Arch Iranian Med 2005; 8 (1): 60 – 62

Case Report

"NUMB CHIN" AS THE FIRST AND SOLE PRESENTING SIGN OF MULTIPLE SCLEROSIS

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Cranial nerve palsies are occasional clinical findings in multiple sclerosis. Sometimes, they are the presenting sign of multiple sclerosis. Facial numbness localized in the region of the distribution of the mental nerve has been labeled 'the numb chin syndrome'. Differential diagnosis in presentations such as these includes cancer that involves the mental nerve.

Here we report a case of numb chin syndrome, a more limited form of trigeminal nerve involvement, as the first and sole presentation of multiple sclerosis.

Archives of Iranian Medicine, Volume 8, Number 1, 2005: 60 – 62.

Keywords: Multiple sclerosis • numb chin

Introduction

Numb chin syndrome is an infrequently recognized neuralgic symptom characterized by hypoesthesia, paresthesia, or thermoalgesic anesthesia of the chin and the lower lip, limited to the region served by the mental nerve. It is more frequently described in association with malignancies than with other systemic diseases. It is most often a forerunner to malignancy progression and relapse, but may also precede the diagnosis of cancer.[1 – 3]

In one case, chin numbness was the first manifestation of HIV infection.[4] Numb chin syndrome of neoplastic origin is usually unilateral, but bilateral presentation may also occur (10%).[5, 6] An important feature is that this syndrome can precede the diagnosis of malignancy in up to 47% of the cases.[7]

Numb chin syndrome may be seen secondary to radiotherapy or exposure to chemical agents.[8] Rarely, it may occur in association with systemic disease such as sickle cell disease,[9] collagen vascular diseases,[10] viral infections,[11] and Lyme disease,[12] or as the first sign of a temporal arteritis.[13] Only two cases of spontaneous remission have been published where both syndromes occurred suddenly. Such a favorable outcome suggests the cause of numb chin syndrome to be of vascular origin, although a viral etiology, as in Bell's palsy, is also possible.

Cranial nerve palsies are occasional clinical findings in multiple sclerosis.[14] Sometimes they are the presenting sign of multiple sclerosis.[16] Isolated pupil-sparing third-nerve palsy, isolated sixth-nerve palsy, sudden hearing loss, and trigeminal nerve involvement have been previously reported as presenting signs of multiple sclerosis.[15 – 20]

Here we report a case of numb chin syndrome, a more limited form of trigeminal nerve involvement, as a first and sole presentation of multiple sclerosis.

Case Report

In September 2002, a 41-year-old woman was admitted to our hospital because of numbness of the chin and lower lip.

Two weeks prior to admission she noted a prickling sensation like pins and needles at the right corner of her mouth. This sensation extended bilaterally to the lower lip and to her chin. Neurologic examination revealed only a superficial hypoesthesia of the chin and lower lip (numb chin syndrome). There was no clinical evidence of palpable regional lymph nodes or other systemic

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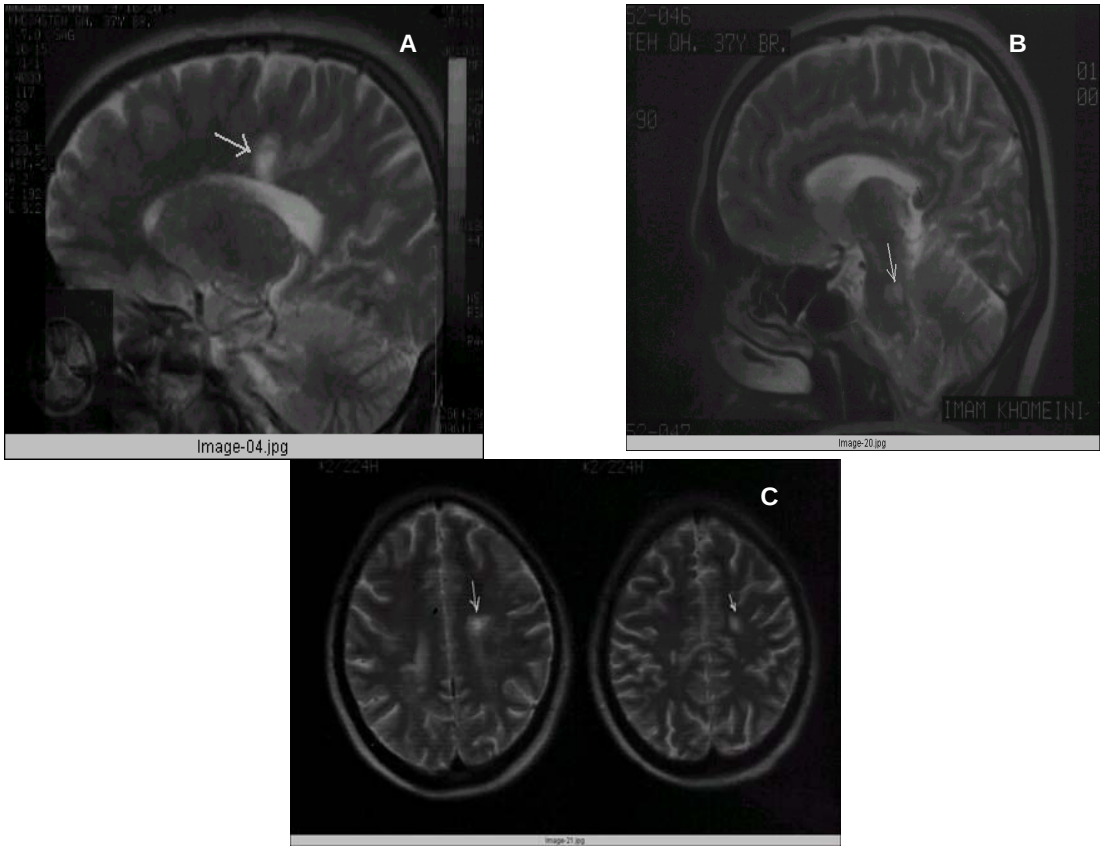

Figure 1. A) T2-weighted MRI showing pericallosal plaques with the pattern of "Dawson fingers"; B) T2-weighted MRI showing a pontine plaque responsible for patient's symptom; C) T2-weighted MRI showing two periventricular plaques.

and neurologic abnormalities.

X-rays of the mandible and a CT scan of the head and neck revealed no lymphadenopathy or masses in the neck or pharynx or lytic lesions to the skull or mandibles. A systemic search for other organ malignancies such as breast cancer, multiple myeloma, and vasculitic disorders revealed no abnormalities. The numbness and hypoesthesia spontaneously disappeared gradually over a few weeks' time.

Four months later, the same patient returned with weakness in her left limbs. Neurologic examination of cranial nerves was normal, except for a mild left central facial weakness and left hemiparesis (grade 3/5).

Brain MRI was performed and revealed multiple hypersignal lesions typical of multiple sclerosis (periventricular and pericallosal with the pattern of "Dawson fingers", in the pons, and centrum semiovale) (Figure1 A, B, C).

After pulse therapy with intravenous methyl-prednisolone, hemiplegia and facial weakness gradually improved. Then the patient was selected as a candidate for interferon therapy.

Discussion

The mechanisms of this syndrome in cancer patients may be some of the followings: metastases or invasion of the mandible, base of the skull lesions, leptomeningeal seeding, perineural or neural invasion, or paraneoplastic syndrome.[21]

According to Nakashima and coworkers, very rare postmortem examinations of the brains of patients with multiple sclerosis who had trigeminal neuralgia have shown demyelination changes in the pons at the root entry zone. They reported 5 MS patients with linear pontine trigeminal root lesions on MRI. All of 5 patients had definite diagnoses of clinical MS and had various types of facial sensory disturbances, such as neuralgia in one patient, hyperesthesia in 2, and paresthesia in 3.[22]

Thomke and colleagues,[14] in a study on 24 patients with isolated cranial nerve palsies, found third and fourth nerve palsies in one patient, sixth nerve in 12 patients, seventh nerve in three patients, and eighth nerve in seven patients. Cranial nerve palsies were the presenting sign in 14 patients and the only clinical sign of an exacerbation in 10 patients. MRI was carried out in 20 patients and results were completely compatible with brainstem lesions (e.g., MRI hyperintense T2-weighted lesions corresponding to the involvement of the ipsilateral intrapontine sixth nerve, indicative of a patient with an isolated left-sided sixth nerve palsy as the presenting sign of MS). Additional abnormal findings of electrooculography, masseter reflex, blink reflex, or combinations of these were found in 20 patients and interpreted in favor of a brainstem lesion at the level of the respective cranial nerve.[16]

Knowledge of the potential of cancers to involve the mental nerve is mandatory, since it may lead to the discovery of an undiagnosed cancer. In Lossos and Siegal's study, this etiology was found in 86% of affected patients. In 64% of these patients the retained etiology was bone metastases either in the mandible (50%) or at the base of the skull (14%). In eight patients (22%) leptomeningeal seeding was the cause of mental neuropathy.[6]

In conclusion, isolated cranial nerve palsies are rare clinical findings in multiple sclerosis, occurring in only about 1.6% of all patients and in 5.2% as the presenting sign.[16] MRI may fail to confirm corresponding brainstem lesions in many patients with isolated cranial nerve palsies. In these patients, electrophysiological tests seem to be more sensitive in disclosing brainstem dysfunction at the level of the affected cranial nerve.

This seems to be the first report of numb chin syndrome as a first and only symptom of multiple sclerosis. We recommend brain MRI, electrophysiological tests, and other investigations for multiple sclerosis in cases presenting with numb chin of unknown etiology, especially when bilateral and remitting.

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