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Case Report / Olgu Sunumu

Intracranial Superficial Siderosis

İntrakranyal Superfisial Siderozis

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Summary

Superficial siderosis of the central nervous system results from hemosiderin deposition in the subpial layers of the brain and spinal cord. The condition is characterized by a classic triad of symptoms, which consists of sensorineural hearing loss, cerebellar ataxia, and myelopathy. This case report provides an account of a woman who presented with sudden vertigo attacks accompanied by sensorineural hearing loss.

Keywords: Intracranial siderosis, hearing loss, vertigo

Öz

Merkezi sinir sistemi süperfisial siderozis hastalığı beyin ve omirilik subpial tabakasında hemosiderin birikmesi nedeniyle oluşmaktadır. İşitme kaybı, serebellar ataksi, miyelopati triadı ile karakterizedir. Biz de işitme kaybı ve vertigo atakları ile tarafımıza başvuran bir siderozis olgusunu sunmak istedik.

Anahtar Kelimeler: İntrakranial siderozis, işitme kaybı, vertigo

Introduction

Superficial siderosis of the central nervous system results from hemosiderin deposition in the subpial layers of the brain and spinal cord. The condition is characterized by a classic triad of symptoms, which consists of sensorineural hearing loss, cerebellar ataxia, and myelopathy (1,2,3). This case report provides an account of a woman who presented with sudden vertigo attacks accompanied by sensorineural hearing loss.

Case Report

A woman aged 52 years presented with a 3-year history of progressive, bilateral sensorineural hearing loss. She had been affected by episodes of vertigo for about 6 months. The first episode occurred one morning after she woke up and ceased about one hour later. In the following 2 months, five more similar attacks occurred during the day with no triggering factors (2,3). Her neurologic examination was notable for bilateral hearing loss and lateral end-gaze nystagmus. The examination of other cranial nerves, motor strength, and sensory tests were normal. There was no abnormality in her coordination or gait. Routine laboratory tests and caeruloplasmin were normal. A cerebrospinal fluid (CSF) study was remarkable for an increase in red blood cells to 220 cells/mm^3 and a protein concentration of 110 mg/dL. Four-vessel cerebral angiography was normal. Magnetic resonance (MR) imaging showed a hypointensity consistent with siderosis along the pial surface of the brain stem and along the folia of the cerebellar vermis, T2 hypointensity appearance surrounding brain stem, and the 7th and 8th cranial nerves exit zones were present, which was evaluated as consistent with superficial siderosis (Figure 1). Audiometry showed bilateral down-sloping mild sensorineural hearing loss, which was slightly worse on the left side.

Discussion

Hemosiderin deposition after chronic subarachnoid hemorrhage (2,3,4,5) results in superficial siderosis of the

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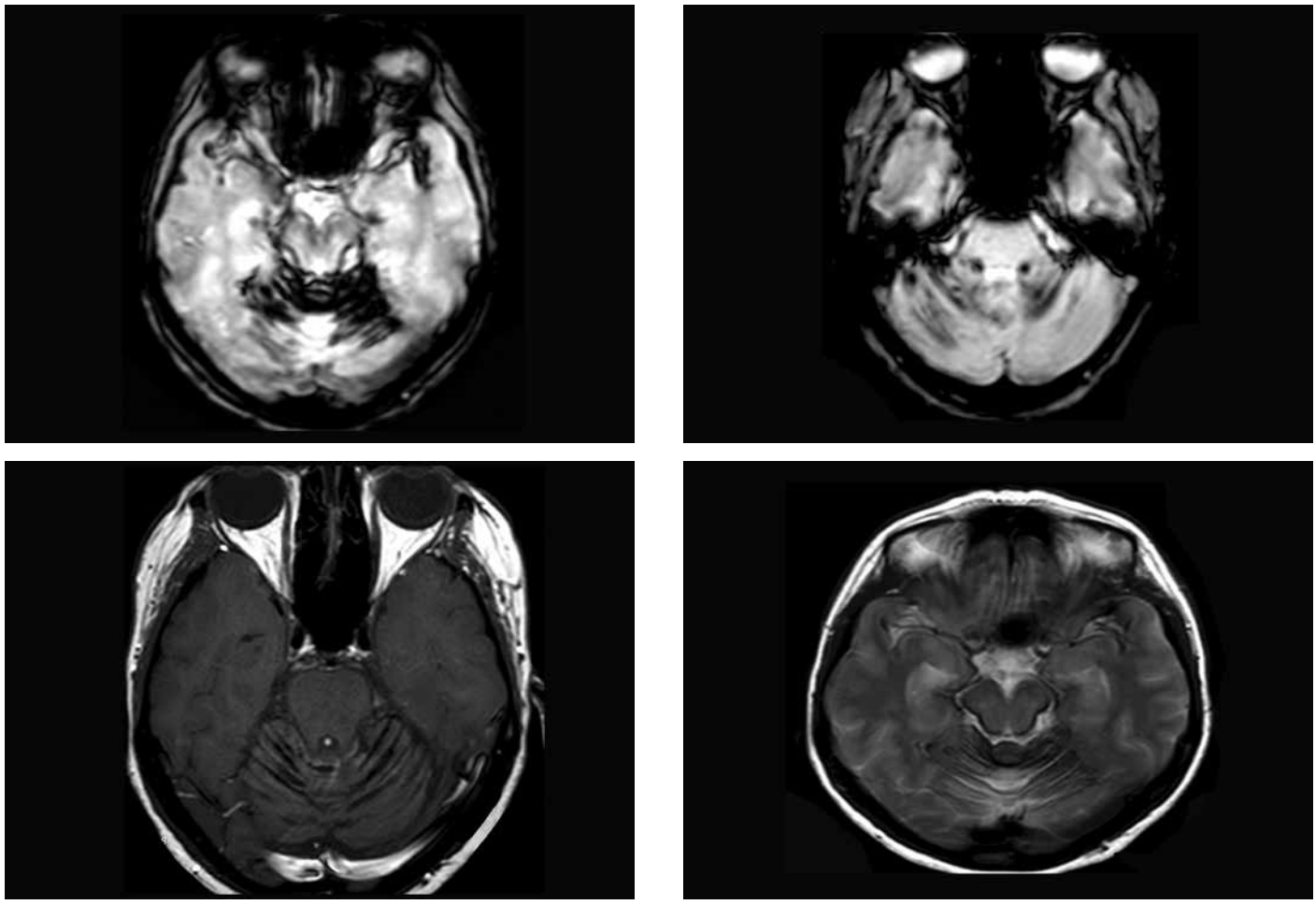

Figure 1. Magnetic resonance imaging showed hypointensity consistent with siderosis along the pial surface of the brain stem and along the folia of the cerebellar vermis, T2 hypointensity appearance surrounding the brain stem, and the 7th and 8th cranial nerves exit zones were present, which was consistent with superficial siderosis

central nervous system (SSCN). A microscopic tear, caused by dural pathology, tumors, vascular malformations, neurosurgical procedures, or trauma (2,6), allows blood into the subarachnoid space. No subarachnoid bleed could be identiŞed in our patient, as well as in one-third of patients with this diagnosis (3,6). This may be because an intraspinal fluid-filled collection is a common accompaniment and may be the likely source of bleeding.

In intracranial siderozis; the pigmentation has a predilection for the superior vermis, crests of the cerebellar folia, basal frontal lobe, temporal cortex, brainstem, spinal cord, nerve roots, and cranial nerves I and VIII. İn this patient; the eighth cranial nerve is particularly susceptible to iron deposition due to its long glial composition (7) thus being the likely cause of vertigo and sensorineural hearing loss. Hearing loss is often unilateral and/or asymmetric (4,8) and worsens over 1 to 15 years, which leaves the patient deaf or with a small island of hearing remaining (4). Our patient presented with sudden severe vertigo attacks accompanied by mild hearing loss. She was diagnosed eventually as having SSCN through brain MR. Vertigo can be the major presentation and may be accompanied by sensorineural hearing loss. In summary, although vertigo is rarely seen in patients with SSCN, it is necessary to consider this diagnosis when patients present with concomitant mild hearing loss.

Authorship Contributions

Informed Consent: A consent form was completed by all participants. Concept: Buket Özkara, Faik Budak, Design: Buket Özkara, Faik Budak, Data Collection or Processing: Buket Özkara, Analysis or Interpretation: Faik Budak, Buket Özkara, Literature Search: Buket Özkara, Writing: Buket Özkara, Faik Budak, Peer-review: Internal peer-reviewed, Conflict of Interest: No conflict of interest was declared by the authors. Financial Disclosure: The authors declared that this study has received no financial support.

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