Case Report

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Trigeminal Neuralgia with Persistent Trigeminal Artery Variant and Schwannomatosis of the Abducens and Lower Cranial Nerves: A Case Report

지속성 삼차동맥 변이로 인한 삼차신경통과 외전신경 및 하부 뇌신경의 신경초종증: 증례 보고

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Multiple cranial neuropathies are complex neurological disorders that present significant treatment challenges due to their intricate and multifaceted underlying causes. Persistent trigeminal artery variant (PTAV) is an unusual anastomosis that connects the internal carotid artery to the cerebellar artery, bypassing the basilar artery. The incidence of PTAVs is approximately 0.18% and neurovascular compression is rarely reported. Patients with schwannomatosis typically have multiple schwannomas without vestibular nerve involvement, which may develop in the spinal nerve roots and, less commonly, in the cranial nerves. We report the case of a 53-year-old female with trigeminal neuralgia associated with PTAV and schwannomatosis in the abducens and lower cranial nerves as dual etiologies for multiple cranial neuropathies.

Index terms Multiple Cranial Neuropathies; Persistent Trigeminal Artery Variant; Abducens Nerve; Lower Cranial Nerves; Schwannoma; Schwannomatosis

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INTRODUCTION

Multiple cranial neuropathies involve the simultaneous impairment of two or more cranial nerves and are relatively uncommon (1). Although the sites and origins of multiple cranial neuropathies vary widely, the most common underlying causes are tumors and vascular diseases (1). While most studies have investigated multiple cranial neuropathies from the perspective of the simultaneous involvement of multiple cranial nerves caused by a single underlying condition or disease, there have been few reported cases of multiple cranial neuropathies resulting from independent causes (2). The persistent trigeminal artery (PTA) and PTA variant (PTAV) are rare remnants of the fetal carotid-basilar anastomosis, with the PTAV uniquely connecting the internal carotid artery to the cerebellar artery and bypassing the basilar artery (BA) (3). The incidence of PTAV is approximately 0.18%, and there have been few case reports of neurovascular compression syndrome associated with PTAV terminating at the anterior inferior cerebellar artery (AICA) (3-5). Patients with schwannomatosis typically have multiple schwannomas without vestibular nerve involvement, which may develop in the spinal nerve roots and, less commonly, in the cranial nerves. This paper presents a case of multiple cranial neuropathies resulting from two distinct causes: neurovascular compression between the PTAV and trigeminal nerve, and schwannomatosis of the abducens and lower cranial nerves, which were treated with a single surgical approach.

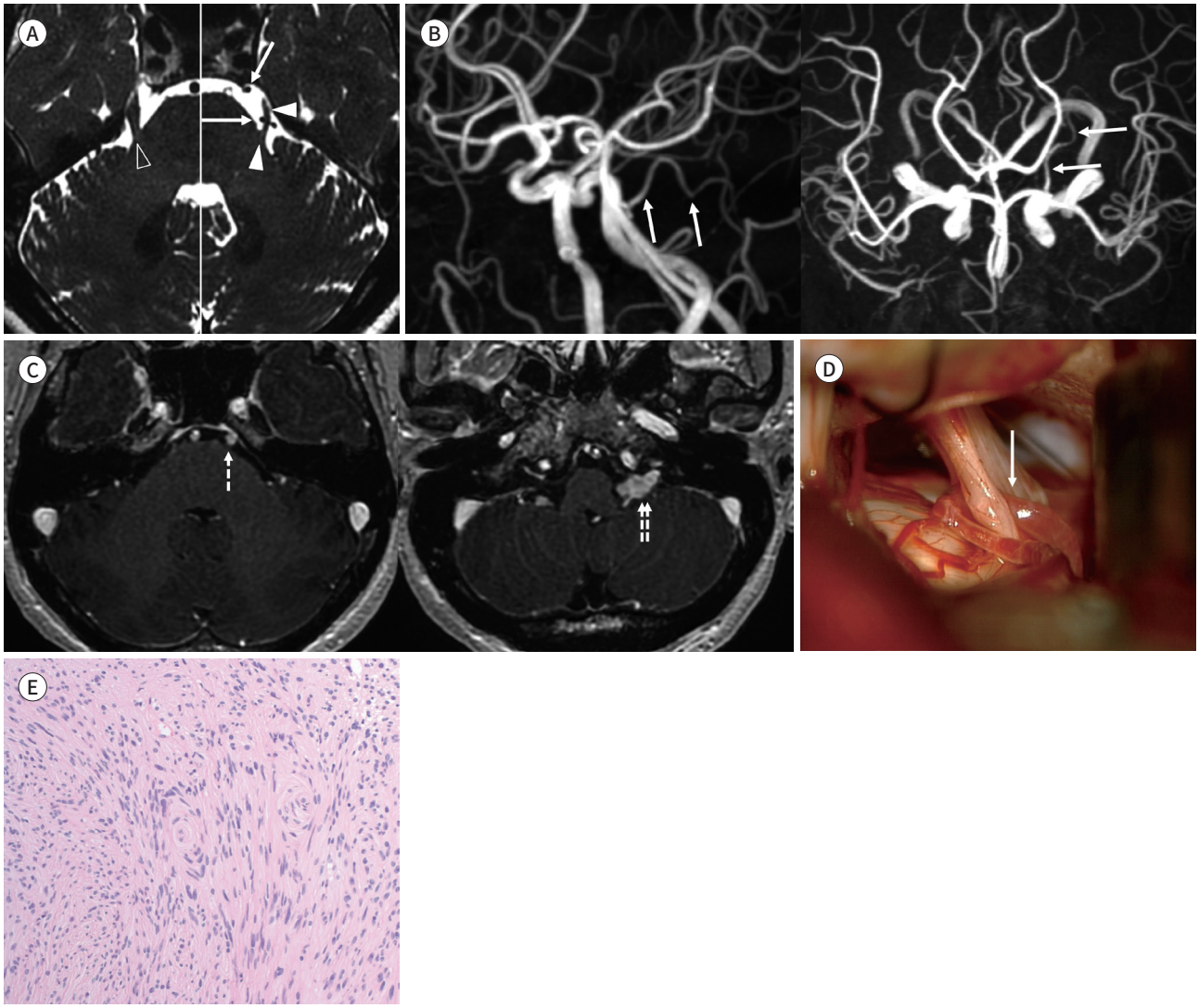
CASE REPORT

A 53-year-old female presented with a four-year history of recurrent sharp, stabbing pain localized in the left upper cheek, upper lip, and upper teeth, corresponding to the left trigeminal nerve-maxillary branch area. The patient underwent MR examination. Axial 3D fast imaging employing steady-state acquisition (Fig. 1A) and MR angiography (Fig. 1B) images revealed a vascular loop of the PTAV (Fig. 1A, B; arrows) passing through the cisternal segment of the left trigeminal nerve (Fig. 1A; arrowheads), causing displacement and thinning of the nerve compared with the normal-appearing right trigeminal nerve (Fig. 1A; empty arrowhead). This also demonstrated that the PTAV originated from the left cavernous internal cerebral artery (ICA), entered the posterior fossa along the medial wall without forming an isolated dural opening, and terminated at the AICA (Fig. 1B; arrows) without joining the BA. Contrast-enhanced T1-weighted images (Fig. 1C) revealed schwannomas in the left abducens nerve (Fig. 1C; dashed arrow) and lower cranial nerves (glossopharyngeal, vagus, or accessory nerves) (Fig. 1C; double dashed arrow). A left retrosigmoid approach was performed for microvascular decompression and tumor removal. During surgery, the PTAV (Fig. 1D; arrow) was observed passing through the left trigeminal nerve in the surgical field, and two masses were identified in the left prepontine and cerebellomedullary cisterns. Histopathological analysis of the specimens (Fig. 1E) revealed a pattern consistent with spindle cells arranged in fascicles, along with strong positivity for the S-100 protein, which is a characteristic feature of schwannomas. After surgery, the patient's neurological symptoms, including facial pain, dysphagia, and diplopia were alleviated. Postoperative MRI demonstrated successful decompression of the trigeminal nerve from the PTAV and gross total removal

of the intracranial tumors.

This study was approved by the Institutional Review Board (IRB No. 2022-10-060) and the requirement for informed consent was waived.

Fig. 1. A 53-year-old female with neurovascular compression between the trigeminal nerve and a PTAV and schwannomatosis of the abducens and lower cranial nerves.
A. Axial 3D fast imaging employing steady-state acquisition sequence of the normal appearing right trigeminal nerve (arrowhead) and a vascular loop of the PTAV (arrows) passing through the cisternal segment of the left trigeminal nerve (arrowheads), causing deformity and thinning of the nerve compared to the contralateral side.
B. MR angiography shows the PTAV originating from the left internal cerebral artery, entering the posterior fossa along the medial wall without forming an isolated dural opening, and terminating at the anterior inferior cerebellar artery (arrows) without joining the basilar artery.
C. Contrast-enhanced T1-weighted images show a schwannoma in the left abducens nerve (dashed arrow) and an additional schwannoma involving the lower cranial nerves (glossopharyngeal, vagus, or accessory nerves) (double-dashed arrow).
D. Intraoperative view shows neurovascular compression of the cisternal segment of the left trigeminal nerve by the PTAV (arrow).
E. Microscopic examination of the lower cranial nerve mass (hematoxylin and eosin stain, ×100) reveals spindle cells arranged in fascicles, demonstrating nuclear palisading, a characteristic feature of schwannomas.
PTAV = persistent trigeminal artery variant



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DISCUSSION

Herein, we report a case of multiple cranial neuropathies with dual etiologies of neurovascular compression associated with PTAV and schwannomas in the abducens and lower cranial nerves. This case emphasized the importance of considering the multiple concurrent causes of complex cranial neuropathies.

Multiple cranial neuropathies involve the simultaneous impairment of two or more cranial nerves and are relatively uncommon (1). The major causes of multiple cranial neuropathies are tumors (30%), vascular disease (12%), trauma (12%), and infection (10%) (1). Most studies have investigated multiple cranial neuropathies from the perspective of simultaneous involvement of multiple cranial nerves caused by a single underlying condition or disease (2).

PTA and PTAV are rare remnants of fetal carotid-basilar anastomosis, with PTAV uniquely connecting the internal carotid artery to the cerebellar artery, bypassing the BA (3). The prevalence of PTAV is approximately 0.18%, and its most common course is through the AICA (3, 5). The prevalence of PTA among patients with trigeminal neuralgia is 2.2% (6), which is higher than that in the general population. Moreover, PTAV is more likely to cause trigeminal neuralgia than PTA (6). Morita et al. (7) explained why a PTAV is more likely to cause trigeminal neuralgia than a PTA by stating that the PTAV courses dorsally along the cerebellar surface and runs near the root entry zone of the trigeminal nerve without joining the BA. In our patient, the PTAV coursed dorsolaterally from the cavernous ICA, causing compression and deformity in the cisternal segment of the trigeminal nerve, which might have contributed to trigeminal neuralgia. Treatment options vary from conservative management to surgical intervention, with microvascular decompression surgery offering immediate pain relief in 90%–95% of patients (4, 5).

Schwannomas are tumors of the peripheral nervous system that originate from Schwann cells (8). Patients with schwannomatosis typically have multiple schwannomas without involvement of the vestibular nerves, which may develop in the spinal nerve roots and, less commonly, in the cranial nerves (8). It is important to note that the patient did not undergo a neurofibromastosis type 2 (*NF2*) gene test; however, this patient may have been diagnosed with schwannomatosis. The essential diagnostic criteria for schwannomatosis are two or more schwannomas (non-intradermal and pathologically confirmed) and no bilateral vestibular schwannomas on high-quality MRI (8). A diagnosis of schwannomatosis can be appropriately suggested by a radiologist in patients with multiple nonvestibular schwannomas (9). Although somatic mutations in *NF2* are often detected in tumor tissues of patients with schwannomatosis, research has shown that *NF2* is not responsible for the germline transmission of schwannomatosis (8). According to the literature, both symptomatic lower cranial and abducens nerve schwannomas can be treated surgically via the retrosigmoid approach; in some cases, radiation treatment may be considered (10).

In conclusion, we report the coexistence of neurovascular compression associated with PTAV and schwannomatosis in the abducens and lower cranial nerves as dual etiologies for multiple cranial neuropathies. This case emphasized the importance of considering the multiple concurrent causes of multiple cranial neuropathies.

Author Contributions

Conceptualization, C.H.J.; data curation, K.J.Y.; formal analysis, K.J.Y.; funding acquisition, C.H.J.; investigation, K.S.G., C.K.G., K.J.Y.; methodology, C.H.J.; resources, C.K.G.; supervision, C.H.J., K.S.H.; validation, K.J.Y., K.S.H.; writing—original draft, K.S.G.; and writing—review & editing, C.H.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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지속성 삼차동맥 변이로 인한 삼차신경통과 외전신경 및 하부 뇌신경의 신경초종증: 증례 보고

김성광[1] · 최혜정[1*] · 조경기[2] · 김정윤[1] · 김상흠[1]

다발성 뇌신경 장애는 복잡하고 다면적인 원인으로 인해 상당히 도전적인 신경학적 장애이다. 지속성 삼차동맥 변이는 기저동맥을 우회하여 내경동맥과 소뇌동맥이 연결되는 드문 발생학적 변이로, 발생률은 약 0.18%이며 이 상태에서 신경혈관 압박은 드물게 보고된다. 신경초종증은 전정 신경을 제외한 다른 신경에 다발성 신경초종이 발생하는 것을 의미하며 뇌신경에 발생하는 경우는 매우 드물다. 우리는 53세 여성이 지속성 삼차동맥 변이 관련 삼차신경통과 외전신경 및 하부 뇌신경의 신경초종증이 동시에 존재하는 다발성 뇌신경 장애 증례를 보고하고자 한다.

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