Acute Severe Headache Associated with Rapid Tumor Growth in a Patient with Vestibular Schwannoma

Hoon Oh, Kyung Soo Kim

Abstract

Purpose: Although headache in vestbibular schwannoma (VS) is uncommon as an initially presenting symptom, headache in VS is usually associated with tumor growth due to various factors, especially abrupt increase in size. We discuss various factors which influence rapid tumor growth including intratumoral hemorrhage and cystic change of tumor.

Case Report: A 68-years old female presented with sudden acute onset of occipital headache and severe dizziness. Brain MRI revealed 28 × 18 × 22 mm sized vestibular schwannoma with cystic change and heterogeneous enhancement (intratumoral hemorrhage) in the left cerebellopontine cistern with intracanalicular extension.

Conclusion: This is a rare case of vestibular schwannoma with cystic change due to intratumoral hemorrhage with atypical apoplectic presentation on the background of hearing loss of 3 year duration. This highly unusual case highlights the need for careful clinical and radiological follow-up, in patients with vestibular schwannoma. In addition, although the predictive value may be limited, we must keep in mind that acute severe headache may be an alarming feature that predicts rapid tumor growth.

Key Words: vestibular schwannoma, severe headache, tumor growth, intratumoral hemorrhage, cystic change

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INTRODUCTION

The vast majority of acoustic schwannomas arise from the Schwann cell investment of the vestibular division of the eighth cranial nerve and only few (less than 5%) from the cochlear division[1]. So, the term "vestibular schwannoma (VS)", reflecting the cell and nerve of tumor origin is preferred and is recommended by the National Institute of Health Consensus Statement[2]. Symptoms and signs associated with VS(s) vary depending on the size at diagnosis, direct tumor compression, invasion or vascular compromise of the surrounding structures and whether there is any obstruction to cerebrospinal fluid pathways, such as slowly progressive sensorirneural hearing

From the Department of Otorhinolaryngology-Head and Neck Surgery, Chung-Ang University College of Medicine.
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Correspondence to: Kyung Soo Kim, M.D., Ph.D. Department of Otorhinolaryngology-Head and Neck Surgery, Chung-Ang University College of Medicine, 224-1, Heukseok-dong, Dongjak-gu, Seoul, Korea,
E-mail: 99-21045@hanmail.net

loss (most common), tinnitus, vestibular disturbances (balance disturbances), vertigo, cerebellar dysfunction, cranial nerve dysfunction, and secondary obstructive hydrocephalus[3-5]. Although headache in VS is uncommon as an initially presenting symptom[6], headache in VS is usually associated with tumor growth due to various factors such as intratumoral hemorrhage[7].

Herein we report a patient of vestibular schwannoma with cystic change due to intratumoral hemorrhage presenting with a sudden acute onset of occipital headache. This highly unusual case highlights the need for careful clinical and radiological follow-up, in patients with vestibular schwannoma.

CASE REPORT

A 68-years old woman presented with a sudden acute onset of occipital headache and severe dizziness. Her headache character was described as like 'being hit over the head with a hammer'. She originally presented with a history of left-sided hearing loss and tinnitus for 3 years. At the time of her visit, she did not have any signs of headache or dizziness, and there was no significant past medical history. Otoscopy and cranial nerve examination were otherwise unremarkable. Pure tone audiometry and auditory brain stem evoked response confirmed left unilateral hearing loss (deafness). Her tinnitus was intermittent and of high pitch. Brain magnetic resonance imaging (MRI) revealed a 16 × 11 × 12 mm sized tumor in the left cerebellopontine cistern, showing T1 hypointense, T2 heterogeneous intensity-irregular enhancement with extension to the internal auditory canal, suggesting vestibular schwannoma (Figure 1). The vestibular function test revealed that there was left canal palsy of 81%, but her dizziness symptom was not severe. Due to the patient's old age, both the patient and her guardian chose not to undergo surgery and decided to closely observe the size of the tumor with MRI every six months. After 2.5 years of follow up, an MRI revealed a slightly marginal increase in size which measured 18 × 12 × 12.5 mm, but there was no cystic change or intratumoral hemorrhage (Figure 1).

The patient agreed to regularly follow up the size of the tumor every year. However, she did not visit the hospital thereafter during 2 years. Then, one day she

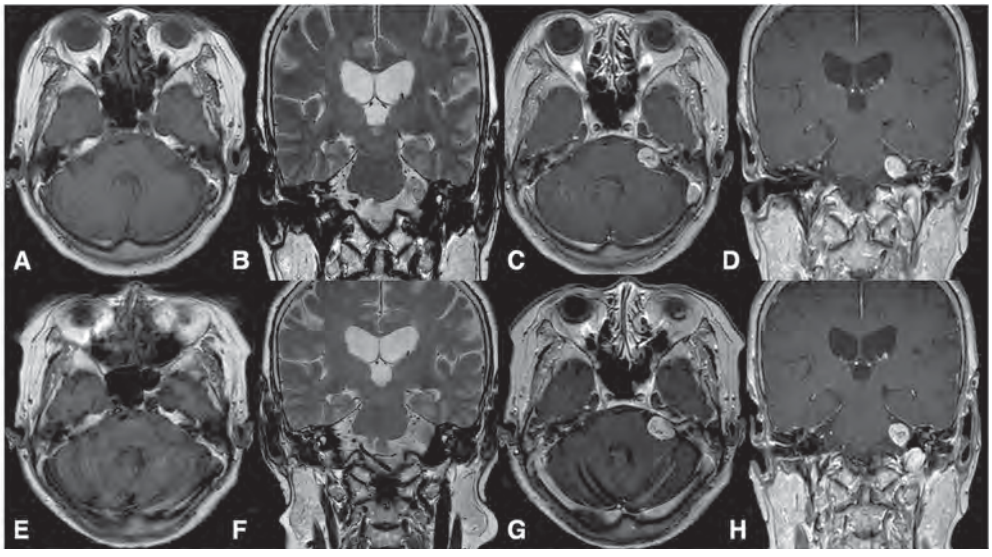

Figure 1. Brain MRI at the time of diagnosis revealed a 16 × 11 × 12 mm sized tumor in the left cerebellopontine cistern, showing T1 hypointense, T2 heterogeneous intensity-irregular enhancement with extension to IAC, suggesting vestibular schwannoma. (A: T1 weighted axial, B: T2 weigted coronal, C & D: T1 weighted axial and coronal view with contrast enhancement) After 2.5 years of follow up, MRI revealed a slightly marginal increase in size which measured 18 × 12 × 12.5 mm, but there was no cystic change or intratumoral hemorrhage. (E: T1 weighted axial, F: T2 weighted coronal, G & H: T1 weighted axial and coronal view with contrast enhancement)

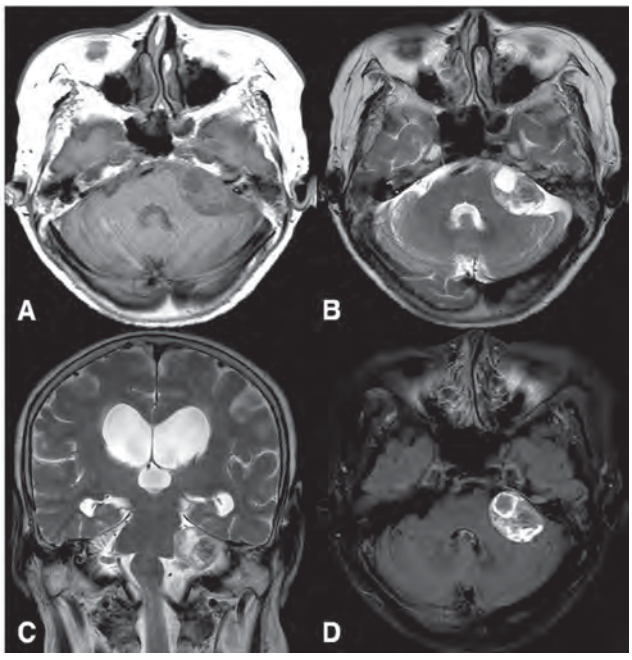

Figure 2. Brain MRI revealed 28 × 18 × 22 mm sized vestibular schwannoma with cystic change and heterogeneous enhancement (intratumoral hemorrhage) in left cerebellopontine cistern with intracanalicular extension when the patient came to emergency room with severe headache. (A: T1 weighted axial, B: T2 weighted axial, C: T2 weighted coronal, D: Fat-suppressed, T1 weighted axial)

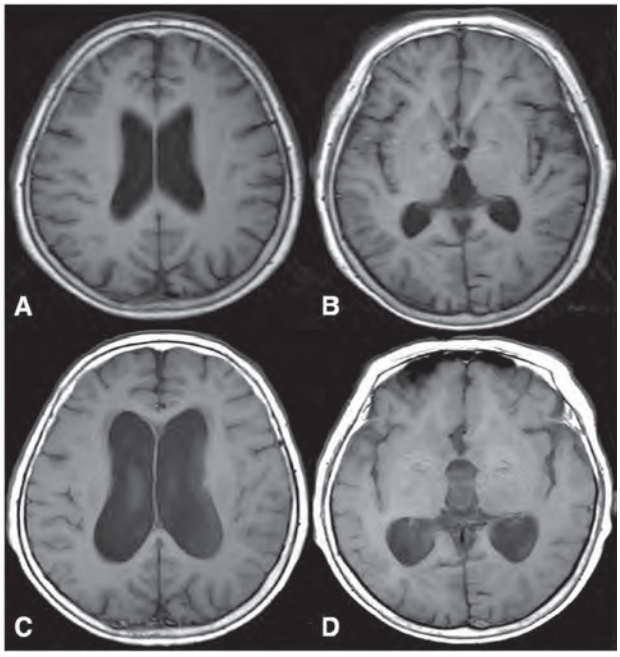

Figure 3. T1-weighted axial brain MRI showed increased volume of lateral and 3rd ventricles, suggestive of obstructive hydrocephalus. (A & B: at the time of diagnosis, C & D: aggravated state)

came to the emergency department due to an acute onset of severe headache. The headache was characterized as continuous, stabbing, and as if being hammered in the occipital area. She also complained of severe dizziness with no other associated symptoms. There were no significant neurologic symptoms. Emergent brain CT and MRI were performed. Both brain CT and brain MRI revealed that the size of the vestibular schwannoma had grown to 28 × 18 × 22 mm and also showed some newly developed cystic change and heterogeneous enhancement (intratumoral hemorrhage) in left cerebellopontine cistern with intracanalicular extension (Figure 2). In addition, it showed increased volume of the lateral and 3rd ventricles, suggestive of obstructive hydrocephalus (Figure 3). After relieving the symptoms with conservative management, she was admitted for surgical treatment.

DISCUSSION

Headache associated with vestibular schwannoma (VS) may vary greatly in location, duration and intensity. The pain may occur early in the clinical course and progress in severity as the tumor grows. In one study, headache was present in 19% of the patients and the frequency increased as the tumor size increased. Patients with a tumor less than 1 cm had no headache, 20% of patients with a tumor size of 1 to 3 cm had headache, and when the tumor was larger than 3 cm, 43% of the patients complained of headache[8]. There is a predilection for the frontal and occipital regions, but the pain may be located in any region of the head. The headache is unilateral, dull or aching in quality and may radiate to the neck, top of the head or front of the head. It is particularly bad in the morning and at night. It tends to get aggravated while lying down, standing up, coughing and sneezing[9].

There are three important mechanisms of the headache development in VS patients: 1) the increase of tumor

size, 2) obstructive hydrocephalus associated with a large tumor blocking the flow of cerebrospinal fluid, and 3) as the tumor grows, the capsule of the tumor presses upon the lining of the inside of the skull (the dura), which has sensory fibers that can transmit the sensation of pressure[10].

Of these, the most important point in this present case is abrupt increase in tumor size. The natural history of VS remains variable with some tumors showing growth, some remaining stationary and a few even shrinking[11]. Because there are no parameters known that predict which tumor will grow and to what extent, the patient is evaluated periodically for symptom assessment, and follow-up MRI scans are obtained to monitor the tumor for signs of growth[12]. VSs exhibit slow and various growth rates as reported previously in the literature, with an average growth rate of 1.9 mm/year in a recent meta-analysis[13]. Furthermore, many consider a 2 mm increase in the greatest diameter clinically significant to intervene with a 3 mm difference in two consecutive MR studies being an evident sign of tumor progression[5]. In the present case, the growth pattern may be slow because the tumor grew about 2mm during 2.5 years on sequential imaging studies.

Various factors which influence rapid tumor growth include rates of cellular proliferation, hemorrhage, infarction, alterations in blood supply, and cystic degeneration or presence of cystic tumors[14]. Even if VS is a slow-growing tumor usually accompanied by gradual onset of symptoms and signs, rapid tumor growth has been associated with apoplectic clinical presentation, including acute neurologic changes and deterioration[5,7,15]. Severe headache, acute vertigo, unilateral hearing loss, nausea or vomiting, and depressed consciousness have been the most common symptoms associated with an acute event[7,15].

In the present case, cystic degeneration with intratumoral hemorrhage in VS influenced rapid tumor growth which led to an acute severe headache similar to the headache character in subarachnoid hemorrhage.

VSs are predominantly solid tumors but cystic degeneration in these tumors can give rise to problems of differentiation. Small cystic changes in VS are not uncommon and have been observed in 9.6-20.5% of cases. Degenerative changes appear to be the principal cause of cyst formation[16]. In addition, there are a number of possible factors which lead to cystic development in VSs: microinfarction, hemorrhage, or radiation related antineoplastic effects with central necrosis[7]. Recently, enzymes like matrix metalloproteinase-2 have been thought to play a role in cyst enlargement[17].

Intratumoral hemorrhage in VS, which usually has an apoplectic onset, remains a less common (<1%) but important clinical event[7]. Mechanism of intracranial tumor bleeding is still not entirely understood. The tumor hemorrhage is thought to result from rupture of the abundant dilated vessels within the tumor. Most reported cases have been spontaneous. Some cases, however, have been linked to a variety of clinical situations, including chronic hypertension, anticoagulation therapy, gamma knife therapy (radiosurgery), previous subtotal resection, pregnancy, voiding, heavy exertion, and minor head trauma[18]. The risk of hemorrhage has been associated with tumor size, certain morphological or histologic features. Larger schwannomas (>2cm) and tumors that contain large or multiple intratumoral cysts are more likely to hemorrhage[19]. Moreover, the "liquefactory nature" of the Antoni B tumor, in close proximity to abnormal vascularity, makes this type of tumor susceptible to hemorrhage[18]. In the present case, the patient-related factor of the rapid tumor growth was not found in any clinical situations.

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