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An unusual cause of superficial siderosis of central nervous system: A case report of a vestibular schwannoma

Santral sinir sistemi yüzeyel siderozisin sıradışı bir nedeni: Vestibüler schwannoma olgusu

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Abstract
Vestibular schwannomas are relatively common benign tumors that account for 7-8% of all intracranial tumors. There is a limited number of cases of subarachnoid hemorrhage associated with vestibular schwannoma in the literature. The incidence of symptomatic tumor bleeding is historically reported less than 1%. Susceptibility-weighted imaging (SWI) is a relatively new Magnetic resonance imaging (MRI) sequence that is highly sensitive to compounds which distort the local magnetic field such as blood products and hemorrhage. We herein report the rare MRI findings of a 53-year-old man with unilateral vestibular schwannoma and hearing loss who was followed for 4 years. Initial diagnosis of the vestibular schwannoma was made on prior conventional MRI. We included SWI sequence in routine brain MRI protocol. During follow-up, SWI demonstrated focal superficial siderosis due to recurrent subarachnoid hemorrhage seen as hypointense blooming artifacts on the middle cerebellar peduncle-pons junction adjacent to the mass. We think that including SWI in routine MRI protocol will contribute to the initial diagnosis of acoustic schwannoma and detection of its complications such as subarachnoid hemorrhage and subsequent superficial siderosis.
Keywords: Vestibular schwannoma, Susceptibility weighted image, Superficial siderosis, MRI, Hemorrhage

Öz
Vestibüler schwannomlar, tüm intrakranial tümörlerin %7-8'ini oluşturan nispeten yaygın görülen iyi huylu tümörlerdir. Literatürde vestibüler schwannoma ile ilişkili az sayıda subaraknoid kanama vakası vardır. Semptomatik tümör kanaması insidansı %1'den daha az olarak rapor edilmiştir. Duyarlılık ağırlıklı görüntüleme (DAG), kan ürünleri ve kanama gibi lokal manyetik alanı bozan bileşiklere karşı oldukça hassas olan yeni bir Manyetik rezonans görüntüleme (MRG) sekansıdır. Bu yazıda, 4 yıllık takipteki tek taraflı vestibüler schwannoma ve işitme kaybı olan 53 yaşında erkek hastanın nadir MRG bulgularını bildirmeyi amaçladık. Olgunun vestibüler schwannoma tanısı daha önceki konvansiyonel MRG bulguları ile koyuldu. Takipte kullanılan rutin MRG protokolüne DAG sekansını dahil ettik. Takip sırasında DAG, orta serebeller pedinkül-pons bileşkesinde kitle komşuluğunda hipointens artefaktlar şeklinde izlenen, rekürren subaraknoid kanamaya bağlı fokal yüzeyel sideroz odağını gösterdi. DAG'nin rutin beyin MRG protokolüne dahil edilmesinin, akustik schwannomanın ilk teşhisine, ayrıca subaraknoid kanama ve buna bağlı yüzeysel siderosis gibi komplikasyonların tespitine katkıda bulunacağını düşünüyoruz.
Anahtar kelimeler: Vestibuler şvannom, Duyarlılık ağırlıklı görüntüleme, Yüzeyel siderozis, MRG, Hemoraji

Introduction

Vestibular schwannomas are relatively common benign tumors that account for 7-8% of all intracranial tumors [1]. Hemorrhage due to vestibular schwannoma is a very rare condition and the incidence of symptomatic tumor bleeding is historically reported less than 1% [2,3]. Bleeding may range from subarachnoid hemorrhage to massive intratumoral hemorrhage [4]. On the other hand, apart from the etiological reason, recurrent bleeding into subarachnoid space results in hemosiderin deposition along the leptomeninges in the superficial layers of the brain. This rare process is defined as superficial siderosis [5].

We herein report the rare Magnetic Resonance Imaging (MRI) findings of a 53-year-old man with unilateral vestibular schwannoma with focal superficial siderosis due to recurrent subarachnoid hemorrhage, who was followed up for 4 years.

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Case presentation

A 53-year-old man was admitted to the otorhinolaryngology department in our hospital due to progressive hearing loss on the left side. The patient did not suffer from ear discharge, pain, or fever. He and his family had no history of neurofibromatosis. Otoscopic examination was normal. The audiogram revealed sensorineural hearing loss with pure tone threshold of 78 dB in the left ear consistent with grade III (non-serviceable) hearing loss according to Gardner-Robertson Hearing Scale (Figure 1). Contrast-enhanced MRI demonstrated a vividly enhancing, lobulated left cerebellopontine lesion with extension into the intracanalicular segment of the left internal auditory canal. The mass lesion measured 23x18 mm in size and displaced and compressed the pons and the fourth ventricle (Figure 2). Susceptibility weighted imaging (SWI) was not included in our brain MRI protocol at that time. The diagnosis of vestibular schwannoma was made with conventional MRI findings. After the initial diagnosis, gamma-knife therapy was performed, and the patient was included in a follow-up program, which he dropped out of at the end of the first year to apply to another clinic. In the fourth year, he came to our department for follow-up MRI. The audiogram tests did not show any change. There was no sign of facial or trigeminal nerve paralysis. The MRI protocol included the following sequences: Axial, coronal, sagittal turbo-spin echo T2-weighted and post-contrast T1-weighted, axial gradient-echo T1-weighted, Fluid attenuation inversion recovery and SWI. A 18x15 mm residual mass lesion in the left cerebellopontine angle was observed. Findings of the pons and fourth ventricle compression were significantly regressed compared to the previous study. On SWI, there were hypointense blooming artifacts which represented focal superficial siderosis in the middle cerebellar peduncle-pons junction adjacent to the mass (Figure 3). No hypointensity compatible with an intratumoral hemorrhage was observed in the mass lesion.

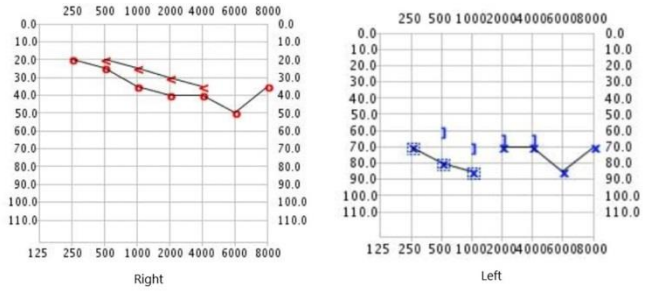

Figure 1: The audiogram of the patient

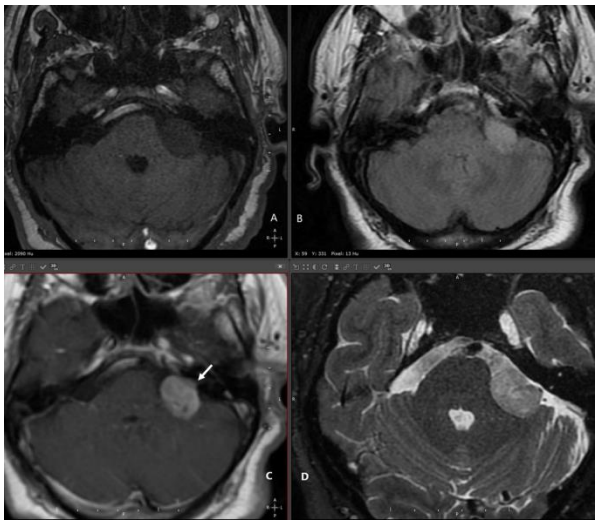

Figure 2: The first MR examination. Axial T1A (A), FLAIR (B), postcontrast T1A (C) and TSE T2A (D). Extension into the intracanalicular segment of the left internal auditory canal (white arrow)

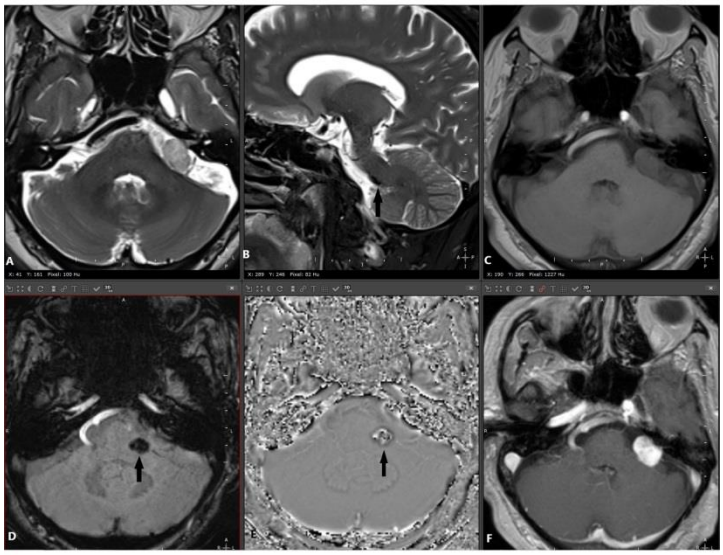

Figure 3: The last MR examination. Axial and sagittal TSE T2A (A, B), T1A (C), axial SWI and phase image (D, E), postcontrast axial T1A (F). Hypointense blooming artifacts which represent focal superficial siderosis in the middle cerebellar peduncle-pons junction adjacent to the mass (black arrows)

Discussion

Brain tumors constitute 0.4-5% of all subarachnoid hemorrhage etiologies [6,7]. Although vestibular schwannomas constitute 8% of all intracranial tumors, there is a limited number of subarachnoid hemorrhage cases associated with vestibular schwannoma in the literature. Despite the lack of an intratumoral hemorrhage finding on MRI, our patient had a focal superficial siderosis on middle cerebellar peduncle-pons junction adjacent to the vestibular schwannoma which was thought to be as a result of recurrent subarachnoid hemorrhage. In an extensive review by Kim SH et al. [2], the authors found 15 consecutive cases of subarachnoid bleeding from vestibular schwannoma. Similar to our patient, there was no intratumoral hemorrhage in 5 of 15 cases in this series.

Although the mechanisms of hemorrhage are not yet clarified, thin-walled dilated vessels, rapid tumoral growth, and cyst formation within a tumor were blamed for vascular fragility with subsequent rupture into subarachnoid space [3,8].

Clinical symptoms of hemorrhagic vestibular schwannomas depend on the size of the tumor and the type of hemorrhage. Larger tumor size (>2 cm) and presence of intratumoral hemorrhage tend to be associated with more

significant neurologic symptoms such as severe headaches, cranial nerve palsy, and even loss of consciousness due to hydrocephalus [2,9]. Our patient did not exhibit any of these symptoms.

Contrast-enhanced MRI is a valuable tool for identification of lesions in the cerebellopontine angle. Most frequent tumors of this area were meningioma and schwannoma, which account for 85–90% of all cerebellopontine angle tumors [10]. Differentiating between these two is critical because different treatment strategies need to be implemented. In their study, Mishra et al. [11] state that differentiating cerebellopontine meningioma from vestibular schwannoma with conventional imaging may be difficult. Presence of hypointense blooming artifact which represents microhemorrhages, an established histologic feature of vestibular schwannomas, is detected well by susceptibility-weighted imaging (SWI) and allows an accurate diagnosis. MRI should be the preferred modality for serial follow-up imaging in acoustic schwannomas. On MRI, residual tumor is best assessed with fat-suppressed contrast-enhanced T1-weighted images [12]. Since our cases had a history of repetitive subarachnoid bleeding with subsequent superficial siderosis, which was seen as blooming hypointensity near the middle cerebellar peduncle adjacent to the tumor on SWI, this imaging technique was thought to be one of the useful and feasible sequences for follow-up MRIs in this patient.

The treatment strategy is not different for hemorrhagic and non-hemorrhagic vestibular schwannomas, and it includes observation, surgical resection, and stereotactic radiotherapy. Especially in geriatric patients or those with small sized vestibular schwannomas with subclinical symptoms, observation is the suggested patient management strategy instead of surgical removal or stereotactic radiotherapy. Serial MRI imaging every 6-12 months is offered for observation in these patient group [3,13]. Follow-up imaging objectives could be listed as identification of recurrent or residual tumor, response to stereotactic radiotherapy, and assessment of tumor size and possible post-therapeutic complications [12]. Since our patient had grade III sensorineural hearing loss, gamma-knife radiotherapy was performed after the initial diagnosis. Serviceable hearing was preserved in first following year. The patient did not show any significant clinical symptoms or progressive hearing deterioration after stereotactic radiotherapy. MRI was chosen as the follow-up imaging modality. Unfortunately, we lost trace on him for 5 years. Six years after stereotactic radiotherapy, the patient still had a serviceable hearing. Routine follow-up MRI was suggested to the patient.

Conclusion

We believe that follow-up MRI in patients with acoustic schwannoma, SWI, is a feasible technique. It would contribute to the initial diagnosis of acoustic schwannoma and its complications, such as subarachnoid hemorrhage and subsequent superficial siderosis.

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