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Numb chin syndrome: A case report and review of the literature

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nbness of the lower lip, the skin of the chin, or the gingiva of the lower anterior teeth ndary to various dental diseases is a common manifestation that is largely underappreciated. association of numb chin syndrome (NCS) with serious disease like metastatic malignancy systemic conditions is frequent enough to warrant a search for these possible etiologies. In paper we report a case of NCS that occurred secondary to malignant disease; we discuss our ings with special emphasis on metastatic malignancies leading to NCS, when the survival nly for a few months.

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Numb chin syndrome (NCS) or mental nerve neuropathy (MNN) is 'an infrequently recognized neuralgic symptom characterized by hypoesthesia, paresthesia, or thermalgesic anesthesia of the skin of the chin and lower lip, limited to the region served by the mental nerve.' The first description of this syndrome was by Charles Bell in 1930 who noted mental anesthesia among patients with jaw trauma and retromandibular tumors.^[1] Later, Roger and Pillas stressed the need to search for visceral neoplasms and malignant hemopathies in such cases after local causes have been ruled out.^[2] Since then, the association of NCS with metastatic malignant disease has been called Roger's sign. NCS is most commonly seen as a manifestation of local dental disease but it becomes clinically more significant when it is associated with neoplastic processes, particularly with breast cancer and malignant lymphomas, as any malignant metastasis to the maxillofacial region shows poor prognosis.

CASE REPORT

A 45-year-old Indian male reported to the department of Oral and Maxillofacial Surgery, Punjab Government Dental College and Hospital, Amritsar with the chief complaint of an intraoral swelling in the left lower jaw for the past 6 months [Figures 1 and 2]. It had slowly increased in size and there was numbness of the skin over the left chin. The patient's medical history was unremarkable. His vital signs were normal. Local examination showed facial asymmetry due to the presence of a nontender, firm extraoral swelling in the left mandibular body region. Intraorally [Figure 3],

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two swellings were present: One was in the left lower lateral incisor region and the other was a large swelling situated posteriorly in the premolar region; both swellings were soft, nontender, lobulated, and attached to the underlying bone. The swellings were noninflammatory in nature. The surfaces were uneven yet smooth. The superior borders were well demarcated and the inferior borders merged with the alveolar mucosa up to the bottom of the vestibule, with obliteration of sulcus. There was anesthesia of the left lower lip, the skin of the chin, and the gingivae. The teeth were slightly mobile due to poor periodontal condition. The submandibular lymph nodes were palpable bilaterally and were firm in consistency and mobile. Radiographs showed no bony involvement. Fine needle aspiration cytology (FNAC) of the lesion and the lymph nodes indicated non-Hodgkin's lymphoma; this was confirmed by incisional biopsy of the lesion under local anesthesia. No other site of disseminated disease was detected at this stage.

These cases are best managed with radiotherapy and chemotherapy and, therefore, the patient was referred to the Department of Oncology and Radiotherapy, Sri. Guru Teg Bahadur Hospital, Amritsar, for further treatment.

DISCUSSION

NCS is most often a symptom caused by dysfunction of the terminal sensory branch of the mandibular division of the trigeminal nerve. It can be easily overlooked by an unsuspecting clinician. Most often, local dental causes are responsible for NCS, but when they have been ruled out metastatic malignancy is seen to be the second most common cause, ahead of other systemic conditions.

Numb chin syndrome



Figure 1: Extra oral view showing swelling in left mandibular body region



Figure 2: Swelling with absence of inflammatory signs



Figure 3: Intra oral view showing masses extending from alveolar ridge

Etiology

Dental diseases that can cause NCS include periapical inflammation, cysts, benign tumors, and fractures in the craniofacial region.

NCS is a potentially significant symptom because of its association with metastatic malignancies. Less than 1% of all tumors metastasize to the maxillofacial region and the mandible is the most commonly affected bone. In adults, metastatic breast cancer (64%) and lymphoma (14%) account for most cases of NCS^[3] and acute lymphoblastic leukemia accounts for a significant number of cases in children.

Signs and symptoms

Sensory neuropathy is usually unilateral, but when neoplasia is the cause bilateral involvement may occur in 10% of cases. Pain and swelling may be present in cases of locally destructive processes such as malignancy or infection. Percussion-induced pain, loosening of teeth, presence of sequestra, and mobility of fractured segments are additional symptoms that may be seen in these patients, depending upon the etiology. If metastatic malignancy is present, constitutional symptoms may be seen. Lossus and Sigel found a metastatic etiology in 89% of their patients with NCS.^[3] When the mandible is involved by metastasis, the primary tumor is commonly seen in the breast, lung, kidney, ovary, or testes.^[4] In 47% of cases MNN precedes the diagnosis of the primary tumor, while in patients with a history of cancer MNN often indicates recurrence of disease.^[3] In one large study of NCS patients, 50% of patients had mandibular metastasis and 36% had either a tumor mass or leptomeningeal involvement in the area of the trigeminal root.^[3] Thus, MNN is most often a forerunner to malignancy progression and relapse, but it may also precede the diagnosis of cancer. Patients with AIDS may develop NCS because of the presence of high-grade lymphoma involving the central nervous system or the mandible.

Pathophysiology

Compression, retraction, or even severance of the mandibular nerve or its branches are responsible for the cases of NCS that result from local dental causes. The exact mechanisms underlying NCS in malignant diseases are not well known and a number of hypotheses have been proposed, such as direct compression of the nerve; perineural invasion; leptomeningeal seeding; paraneoplastic syndrome; and invasion of the dura mater, cavernous sinus, or skull base by the malignancy, which then involves the trigeminal ganglion.^[5]

Diagnosis

Clinical diagnosis is largely based on a detailed case history along with thorough systemic and local examination. Panoramic radiography is a useful starting point. Computerized tomography (CT) scan of the head and neck should be done if possible. Nuclear bone scintigraphy (bone scanning) may also help in identifying mandibular bone disease such as metastasis or osteomyelitis. FNAC and biopsy of the lesion are essential for confirmation of the diagnosis. Lumbar puncture and cytologic analysis of cerebrospinal fluid can be of use in cases where radiography is negative. [Downloaded free from http://www.ijdr.in on Tuesday, July 24, 2012, IP: 125.16.60.178] || Click here to download free Android application for this journal

Numb chin syndrome

Management

Identification of the underlying pathology and its treatment is the basic principle for management of these cases. In NCS resulting from dental etiologies (e.g., after enucleation of a cyst or tumor), the prognosis is usually good. Treatment of any systemic etiology helps in resolution of NCS. Where NCS is caused by metastasis, the prognosis is grim, the average survival rate after diagnosis being approximately 5 months if caused by mandibular metastasis and 12 months if leptomeningeal metastasis is present. Treatment does little to affect the outcome in these cases.

Conclusion

MNN is mostly a benign symptom; however, any numbness in the lower lip or the mental or gingival region must alert the dentist to the possibility of serious disease. The etiology must be established definitely, using the necessary diagnostic aids to rule out any metastatic malignant disease.

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