Case Report

Numb Chin Syndrome as a Primary Symptom of Burkitt Lymphoma

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ABSTRACT: Numb chin syndrome (NCS) is a sensory neuropathy characterized by numbness involving the distribution of the mental nerve. Although rare, NCS is very important because it could be the first and only symptom of an underlying malignancy. Therefore, primary care physicians should pay attention to middle-aged persons or cancer-treated persons who complain of unilateral numbness of the lip or chin, and consider malignant etiology until proven otherwise. In this article we describe a 54-year-old man complaining of numbness of the right side of his chin due to peripheral spread of Burkitt lymphoma, and provide review of several articles.

KEY WORDS: Numb chin syndrome, Mental nerve neuropathy, Lymphoma

Gen Med: 2010; 11: 35–38

INTRODUCTION

Numb chin syndrome (NCS) or mental nerve neuropathy is a sensory neuropathy characterized by numbness, paresthesia, and very rarely pain in the distribution of the mental nerve. It is uncommon but very important because it could be the first and only symptom of an underlying malignancy. In this article, we report a patient with NCS occurring as the initial manifestation of Burkitt lymphoma.

CASE REPORT

A 54-year-old man consulted us (Department of General Medicine) complaining of numbness and slight pain in the right side of his chin, ongoing for one month. About one month before consultation he visited his dentist complaining of numbness for a few days in the right side of his lower jaw, but no any abnormality was pointed out. Evaluation at another hospital included brain magnetic resonance imaging (MRI) scan and MR angiography, both of which were
reported to be normal. Two weeks later he was admitted to the emergency department of the hospital for exacerbation of numbness, headache and transient drowsiness when he was drinking. He recovered within a few hours completely, and there was no abnormality including blood chemical panel, chest X-ray, electrocardiogram, and electroencephalogram findings, so he discharged next morning. One week later he complained of left shoulder and leg pain following chest pain treated with nonsteroidal anti-inflammatory drugs, but his complaints were not relieved. Considering this clinical course, his physician referred him to us.

His medical history included lumbar disk herniation, acute pancreatitis and benign mandible tumor extirpated ten years ago. His medication was only pravastatin sodium for hypercholesterolemia.

At the initial examination he presented without fever, anemia or body weight loss. Neurological examination revealed hypesthesia below the lower lip on the right side, the location of the distribution of the third division of the trigeminal nerve. Other cranial nerves, including the facial nerve and the central nervous system, and a musculoskeletal examination were normal. Cervical lymph nodes were not swollen. No supraclavicular or axillary lymph nodes were found. Cardiovascular, respiratory or gastrointestinal systems were also normal.

Initial investigation showed leukocytosis 17940/mm³ with a few immature neutrophils, elevated C reactive protein 9.3 mg/l and Lactate dehydrogenase 593 IU/l. We consulted our dentist, and she replied that inflammation of dens molaris was suspicious, but it could not explain the accompanying osteolysis and aberrant value of laboratory data (Fig. 1). Three days later, the subject was hospitalized urgently for further examination because his peripheral blood leukocyte count was elevated to 22,920/mm³, with 14% myelocytes, 8% metamyelocytes, 1% blastic cell, 56.5% neutrophils, 1.5% eosinophils, 0.5% basophils, 5.5% monocytes, 10.5% lymphocyte and 2.0% atypical lymphocytes. A bone marrow examination demonstrated abnormal lymphoblasts with the monoclonal expression pattern of CD10 (+), CD20 (+), CD3 (−), CD5 (−), CD7 (−), bcl-2 (−), c-myc (+). Biopsy specimen from the right lower gingiva showed the dens medium-sized lymphocytes with macrophages, so called starry sky appearance (Fig. 2). Cerebrospinal fluid was also positive for malignant cells. These findings confirmed the diagnosis of Burkitt lymphoma with metastasis to the right gingiva and leptomeningeal space.

DISCUSSION

The classic illness script of NCS due to malignancy is as follows: Most patients are around 40–50 years, with females more common. A typical complaint is unilateral numbness of chin and lower lip. This numbness is often accompanied by an abnormal sensation of “thickening” of the lip similar to the experience of dental anesthesia. Pain is an infrequent symptom, and motor function of lower face remains intact because the inferior alveolar nerve has no motor fibers. A numb chin may represent the first symptom of underlying malignancy, but more frequently signals...
progression or relapse of known disease (27.7% and 37.7%, respectively)\(^5\). In exceptional cases, NCS by acute leukemia can occur in teenagers\(^6\).

Although Charles Bell reported the first case in 1830\(^7\), various malignant etiologies of NCS have been reported since 1963, when Calverly et al. introduced the term “syndrome of numb chin”\(^8\). Gil et al. reported in their review of 136 cases that breast cancer was the leading cause (40.4% of cases), followed by lymphoma in 20.5%, prostate cancer in 6.6%, Leukemia 5.1%, Myeloma 3.8%, Lung cancer 2.9%. Miscellaneous causes were Sarcoma, Myeloma and Melanoma. Other literature reports cancers of the head and neck, thyroid, nasopharynx, gastrointestinal system, kidney and melanoma as causative malignancy\(^8,9\).

The mechanisms of NCS as a presence of malignancy are not well known but may be peripheral spread (i.e., metastasis or invasion of mandible), central spread (i.e., located at the base of the skull, leptomeningeal seeding, perineural or neural invasion) or paraneoplastic syndrome\(^2\).

Numb chin syndrome due to malignancy is also associated with an ominous prognosis with a weighted mean survival of 6.9 months\(^5\). Our case responded to systemic chemotherapy at first and discharged as cytogenetic remission. However, he suffered a relapse four months later and died by sepsis, 11 months after the initial presentation. Every recent case report on NCS by Burkitt lymphoma also shows adverse prognosis\(^4,10\).

NCS or mental nerve neuropathy is well recognized to occur in association with various conditions besides malignancies: inflammatory disorders, traumas, cysts, syphilis, arachnoiditis, sarcoidosis, connective tissue diseases, vasculitis, multiple sclerosis, vertebro–basilar insufficiency, systemic infections, age–related neuropathy, drugs, and toxic chemicals\(^6\). However, the most common cause of NCS or mental nerve neuropathy is invasive dental treatment procedures (e.g., extractions, implants) followed by malignant metastasis\(^3\). Therefore, when we doubt the NCS we should exclude a dental cause at first, then consider the symptom with systemic malignancies, especially breast cancer in women, prostatic cancer in men, and malignant lymphoma in both.

Imaging study and biopsy is necessary to diagnose the underlying cause. For imaging study, authorities recommend panoramic radiographs and CT scans or MRI of the mandible, skull base, and brain. Other valuable tests, such as chest x-rays, lumbar puncture, bone scintigraphy, careful view of blood smear, bone marrow aspiration and biopsy should be considered\(^1,5\), depending on the situation. Mammography and measuring of prostate specific antigen should be added to the list, considering the frequency. Finally, the most important step in the diagnosis is to recognize the potential clinical significance of unilateral chin or lip numbness\(^8\).

CONCLUSION

Numb chin syndrome is rare, but this impressive symptom could be the only clue to systemic malignancy. Primary physicians should pay attention to middle aged person or cancer–treated person who complains unilateral numbness of the lip or chin, and consider malignant etiology until proven otherwise.

References


