Acute Unilateral Hearing Loss as an Early Symptom of Lateral Cerebral Sinus Venous Thrombosis

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Background: Increasing availability of neuroimaging has facilitated the diagnosis of cerebral sinus venous thrombosis (CSVT). However, CSVT may also present with unspecific or atypical symptoms, resulting in diagnostic delay. Single reports suggested otologic symptoms as such pitfalls.

Objective: To screen patients with CSVT for otologic symptoms.

Design: Ten-year retrospective case series.

Setting: Primary and tertiary care university clinic.

Patients: Thirty-eight patients with CSVT.

Results: Of 38 patients with CSVT, 3 individuals had acute unilateral hearing loss, 2 of which also had comitant tinnitus and headache, and were initially treated at the ear, nose, and throat department. Magnetic resonance imaging after hospital discharge showed ipsilateral thrombosis of the lateral venous sinus. Two female patients took oral contraceptives, 1 of whom also had a heterozygous factor V Leiden mutation.

Conclusions: Cerebral sinus venous thrombosis may present with unspecific symptoms such as acute unilateral hearing loss. If in conjunction with headache or risk factors for venous thrombosis, the suspicion of ipsilateral lateral CSVT should prompt rapid imaging including venography.


Cerebral sinus venous thrombosis (CSVT) accounts for approximately 0.5% of all cerebrovascular diseases, mainly affecting young female patients.\(^1\) Presenting symptoms may show great diversity, depending on the location, extension, and prevailing pathophysiology of the process (eg, they may be related to elevated intracranial pressure such as headache or to focal ischemia or hemorrhage such as hemiparesis and aphasia).\(^1,\)\(^2\)

Noninvasive imaging techniques have largely facilitated the diagnosis of CSVT and may also increase its detection without classic symptoms such as impaired consciousness, focal neurologic deficits, and generalized seizures. Indeed, case reports have called attention to unexpected symptoms such as isolated headaches, visual loss, auditory hallucinations, vertigo, confusion, cognitive deficits, and psychiatric disturbances.\(^1,\)\(^5\)

Previously, it was suggested that otologic symptoms such as hypacusis or tinnitus may represent another mode of atypical clinical presentation, particularly of lateral CSVT.\(^6,\)\(^9\) This prompted us to screen presenting symptoms in a consecutive series of patients with CSVT focusing on such symptoms. Here we report clinical and neuroimaging features including thorough follow-up information of 3 CSVT cases in whom acute hearing loss preceded the diagnosis of an ipsilateral lateral CSVT.

METHODS

We identified 38 patients diagnosed as having CSVT at our clinic from January 2000 to October 2010 based on the in-hospital radiological database.

After careful review of medical records for demographics, presenting symptoms, and risk factors for venous thromboembolic events, we identified 3 patients who had acute unilateral hearing loss and were all first seen at the local ear, nose, and throat department. All patients had brain computed tomographic (CT) or MRI scans including venography.
REPORT OF CASES

PATIENT 1

A 48-year-old woman had vertigo and nausea, and she was vomiting. She also described having left ear acute hearing loss, with tinnitus and bifrontal stabbing headache. Audiometry confirmed unilateral moderate sensorineural deafness. Results from neurologic examination and lumbar puncture were otherwise normal, and brain CT was interpreted as normal. She was treated with hydroxyethyl starch and pentoxifylline for 7 days. An audiogram at hospital discharge showed no improvement. Outpatient brain MRI 15 days after initial presentation showed a lateral CVST, affecting the left sigmoid and transverse sinuses and the internal jugular vein, without parenchymal involvement (Figure, A, B, and C). At that time, she still reported left ear hypacusis and tinnitus; her headache had subsided. Retrospective analysis revealed a thrombus formation (hyperdense signal) also in the initial CT scan (Figure, A). Identified risk factors comprised oral contraception for 30 years and a heterozygous factor V Leiden mutation. After intravenous heparin therapy following oral phenprocoumon, she was discharged from the hospital after 12 days free of otologic symptoms. Follow-up MRI after 6 months showed partial recanalization of the sigmoid sinus. Phenprocoumon treatment was discontinued after 12 months. There were no further events during 24 months of follow-up.

PATIENT 2

A 33-year-old woman had acute right ear hearing loss, associated with diffuse frontal and lateral right-sided headache. The audiogram showed mild sensorineural hearing loss. Symptoms subsided following 5-day treatment with hydroxyethyl starch, pentoxifylline, and corticosteroids. Outpatient brain MRI 5 days after hospital discharge showed a right-sided thrombosis of the transverse and sigmoid sinuses and internal jugular vein (Figure, D, E, and F). Results from neurologic examination and thrombophilia screening were normal. The only risk factor identified was oral contraception use. She was treated with intravenous heparin, and she was subsequently prescribed an oral antiocoagulant for 6 months. At discharge from the neurology department, the audiogram was normal, parallel to recovered hearing function. During 32 months of follow-up including an uneventful pregnancy, there were no further events.

PATIENT 3

A 48-year-old man had left ear acute hearing loss and ipsilateral tinnitus. Eight years prior, he had bilateral hearing loss. The audiogram demonstrated bilateral sensorineural deafness. He received hydroxyethyl starch and pentoxifylline infusions for 10 days and corticosteroids for 3 days. A control audiogram objectified improvement of 10 dB on the left side; the tinnitus diminished. Forty-three days later, he was admitted to the neurology department because of the diagnosis of thrombosis of the left sigmoid and transverse sinuses on MRI (Figure, G, H, and I). At this time, hypacusis and tinnitus were still present. No risk factors could be detected. He was prescribed an oral anticoagulant for 6 months. Follow-up MRI after 3 months showed partial recanalization of the left transverse sinus. During follow-up of 92 months, he had no further neurologic events except for 1 acute exacerbation of the left-sided tinnitus.

All 3 reported patients had an excellent prognosis (modified Rankin scale score of 0 at follow-up), and none have experienced other thromboembolic events so far (eg, deep vein thrombosis or pulmonary embolism).

COMMENT

Reviewing 38 patients with CSVT, we identified 3 individuals with acute hearing loss who were initially treated at the ear, nose, and throat department. Two of them had concomitant ipsilateral tinnitus and 2 had severe headaches. After confirmation of acute sensorineural hearing loss by audiogram, all 3 patients received rheologic infusion therapies. After hospital discharge, ambulatory brain MRI showed lateral CSVT ipsilateral to the side of hypacusis. At the neurology department, anticoagulation was initiated and prothrombotic conditions were detected in the 2 female patients, but none had local otogenic processes such as tumors, middle ear infections, or mastoiditis.

Diagnostic delay or misdiagnosis due to uncharacteristic presentation postpones appropriate therapy. Withholding early anticoagulation may promote thrombus progression, potentially leading to elevated intracranial pressure and secondary parenchymal involvement. Thus, characterizing CSVT cases with atypical symptoms is important to increase the chance of capturing them in an increasing number of patients with unspecific symptoms presenting at outpatient and emergency departments.

Thrombosis of the lateral sinus system poses particular diagnostic challenges, as headache is frequently isolated. In a large series of 62 patients with isolated lateral sinus thrombosis, hearing loss was found in 2 patients (3%), but no information about laterality of the symptoms and thrombosis was presented in this article. To our knowledge, we thus provide the first report of 3 stereotyped cases with acute unilateral hearing problems (also supported audiometrically) as the presenting symptom of an ipsilateral lateral CSVT. There also exist a few single case reports relating acute hearing loss to lateral sinus thrombosis.

These and our cases share several similarities. First, they all were relatively young, following the typical age pattern of CSVT. Second, all but 1 (patient 3) had concomitant headache. Third, prothrombotic risk factors or thrombophilia were present in all patients except patient 3 and 1 patient with a local trauma. Fourth, none showed parenchymal involvement, potentially explaining excellent prognosis. Finally, hearing function improved with anticoagulation therapy, and neuroimaging confirmed (partial) recanalization in all individuals.

There may be 2 different mechanisms linking lateral CSVT with acute sensorineural hearing loss. (1) The cochlear venous blood drains through the cochlear veins and
the labyrinthine vein into the petrosal sinus or directly into the transverse sinus. Thus, lateral CSVT might increase inner-ear pressure, induce anoxia, and impede cochlear function. Supportive evidence comes both from animal experiments and histopathology.6,8,12-14 (2) Alternatively, elevated intracranial pressure could be transmitted to the endolymph, disturbing hearing function via increased inner-ear fluid pressure or directly leading to compression and dysfunction of the vestibulocochlear nerve.8,15 However, this appears unlikely in our setting given the lack of neuroimaging evidence for intracranial hypertension in these patients.

Figure. Neuroimaging studies of patients demonstrating lateral cerebral sinus venous thrombosis (arrows) ipsilateral to acute hypacusis. Computed tomographic scan (A); axial fluid-attenuated inversion recovery images (B and E); sagittal, axial, and coronal T1-weighted images (D, G, and H, respectively); and venous magnetic resonance angiographic images (C, F, and I).
Our observations reflect experience from a single tertiary care university center obtained by retrospective analysis, thus they may be biased. Consequently, we acknowledge that no conclusion on causation can be made from this work. Moreover, it is not unlikely that otologic symptoms might be neglected in the presence of severe headaches, focal deficits, or other more striking symptoms. Thus, it is tempting to hypothesize that otologic symptoms might have been underestimated in previous CSVT studies. Our observations may also be relevant for otolaryngologists testing patients for acute idiopathic sensorineural hearing loss.

In summary, we suggest acute otologic symptoms as potential early warning signs of lateral sinus thrombosis that should be considered in future prospective CSVT studies. In particular, in conjunction with headache or prothrombotic risk factors, rapid neuroimaging including venography appears warranted to rule out CSVT.

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