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Case Report

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Isolated Bilateral Cerebral Peduncular Infarction Manifesting Pseudobulbar Palsy and Quadriparesis: a Case Report

OPEN ACCESS

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HIGHLIGHTS

- Isolated bilateral cerebral peduncular infarction (BCPI) is extremely rare disease.
- Clinical manifestations are dependent on the extent of infarction within the peduncle.
- Isolated BCPI may present a severe pseudobulbar palsy with preserved limb function.

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Conflict of Interest
The authors have no potential conflicts of interest to disclose.

ABSTRACT

Bilateral cerebral peduncular infarction (BCPI) is a very rare disorder among stroke patients. The main clinical manifestations in the previously reported BCPI case reports was associated with locked-in syndrome or persistent vegetative state. Here, we present a 51-year-old woman who had pseudobulbar palsy and quadriplegia. Magnetic resonance imaging showed an acute infarction in the middle areas of the cerebral peduncle with a unique "Mickey Mouse ears" sign. Diffusion tensor imaging and tractography showed relatively preserved corticospinal tracts, but the corticobulbar tracts were not detected. Magnetic resonance angiography showed posterior cerebral artery and vertebrobasilar artery occlusion. Cerebral perfusion insufficiency due to stenosis or occlusion of the vertebrobasilar artery and its branches may lead to BCPI. The prognosis and clinical manifestations of BCPI are related to the extent of the infarction in the involved cerebral peduncle and whether other territories are involved. Isolated BCPI may present a severe pseudobulbar palsy with relatively preserved limb function depending on the involvement pattern.

Keywords: Cerebral Peduncle; Brain Stem Infarctions; Vertebrobasilar Insufficiency; Pseudobulbar Palsy

INTRODUCTION

Bilateral cerebral peduncular infarction (BCPI) is a very rare disorder among stroke patients. Pure midbrain infarction has been reported to occur in 0.7% to 2.3% of patients with acute ischemic stroke, and the prevalence of isolated bilateral midbrain infarction accounted for 0.12% of total ischemic infarction [1]. According to previous study, BCPI has been reported to occur in 0.26% of all ischemic infarctions [2]. Isolated BCPI cases identified by magnetic resonance imaging (MRI) are extremely rare, and to the best of our knowledge, only a few cases have been reported previously [2-4]. It can be identified in MRI as a unique "Mickey Mouse ears" sign [5]. In many cases, infarctions have also been found in areas other than the cerebral peduncle including the thalamus, pons, and other vertebrobasilar artery territories [2,6]. According to previous case reports, a locked-in syndrome (LIS) or a persistent vegetative state were the main clinical manifestations of BCPI [6,7]. Here, we describe a case report of an isolated BCPI patient manifesting pseudobulbar palsy and mild quadriplegia.

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CASE REPORT

A 51-year-old female patient was admitted to the intensive care unit in April 2020 with dizziness, dysarthria, pseudobulbar palsy, and motor weakness but without diplopia or limitation of extraocular movement. Her past history included 10 years of hypertension and 5 years of diabetes mellitus (DM). Upon admission, the patient had dysarthria, dysphagia, quadriparesis, and ataxia in all extremities, however this was found to be more pronounced on the right side on neurological examination. The patient showed pathological reflexes including the Babinski sign, ankle clonus and Hoffman's sign. There were no limitations of extraocular movements (vertical or horizontal), nystagmus, ptosis, or visual disturbance. She also showed intact pupillary reflexes.

Due to pseudobulbar palsy, the patient had pathological laughing and crying on examination. She underwent brain MRI. There were abnormal findings in the bilateral cerebral peduncle with hyperintense signals on the diffusion-weighted imaging (DWI) and hypointense signals on the apparent diffusion coefficient (ADC; Fig. 1). Occlusion of the bilateral posterior cerebral artery (PCA) and vertebrobasilar artery and focal stenosis of the right M1 artery were found in the magnetic resonance angiography (MRA; Fig. 2). The dual antiplatelets—aspirin and clopidogrel—were administered for the improvement of circulation. However, the patient's dysarthria worsened on the 10th day of hospitalization. The brain MRI showed an increased extent of diffusion-restricted lesions at the bilateral midbrain on DWI/ADC (Fig. 3). MRI findings revealed no involvement of the thalamus, cerebellum, or other PCA territories. After the platelet drug response assay, the clopidogrel was changed to cilostazol. Diffusion tensor imaging (DTI) and tractography were performed in this patient to assess the affected corticospinal and corticobulbar tracts. The results showed relatively preserved corticospinal tracts, but the corticobulbar tracts were not delineated (Fig. 4). The DWI were obtained on the Philips IngeniaElitionX scanner using a diffusion sequence (DTI 45d 2 mm; the echo time = 72 ms and the repetition time = 8,955.32 ms).

A DTI diffusion scheme was used, and a total of 33 diffusion sampling directions were acquired. The in-plane resolution was 1.91071 mm, the b-value was 600 s/mm^2, and the

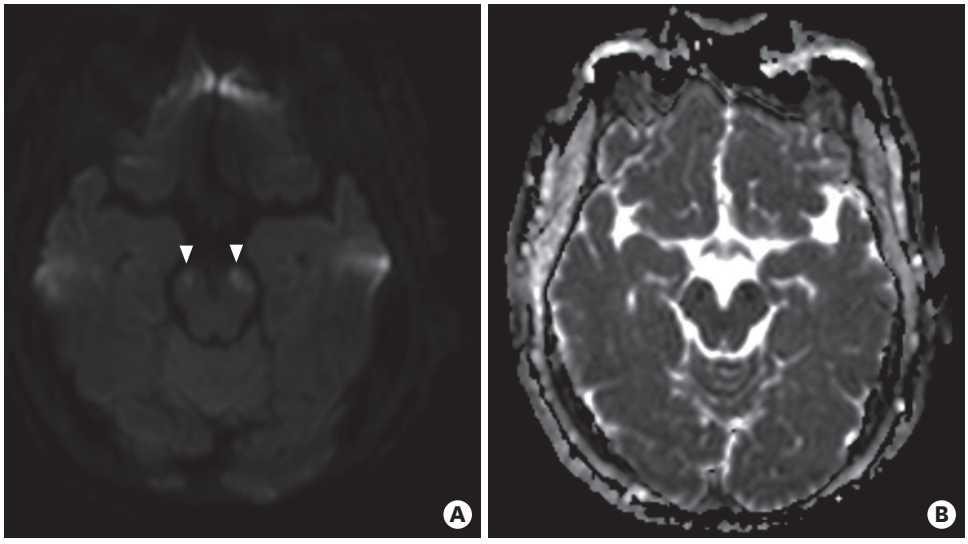

Fig. 1. (A) High signal intensity in bilateral cerebral peduncles (arrowhead) on the diffusion-weighted image. (B) Apparent diffusion coefficient image showing corresponding hypointensity in bilateral cerebral peduncles (April 2020).

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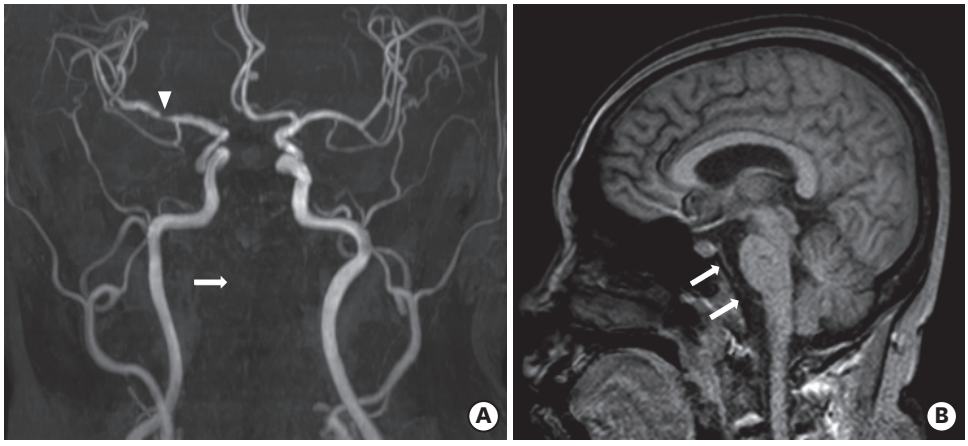

Fig. 2. (A) Magnetic resonance angiography showing vertebrobasilar artery and bilateral posterior cerebral artery occlusion (arrow) and right M1 artery focal stenosis (arrowhead). (B) Parasagittal T1-weighted magnetic resonance imaging demonstrates a linear structure (arrows) in the pontine cistern, suggesting basilar artery thrombosis (April 2020).

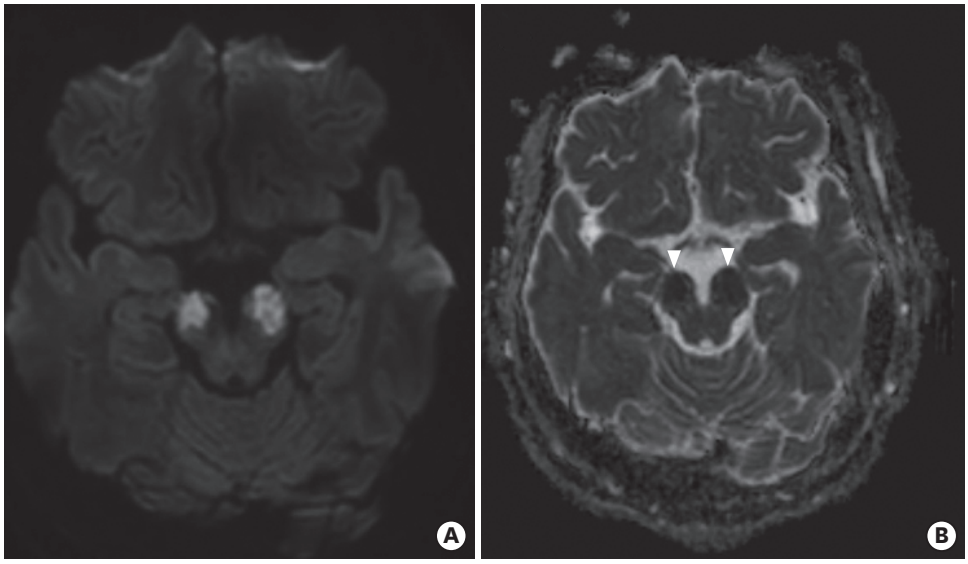

Fig. 3. (A) Follow-up brain magnetic resonance imaging showing an increased extent of diffusion-restricted lesions at bilateral cerebral peduncles on diffusion-weighted image. (B) Apparent diffusion coefficient image showing corresponding increased extent of lesions in bilateral cerebral peduncles (arrowhead) (May 2020).

slice thickness was 2 mm. The b-table was checked by an automatic quality control routine to ensure its accuracy. We reconstructed the diffusion data using q-space diffeomorphic reconstruction [8] to obtain the spin distribution function [9] in the Montreal Neurological Institute space. A diffusion sampling length ratio was 1.25, and 2-mm isotropic resolution was used. The restricted diffusion was quantified using restricted diffusion imaging [10].

At 3 weeks after onset, the patient started comprehensive rehabilitation therapy. A neurological examination revealed severe dysarthria and dysphasia. The patient was fed through a nasogastric tube due to dysphagia. In the Korean version of the Mini-Mental State Examination, the patient's score was 28 points. On the manual muscle test (MMT), the patient showed MMT grade 2 on the right side and MMT grade 3 on the left side. On the Videofluoroscopic Swallowing Study (VFSS), decreased tongue movement, delayed

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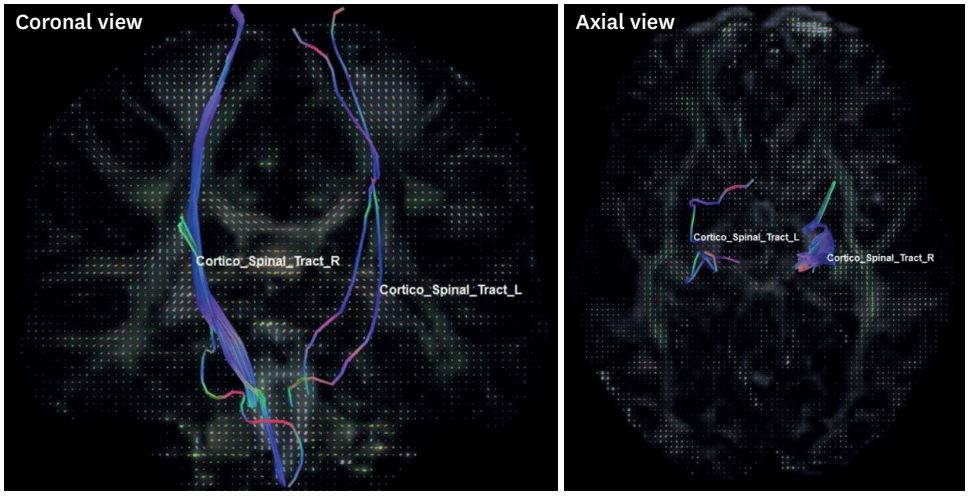

Fig. 4. DTI and tractography showed partially preserved corticospinal tracts. The corticobulbar tract was not detected on DTI and tractography (June 2020).
DTI, diffusion tensor imaging.

swallowing reflex, incomplete pharyngeal peristalsis, and incomplete laryngeal elevation were detected. The VFSS findings showed a penetration aspiration scale (PAS) score of 4 for a small amount of liquid (5 mL) and a PAS score of 8 for a large amount of liquid (15 mL). The patient underwent intensive rehabilitation treatment for 8 weeks and showed improvement in her sitting and standing balance, hand function, and ambulatory function, with the Functional Ambulation Classification score improving from 1 to 3 compared with her condition upon admission. The MMT grade improved one grade on both sides. She also showed improvement in activities of daily living including personal hygiene, dressing, chair/bed transfer and ambulation and her Modified Barthel Index score improved from 14 to 33. After VFSS, the patient started oral feeding. The patient's diet was gradually built up from dysphagia diet to general diet with food thickener. The patient continuously received dysphagia rehabilitation including neuromuscular electrical stimulation. The follow-up VFSS showed persistent delayed swallowing reflex, incomplete pharyngeal peristalsis, incomplete laryngeal elevation, and a PAS score of 4 for a small amount of liquid and a PAS score of 7 for a large amount of liquid. The patient continued using food thickener.

The patient provided signed informed consent.

DISCUSSION

To the best of our knowledge, BCPIs identified by MRI were first reported by Park et al in 1997 [6], and their unique imaging findings were first described as the “Mickey Mouse ears” sign by Asakawa et al. [5]. According to the previous reports on BCPI, it is associated with several clinical manifestations, mainly LIS or disturbance of consciousness. In general, LIS is responsible for lesions of the bilateral ventral pontine, and LIS due to pure midbrain lesions is very rare. Previously reported cases of LIS due to midbrain lesions involved medial and central portions of the peduncle, accompanied by other infarction sites including the thalamus, pons, and other vertebrobasilar artery territories [2,6]. However, in the present case, the patient mainly showed severe pseudobulbar palsy, mild quadriparesis, ataxia, intact pupil light reflexes, and full eye movement. The mental and consciousness state was alert.

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MRI findings showed isolated BCPIs, and the lesions were limited to almost the middle area of the peduncle and relatively sparing of the lateral and medial portions, which included the efferent nerve fibers responsible for the voluntary eye movement. DTI tractography imaging showed relatively preserved corticospinal tracts, but the corticobulbar tracts were not detected by tractography (Fig. 4). These findings were correlated with the patient's symptoms of pseudobulbar palsy. Considering this, we could conclude that the clinical manifestations of BCPI are related to the extent of the infarction within the cerebral peduncle as well as the area of the involvement in other vertebrobasilar artery territories. Isolated BCPI may present a severe pseudobulbar palsy with relatively preserved limb function depending on the involvement pattern.

The midbrain vascular system is mainly supplied by the posterior circulation containing the vertebrobasilar artery, posterior cerebral arteries and superior cerebellar arteries [11]. According to the midbrain vascular system, the blood supply of the cerebral peduncle primarily comes from small branches of the posterior choroidal artery, superior cerebellar artery, and posterior communicating artery, and all branches originate from the vertebrobasilar artery [3,11]. The etiology and risk factors of BCPI are similar to those of cerebral infarction of posterior circulation [11,12]. Of the 23 patients with mesencephalon infarct, the etiological factor in 11 (48%) patients was a large artery atherothrombosis (LAA), while a cardioembolic etiology was found in 6 (26%) and only 2 (8.5%) patients were due to small vessel occlusion [12]. Chen et al. [2] reported 14 patients of BCPI. Of these, 12 (85.7%) involved vertebrobasilar artery occlusion or stenosis, 11 (78.6%) were caused by LAA, 1 was found to have artery-to-artery embolism, and 2 were diagnosed with cardiac embolism [2]. Concerning vascular risk factors, 13 (92.9%) patients had hypertension and 8 (57.1%) had DM [2]. Our patient had a 10-year history of hypertension and 5-year history of DM. Neither the basilar artery nor PCA was seen on the MRA, but right M1 stenosis was observed (Fig. 2A). The linear structure in the pontine cistern in the parasagittal view on MRA suggested basilar artery thrombosis (Fig. 2B). The patient had no evidence of embolus.

The main mechanism of BCPI caused by LAA is cerebral perfusion insufficiency [11,12]. We could conclude that the main mechanism of isolated BCPI in our patient may have been related to hypoperfusion. However, despite vertebrobasilar artery occlusion and stenosis, there was no involvement of the thalamus, cerebellum, or occipital lobe. We hypothesized that the terminal small branches supplying the cerebral peduncle are hemodynamically more easily influenced than relatively large branches supplying the other territories of the posterior circulations are.

Although the infarct area is small in BCPI, the anatomical complexity of multiple nerve fiber pathways can lead to a poor prognosis. According to previous study, many cases of acute BCPI have shown progressive worsening. The most common symptoms were nausea, vomiting, dizziness, and paralysis, followed by gradually developing LIS, a persistent vegetative state, or even death [11]. In such cases with poor prognosis, infarctions have also been found in areas other than the cerebral peduncle including the thalamus, pons, and other vertebrobasilar artery territories. Our case patient had dysarthria progression after the stroke onset, and we confirmed an increased infarct size in cerebral peduncle, but no other sites involvement on MRI. The DWI sequence showing isolated bilateral cerebral peduncle hyperintensity signals, described as a Mickey Mouse ears sign, is characteristic of the diagnosis [5]. Isolated BCPI is expected to have relatively preserved motor function except pseudobulbar palsy depending on the involvement pattern. The patient received prompt standardized treatment for cerebral

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infarction and early rehabilitation treatment within 3 weeks after the onset. Although she showed persistent dysphagia and dysarthria, functional recovery was achieved through continuous rehabilitation treatment for 2 months.

In conclusion, cerebral perfusion insufficiency due to occlusion or stenosis of the vertebrobasilar artery and its branches may lead to BCPI. The specific imaging findings of the "Mickey Mouse ears sign" are indicative of this. The clinical manifestations and prognosis of BCPI are related to the extent of the infarction in the involved cerebral peduncle and whether other sites, such as thalamus, pons, or other vertebrobasilar artery territories, are involved. Isolated BCPI may present a severe pseudobulbar palsy with relatively preserved limb function depending on the involvement pattern.

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