Vernet's Syndrome Associated with Internal Jugular Vein Thrombosis

Myungah Lee, MD, Youngjae Heo, MD, and Taewon Kim, MD, PhD

Our objective is to present a case of Vernet's syndrome (cranial nerve (CN) IX, X, and XI palsy) associated with cerebral venous thrombosis (CVT) in an internal jugular vein. The patient presented with acutely developed dysphagia. The weakness of the left sternocleidomastoid and trapezius muscles was observed. The initial magnetic resonance imaging and computed tomography (CT) with contrast enhancement showed contrast-filling defect in the left internal jugular vein inside the jugular foramen. The magnetic resonance venography with contrast enhancement revealed a partial filling defect in the left sigmoid sinus and total occlusion of the left internal jugular vein. Under the diagnosis of CVT associated with CN IX, X palsy, anticoagulation therapy with low-molecular-weighted heparin was initiated. Despite the continued anticoagulation therapy for 3 months, neither the burden of thrombosis in the left sigmoid sinus and internal jugular vein on neck CT nor dysphagia symptoms improved. Clinicians need to be aware of internal jugular venous thrombosis as one of the differential diagnoses in Vernet's syndrome in patients in a hypercoagulable state. Further reporting of similar cases is needed to confirm the association between CVT and Vernet's syndrome.

Keywords: Cerebral venous thrombosis—Vernet's syndrome—jugular foramen

Introduction

Multiple lower cranial nerves (CN) palsies caused by various pathologic lesions in the jugular foramen are well-known conditions. These conditions are known as specific syndromes according to the cranial nerves involved, such as Vernet's syndrome (CN IX, X, and XI), Collet-Sicard syndrome (CSS; CN IX, X, XI, and XII), Villaret's syndrome (CN IX, X, XI, XII, and sympathetic chain), etc. These syndromes are caused by various etiologies including malignancy, infection, trauma or fracture, as well as vascular events such as dissection, thrombosis, or aneurysm.[1]

The clinical manifestation of cerebral venous thrombosis (CVT) is diverse, and a spectrum of neurological symptoms or signs has been reported. However, jugular foramen syndrome caused by CVT is very rare. Only a few cases of CSS and Vernet's syndrome as a result of CVT have been reported in the current literature.[1-5] We describe a rare case of Vernet's syndrome associated with CVT.

Case Presentation

A 77-year-old man presented with abruptly developed dysphagia. He had been experiencing hypertension, dyslipidemia, and nonvalvular atrial fibrillation for which he had been taking amlodipine, atorvastatin, and rivaroxaban. Four days before developing dysphagia, he had discontinued the rivaroxaban because he was scheduled to undergo a tooth extraction on the following day. Over the 3 days after the dental procedure, the bleeding continued and he developed acute swallowing difficulty. He also experienced a gradually aggravating orthostatic headache thereafter. A neurologic examination revealed the deviation of the pharyngeal wall and uvula to the right side with loss of left-sided gag reflex and decreased movements of the left soft palate. The weakness of the left

From the Department of Neurology, College of Medicine, Incheon St Mary's Hospital, The Catholic University of Korea, Seoul, Korea.

Received July 17, 2018; revision received September 17, 2018; accepted October 5, 2018.

Disclosure: Nothing to report.

Address correspondence to Taewon Kim, MD, PhD, Department of Neurology, Incheon St's Mary's Hospital, Catholic University of Korea, 665 Bupyeong-6-dong, Bupyeong-gu, Incheon 403-720 Korea. E-mail: kimtaewon@catholic.ac.kr.

1052-3057/$ - see front matter

https://doi.org/10.1016/j.jstrokecerebrovasdis.2018.10.008

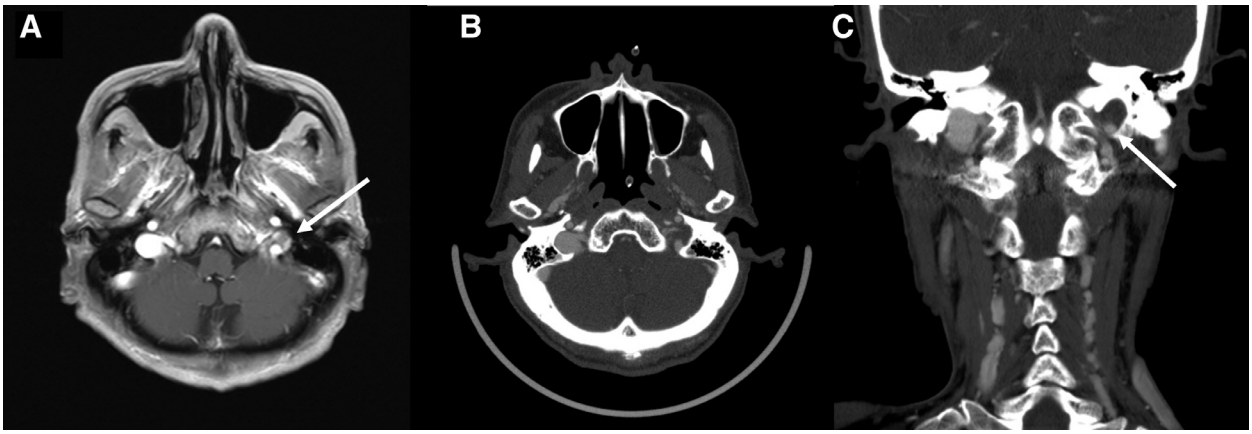

Figure 1. *Initial magnetic resonance imaging (MRI) (A) and computed tomography (CT) with contrast enhancement (B, C). Contrast-filling defect in the left internal jugular vein inside the jugular foramen.*

sternocleidomastoid and trapezius muscles was observed. The dysfunction of the left cranial nerve IX, X, XI was suspected. The initial magnetic resonance imaging and computed tomography (CT) with contrast enhancement showed a contrast-filling defect in the left internal jugular vein inside the jugular foramen (Fig 1A-C). The magnetic resonance venography with contrast enhancement revealed a partial filling defect in the left sigmoid sinus and total occlusion of the left internal jugular vein (Fig 2). The C-reactive protein level was 43.69 mg/L (normal <5 mg/L). Other laboratory results were all normal, including platelet count, prothrombin time, partial thrombin time, antithrombin III Level, fluorescent antinuclear antibody, rheumatoid factor, lupus anticoagulant antibody, anticardiolipin antibody, antineutrophil cytoplasmic antibodies, anti-Sjogren's syndrome-related antigen type A or B, human leukocyte antigens B27 and B51, Complement 3 and C4, C-reactive protein, factors V, VII, and VIII levels, factor V, Leiden gene, protein C activity, protein S activity, peripheral blood morphology, and blood culture. Chest and abdomen CT with iodine contrast enhancement for evaluating latent cancer revealed no abnormal findings.

Under the diagnosis of CVT associated with CN IX, and X palsy, anticoagulation therapy with low-molecular-weighted heparin was initiated. The C-reactive protein level gradually decreased from 43.69 to 2.91 mg/L for 1 month with anticoagulation. However, despite the anticoagulation therapy with low-molecular-weighted heparin followed by nonvitamin K oral anticoagulant with rivaroxaban for 3 months, neither the burden of thrombosis in the left sigmoid sinus and internal jugular vein on neck CT nor the dysphagia symptoms improved.

Discussion

In this study, we reported a patient with Vernet's syndrome associated with CVT in the left sigmoid sinus and internal jugular foramen. The patient's dysphagia symptom developed abruptly after both the discontinuation of anticoagulant agent and a dental procedure with ensuing bleeding complication.

The clinical characteristics of CVT manifest various symptoms and signs including headache, increased intracranial pressure, seizure, venous infarction, or hemorrhage. Only a few cases of CVT in the jugular foramen manifesting cranial nerve palsy have been reported in the literature. While CSS and Vernet's syndrome as a result of CVT have been reported, such cases are very rare.[1-5] While various etiologies have been known to cause CVT, 20%-25% of CVT have no known etiology.[5] In our case, both the abrupt discontinuation of anticoagulants and a dental procedure with sustained bleeding complication can be causal or at least contributing factors to the development of CVT. Involvement of CN IX, X, and XI in our

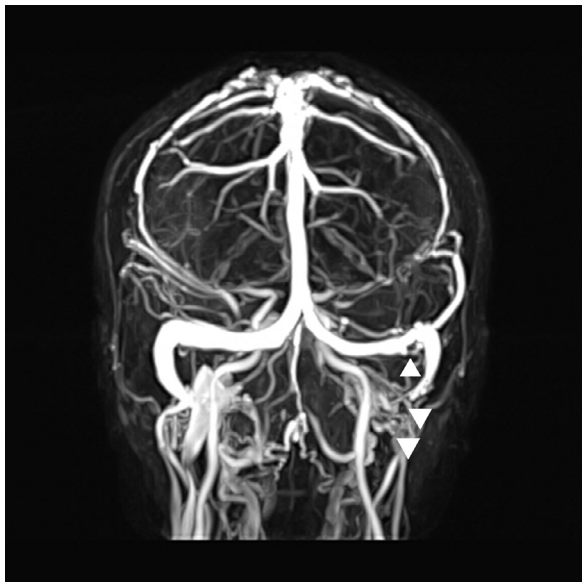

Figure 2. *MR venography with contrast enhancement revealed a partial filling defect in the left sigmoid sinus and total occlusion of the left internal jugular vein (arrow heads).*

case suggested a pathologic lesion located at the jugular foramen. This foramen consists of a smaller anteromedial portion (pars nervosa) and a larger posterolateral portion (pars vascularis) that are separated by a complete or incomplete fibrous or bony septum. The pars nervosa contains CN IX-XI, the inferior petrosal sinus, and the meningeal branch of the ascending pharyngeal artery. The pars vascularis contains the sigmoid-jugular complex.[5] The plausible mechanism of cranial nerve palsies resulting from CVT in the jugular foramen might be explained by the impingement of the cranial nerves between the bony structure of the jugular foramen and the swollen thrombosed vein. Alternatively, local inflammation around the thrombosed vein leading to cranial nerve dysfunction could be a possible explanation.

Despite the anticoagulation therapy for a fairly substantial duration, the burden of thrombosis did not decrease and the patient's dysphagia continued. Considering the prolonged duration of the life span of thrombosis and the failure to remove or resolve the thrombosis swiftly for a number of days with anticoagulation therapy in most CVT cases, this temporal aspect of CVT duration might indicate an important cause of the prolonged cranial nerve dysfunction, implying significant or even irreversible nerve damage. In this regard, we suggest endovascular thrombectomy might be considered an optional management in terms of a rapid removal of thrombus and consequently decompressive relief of nerve impingement damage. However, this approach has poor evidence and it is only a hypothetical suggestion based on our experience of this patient, the dysphagia of whom we had failed to improve, despite performing the proper anticoagulation therapy under the current guideline.

In conclusion, we report a rare case in which CVT in the jugular foramen was associated with Vernet's syndrome presenting with CN IX, X, and XI palsy. CVT could be considered a differential diagnosis in the setting of jugular foramen syndrome and a hypercoagulable state. Further reports of similar cases are needed to confirm the association between CVT and Vernet's syndrome.

References

1. Shima K, Iwasa K, Yoshita M, et al. Vernet's syndrome induced by internal jugular vein thrombosis. J Neurol Neurosurg Psychiatry 2016;87(11):1252-1253.
2. Otto M, Otto V, Gotzinger R, et al. Collet-Sicard's syndrome as a result of jugular vein thrombosis. J Neurol 2001;248:143-144.
3. Moon TS, Sung KB, Shin DJ. A case of Collet-Sicard syndrome resulting from jugular vein thrombosis. J Korean Neurol Assoc 1994;12:348-353.
4. Malin J-P, Haas J, Schliack H, et al. Zur Ätiologie des Foramen jugulare-und des Collet-Sicard-Syndroms. Aktuelle Neurologie 1984;11:50-53.
5. Handley TP, Miah MS, Majumdar S, et al. Collet-Sicard syndrome from thrombosis of the sigmoid-jugular complex: a case report and review of the literature. Int J Otolaryngol 2010;2010:1-5. https://doi.org/10.1155/2010/203587.