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Numb Chin Syndrome in Acute Lymphoblastic Leukemia

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Numb chin syndrome is a sensory neuropathy of the inferior alveolar branch of the trigeminal nerve, characterized by unilateral numbness of the chin, the lower lip and the buccal and gingival mucosa. We report a girl with acute lymphoblastic leukemia of B-cell type who initially presented with numb chin syndrome resulting from skull base infiltration.

Key words: *B-ALL, Complications, Cranial nerve involvement, Numb chin syndrome*.

Numb chin syndrome (NCS) is a sensory neuropathy of the inferior alveolar nerve, a branch of the trigeminal nerve. In adults, NCS may be associated with malignancies of breast, lung and prostate [1]. It has also been described in Non-Hodgkin lymphoma and Burkitt lymphoma. Numb chin syndrome is uncommonly reported in children [2-5]. We report this syndrome as one of the initial symptoms of acute lymphoblastic leukemia in a child.

CASE REPORT

A 9-year-old girl presented with a two week history of increasing fatigue, fever, night sweat and weight loss. Physical examination revealed firm and painless right-sided fronto-parietal swellings. There was no lymphadenopathy or hepatosplenomegaly. The girl also reported unilateral hypoesthesia of the left chin, the lower lip, and the gingival and buccal mucosa for one month. There were no other neurological symptoms. MRI of the head showed multiple bone lesions with contrast-enhancement of the extra- and intracranial soft tissue at different locations (frontal, right temporo-parietal, left sphenoid and left mastoid). Complete blood count was within normal limits, but LDH (2315 U/L) and uric acid (16.9 mg/dL) were highly elevated. Typical Burkitt blasts were seen in the blood smear and the bone marrow was completely replaced by blasts. Cerebrospinal fluid showed no evidence of leukemic cells. The patient was treated according to the prospective multicenter trial B-NHL BFM 04 with

intensified central nervous system (CNS) treatment including repeated applications of intrathecal chemotherapy with prednisone, methotrexate, and cytosine arabinoside. She achieved complete remission after the first course of chemotherapy and the symptoms improved. Numb chin syndrome resolved completely after three courses of chemotherapy. Cranial lesions also decreased in size on follow-up MRI. The child achieved complete remission.

DISCUSSION

Numb chin syndrome in malignancies results from invasion of the mandibular canal or mental foramen by mandibular metastases or from invasion of the mental nerve and/or the alveolar nerve itself. In addition, subcutaneous mental metastasis or enlarged lymph nodes can cause compression of the inferior alveolar nerve before its entry into the mandibular canal. Metastatic infiltration of the proximal root of the mandibular nerve at the skull base and intracranial leptomeningeal invasion may also lead to NCS. "Paraneoplastic" NCS has rarely been described resulting from antibody production against the Gasserian ganglion [1].

The association of NCS with malignancies of B-cell type, as seen in our patient, is an interesting phenomenon. Fenaux, *et al*. [6] described 10 of 18 adult patients with B-ALL presenting with NCS. The authors considered that the presence of isolated NCS, which was observed in four patients, was diagnostic of CNS involvement and recommended additional CNS treatment for these patients. Kuroda, *et al*. [7] reported on the involvement of the trigeminal nerve root in a 57-year old man with Burkitt cell acute leukemia. They demonstrated heavy infiltration of leukemic cells and destruction of axons and myelin by leukemic cells in a post-mortem study of the trigeminal nerve.

Skull base infiltration is frequently observed in children with malignancies of B-cell type [5, 8-10]. Choi, *et al*. [10] reported four children with B-cell lymphoma experiencing visual disturbances or visual loss due to central skull base (sphenoid bone) involvement. Burkitt lymphoma involving the clivus has also been reported in three children; with two of them having sixth cranial nerve palsy [8]. Seixas, *et al*. [5] recently described the first case of skull base infiltration in a child with Burkitt leukemia and clinical signs of NCS. Cavernous sinus involvement led to paresis of the right oculomotor nerve and ultimately to NCS. Tumors of the clivus commonly present with diplopia and cranial nerve palsies, especially of the sixth cranial nerve. Other cranial nerves such as the trigeminal nerve exit the skull through the foramen ovale and might be damaged by skull base infiltration, giving the symptoms of NCS, as seen in our patient. Cranial MRI revealed leukemic infiltration of the sphenoid in our patient explaining the symptom of numb chin syndrome. This was interpreted as cranial nerve infiltration. Therefore, a more intensive CNS therapy was administered in this patient.

We conclude that NCS in children with malignancies of B-cell type, with or without detectable infiltrations of the skull base, should be interpreted as cranial nerve involvement and consequently treated with intensified CNS therapy.

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Hopkins Syndrome and Phantom Hernia: A Rare Association

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Acute flaccid paralysis (AFP), other than paralytic poliomyelitis, are usually due to demyelination like Guillian Barre syndrome (GBS), transverse myelitis and traumatic neuritis. Poliomyelitis like illness, Hopkin's syndrome or Post Asthmatic Amotrophy, associated with bronchial asthma and hyperIgEemia has been reported in literature. We present a two and a half year old child who developed AFP with phantom hernia following an episode of bronchial asthma.

Key words: *Acute Flaccid Paralysis, Phantom hernia, Hopkin's syndrome, HyperIgEemia.*

Subsequent to decline in cases of paralytic poliomyelitis, demyelination is the most common cause of AFP followed by traumatic neuritis. AFP has also been reported with asthmatic attack and hyperIgEemia (Hopkin's syndrome or Post asthmatic amotrophy) and following ICU admission for critical illness, called 'critical illness polyneuropathy/myopathy [1-3]. Phantom hernia refers to unilateral bulging of abdominal wall due to patchy paralysis of abdominal wall muscles. It was first described in paralytic poliomyelitis by Achar [4]. It is derived from the word 'phatasm' meaning mental imagery produced by fantasy [5]. It has also been reported with non-polio conditions like hypokalemia complicating gastroenteritis [6].

We report a case of AFP with phantom hernia associated with an asthmatic attack and hyper-IgEemia. Several cases of poliomyelitis *like* illness following bronchial asthma have been reported [7-10], but associated phantom hernia has not been documented.

CASE REPORT

A two-and-a-half year old male child with normal growth and development was admitted in PICU with acute severe asthma. He had intermittent asthma for the past one and a half years. He was fully immunized including pulse polio immunizations. He had three days of ICU stay and was given nebulization with salbutamol, ipratropium and steroids, ampicillin, IV methyl prednisolone and $MgSO_4$ infusion. He responded and did not require any ventilator support or muscle relaxants.

On the 4th day of hospital stay, he developed acute flaccid paralysis of the left lower limb, which progressed and involved the right lower limb in the next two days. His higher mental function and upper limbs were normal. He had hypotonia, grade 0 power, areflexia and down going plantars in both lower limbs. He had no sensory involvement except for myalgia. Abdominal reflex on the left lower quadrant was absent. There was transient bladder involvement also. Blood counts were within normal limits and there was no eosinophilia. CSF study one week after paralysis showed 0-2 lymphocytes/mm^3 and protein of 20 mg/dL and sugar of 80 mg/dL. Mantoux test was negative. Chest *X*-ray showed increased bronchovascular markings. Serum CPK and LDH were normal. Mycoplasma antibody was