

Single Case – General Neurology

A Unique Constellation of Multiple Cranial Neuropathies in a Patient with Preeclampsia

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Keywords

Cranial neuropathy · Hypertension · Hypoglossal nerve · Multiple cranial neuropathies · Preeclampsia · Pregnancy

Abstract

A 36-year-old nullipara at 35 weeks of gestation woke up with slurred speech and dysphagia. The next day, she developed abruption of the placenta, underwent an acute cesarean, and was diagnosed with severe preeclampsia. Neurologic examination revealed flaccid dysarthria, bilateral soft palate palsy, reduced taste of the left posterior tongue, left-sided tongue deviation, and paralysis of the left sternocleidomastoid and trapezius muscles. MRI revealed left-sided tongue edema compatible with acute left hypoglossal nerve denervation and electromyography of the left trapezius and glossal muscles showed profuse denervation potentials. In conclusion, multiple cranial neuropathies may occur in and even be a presenting symptom of preeclampsia. In this study, we report the first case of multiple cranial neuropathies involving cranial nerves IX, X, XI, and XII in a patient with preeclampsia. Possible pathogenic mechanisms of cranial neuropathy in preeclampsia include immune-mediated neuropathy with or without demyelination, microvascular thromboses, and perineural edema.

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Introduction

Preeclampsia is a multisystem hypertensive disorder of pregnancy that typically develops after 20 weeks' gestation and up to 6 weeks postpartum [1]. Idiopathic cranial nerve (CN) palsies, particularly CN VII, occur at a higher frequency during pregnancy [1, 2] and appear to be associated with preeclampsia [1, 3]. This case report is the first to describe multiple cranial neuropathies involving CN IX, X, XI, and XII in a woman with preeclampsia.

Case Report

A 36-year-old woman, gravida 3, para 0, was diagnosed with gestational hypertension at 29 weeks and 2 days of gestation (29+2). Three days prepartum (35+4), the patient developed severe localized pain behind the left mastoid process, and 1 day prepartum (35+6), the patient woke up with slurred speech and dysphagia of solid foods with nasal regurgitation. The following day (36+0), she developed abruptio of the placenta and was admitted for an acute cesarean using spinal anesthesia. Severe preeclampsia was diagnosed with hypertension up to 175/113 mm Hg and a spot urine albumin of 366 mg/dL (ref. <20 mg/dL). Blood tests revealed hemoglobin 7.73 g/dL (ref. 12.0–15.5 g/dL), platelet count $125 \times 10^9/L$ (ref. $145\text{--}390 \times 10^9/L$), creatinine 170 $\mu\text{mol/L}$ (ref. 50–90 $\mu\text{mol/L}$), lactate dehydrogenase 410 U/L (ref. 105–205 U/L), and urate 0.47 mmol/L (ref. 0.16–0.35 mmol/L). Liver function tests were normal.

Neurologic examination postpartum revealed: flaccid nasal dysarthria, bilateral soft palate palsy, reduced taste of the left posterior tongue, left-sided tongue weakness and deviation to the left, and paralysis of the left sternocleidomastoid and trapezius (Fig. 1). Together, these findings indicate a paresis of the left CN IX, XI, and XII and bilateral CN X (Fig. 1, 2).

MRI of the brain and cervical spine with gadolinium showed normal parenchyma without enhancement of the CNs. MRI short tau inversion recovery showed mild left-sided tongue edema, compatible with acute hypoglossal denervation (Fig. 2). CT angiography of the head and neck arteries and cerebral venography were normal. CSF analyses performed 4 days after the onset of symptoms showed normal cell count and protein concentration and were without oligoclonal bands. Serological screening was negative for syphilis, HIV and an exhaustive panel of autoimmune autoantibodies. Electromyography 23 days after symptom onset showed profuse denervation potentials without voluntary activity in the left trapezius and complex repetitive discharges and denervation potentials in the left glossal muscles. The left sternocleidomastoid could not be voluntarily activated or palpated. Despite empirical corticosteroid therapy, the symptoms and repeated electromyography remained largely unchanged six weeks after the onset.

Discussion

Cranial neuropathies have been reported in association with preeclampsia, most commonly involving CN VII [2, 3]. Sporadically, involvement of CN III, V, VI, and XII have also been reported [4, 5]. This report presents the first description of multiple cranial neuropathies involving CN IX, X, XI, and XII in a patient with preeclampsia.

Multiple cranial neuropathies may arise due to a variety of causes including brainstem stroke (ischemic or hemorrhagic), neoplastic disease, neuroinfection, neurosarcoidosis, Guillain-Barré syndrome and variants, vasculitis, diabetes mellitus, idiopathic cranial polyneuropathies, carotid artery dissection, caroticocavernous fistula, cavernous sinus thrombosis, idiopathic cavernous sinusitis, and traumatic head injury [6].



Fig. 1. Neurologic examination demonstrating CNs XI and XII paresis. **a** Left-sided weakness and deviation of the tongue upon protrusion, indicating a left CN XII paresis. **b** No visible contraction of the left sternocleidomastoid muscle upon head rotation to the right, indicating a left CN XI paresis. **c** Scapular winging upon pushing the left arm forward. The lateral scapular winging from a CN XI paresis can be distinguished from a serratus anterior muscle paresis. In the latter, the inferomedial angle rotates medially, rather than laterally, and the winging angle is less pronounced.

The absence of medullary lesions on MRI, as well as signs or symptoms implicating long sensory or motor tracts, indicates that the nerve lesions were located outside the brainstem. Neurophysiologic examinations showed denervation of the trapezius and glossal muscles. CSF analysis did not suggest an infectious or inflammatory etiology; however, lumbar puncture was performed only 4 days after symptom onset and could potentially be higher in inflammatory conditions if repeated later. While the pathogenesis in the present case remains unclear, preeclampsia has previously been suggested to predispose to cranial neuropathy through different pathogenic mechanisms [4]. Amongst these are an activation of the immune response, causing an immune-mediated neuropathy which may be demyelinating, microvascular thromboses of the vasa nervorum and compression due to perineural edema [1, 2, 4, 7].

In the context of previous studies, we hypothesize that cranial neuropathies in association with preeclampsia may be more than a mere coincidence and encourage our colleagues to consider similar presentations as possible warning signs of preeclampsia.

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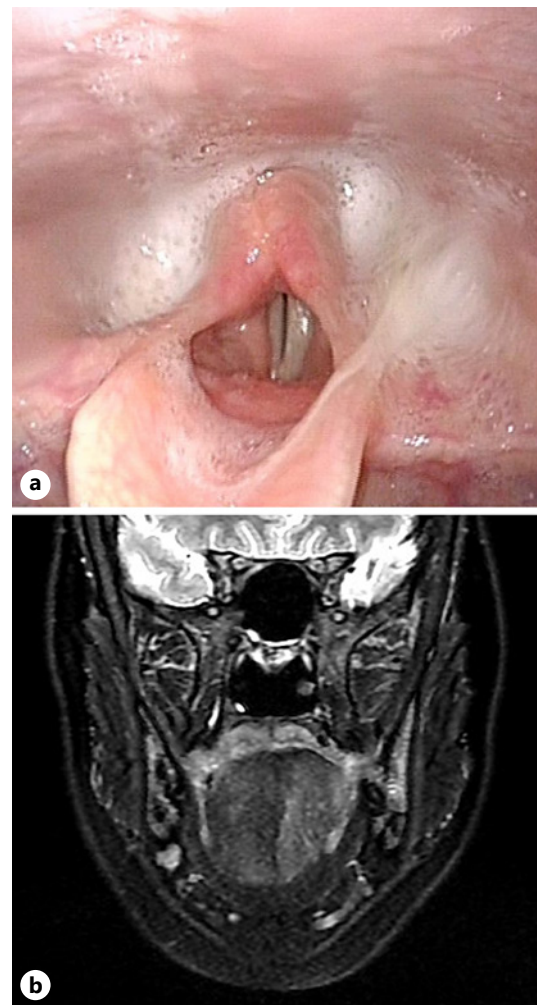


Fig. 2. Fiberscopy and coronal MRI STIR. **a** Fiberscopy revealed stagnated saliva in the pyriform sinuses which could not be cleared by coughing or swallowing. **b** Coronal MRI STIR shows edema of the left side of the tongue. The findings are compatible with acute hypoglossal denervation. STIR, short tau inversion recovery.

Statement of Ethics

Written informed consent was obtained from the patient for the conduction and publication of this case report, including all photographs, scans, and endoscopy. Ethical approval was not required for this study in accordance with national guidelines.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Pardis Zarifkar: (1) obtained patient consent, (2) study conception, (3) data acquisition and interpretation, and (4) formulation of the first draft of the manuscript. Klaus Hansen: (1) study conception, (2) guidance on relevant investigations and treatment, (3) data acquisition and interpretation, and (4) manuscript revision for important intellectual content. Clarissa Crone: (1) data acquisition and interpretation and (2) manuscript revision for important intellectual content. Kirsten Svenstrup: (1) data acquisition and interpretation and (2) manuscript revision for important intellectual content. Vibeke André Larsen: (1) data acquisition and interpretation and (2) manuscript revision for important intellectual content. William Kristian Karlsson: (1) study conception, (2) data acquisition and interpretation, and (3) manuscript revision for important intellectual content.

Data Availability Statement

All data that supports the findings of this study are included in this article. Further inquiries can be directed to the corresponding author.

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