An unusual case of Epstein-Barr virus complicated by multiple cranial nerve neuropathy

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 Epstein-Barr virus (EBV) is a deoxyribonucleic acid (DNA) virus of the herpesvirus family that infects an estimated 90 percent of humans worldwide. It affects many organ systems; however, involvement of the central nervous system is uncommon. An estimated 0.37 percent of patients with infectious mononucleosis develop neurologic complications. There has only been one previous report of synchronous vocal fold and tongue paresis secondary to EBV in the literature, hence the importance of recognizing the disease presentation and possible complications.

We present a case of hypoglossal nerve and left recurrent laryngeal nerve palsy in a 17-year-old man. He initially presented to his local medical officer with odynophagia and “hot potato” voice. He was admitted to the hospital due to inadequate oral intake. He had congenital adrenal hyperplasia for which he was being treated with dexamethasone 0.25 mg nightly, fludrocortisone acetate 0.1 mg twice daily, and hydrocortisone 10 mg each morning. After consultation with pediatric endocrinology, he was commenced on 8 mg of dexamethasone twice daily with the aim of improving the obstructive swelling. His initial investigations revealed a white cell count of 15.7 × 10^9/L, neutrophils 9.1 × 10^9/L, C-reactive protein 2.2 mg/L, and positive immunoglobulin G and immunoglobulin M for EBV.

Although clinically improving, on day 2 after admission he developed hypoglossal nerve palsy, evidenced by tongue deviation to the left, with decreased tongue movement. Given that he was symptomatically improving otherwise, he was discharged on a weaning dose of dexamethasone in consultation with endocrinology. At review one week post admission, he had developed a weak hoarse voice, different from his “hot potato” voice. Nasendoscopy revealed a left vocal cord palsy. The patient underwent magnetic resonance imaging (MRI) and computed tomography (CT), which excluded underlying structural abnormalities. Less than one month after the onset of his symptoms, the patient’s vocal cord palsy had completely resolved. At review seven months post admission, all of his symptoms had completely resolved.

Discussion

Cranial neuropathies are uncommon in the setting of acute EBV infection, and the mechanism by which they develop is poorly understood. It is proposed that the virus causes direct injury to neural tissue, to which the host mounts an immunologic response that subsequently causes denervation of the neural tissue. It could be postulated that a similar etiology occurs with herpes simplex virus etiology and cranial neuropathies (e.g., Ramsey-Hunt syndrome or Bell’s palsy).

Neurologic complications associated with infectious mononucleosis are rare and clinical severity varies widely. Involvement of a large proportion of cranial nerves has been reported in association with EBV, the facial nerve being the most common. Hypoglossal paralysis has been reported both in isolation and in conjunction with other neuropa-thies. Across the spectrum of clinical involvement there appears to be consistency in the timing of resolution of symptoms, typically at two to three weeks post-onset. Full resolution occurs at anywhere from eight weeks to seven months.

In settings where the clinical picture suggests a viral neuritis the possibility of a structural cause needs to be excluded. CT scan from skull base to below the aortic arch has been recommended. The reported role of MRI in the context of EBV infection and associated cranial nerve lesions is twofold, firstly for detection of virus-associated cranial nerve enhancement and secondly for the exclusion of malignancy. It is postulated that cranial nerves affected by EBV infection frequently enhance with contrast medium due to hypervascularity of the nerve and/or disruption of the blood-brain barrier.

The management of EBV-associated neurologic complications can be considered in the context of nonmedical, medical, and surgical management. Nonmedical management involves general education, particularly on the risk of splenic rupture. Medical management of EBV without neu-
rologic complications is supportive: rest and rehydration. Variation exists in the reported administration and efficacy of steroids in this setting, from improvement of symptoms within one month\textsuperscript{4} to partial resolution at 11 weeks.\textsuperscript{1} Other published reports support the notion that no specific therapeutic intervention will influence the clinical course of the disease.\textsuperscript{2,3}

Cranial nerve paralysis associated with EBV infection is typically managed by an otolaryngologist; however, surgical intervention is rare. Careful follow-up by an otolaryngologist is highly recommended,\textsuperscript{2} although resolution over time is expected.

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References
