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# NUMB CHIN SYNDROME AS AN INITIAL PRESENTATION OF ACUTE LYMPHOBLASTIC LEUKEMIA.

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## Abstract:

Paresthesia and anesthesia in the territory of the mental nerve (Numb Chin Syndrome) may result from a variety of conditions. When not caused by dental disease, the findings may be the heralding symptom of a malignant disease. Its recognition should prompt a search for malignancy. We describe a case of Numb Chin Syndrome occurring as the initial manifestation of Acute Lymphoblastic Leukemia. We discuss pathogenesis, diagnosis and treatment of the syndrome.

**MeSH words:** mental neuropathy, leukemia, malignancy, numb chin

## Introduction

Numb Chin Syndrome (NCS) is a rare and under appreciated sensory neuropathy. Also called mental neuropathy, it is usually associated with malignant neoplasia (1). NCS is characterized by numbness or sensory loss in the distribution of the mental branch of the mandibular nerve. It can appear together with other signs of neoplastic dissemination or constitute the presenting symptom of the disease (2).

We describe a patient with NCS as the presenting symptom of acute Lymphoblastic Leukemia.

## Case report

A 45-year-old woman, usually healthy, was referred to the hospital because of numbness in the left lower lip and chin for 5 days. The patient had been in good health until 2 weeks earlier, when there was the onset of generalized myalgia and a subfebrile illness that was diagnosed as viral infection. Five days before referral started feeling numbness and hyposthesia in the left side of the chin and left lower lip. There was no dental pain, local swelling or other sensory or motor abnormalities. The temperature that was subfebrile 2 weeks earlier returned to normal after a few days. The patient complained of a mild generalized headache, but reported no

vomiting, visual changes, swallowing or speech difficulties.

Laboratory tests showed an increased erythrocytation rate, leukocytosis with monocytosis and thrombocytopenia with normal hemoglobin levels. She was then referred to hospital.

On arriving to the Emergency Medicine Department the patient appeared well. The vital signs were: blood pressure of 125/84 mmHg, pulse of 98 beats/minute, an oral temperature of 36.6<sup>0</sup> C and 98% O<sub>2</sub> saturation.

There was no facial asymmetry. No rash, petechiae, echymosis or icterus were found. No pallor, cyanosis, clubbing or peripheral edema were noted. An ENT examination showed normal gums and teeth. No local swelling or pain was present. No lymphadenopathy or masses were found in the neck or elsewhere.

The lungs and heart were normal. The abdomen was soft and no organomegaly was present. On neurological examination the pupils were equal and reactive. The retinas were normal. Extraocular movements were full and no nystagmus was elicited. There was a patch of impaired sensation to touch and pinprick in the left side of the chin and the left lower lip. The rest of facial sensation and facial movements were normal. The remaining cranial nerve functions were preserved. The strength of the limbs was normal. Sensation of all types and deep tendon reflexes were normal and symmetric throughout. The plantar responses were flexor. The coordination was normal and the Romberg's test negative.

A chest X-rays revealed a large and well-delineated mass in the anterosuperior mediastinum. The lungs, heart and bones were normal. Panorax radiographs of the mandible disclosed a mild bony reabsorption area of the mandible in the left side. The bony cortex was intact and no soft tissue mass was identified.

The blood cell count showed that the hemoglobin was 12.7 gr/dl; the hematocrit 37.7%; the white cell count was 17.000/mm<sup>3</sup>, and a platelet count of 23.000/mm<sup>3</sup>. The peripheral blood smear demonstrated leukoerythroblastic changes with 2% blasts, 1% promyelocytes, 1% metamyelocytes, 2%

myelocytes, 10% bands forms, 19% segmented neutrophils, 48% lymphocytes, 14% large variant lymphocytes and 3% monocytes. There were also 5% of normoblasts.

The levels of glucose, urea nitrogen, creatinine, calcium, electrolytes, bilirubin, alkaline phosphatase and liver enzymes were within normal limits. The uric acid was elevated to 11 mg/dl and the LDH levels increased to 3590 U/L (normal 318-618 U/L). The ECG was normal.

Because the clinical, X-ray and blood cell count findings an urgent hematology consultation was requested. The patient was admitted and a bone marrow aspiration was done that showed that the marrow was infiltrated with lymphoblasts and the diagnosis of Acute Lymphoblastic Leukemia was done. The next morning the patient was transferred to a tertiary medical center where the cranial CT scan and CSF examination were normal and started chemotherapy. The immunophenotyping was compatible with Acute Lymphoblastic Leukemia T cell type.

## Discussion

Numb Chin Syndrome (NCS), or mental nerve neuropathy is a rare and seemingly harmless symptom. The symptoms include numbness of the skin of the chin, the lower lip and sometimes the gingiva (2). The presentation is usually unilateral but may be bilateral. The motor function of the face is intact.

In most cases the syndrome is caused by metastatic involvement of the mental nerve or the lower alveolar nerves of the jaw (50%) and in a smaller proportion of patients by involvement of the proximal mandibular root of the trigeminal nerve at the base of the skull (14%) or by intracranial leptomeningeal spread (22%) (1).

In the presence of a destructive process such as malignancy or infection, pain and swelling may be present. If metastatic malignancy exists, then constitutional symptoms as fever, weight loss and fatigue may also be present. The NCS may also precede any other symptoms of malignancy (3,4)

The most common source of metastases is the breast and lymphoma in adults and acute

lymphoblastic leukemia in children (2). Other malignancies less commonly associated with NCS includes lung, prostate, melanoma, sarcomas, renal carcinoma, multiple myeloma and neck cancers.

Non-malignant etiologies include dental abscess or osteomyelitis, dental and facial trauma and benign tumors (2). Systemic non-malignant diseases are very rare etiologies as amyloidosis, sickle cell anemia, diabetes mellitus syphilis and systemic vasculitis.

The diagnosis of NCS is clinical but imaging and laboratory studies are needed to confirm the diagnosis and establish the place of nerve involvement and the etiology. Because a large number of NCS presentations are due to metastases to the mandible the panoramic radiography of the jaw is a good starting point (3). Imaging may show osteoblastic or osteolytic lesions along the mandible. CT of the mandible may also be necessary. CT of skull base and brain may reveal a mass lesion, brain metastases or leptomeningeal invasion.

If the routine imaging studies had been completed and non-diagnostic, MRI or nuclear bone scanning may identify metastases or osteomyelitis in the jaw or other areas in patients with widespread malignancy (2).

Lumbar puncture and cytology of CSF may help in carcinomatous meningitis or leptomeningeal metastases if imaging studies fail to reveal an anatomic lesion (1).

The treatment is of the underlying disease and the prognosis is poor in malignant disease. In cases of dental source, an oral surgery consultation is indicated.

### Conclusion

The appearance of NCS may be the heralding symptom of a malignant disease or the first manifestation of relapse. Its recognition should prompt a search for malignancy. Because at first glance the syndrome looks innocuous, after recognizing the syndrome a high index of suspicion will help avoid misdiagnosis.

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