic connections from the left internal maxillary artery (long arrow) and left middle meningeal artery (short arrow) to the left ICA branches

Conclusions

Subcortical hemorrhage due to MMD is a potential cause of bilateral SSNHL, and early brain imaging is recommended. Bilateral damage of the auditory pathways is the likely cause of cortical deafness in this clinical scenario.

Abbreviations

MMD	Moyamoya disease
SSNHL	Sudden sensorineural hearing loss
GCS	Glasgow coma scale
MCA	Middle cerebral artery
ACA	Anterior cerebral artery
PCA	Posterior cerebral artery
PICA	Posterior inferior cerebellar artery
SCA	Superior cerebellar artery
ECA	External carotid artery
AICA	Anterior inferior cerebellar artery
ICA	Internal carotid artery
VA	Vertebral artery

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Ethics approval and consent to participate
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Consent for publication
Not Applicable.

Competing interests
The author declares that he has no conflict of interest.

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