LETTER TO THE EDITOR

To the Editor

Facial Nerve Palsy from Cerebral Venous Sinus Thrombosis: An Effect of Increased ICP

A 20-year-old right hand–dominant previously healthy female on an oral contraceptive developed new-onset persistent, holocephalic, pressure-like headache. This was associated with nausea, vomiting, and pulsatile symptoms. She initially was diagnosed as having tension headache by her family doctor and then migraine by an emergency physician at her local hospital. Her headache remained unchanged despite treatment with both oral and intravenous analgesics. Ten days after the onset, she began to develop bilateral horizontal diplopia. She finally presented to our emergency department 17 days after onset, when she developed a new left facial weakness.

Her examination showed bilateral papilledema, bilateral lateral rectus palsies, and a left lower motor neuron facial weakness without involvement of taste. A nonenhanced computed tomography scan of the head showed suspicion of cerebral venous sinus thrombosis (CVST). An magnetic resonance imaging/magnetic resonance venography (MRV) study was completed (Figure 1) the next morning and confirmed the presence of a left transverse and sigmoid sinus thrombosis that extended into the left internal jugular vein. She was started on intravenous anticoagulation treatment. Initial workup including a venous hypercoagulable panel (proteins C and S, lupus anticoagulant, anticardiolipin antibody, activated protein C, and antithrombin III) was negative. Seven days after treatment with intravenous heparin, the patient's deficits remained unchanged; therefore, a lumbar puncture (LP) was performed to rule out infectious and inflammatory causes as well as to allow for the prospect of a therapeutic response from reducing increased intracranial pressure (ICP).

The resolution of papilledema may be relatively rapid in the presence of interventions to rapidly relieve raised ICP,[1] whereas spontaneous resolution of CVST-associated cranial nerve (CN) VII paresis in the presence of retained anticoagulation has been shown to have a more prolonged course.[2] The presence of associated bilateral papilledema influenced the decision to pursue a diagnostic and potentially therapeutic LP, necessitating a brief temporary cessation of anticoagulation.

The LP demonstrated an opening pressure of 46 cm H_2O and, after withdrawing 25 ml, had a closing pressure of 14.4 cm of H_2O. The cell count, glucose, and protein studies of her cerebrospinal fluid (CSF) were within normal limits. Before the end of the procedure, while CSF was being drained, the patient felt that her headache had improved. We reassessed her within minutes after completion of the LP and found that her facial weakness, diplopia, and headache had resolved. While in the hospital, her headaches returned without any cranial neuropathy. She was initially started on acetazolamide and titrated to a maintenance dosing of 1000 mg/day, but required additional analgesia. Her headaches gradually resolved over the next 10 days.

She was discharged home after 11 days on oral anticoagulation, her oral contraceptive was discontinued, and she had a follow-up from the Neurology and NeuroOphthalmology department. On subsequent follow-up over the course of the next year, her headache recurred and gave way to chronic headache without any recurrence of her CN symptoms. A follow-up MRV 3 months after her initial presentation demonstrated some resolution but persisting occlusion of the left transverse sinus. Repeat MRV at 6 months and 1 year showed no change despite continuation of her therapeutically dosed anticoagulation regime.

Given her absence of recurring papilledema or cranial neuropathy, the alternative of chronic migraine was considered. Her acetazolamide was titrated down to discontinuance, whereas topiramate was introduced and titrated up to a maintenance dosing of 100 mg/day. Given that her initial response was suboptimal, a confirmatory test to assess for persisting raised ICP was necessary. Thus, after a decremental titration to discontinuance and washout period that exceeded 4 weeks, a repeat LP was performed that demonstrated a normal opening pressure. Anticoagulation and acetazolamide was discontinued and she was treated thereafter as having chronic migraine with topiramate and Botox for prophylaxis. Prochlorperazine and ibuprofen were used as an abortive strategy. Her headache reverted to an infrequent episodic character and she was subsequently titrated off of all prophylactic medications.

CVST is an uncommon vascular disease that classically presents with headache, seizures, and focal neurological deficits with progression to coma and death.[3] Our case represents an extremely rare presentation of CVST with CN involvement. CN pathology can be associated with CVST, but is most commonly involving the abducens (VI) nerve and rarely others. There have rarely been reported cases of involvement of the facial (VII) nerve, which can occur with or without nerve VI involvement.[2,6] It has been debated as to why this nerve is involved pathophysiologically. Some have theorized that it is a purely pressure-related effect[4,5]; however, because of its short intracranial course and relative protection against the effects of pressure, others have argued that it is due to direct venous clot burden affecting adjacent venous sinuses. Clot burden is theorized to either create localized meningeal inflammation or venous congestion that would directly involve local CNs.[6,7] The most common locations for presentation with facial nerve paralysis are the ipsilateral transverse sinus, inferior petrosal sinus, and sigmoid sinus.[6,7] The assertion that this phenomenon is pressure-related is further supported by the relatively more robust literature in the case of idiopathic intracranial hypertension (IIH) and the presence of nerve VII involvement. IIH is a relatively more common disorder, which, as the name suggests, is characterized by an idiopathic increase in ICP presenting classically with headache, visual changes, and occasionally CN involvement. Again, the most common CN to be involved is the VI, in approximately 20% to 47% of patients.[8] Nerve VII involvement is rare, but has been reported either as unilateral or bilateral. In all reported cases of IIH through 1997 and thereafter, CN involvement was temporally associated with increased ICP and subsequently significantly improved or resolved with management of the underlying pressure.[9-11] Further evidence for this hypothesis is found in a case of a child with a ventriculoperitoneal shunt who presented with facial diplegia secondary to shunt

https://doi.org/10.1017/cjn.2016.446 Published online by Cambridge University Press

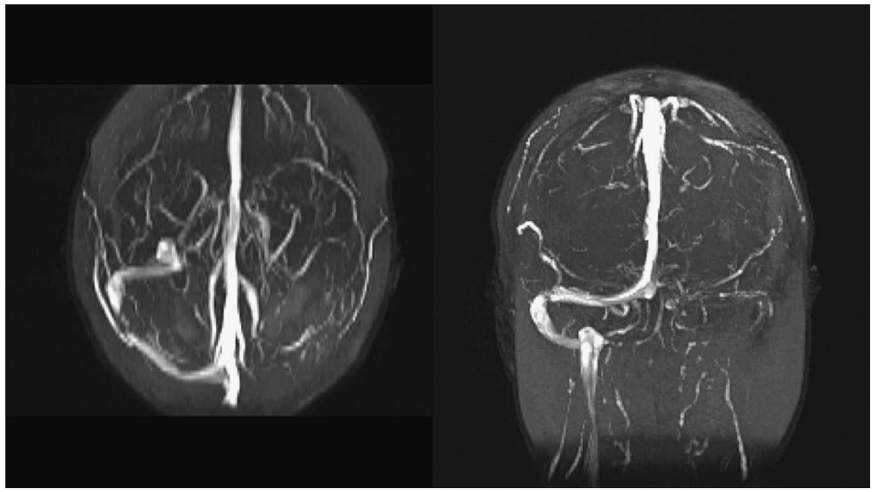

Figure 1: *Axial and Coronal MRV demonstrating occlusion of left transverse and sigmoid sinus extending into the internal jugular vein.*

malfunction and saw complete resolution of their facial diplegia with correction of ICP.[12] Experimental studies have shown a close relationship between pressure in the CSF and pressure on the CNs[13] adding further evidence to this effect.

We believe that our case demonstrates further evidence for CN involvement in CVST may be related to direct ICP effects rather than direct clot burden. This was demonstrated clinically by the patient's facial weakness completely resolving with reduction of ICP through a lumbar puncture, which occurred after demonstration of the clot on MRV and that was still present on the subsequent follow-up MRV 3 months later. Thus, the resolution of CN symptoms took place in the presence of clot by alleviating raised ICP. Furthermore, our patient went on to have chronic headaches without any CN features despite continuing to show evidence of clot burden and a normal opening pressure on subsequent LP. This may also indicate that relief from increased ICP from various causes may reverse cranial neuropathies in the appropriate clinical setting.

Disclosures

MWH and MS do not have anything to disclose.

Mohammed Wasif Hussain
Division of Neurology, Department of Medicine
University of Alberta
Edmonton, AB, Canada
Email: mhussain@ualberta.ca

Muzaffar Siddiqui
Division of Neurology, Grey Nuns Hospital
University of Alberta
Edmonton, AB, Canada

References

1. Friedman D. Papilledema. In: Miller N, Newman N, Biousse V, Kerrison J, editors. Walsh and Hoyt's clinical neuro-ophthalmology, 6th ed. Philadelphia, PA: Lippincott Williams and Wilkins; 2005, P. 237-81.
2. Kulkarni GB, Ravi Y, Nagaraja D, Veerendrakumar M. Lower motor neuron facial palsy in cerebral venous sinus thrombosis. J Neuroscie Rural Pract. 2013;4:224-6.
3. Ameri A, Bousser MG. Cerebral venous thrombosis. Neurol Clin. 1992;10:87-111.
4. Lefkowitz D. Cortical thrombophlebitis and sinovenous disease. In: Vinken PJ, Bruyn GW, Klawans HL, Toole JF, editors. Handbook of clinical neurology, Vol. 54. Amsterdam, Netherlands: Elsevier Science Publishers; 1989, p. 395-423.
5. Straub J, Magistris MR, Delavelle J, Landis T. Facial palsy in cerebral venous thrombosis: transcranial stimulation and pathophysiological considerations. Stroke. 2000;31:1766-9.
6. Kuehnen J, Schwartz A, Neff W, Hennerici M. Cranial nerve syndrome in thrombosis of the transverse/sigmoid sinuses Brain. 1998;121:381-8.
7. Kalbag RM. Cerebral venous thrombosis. In: Kapp JP, Schmidek HH, editors. The cerebral venous system and its disorders. Orlando, FL: Grune & Stratton; 1984, p. 505-36.
8. Skau M, Brennum J, Gjerris F, Jensen R. What is new about idiopathic intracranial hypertension? An updated review of mechanism and treatment. Cephalalgia. 2006;26:384-99.
9. Capobianco DJ, Brazis PW, Cheshire WP. Idiopathic intracranial hypertension and seventh nerve palsy. Headache. 1997;37:286-8.
10. Kearsey K, Fernando P, Hani TS, Benamer HTS, Buch H. Seventh nerve palsy as a false localizing sign in benign intracranial hypertension. J R Soc Med. 2010;103:412.
11. Bandyopadhyay S, Jacobsen DM. Clinical features of late-onset pseudotumor cerebri fulfilling the modified Dandy criteria. J Neuroophthalmol. 2002;22:9Y11.
12. Thines L, Vinchon M, Lahlou A, Pellerin P, Dhellemmes P. Facial diplegia revealing ventriculoperitoneal shunt failure in a patient with Crouzon syndrome. J Neurosurg. 2007:(1 Suppl Pediatrics): 107:46-8.
13. Takeda T, Takeuchi S, Kishimoto S, Saito H. Interstitial fluid pressure in the facial nerve: relationship between facial nerve pressure and cerebrospinal fluid pressure. Am J Otolaryngol. 1989;10:345-50.

https://doi.org/10.1017/cjn.2016.446 Published online by Cambridge University Press