Numb Chin Syndrome: Atypical presentation of metastatic breast cancer

Britta L. Bureau MD[1], Jennifer M. Connelly MD[1], Paul E. Barkhaus MD[1], Ryan Brennan DO[1]

[1]Department of Neurology, Medical College of Wisconsin, Milwaukee, WI

Introduction

In 1830, Charles Bell described two cases of "Numb Chin Syndrome" (NCS) as a mental neuropathy manifested by numbness of the chin and lip.[1] One case had breast cancer and presented with normal lip movement but no sensation on the left half of her lip. Ultimately, it was discovered she had a glandular tumor at the jaw angle where the alveolar branch of the trigeminal nerve courses.[1]

We report a variant of the NCS. Traditionally, NCS is defined by ipsilateral loss of chin sensation. This case is unique in that she had the expected unilateral loss of chin sensation, but her "numb chin" rapidly expanded to include the lower distribution of the ipsilateral maxillary branch of the trigeminal nerve. In addition, she also presented with isolated weakness of the depressor labii inferioris which is innervated by the buccal branch of the facial nerve.

Clinical presentation

A 53-year-old female with past medical history of estrogen receptor positive invasive ductal carcinoma of the right breast underwent partial mastectomy, chemotherapy, and radiation. She was in remission for 6 years when she was diagnosed with new lung and liver metastatic lesions and subsequently resumed chemotherapy. She then developed left chin numbness followed by progressive difficulty retaining food and drink in her mouth. Her left lower lip then became weak. MRI with contrast of the brain/face and dental x-rays were negative eight weeks prior to symptom onset.

Neurological exam revealed left lower lip weakness that appeared to "droop" when attempting to smile (Figure 1). The area of decreased sensation to sharp and light touch over her left lower face had enlarged to between the naso-labial fold and the inferior mandibular border. There was slight extension of reduced sharp touch in the right medial chin that was due to midline overlap of sensory fibers. The remainder of her neurological examination was unremarkable, particularly for any sensory or motor deficits.

The contrast-enhanced MRI was repeated which revealed a 1.9 cm enhancing left parotid mass (Figure 2), not previously seen in the MRI Brain and face completed eight weeks prior at symptom onset. The patient was subsequently referred for palliative radiation therapy, therefore electrodiagnostic studies were deferred. The patient deceased 5 weeks post-onset of her cranial neuropathies.

Discussion

This case is unique because her "numb chin" rapidly expanded to include the lower distribution of the ipsilateral maxillary branch of the trigeminal nerve. In addition, she also presented with isolated weakness of the depressor labii inferioris which is innervated by the buccal branch of the facial nerve. Our primary differential diagnoses included atypical NCS due to metastasis, infection, or trauma.

What began as an uncomplicated NCS that rapidly expanded to involve a greater portion of the trigeminal nerve distribution in addition to a motor branch of an additional cranial (facial) nerve. Both can be accounted for by the parotid metastasis based on their anatomy. Noteworthy is the pseudo-localization because the apparent focal deficits in both cranial nerves did not signify a distal lesion in each, but rather partial lesions of their proximal segments. Thus, her focal neurologic deficits corresponded with the findings on imaging.

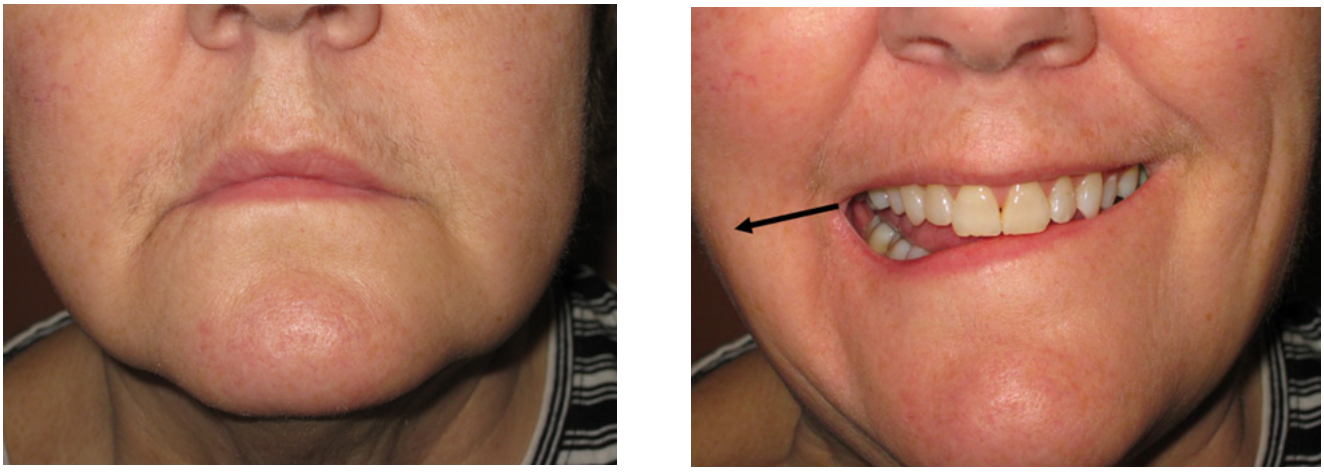

Figure 1: Left lower facial weakness, most likely explained by lack of Buccal nerve innervation to the Depressor Labii Inferioris (© R. Brennan, DO, with permission)

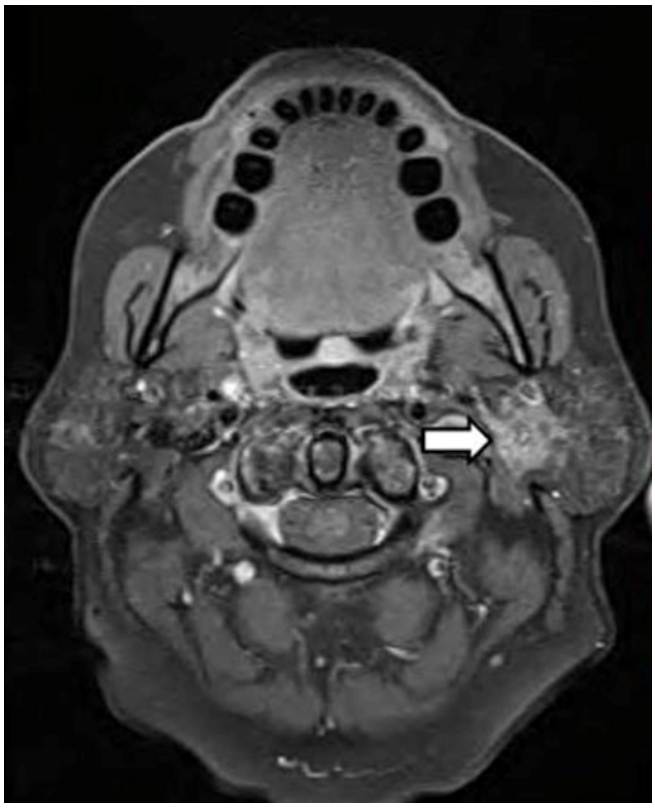

Figure 2: T1-weighted MRI with gadolinium illustrates 1.9 cm enhancing left parotid gland mass in the expected location of the facial nerve (© R. Brennan, DO, with permission)

In typical NCS, the most likely etiology is cancer if there is no history of trauma or dental injury to the inferior alveolar nerve, then the most likely etiology is cancer. NCS may precede diagnosis of malignancy in 47% of patients.[2] Kuroda et al. describe a case of NCS as the initial presentation of Burkitt's cell acute lymphoblastic leukemia.[3] As standard MRI sequences do not usually include the oromandibular region, a mandibular or facial MRI should be performed in NCS.[2] Lossos et al. report in their study that 67% of NCS was a late manifestation of cancer associated with disease progression and 31% NCS signified relapse.[4] The majority of NCS cases described are isolated sensory deficits. Combined cranial neuropathies with additional neurologic symptoms have been reported.[4] Approximately 10% of NCS present with bilateral symptoms.[5] Brazis et al. describe a case of recurrent squamous cell carcinoma where the mental foramen and the infraorbital foramen were involved which resulted in simultaneous, progressive numb chin and cheek syndromes respectively.[6] Their patient had unilateral lip paresis similar to our patient.[6]

Mechanisms of NCS include compression of the mental or inferior alveolar nerves by mandibular metastases [50%], intracranial involvement of the mandibular nerve by skull base lesions such as Meckel's cave tumor [14%], leptomeningeal metastasis [22%], or malignant infiltration of the mental nerve.[3,4,7] In females, the most common malignancy is breast due to metastatic lesions along the sheath of the mentalis branch of the inferior alveolar nerve, often at the mental foramen, or metastatic involvement of the jaw. NCS syndrome is also a common presentation in lymphoma and less often in leukemia, prostate, and lung cancer.[2,3,4] Nonmetastatic causes of NCS are rare including Sjogren's, multiple sclerosis, diabetes mellitus, temporal arteritis, systemic amyloidosis, HIV, drug toxicity, local infection, and natural aging.[2,4,8]

NCS is clinically significant as it is an ominous sign for underlying malignancy and often associated with a poor prognosis as it can be a late manifestation of a systemic malignancy.[3,4,9] Brady et al. found that mean survival of NCS was 6.9 months, having only 15% of patients who present with NCS survive more than nine months.[2]

Conclusion

We present a unique case of NCS which resulted from metastatic breast cancer. This case is unique because it evolved quickly from a "simple" clinical left NCS to proximal left partial fifth and partial cranial neuropathies. Our case reinforces the point that a seemingly harmless NCS should be considered an ominous sign until malignancy is excluded. Awareness and early localization of NCS are critical to efficiently develop a targeted treatment plan to prevent tumor spread and potentially improve clinical outcomes and survival.

Corresponding Author

Britta L. Bureau, MD
Medical College of Wisconsin
8701 W Watertown Plank Road
Wauwatosa, WI, USA 53226
bbureau@mcw.edu

Disclosures

Ryan Brennan DO has received speaking and consulting fees from Abbvie. Also has received consulting fees from Medtronic.

Britta L. Bureau BS has nothing to disclose.

Jennifer M. Connelly MD has nothing to disclose.

Paul E. Barkhaus MD has nothing to disclose.

Poster Presentations

1. Society for Neuro-Oncology Annual Scientific Meeting in November 2022 in Tampa, FL
2. American Association of Neuromuscular & Electrodiagnostic Medicine Annual Meeting in September 2022 in Nashville, TN.

Oral Presentation

Wisconsin Neurological Society Annual Meeting. October 2022 in Wisconsin Dells, WI.

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