LETTER TO THE EDITOR

To The Editor

Painful Trigeminal Neuropathy and Horner's Syndrome as Manifestations of Cervical Myelopathy

Keywords: Neurology - Clinical, Metastatic tumors, Facial pain, Spinal cord injury

A 54-year-old male presented with a 2-week history of constant left-sided burning, tingling facial pain in the territory of the trigeminal nerve (CNV) with radiation to the neck and shoulder. He had moderate respiratory distress upon arrival at the hospital. Laboratory investigations revealed salicylate poisoning. He had been taking high doses of acetylsalicylic acid (ASA) daily to cope with his severe facial pain. He was stabilized with oxygen supplementation and treated via alkaline diuresis for the salicylate toxicity. He became unable to walk within a day of hospital admission. Neurology was consulted for assessment of this heterogeneous symptom complex.

The patient's neurological examination was significant for subtle left-sided Horner's syndrome with miosis and ptosis. Additionally, he had mild left hemiparesis. Sensory examination was negative for sensory loss to touch or pinprick. However, the patient had continuous left-sided pain in the forehead, maxilla, and mandibular areas. The pain radiated to the neck and shoulder ipsilaterally. The pain was not aggravated by sensory stimulation in the distribution of CNV. No contralateral pain or sensorimotor deficits were present. Cerebrospinal fluid analysis was normal except for mildly elevated protein. No intracranial abnormality was detected on brain CT, CT angiography, or brain magnetic resonance imaging (MRI). Since a cervical myelopathy could explain his deficits, a cervical spine MRI was ordered. It revealed a C4 compression fracture (Figure 1A, B) with retropulsion of disc into the spinal canal leading to severe canal stenosis. The scan showed a destructive, lytic lesion of the C4 body (Figure 1C) with an additional small lytic bone lesion at C6. Fluid within and widening of the left C3–C4 facet joint were also noted.

To investigate for an underlying malignancy, a CT scan of the chest, abdomen, and pelvis was requested. The scan showed lesions involving the gall bladder, liver, thoracolumbar spine as well as lymphadenopathy. Axillary lymph node biopsy showed a metastatic poorly differentiated carcinoma. Typing of the biopsies demonstrated a metastatic CK7+, CK20+ carcinoma of urothelial, gastric, or biliary origin. The patient's fracture was stabilized surgically with a C4 vertebrectomy and C3–C5 fusion with a fibular structural allograft to prevent progression of the cervical myelopathy. Postsurgical imaging showed the allograft partially extruded. More surgery was considered, but the patient's condition deteriorated further, and he was placed under palliative care.

CNV is responsible for sensory innervation of the face. Painful trigeminal neuropathy (PTN) is a form of craniofacial pain occurring in the territory of one or more branches of CNV: Ophthalmic (V1), maxillary (V2), and mandibular (V3). It is distinguished from trigeminal neuralgia (TN) in that TN is characterized by brief, paroxysmal, severe episodes of pain akin to electric shock triggered by innocuous stimulation.[1] PTN is characterized by spontaneous continuous burning, tingling, or pins-and-needles-type pain typically with allodynia/hyperesthesia or hypoalgesia.[1] PTN can be associated with nerve injury from dental procedures, facial trauma, neoplasms, or infectious diseases.[2] The PTN in the present case likely relates to injury of the spinal trigeminal tract. Afferent trigeminal fibers have been postulated to descend into the upper levels of the spinal cord before synapsing and ascending through the spinothalamic tract.[3] The PTN on the left side was compounded by neck and shoulder pain, which could, in part, reflect injury to cervical facet joints. Hypo or hypermobility of the facet joints can cause intervertebral instability leading to root irritation and cervical radiculopathy with associated pain.[4] In our case, there was fluid within and widening of the C3–C4 facet joint, producing greater instability at this interspinal space, consistent with the dermatomal findings of neck and shoulder pain.

Sympathetic innervation to the head and neck region is provided by the oculosympathetic pathway.[5] First-order neurons in the posterolateral hypothalamus descend into the cervical spinal cord and synapse with preganglionic neurons in C8–T1.[5] Preganglionic fibers exit the spinal cord and ascend through the stellate and middle cervical ganglia before synapsing in the superior cervical ganglion.[5] Horner's syndrome involves disruption of this pathway resulting in miosis, ptosis, and anhidrosis.[5] Associated neurologic and systemic signs can help in the localization. Lesions of the first-order neurons can be associated with vertigo, cranial nerve palsies, sensorimotor deficits, or radiculopathy.[6] Symptoms suggestive of pulmonary disease can occur with lesions of second-order neurons.[6] Third-order neuron Horner's syndrome can present with ischemic manifestations or with a sixth nerve palsy.[6]

The combination of CNV abnormalities and Horner's syndrome is classic for Raeder's syndrome, which can be caused by lesions of the middle cranial fossa or the internal carotid artery (ICA).[7] Postganglionic oculosympathetic fibers ascend through the adventitia of the ICA and enter the cranium, where the fibers leave the ICA and join CNV.[7] However, in the present case, brain MR and CT angiography revealed no evidence of dissection or intracranial abnormalities. Hence, unlike Raeder's, the likely mechanism for Horner's syndrome here is a direct disruption of the first or second-order neurons. Lesions of the first-order neurons may result in Horner's syndrome with contralateral hemiparesis if the diencephalon or brainstem are injured.[8] This is because decussation of the medial lemniscus, corticospinal, and reticulospinal pathways occurs within the brainstem. In the present case, hemiparesis was on the ipsilateral side, given that the lesion did not involve supraspinal regions.

This case highlights the need for a broad differential when presented with heterogeneous neurological presentations. The patient's craniofacial pain led to a diagnosis of cervical myelopathy, and, ultimately, metastatic carcinoma. Our case report has limited generalizability because it involves a single patient. The neural mechanisms proposed here are speculative. Further research including neuroanatomical tracing studies of the spinal trigeminal tract with immunohistochemical identification of synaptic boutons would help elucidate the relationship between craniofacial pain and the cervical spine.

https://doi.org/10.1017/cjn.2020.258 Published online by Cambridge University Press

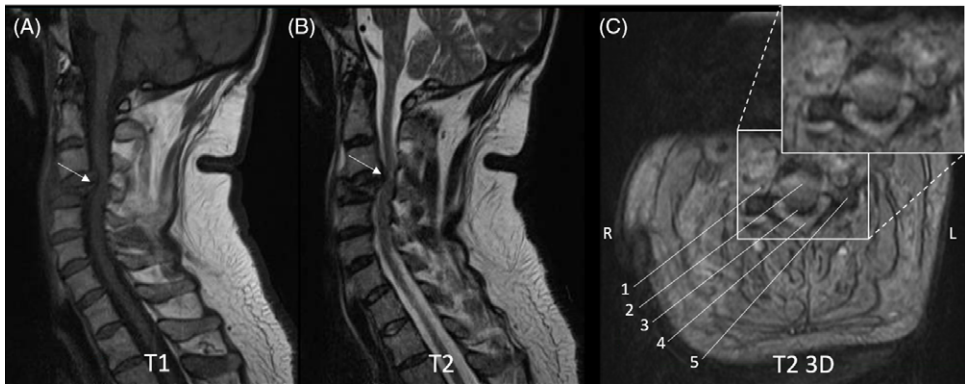

Figure 1: *MRI showing a C4 compression fracture. This 54-year-old with painful trigeminal neuropathy and Horner's syndrome exhibited a C4 spinal cord compression fracture on both T1-weighted and T2-weighted sagittal MRI images (A, B). Point of maximal spinal cord compression (arrows). The vertebral anatomy is distorted due to collapse of the C4 spinal body onto the spinal tissue on a T2-weighted axial MRI image composite (C, box). Vertebral artery (1). C4 spinal body collapsed onto the spinal cord (2). Spinal cord (3). Cerebrospinal fluid (4). Pedicle (5).*

Despite its limitations, our case report demonstrates the unusual ways in which cervical myelopathy and metastatic disease may present. Therefore, disease of the cervical spine should be considered as a potential localization when examining patients presenting with PTN and/or Horner's syndrome.

Acknowledgments

We thank the patient's family for sharing their family member's medical journey.

Disclosures

The authors have no conflicts of interest to declare.

Authors' Contributions

Z.M. wrote the initial manuscript draft and created the figure. E.A. wrote significant portions and edited final versions of the manuscript. Z.S. is the staff neurologist who oversaw writing of the report.

Zacnicte May
Doctor of Medicine Program (YR4), Department of Medicine, University of Alberta, Edmonton, Alberta, Canada

Ekhlas Assaedi
Adult Neurology Residency Program (PGY-4), Department of Medicine, Division of Neurology, University of Alberta, Edmonton, Alberta, Canada

Zaeem Siddiqi
Department of Medicine, Division of Neurology, University of Alberta, Edmonton, Alberta, Canada

Correspondence to: Dr. Zaeem Siddiqi, Department of Medicine, Division of Neurology, University of Alberta, 7-112P Clinical Sciences Building, Edmonton, Alberta, Canada. Email: zsiddiqi@ualberta.ca

References

1. International Headache Society. Part III: Neuropathies & facial pains and other headaches [Internet]. IHS Classification ICHD-3. 2019. Available from: ichd-3.org4.
2. Peñarrocha M, Cervelló M, Martí E, Bagán J. Trigeminal neuropathy. Oral Dis. 2007;13(2):141–50.
3. Shinozaki T, Sakamoto E, Shiiba S, et al. Cervical Plexus Block Helps in Diagnosis of Orofacial Pain Originating from Cervical Structures. Tohoku J Exp Med. 2006;210(1):41–7.
4. Steilen D, Hauser R, Woldin B, Sawyer S. Chronic Neck Pain: Making the Connection Between Capsular Ligament Laxity and Cervical Instability. TOORTHJ. 2014;8(1):326–45.
5. Miller N, Kanagalingam S. Horner syndrome: clinical perspectives. Eye Brain. 2015;7:35–46.
6. Davagnanam I, Fraser CL, Miszkiel K, Daniel CS, Plant GT. Adult Horner's syndrome: a combined clinical, pharmacological, and imaging algorithm. Eye. 2013;27(3):291–8.
7. Murnane M, Proano L. Raeder's Paratrigeminal syndrome: a case report. Acad Emerg Med. 1996;3(9):864–7.
8. Rossetti AO, Reichhart MD, Bogousslavsky J. Central Horner's syndrome with contralateral ataxic hemiparesis: a diencephalic alternate syndrome. Neurology. 2003;61(3):334–8.

https://doi.org/10.1017/cjn.2020.258 Published online by Cambridge University Press