Case Report
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대한영상의학회지

Bilateral Wallerian Degeneration of the Middle Cerebellar Peduncle and Unilateral Hypertrophic Olivary Degeneration Secondary to Pontine Hemorrhage: A Case Report

뇌교출혈 후 이차적으로 발생한 양측 중간소뇌다리의 월러변성과 편측 비대성올리브핵변성: 증례 보고

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The two distinct types of axonal degeneration that occur after neuronal injury include Wallerian degeneration (WD) and transneuronal degeneration. The most commonly recognizable cause of secondary degeneration is cerebral infarction, but may also include a variety of conditions including hemorrhage, trauma, necrosis, and focal demyelination. Herein, we present a rare case of WD of the cerebellar peduncles accompanied by unilateral hypertrophic olivary degeneration following pontine hemorrhage.

Index terms
Intracranial Hemorrhages
Wallerian Degeneration
Olivary Nucleus

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INTRODUCTION

There are two distinct types of axon degeneration that occur after neuronal injury: Wallerian degeneration (WD) and transneuronal degeneration (1). WD has been referred to as distal axonal degeneration following axon transection or neuronal damage. Transneuronal degeneration refers to trans-synaptic degeneration of axons and their accompanying myelin sheaths, and is associated with a lesion in either the afferent or efferent connection of the involved neuron (2). Both forms of degeneration are pathological concepts and terms in that they represent cellular level changes. However, when these degenerative changes occur along known neural pathways, they can be detected by magnetic resonance imaging (MRI). There are several reports depicting WD on MRI (3-6). However, reported cases of hypertrophic olivary degeneration (HOD), a unique type of trans-synaptic degeneration caused by lesions in the dentato-rubro-olivary pathway accompanied by WD are exceedingly rare. Here, we present a case of WD of pontocerebellar fibers accompanied by HOD after pontine hemorrhage.

CASE REPORT

A 49-year-old man was admitted to our emergency room with sudden mental deterioration. He had a history of hypertension. On neurologic examination, he demonstrated dysarthria, right hemiparesis, and right eyeball deviation. His mental status was drowsy at initial presentation, but subsequently worsened to stupor. Brain computed tomography (CT) imaging was performed and showed a large pontine hemorrhage (Fig. 1A).

He was admitted to the intensive care unit and received conservative treatment. The hemorrhage in the pons was reduced in size on follow-up CT, and his mental status also improved to alert. After approximately nine weeks of conservative care and rehabilitation, he was discharged with mild dysarthria, diplopia, dizziness, and right-sided weakness.

Seven months after pontine hemorrhage, he was admitted to our hospital because of the worsening of dizziness over the

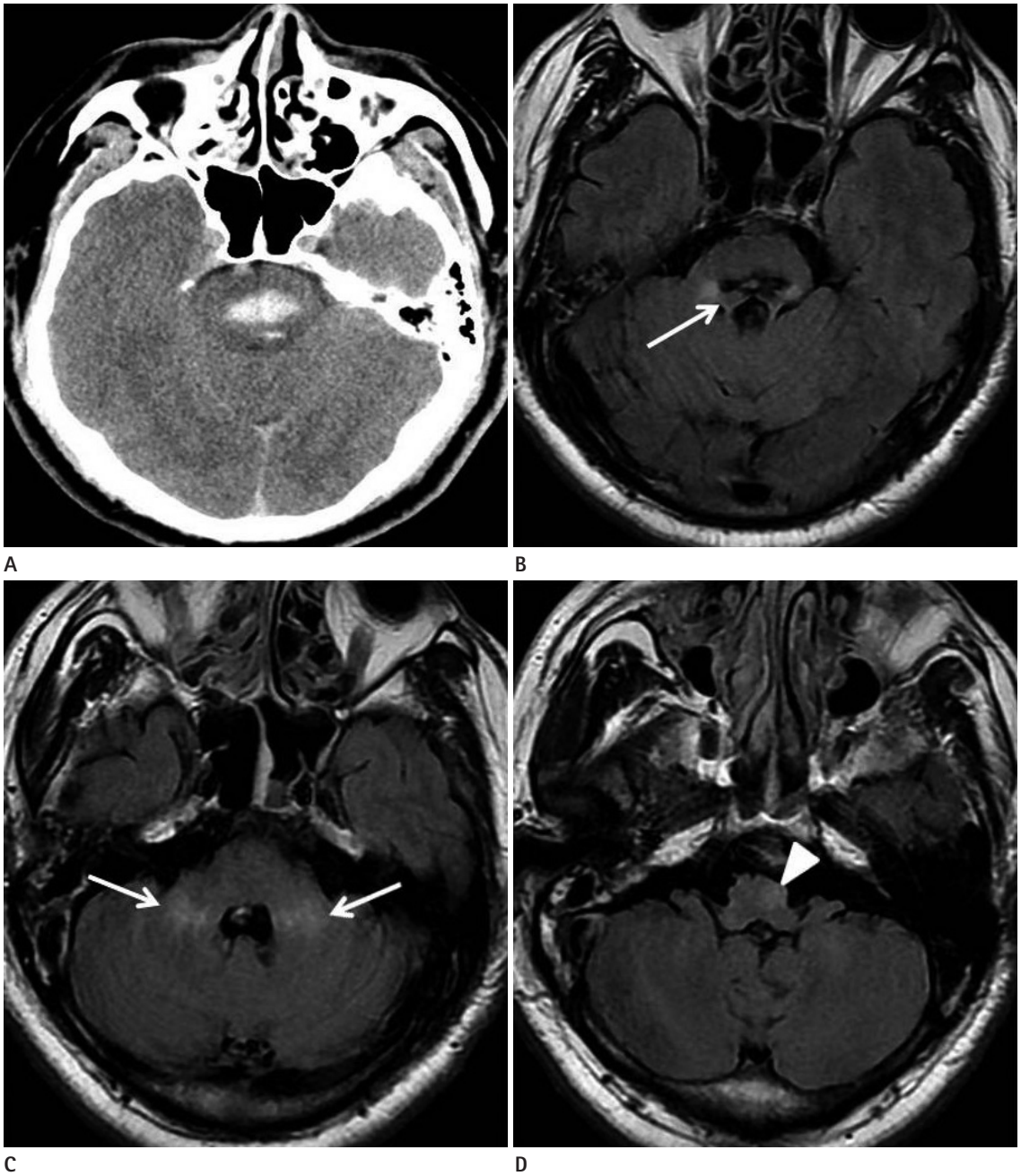

Fig. 1. A 49-year-old man with Wallerian degeneration of the pontocerebellar tract and hypertrophic olivary degeneration after pontine hemorrhage.
A. Axial unenhanced brain computed tomography image obtained at the time of onset shows a large pontine hemorrhage.
B. FLAIR image obtained 7 months after onset shows sequelae of hemorrhage in the bilateral aspects of the central pons, which left the right central tegmental tract (arrow) relatively spared.
C, D. Axial FLAIR images show a faint high signal intensity lesion bilaterally in the middle cerebellar peduncles (arrows; **C**). In addition, a hyperintense lesion with mild enlargement is seen in the left inferior olivary nucleus (arrowhead; **D**).
FLAIR = fluid attenuated inversion recovery

month preceding the second admission. Examination revealed postural vertigo and gait disturbance. Other symptoms such as hearing loss, ear fullness, and tinnitus were absent. Electrocardiography and carotid ultrasonography studies were normal. MRI showed a dark signal intensity lesion in the pons on fluid attenuated inversion recovery (FLAIR) imaging, compatible with the previous finding of pontine hemorrhage (Fig. 1B). In addition, bilateral hyperintense lesions were seen along the middle cerebellar peduncle (MCP) (Fig. 1C). The left inferior olivary nucleus was mildly enlarged, with high signal intensity (Fig. 1D).

DISCUSSION

Our case demonstrated two characteristic imaging findings on MRI performed 7 months after initial injury. First, bilateral and symmetrical hyperintensities were visible along the MCPs on FLAIR images. Such lesions of the MCPs have been attributed to bilateral WD of the pontocerebellar tracts (4). WD is most frequently observed in the corticospinal tract following infarction of the motor cortex or internal capsule. Descriptions of WD of the cerebellar peduncles are rare, but when it occurs, WD usually involves the MCPs because they are the largest and the main path for the pontocerebellar tracts (3). Histologic and metabolic features can be correlated with specific findings on MRI (7).

In our case, bilateral MCPs showed hyperintensity on FLAIR images at 7 months of follow-up, which correlated with stage 3 WD. This stage is characterized by myelin lipid breakdown, gliosis, and changes in water content and structure, due to which the tissue becomes hydrophic and thus hyperintense on T2-weighted and FLAIR images. Symmetric lesions of the MCPs can also be seen in other conditions, such as Wilson's disease, hepatic encephalopathy, extrapontine myelinolysis, acute disseminated encephalomyelitis, and neurodegenerative disorders (8). The diagnosis of these diseases can be made relatively easily on the basis of clinical data. As in our case, an associated pontine hemorrhage is the key to a definitive diagnosis of WD of the pontocerebellar tracts. Moreover, MRI findings of WD after pontine hemorrhage or infarction include faint symmetrical hyperintensities in the central portion of the MCP (8).

A second main finding on follow-up FLAIR imaging in our patient was an enlargement of the left inferior olivary nucleus, with high signal intensity, indicative of HOD. HOD involves the dentato-rubro-olivary tract, also called the Guillain-Mollaret triangle, which is composed of the contralateral dentate nucleus, the ipsilateral red nucleus, and the ipsilateral inferior olivary nucleus (9). Typically, HOD appears as an increase in signal on T2- and proton density-weighted images, and an increase in the size of the olivary nucleus is also observed, as transneuronal degeneration results in hypertrophy of this region (5). Olivary hypertrophy typically develops around six months after the event and resolves after three to four years (4). In our case, lesions were seen on MRI 7 months after the initial attack, which falls within the typical range of disease development.

HOD rarely coexists with WD of pontocerebellar tracts secondary to pontine lesions, because pontine lesions often also involve areas in the Guillain-Mollaret triangle. To our knowledge, this is the second case depicting MR imaging of WD of the pontocerebellar fibers accompanied by HOD after the O'Uchi's report (10). Even though HOD and WD are well known degenerations in the central nervous system, it may be meaningful to report our observations because there may be relatively few reports on combined lesions in one subject. O'Uchi reported 5 cases with WD of the pontocerebellar tracts after pontine hemorrhage. In one of the five cases, follow-up MRI showed high signal intensities in bilateral MCPs and bilateral olivary nuclei following left pontine hemorrhagic infarct. In contrast to the case series reported by O'Uchi., our case demonstrated unilateral HOD following pontine hemorrhage. Although the patient experienced bilateral central pontine hemorrhage, sequelae of the hemorrhage left the right central tegmental tract in the upper pons relatively intact, explaining why HOD was observed only on the left. HOD can be found incidentally on MRI scans because it is not associated with symptoms or signs. In almost half of patients with HOD, however, present with dyskinesia such as palatal myoclonus which is rhythmic involuntary movements of the oropharynx and a dentato-rubral tremor (9). Our patient presented with dizziness and gait disturbance, which may be associated with WD of cerebellar peduncles, but he did not have symptoms relevant to HOD.

In conclusion, the case presented herein demonstrated WD of bilateral MCPs combined with unilateral HOD following pontine hemorrhage. In a patient with the appropriate clinical

presentation, the observation of a pontine lesion with signal intensity abnormalities of the MCPs and olivary nucleus makes it possible to recognize WD of the pontocerebellar fibers and transneuronal degeneration in the dentato-rubro-olivary pathway, which should not be mistaken for a primary independent lesion.

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뇌교출혈 후 이차적으로 발생한 양측 중간소뇌다리의 월러변성과 편측 비대성올리브핵변성: 증례 보고

윤재홍 · 김상현* · 최순섭 · 강명진 · 조 은

뉴런의 손상 후에는 월러변성과 신경원통과 변성이라는 두 가지 유형의 축삭변성이 발생한다. 이차 변성의 가장 흔한 원인은 뇌경색이지만 출혈, 외상, 괴사, 국소적 탈수초 등에 의해서 이차 변성이 나타날 수 있다. 우리는 뇌교출혈 후에 양측 소뇌다리의 월러변성과 동반된 편측 비후성올리브핵변성이 발생한 드문 증례를 보고하고자 한다.

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