CASE REPORT

Dolichoectatic and Tortuous Vertebrobasillary Arterial System Causing Progressive Left-Sided Hearing Loss in a Patient with Previous Right-Sided Deafness

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Giant dolichoectatic aneurysms in the vertebrobasilar system are rare. They usually manifest themselves when they compress the brainstem or cranial nerves, or when they are complicated by intraluminal thrombi or dissection. A 63-year-old man presented with a 6-month history of progressive left-sided hearing loss and tinnitus, in addition to having hearing loss in the right ear since childhood. In audiological assessment total hearing loss was present in the right ear, while a mild degree of sensorineural hearing loss was noted in the left ear, especially at high frequencies. Speech discrimination was lower than normal, which was inconsistent with the degree of measured hearing loss, suggesting retrocochlear pathology. The tone decay test was positive for the left ear. All these findings, including an auditory brainstem response test, suggested the existence of retrocochlear pathology on the left side. Temporal-cranial MRI and cerebral angiography revealed a dolichoectatic basillar artery aneurysm that was compressing brainstem and bilateral tortuous dolichoectatic vertebral arteries compressing the root entry zones of the 7-8th cranial nerves. No surgical or vascular interventional treatment was recommended because of the high risk of complication based on the location of the lesion. After 1 year of anti-aggregant and antihypertensive treatment, audiological tests revealed total hearing loss in the left ear. Anti-aggregant and antihypertensive treatment was continued.

Dolichoectasia of the vertebrobasilar system is an elongation and dilatation of the major arteries of the posterior fossa [1]. This condition is also referred to as a fusiform aneurysm of the vertebral and basilar arteries [1]. Aneurysms of the vertebrobasilar circulation comprise up to 15% of all intracranial aneurysms [2,3]. Giant aneurysms in the vertebrobasilar system are uncommon, comprising just 8% of all giant intracranial aneurysms [2].

CASE

A 63-year-old man presented with a 6-month history of progressive left-sided hearing loss and tinnitus, in addition to having hearing loss in the right ear since childhood. He had been given an air-conduction hearing aid for the left ear at another medical center, but the device did not improve his hearing. He pointed out that he could hear voices, but could not understand any words. The patient also reported that he began experiencing loss of balance while walking for the last 2 years. There was no vertigo or dizziness, nausea, or vomiting. Past medical history was uneventful, except for hypertension. Ear examination revealed bilateral exocytosis of the external auditory canal and sclerotic tympanic membranes. There was no nystagmus and no paresis or paralysis of the facial nerve or other cranial nerves. The remainder of the ENT examination was within normal limits. An audiological assessment was performed. Total hearing loss was noted in the right ear, while a mild degree of sensorineural hearing loss was noted in the left ear, especially at high frequencies (Figure 1). Speech discrimination was 52% at a sound level of 70 dB for the left ear, which was inconsistent with the mild degree of hearing loss we measured, suggesting retrocochlear pathology. The tone decay test was positive for the left ear, which prompted us to consider left retrocochlear pathology. The left ear's responses to the otoacoustic emission test were positive for the 1-kHz, 2-kHz, and 6-kHz sound levels, and negative for the 4-kHz sound level. All these findings, including an auditory brainstem response test, suggested the existence of retrocochlear pathology on the left side. Temporal and cranial MRI revealed a dolichoectatic vertobasillary arterial system with a right vertebral artery crossing to the left and coursing near the contralateral one. Moreover the basillary artery showed a prominent fusiform aneurysmatic dilatation and compressed the left middle cerebellar peduncle and the brainstem anteriorly on the left. The two tortious vertebral arteries impinged upon the root entry zone of the 7-8th cranial nerves on the brainstem (Figure 2). Cerebral angiography revealed severe dolichoectatic fusiform aneurysmal dilatation of the basilar artery involving the entire basilar artery (Figure 3). The interventional radiology and neurosurgery departments evaluated the patient to determine if any endovascular or surgical interventions were possible, but no interventional treatment was recommended because of the high risk of complication risk due to the location of the lesion. It was decided to follow-up the patient with medical treatment, and anti-aggregant and anti-hypertensive treatments were started. He was encouraged to use a hearing aid device for the left ear. We followed-up the patient about for 1 year without any problem. At the 1-year follow-up visit the patient reported an increase in his left-sided hearing loss. Follow-up audiological assessment was performed, which demonstrated that the left ear hearing loss had progressed (Figure 4). The anti-aggregant and antihypertensive treatment was continued.

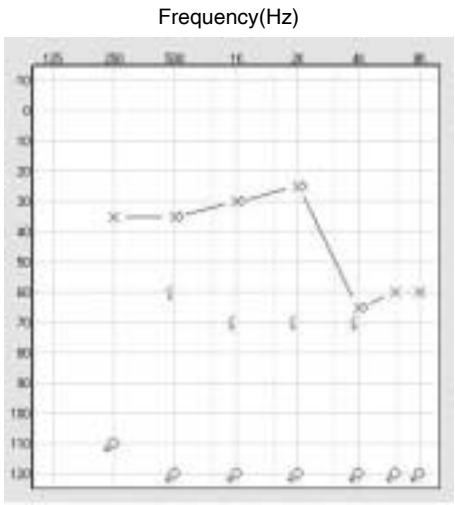

Figure-1: Upon the patient's initial presentation, total hearing loss in the right ear and a mild degree of sensorineural hearing loss in the left ear were detected in the pure tone audiogram.

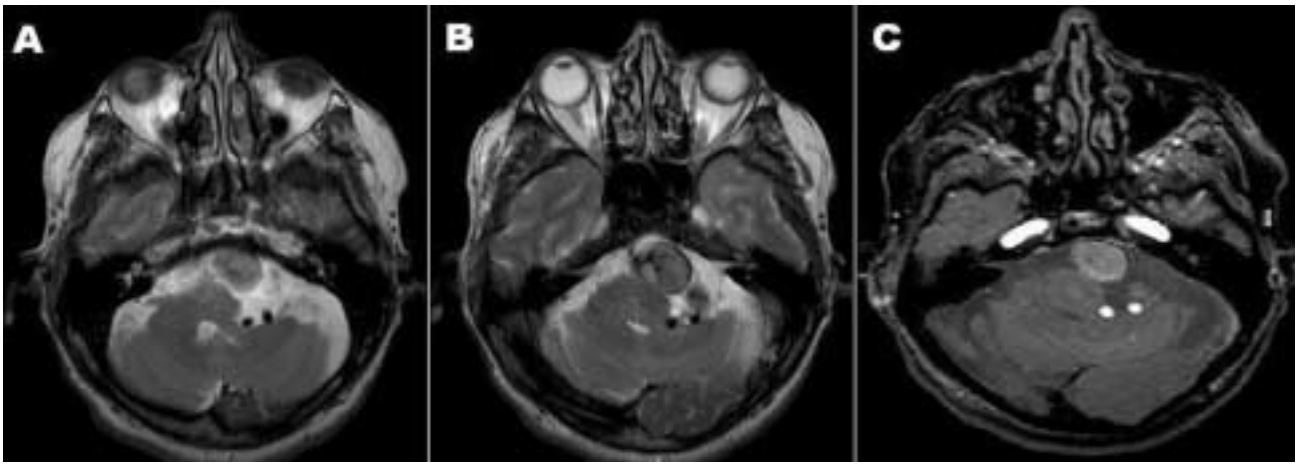

Figure-2: Axial T2-weighted imaging shows two tortuous dolichoectatic vertebral arteries compressing the root entry zone of 7-8th nerve complex and dolichoectatic fusiform basilar artery aneursym with a mass effect on the brainstem and left cerebellar peduncle (A, B). On brain MR angiography, arteries are well depicted (C).

Figure-3: Left vertebral artery injection on cerebral angiography shows the bilateral dolichoectatic vertebral artery (also seen retrograde filling of the right vertebral artery) and severe dolichoectatic fusiform aneurysmal dilatation of the basilar artery, involving the entire basilar artery.

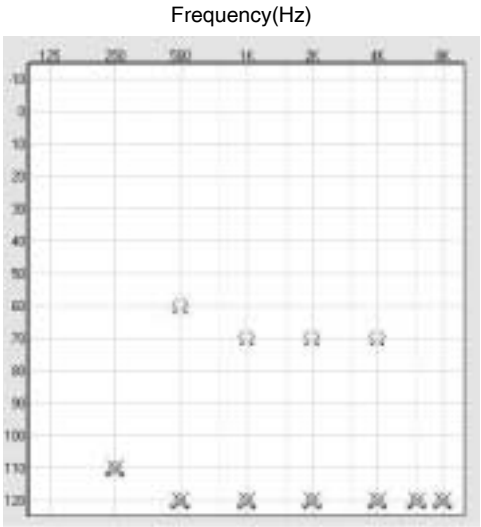

Figure-4: Progression from a mild degree of hearing loss to total hearing loss in the left ear was detected with the 1-year follow-up audiogram.

DISCUSSION

Intracranial fusiform aneurysms can be divided into 2 clinically distinct subtypes. The first is acute dissecting aneurysms, which usually cause subarachnoid hemorrhaging or cerebral ischaemia. The other type, chronic fusiform or dolichoectatic aneurysms, are relatively slow growing, but may lead to serious complications as they progress [4]. When a chronic fusiform aneurysm is giant-sized it is called a dolichoectatic aneurysm. The incidence of intracranial dolichoectasia ranges from 0.06% to 5.8% [5]. According to angiographic and postmortem studies, the vertebrobasilar system is most frequently involved [5]. Most patients are ≥ 50 years old and have arterial hypertension [5]. Giant aneurysms are defined as those with a diameter ≥ 25 mm [2].

While most patients are asymptomatic, they can also present with cranial nerve dysfunction (compression of the cranial nerves or brainstem), transient ischemic attacks, hydrocephalus, subarachnoid hemorrhaging and midbrain infarct, and ischemia in the vertebrobasilar arterial territory [1,6]. Vertebrobasilar dolichoectasia usually manifests itself when it compresses the brainstem or cranial nerves, or when it is complicated by intraluminal thrombi or dissection. Cranial nerve dysfunction involving the trigeminal through the hypoglossal nerves has been reported in aneurysms of the posterior circulation [3]. In 21%-35% of the patients a dilated basilar artery leads to symptomatic brainstem and clinical nerve compression [5]. Hearing loss and vertigo can be presenting symptoms. In such cases audiological testing can prompt the clinician to consider retrocochlear pathology. In particular, one-sided asymmetric hearing loss and decreased speech discrimination are important clues suggesting the presence of retrocochlear pathology. As in the presented case, a tone decay test can still be important for indicating the presence of retrocochlear pathology. If there is suspicion of retrocochlear pathology, an imaging study is required. The lesion may be found incidentally on intracranial imaging studies. Vascular abnormalities of the basilar artery can produce symptoms of vertebrobasilar insufficiency, such as vertigo, by leading to hypoperfusion [7]. Bilateral loss of cranial nerve VIII function has been reported in a rare case of bilateral ischemia of the labyrinthine artery due to basilar dolichoectasia [8].

Treatment options are limited in patients with vertebrobasilar fusiform aneurysms. Most cerebrovascular accidents associated with dolichoectasia are ischemic, although some are hemorrhagic. Most often, patients are treated medically with anti-platelet or anticoagulant agents. Untreated intracranial aneurysms can lead to a number of severe consequences related to mass effect, nerve compression, and thromboembolic stroke, and they can rupture and cause a subarachnoid hemorrhage [9]. Intracranial bleeding in these patients is considered uncommon; only a small percentage of untreated intracranial aneurysms will lead to rupture and subarachnoid hemorrhaging [9]. Passero et al. found a relationship between intracranial bleeding, and maximum diameter of the basilar artery, degree of lateral displacement of the basilar artery, hypertension, use of anti-platelet and anticoagulant agents, and female gender [6]. In the treatment of this condition, neurointerventional endovascular and conventional surgical methods can be used [2]. Management often poses difficult therapeutic problems, and surgery-related morbidity and mortality may be high, depending on the size of the aneurysm [2]. Surgical procedures include end-to-end anastomosis, aneurysmorraphy, aneurysm clipping, proximal ligation, and aneurysm trapping and wrapping [2,10]. The morphology of fusiform aneurysms makes them unsuitable for clipping and surgical access to many posterior circulation aneurysms is difficult, which is especially true for proximal basilar artery aneurysms [2]. Interventional neuroradiology has an expanding role in the treatment of intracranial aneurysms. With endovascular occlusion techniques, temporary arterial occlusions can be performed while the patient is fully conscious, facilitating continuous neurological testing [2]. The incidence of ischaemic complications resulting from endovascular occlusions of the basilar and vertebral arteries range from 5% to 31% for transient, and 0% to 18% for permanent neurological deficits [2]. In addition to anticoagulation or anti-platelet therapy, rigorous management of arterial hypertension is recommended.

The diagnosis of vertebrobasilar aneurysms is traditionally made with conventional catheter angiography, which is considered the gold standard. With this invasive procedure the risk of stroke is estimated to be 0.5%-1.0%; therefore, it is recommended to reserve this invasive procedure only for those patients for whom treatment is planned [9]. Three-dimensional CT angiography and MR angiography can also be used in diagnosing and following-up cerebral aneurysms that are ≥ 5 mm [9]. Unlike MR angiography and catheter angiography, CT

angiography cannot provide information about blood flow in the aneurysm [1]. However two benefits of MR angiography over CT- or catheter angiography are: 1) there is no need to injection of intravenous contrast material i.e. iodinated or gadolinium-based 2) there is no radiation exposure. Vestibular schwannomas, meningiomas, metastases, endolymphatic sac tumors, and other vascular lesions should be considered in the differential diagnosis of the disease. Neuroimaging workup must be tailored individually to provide accurate and optimal follow-up.

Vascular abnormalities including dolichoectatic and tortuous vertebrobasillary arteries and aneursyms can cause severe dysfunction of cranial nerve VIII. One should consider this entity in the differential diagnosis of retrocochlear pathologies, especially in the presence of impaired audiological test results.

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