



## Unilateral sudden hearing loss: a rare symptom of Moyamoya disease

Tek taraflı ani işitme kaybı: Moyamoya hastalığının nadir bir semptomu

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### ABSTRACT

A 38-year-old female patient experienced a sudden onset of unilateral sensorineural hearing loss due to Moyamoya disease. A detailed summary of audiological and neurological findings indicated that the sudden hearing loss might be due to Moyamoya disease resulting in occlusion of posterior and middle cerebral arteries. Intravenous prednisolone and trimetazidine dihydrochloride may improve hearing thresholds and speech understanding. To our knowledge, this is the first article in the literature reporting a case of sudden hearing loss as the first manifestation of Moyamoya disease in a young adult.

**Keywords:** Hearing loss; Moyamoya; sudden; unilateral.

### ÖZ

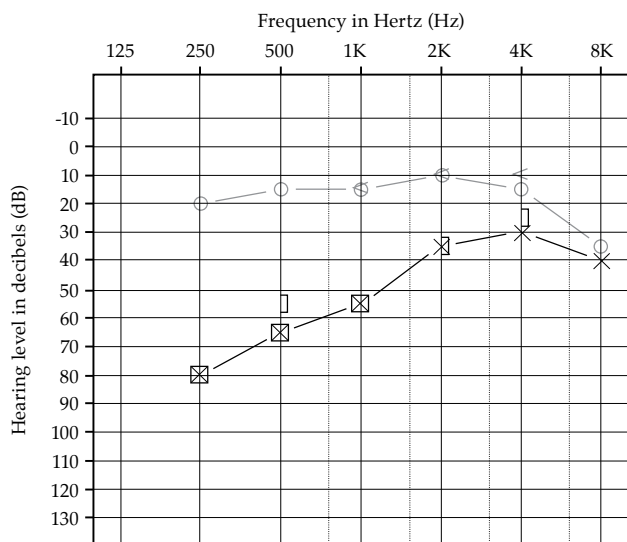
Otuz sekiz yaşında bir kadın hasta Moyamoya hastalığına bağlı olarak tek taraflı ani başlangıçlı işitme kaybı ile başvurdu. Odyolojik ve nörolojik ayrıntılı muayene ve tetkikler, posterior ve middle serebral arterlerin tıkanıldığını ve bunun Moyamoya hastalığından kaynaklanabileceğini gösterdi. İntravenöz prednizolon ve trimetazidin dihidroklorür işitme eşiklerini ve konuşmayı anlama skorlarını düzeltmede yardımcı oldu. Bildiğimiz kadarıyla, bu olgu sunumu Moyamoya hastalığının ilk belirtisi olarak ani işitme kaybı geçiren genç bir erişkinini bildiren ilk yazıdır.

**Anahtar Sözcükler:** İşitme kaybı; Moyamoya; ani; tek taraflı.

Moyamoya disease is a chronic cerebrovascular disorder characterized by progressive stenosis or occlusion of bilateral internal carotid arteries with an extensive network of cerebral collaterals. The disease was first described in 1957 by Takeuchi and Shimizu.<sup>[1]</sup> In 1969, Suzuki and Takaku<sup>[2]</sup> named it Moyamoya which means puff of smoke that represents the characteristic reticulate appearance on angiography.

The cause of Moyamoya disease is not known although there is evidence that the disease is hereditary. Its average annual incidence is 0.54 per 100,000 persons but it is the most common pediatric cerebrovascular disease in Far East Asia.<sup>[3]</sup> The reported incidence in the USA is approximately 0.086 per 100,000 patients.<sup>[4]</sup> The disease has a bimodal age distribution. The first peak occurs in childhood and cerebral ischemic events are more common in children. The second





**Figure 1.** Audiogram showing left-sided low-frequency sensorineural hearing loss with an average of 52 dB.

peak occurs in the fourth decade of life and adults experience hemorrhagic strokes more commonly. This article presents a case of sudden hearing loss (SHL) as the first manifestation of Moyamoya disease in a young woman.

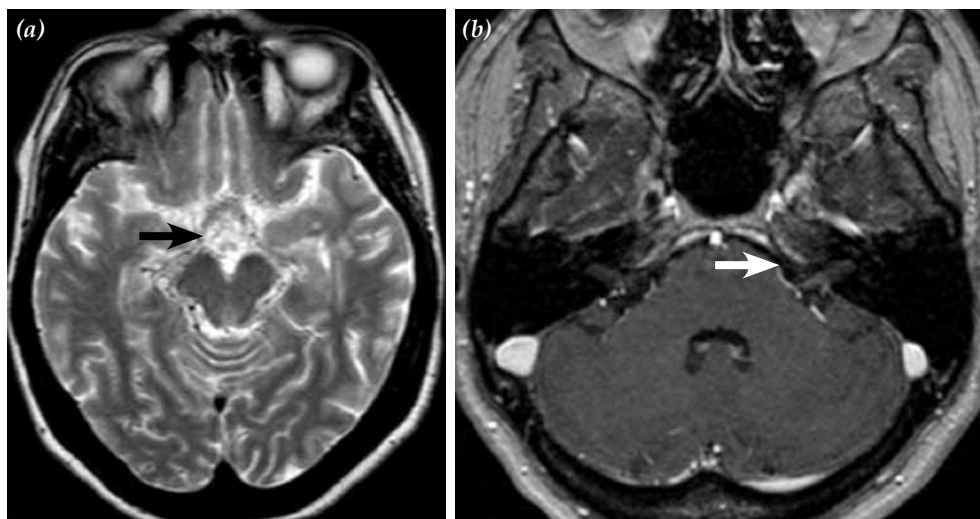
### CASE REPORT

A 38-year-old woman was admitted to our department with the complaint of left-sided SHL accompanied by ipsilateral tinnitus and mild imbalance for one day. She had a history of ischemic cerebrovascular stroke 10 years ago

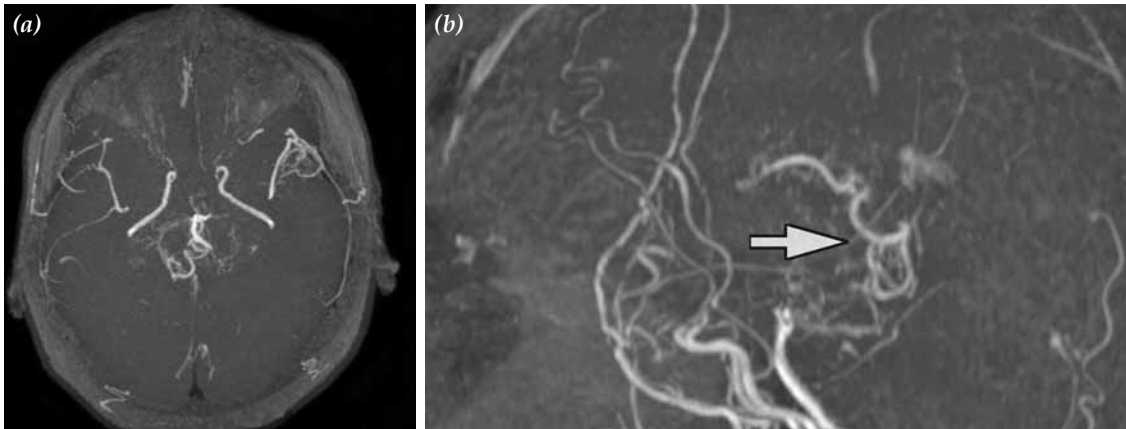
with left central facial paralysis and sudden loss of vision in both eyes. She had been hospitalized for this reason and fully recovered. Since that time, she had been taking aspirin 100 mg per day. She had smoked 20 cigarettes per day for 15 years. There was no history of previous viral illness, flying, diving, strenuous physical activity, or head trauma. Family history was also negative. A written informed consent was obtained from the patient.

On otoscopic examination, her left ear appeared normal, while basic audiometry confirmed left sided sudden sensorineural hearing loss (SSHL). The patient's gait was normal with no signs of cerebellar or vestibulospinal dysfunction. Pure tone audiometry revealed a left-sided low-frequency sensorineural-type hearing loss with an average of 52 dB. The speech recognition threshold was 55 dB, and the speech discrimination score was 68%. Hearing thresholds on the right ear were within normal levels (Figure 1).

A brain magnetic resonance (MR) imaging scan was obtained one week after the SHL, which included 3 mm contiguous, axial T<sub>1</sub>-weighted images, with and without gadolinium, and T<sub>2</sub>-weighted images. Magnetic resonance imaging demonstrated multiple vessels around the mesencephalon. The anterior inferior cerebellar artery was extending into the left internal acoustic channel (Figure 2). Afterwards, radiologists recommended MR angiography for



**Figure 2.** Axial magnetic resonance (a) T<sub>2</sub>-weighted, non-contrast image showing multiple vascular network around mesencephalon; (b) T<sub>1</sub>-weighted, contrast image, showing anterior inferior cerebellar artery extending into the left acoustic channel.



**Figure 3.** Axial and sagittal magnetic resonance angiography. (a) Both internal carotid artery, middle cerebral artery and posterior cerebral artery are not seen. (b) Abnormal vascular network is seen around circle of willis.

evaluation of brain perfusion, due to multiple vessels around the mesencephalon seen in MR imaging. Magnetic resonance angiography revealed severe findings compatible with Moyamoya disease-- both internal carotid arteries, posterior and middle cerebral arteries were not observed and an abnormal vascular network was seen in the basal ganglia (Figure 3).

Results of a complete metabolic panel, coagulation studies and complete blood cell count remained within normal limits. Specific tests for hypercoagulability disorders included Von Willebrand factor, factor 5, 8 and 13, prothrombin time, partial thromboplastin time, international normalized ratio, protein C and S, D-dimer, fibrinogen were normal.

The patient was treated with 1 mg/kg intravenous prednisolone per day in addition to trimetazidine dihydrochloride 35 mg tablet twice a day as a vasodilator. On the fourth day of treatment the patient's hearing loss recovered and audiograms showed a significant improvement in auditory thresholds in the left ear with an average of 22 dB.

## DISCUSSION

Sudden sensorineural hearing loss involves an acute unexplained hearing loss of greater than 30 dB over three contiguous pure-tone frequencies occurring within three days. Most cases are idiopathic, and usually present as unilateral hearing loss. The prognosis depends on the severity of the hearing loss. There are some theoretical possibilities, which include labyrinthine viral infection, labyrinthine vascular

problems, intracochlear membrane ruptures and autoimmune disorders<sup>[5]</sup> that may cause SSHL. Although vascular problems are rare, the cochlea is very sensitive to changes in blood supply. Thrombosis, vasospasm and reduced blood flow can cause SHL.<sup>[6]</sup>

Moyamoya disease may reduce the blood flow of distal intracranial internal carotid arteries. It may result in stroke, transient ischemic attacks, seizures or hemorrhage from fragile collateral vessels. The possible mechanism of our patient's symptoms is vascular occlusion. The inner ear is supplied by the internal auditory artery, which is an end-artery and usually a branch of the anterior inferior cerebellar artery. Occlusion of the internal auditory artery, so-called labyrinthine ischemia, causes loss of auditory function, resulting in hearing loss. Internal auditory artery ischemia mostly occurs due to thrombotic narrowing of the anterior inferior cerebellar artery.<sup>[7]</sup>

Moyamoya collaterals can be visualized with MR angiography. Magnetic resonance angiography demonstrates stenotic or occlusive lesions in the distal internal carotid arteries and proximal circle of Willis vessels.<sup>[8]</sup> Currently there is no definitive medical treatment to reverse or stabilize the course of Moyamoya disease. Aspirin and calcium channel blockers play a supportive role to prevent further ischemic injury. There is no standardized surgical approach for Moyamoya disease.

To the best of our knowledge, this is the first reported case of Moyamoya disease associated with SSHL which resolved completely.

Tseng and Luo<sup>[9]</sup> have reported a case report with unilateral SSHL as the first presenting symptom in Moyamoya disease, but their patient showed only mild improvement.

A study by Narozny et al.<sup>[10]</sup> concluded that hyperbaric oxygen therapy with high doses of glucocorticoids improves the results of conventional SSHL treatment. Our patient was treated with steroids in addition to vasodilators and anticoagulant drugs. However, on the fourth day of treatment our patient's hearing was improved so hyperbaric oxygen therapy was not included in the treatment. Sudden hearing loss associated with Moyamoya disease may be a vascular pathology. Hyperbaric oxygen in addition to steroids, vasodilator and anticoagulant agents may be useful. However, there is no adequate experience on this subject.

In conclusion, we wish to highlight that Moyamoya disease may trigger SSHL, which is a rare condition that is poorly described in literature. We report a novel case of unilateral SSHL, likely due to Moyamoya disease, which was managed successfully with medical treatment. Early recognition of the SSHL appears to be the ultimate key, as it may mean potential reversal of hearing loss with steroid and vasodilator therapy in Moyamoya disease.

#### **Declaration of conflicting interests**

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

#### **Funding**

The authors received no financial support for the research and/or authorship of this article.

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