



## Case Report

## Varicella zoster presenting as cranial polyneuropathy☆☆☆

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## ABSTRACT

Cranial polyneuropathy is commonly caused by Lyme disease. We discuss the case of a man who presented with cranial nerve deficits causing dysphagia, dysphonia and facial weakness. This diagnostic dilemma stemmed from a workup that ruled out Lyme and vascular causes leading to an expanded search for infectious explanations, which revealed varicella zoster in the cerebrospinal fluid. On review, this phenomenon is rarely reported, but has been observed with a number of herpes family viruses. In emergency department settings, clinical suspicion should be raised for VZV infection even in the absence of rash in patients that present with multiple cranial nerve palsies.

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## 1. Introduction

CNS varicella zoster reactivation is a relatively rare and scarcely reported phenomenon, with most of the literature on the topic comprised of literature reviews and case series [1–3]. Diagnosis is a challenge, as clinical suspicion is low and the gold standard for diagnosis is a relatively invasive procedure, cerebrospinal fluid analysis (CSF) via lumbar puncture [4,5]. The most common causes of cranial polyneuropathy deficits are vascular, followed by infectious causes, namely *Borrelia burgdorferi*, especially in Lyme endemic areas [6]. Here, we report the case of a patient who presented with acute onset of dysphagia, hoarseness, hypoacusis, and facial muscle weakness attributed to varicella zoster in his CSF.

## 2. Case

A 59-year-old gentleman with a history of end stage renal disease on hemodialysis presented to our emergency department for acute onset of progressive dysphagia, first to solids, then to liquids, as well as hoarse dysphonia for 3 days. Two days prior to admission, he visited his primary care physician and received treatment for presumed viral pharyngitis. On presentation, he was hypertensive, with systolic blood pressure oscillating between 180 and 200 mm Hg, but afebrile, and with lab values that proved unremarkable. His exam was significant for right sided weakness of the upper and lower face, eyelids, palate and tongue,

in addition to right sided hypoacusis and hoarse dysphonia. Olfaction, visual acuity, extraocular movements and sternocleidomastoid strength remained intact. Given his acute onset neurologic findings and uncontrolled hypertension, a cerebrovascular insult was highest on our differential. This hypothesis was evaluated with Computed Tomography, Angiography, and Magnetic Resonance Imaging, all of which were negative for any vascular phenomena. Next on our differential was an infectious agent. The literature is rife with examples of Lyme disease attacking multiple cranial nerves in haphazard patterns, creating peripheral pattern palsies [7]. Given our medical center's location in a region where Lyme is endemic, we considered this possibility strongly. Unfortunately, our patient's serology was negative for Lyme IgG and IgM. There are reports of false negatives in laboratory assays, thus, infection, especially Borreliosis, remained high on our differential [8]. The decision was made to pursue CSF analysis, which revealed glucose of 61 mg/dL, protein of 118 mg/dL, consistent with viral CSF infection, and, most importantly, PCRs negative for Lyme disease, but positive for Varicella Zoster [9]. He was started on Acyclovir, his exam began to improve after the first dose, and he was discharged home to complete the remainder of his treatment as an outpatient.

## 3. Discussion

Previous literature on varicella zoster virus characterizes various neurologic findings upon reactivation including meningitis, encephalitis, myelopathy, vasculopathy, postherpetic neuralgia, cerebellitis, and ocular disorders [10]. Mentions of polyneuropathy remain conspicuously sparse. Ramsay Hunt Syndrome, the most well-described herpes family virus reactivation, produces cranial neuropathy, but is concomitant with a painful vesicular rash [11]. Further, the vast majority of literature surrounding cranial polyneuropathy pertains to Lyme disease

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[12]. Our patient presented with cranial polyneuropathy in the absence of rash, with negative Lyme titers and PCRs in the serum and the CSF. Given his risk factors, including gender, age, renal disease, diabetes, hypertension, and hyperlipidemia, our team pursued vascular explanations for his findings, which turned out to be negative. Classically, we could have started steroid treatment had we initiated therapy in the first 72 h after symptom onset, however, we did not pursue an infectious work-up early enough in the course of his disease to accomplish this [13]. The patient consistently showed negative imaging findings, and a diagnosis proved challenging. Since this condition is rare, there appeared to be little to no guidelines regarding management. In our patient, we initiated Acyclovir as soon as his CSF PCR resulted [14]. Other treatment options commonly referenced in the literature included Valacyclovir, Ganciclovir, and Famciclovir but we chose Acyclovir given its overall support in the literature [15]. There was no evidence supporting the use of corticosteroids late in disease course.

#### 4. Conclusion

In conclusion, varicella zoster polyneuropathy is a rare condition which should be suspected in patients presenting with multiple cranial nerve palsies. CSF analysis via lumbar puncture is imperative for diagnosis. Work-up for Lyme disease, varicella, and other herpes family viruses, including CSF analysis early on in the course of disease, is reasonable in these patients and can portend favorable outcomes.

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