**Original Article**

# Oral and Maxillofacial Manifestations of Neurological Diseases in Southern Provinces of Iran

**Amanat D.**[a]

[a] Dept. of Oral Medicine, School of Dentistry, Shiraz University of Medical Sciences, Shiraz, IRAN

**KEY WORDS**
Neurological diseases; Orofacial manifestations; Southern Provinces of Iran

***Received Dec.2011;***
***Received in revised form Dec.2011;***
***Accepted Jan.2012***

**ABSTRACT**

**Statement of Problem:** The central nervous system and oral cavity have a close anatomical location. Recognition of the orofacial manifestations of neurological diseases is in direct relation with the responsibilities of a dentist.

**Purpose:** The objective of this study was to familiarize dentists and even physicians with orofacial manifestations of related neurological diseases.

**Materials and Method:** This cross- sectional study was performed on 1284 patients with known cases of neurological diseases referred from all over the Southern Provinces of Iran to Oral Medicine Department of Shiraz Dental School from 1997 to 2010. Orofacial manifestations of 34 different types of neurological diseases were classified in four main groups of cranial nerve disorders, main neurological disorders, neoplastic and tumoral neurological lesions and congenital anomalies of neurological disease. These groups were evaluated with respect to age, sex, signs and symptoms, clinical appearance and location of the lesions. Data were analyzed by using SPSS software version 15, by using relative frequency and frequency quotients.

**Results:** Orofacial manifestations were shown in 532 patients of the first group (41.48%), 386 patients of the second group (30.11%), 243 patients of the third group (18.93%) and 123 patients of the fourth group (9.48%), respectively.

**Conclusion:** According to the results of this 14 years study, cranial nerve disorders showed most of oral signs and symptoms followed by main neurological disorders, neoplastic and tumoral lesions, and congenital anomalies of neurological diseases respectively. So, dentists should be familiar with these manifestations to have a better recognition, diagnosis and correct decision upon treating these manifestations in such patients.

*** Corresponding author.** Amanat D., Address: Department of Oral Medicine, School of Dentistry, Shiraz University of Medical Sciences, Shiraz, IRAN; Tel: 0711-6263193-4; Fax: 0711-6270325; Email: amanatd@sums.ac.ir

## Introduction

The central nervous system (C.N.S) and oral cavity have a close anatomical location. They have common vascular supply and a related embryological origin [1]. Motor and sensory organs in orofacial area have numerous connections with C.N.S. via the cranial nerves [2]. As yet, diseases of C.N.S. have been studied in different branches of medicine such as otology, Ophthalmology, neurology, neurosurgery and dentistry [3]. Recognition of orofacial manifestations of C.N.S. is in direct relation with the responsibilities of a dentist.

Classification and defining of a unique methodology for recognition of the relationship between neurology and dentistry is not easy, because of large volume of data and multiple aspects of them. So, the information selected, should be in a particular directed manner which considers 1) congenital anomalies disturbances of development and growth including phakomatosis and gross head and neck anomalies, 2) neoplasia with neural origin involving orofacial regions, 3) diseases

of cranial nerves and 4) main neurological disorders with orofacial manifestations and considerations [3].

Neurocutaneous syndromes (phakomatosis) include four important diseases: Sturge- Weber syndrome, tuberous sclerosis, neurofibromatosis of Von Recklinghausen and incontinentia pigmenti [4]. These diseases although unnoticeable, have important role in neurology. Their specific importance is that of direct relationship to oral cavity and facial structures [5]. Gross head and neck anomalies are rare and Crouzon and Apert syndrome are two important syndromes of these anomalies having more incidence and neural- oral manifestations [6].

Orofacial area may be affected by nervous system neoplasias in different ways [7]. Three more possible conditions include 1) neoplasias of orofacial nerves and their sheaths (e.g. neurofibroma, neurolemmoma, traumatic neuroma, malignant schwannoma), 2) cranial nerve tumors with orofacial affections (e.g. trigeminal neurinoma, acoustic neurinoma, and olfactory neuroblastoma), and 3) systemic tumors with significant neural and orofacial symptoms (e.g. Gardner syndrome and multiple endocrine neoplasia llb) [7-11].

Cranial nerves V, VII, IX, X and XII disorders are important for a dentist, because all sensory and motor functions in the orofacial region are under the control of these nerves. Bell's palsy, trigeminal neuralgia, vagoglosso-pharyngeal neuralgia, spheno- palatine neuralgia, post herpetic neuralgia, atypical facial pain, hypoglossal nerve paralysis and auriculotemporal syndrome are among common cranial nerve disorders [3,8-9, 12].

Main common neurological disorders with orofacial manifestations include epilepsy, cerebro- vascular accident, multiple sclerosis, Parkinson's disease, temporal arteritis, Guillain- Barr syndrome, Myasthenia Gravis, Huntington Chorea disease and orolingual paresthesia [3, 8-9, 13-17].

Various researches, up to the present time, have studied one or two orofacial manifestations of specific neurological disorders mostly as a case- report. They have not investigated these manifestations in a vast majority of neurological disorders. This present article, seems to be at least the first kind of its own in Iran which studies orofacial manifestations of almost altogether related neurological disturbances with respect to age and sex of the patients. This fourteen years survey tries to make a logical and understandable connection between neurology and dentistry by revealing orofacial manifestations of neurological diseases, enabling dentists to be more familiar with such a connection in order to be capable of better diagnosis of these manifestations in relation to neurological disorders.

**Materials and Method**

Orofacial manifestations of 34 different neurological disorders were evaluated in this cross- sectional study. A total number of 1284 patients (617 males and 667 females) who were medically diagnosed with neurological disorders and had orofacial manifestations were evaluated for these manifestations with respect to age, sex, signs and symptoms, and clinical appearance as well as location of lesions. These patients had an age range of six months to eighty-five years old that referred from all over the Southern Provinces of Iran, to Oral Medicine Department of Shiraz Dental School for their orofacial lesions between February 1997 to April 2010.

Patients who had other systemic diseases and/or oral disturbances interfering with oral manifestations of neurological diseases as well as those with suspicious oral signs and symptoms and clinical appearances were excluded from the study.

Because of large volume and multiple aspects of data collected, classification and defining of a unique methodology was not easy. Hence, for the sake of simplicity, and in order to have almost all possible connections, these 34 neurological diseases were classified in four main categories, each of which containing its related diseases and subgroups. These four categories consist of cranial nerves V, VII, IX and XII disorders, main neurological disorders, neoplastic and tumoral neurological lesions and congenital anomalies of neurological diseases. Their related diseases are shown in table 1 through table 4. Due to the main objective of this study, each patient was exactly examined clinically for the type and characteristics of his or her orofacial manifestations. No radiography or photography was required to be taken from the patients for the purpose of this article.

According to the three main variables of this study, namely, location, signs and symploms and

**Table 1** Orofacial Manifestations in Cranial Nerves V, VII, IX, X and XII Disorders *

| Signs and symptoms | Location | Age range | % in this group | % in all groups | Male | | female | | Total number of patients |
|---|---|---|---|---|---|---|---|---|---|
| | | | | | No. | % | No. | % | |
| Loss of taste | Oral cavity | 21-69 | 11.47 | 4.76 | 24 | 4.51 | 37 | 6.96 | 61 |
| Cluster headache | Head and neck | 22-48 | 12.41 | 5.15 | 47 | 8.83 | 19 | 3.57 | 66 |
| Paresthesia | Oral cavity | 40-79 | 10.33 | 4.29 | 18 | 3.38 | 37 | 6.96 | 55 |
| Inability to eat or speech | Oral cavity | 18-67 | 6.40 | 2.66 | 18 | 3.38 | 16 | 3.05 | 34 |
| Constant unilateral pain | Head and neck | 15-85 | 59.39 | 24.62 | 150 | 28.20 | 166 | 31.20 | 316 |
| Total | --- | 15-85 | 100 | 41.48 | 257 | 48.30 | 275 | 51.70 | 532 |

*** Name of the diseases in this group:** Bellś palsy, Trigeminal neuralgia, vago glossopharyngeal neuralgia, spheno palatine neuralgia, post herpetic neuralgia, causalgia, hypoglossal nerve paralysis, auriculotemporal syndrome, Amat syndrome, atypical facial pain.

clinical appearance of oral lesions, diagnosis was based on visualization (for location of lesions), taking case-on visualization (for location of lesions), taking case-history and visualization (for signs and symptoms), and visualization, case-history and physical examination (for clinical appearance) as well as radiography for diagnosis of central bony lesions and histo pathology (for just oral lesions of congenital anomalies) diagnosis of these main variables was done by the author who is a specialist in oral medicine and final confirmation of some diagnoses (e.g. cluster headache and loss of taste) was done by cooperation of a neurologist.

Data with respect to age, sex, signs and symptoms and clinical appearance as well as location of lesions were collected for each patient. Informed consent was obtained from all the patients assuring them that name and / or address of them will be remained confidentially.

#### Statistical Analysis

Data obtained, were analyzed by using SPSS software version 15 and descriptive results were shown in related tables including relative frequency and frequency quotients of each group of neurological diseases.

## Results

In first group (cranial nerve disorders), 257 males and 275 females with the age range of 15-85 years, comprising 41.48% of all patients , showed loss of taste (11.47%) , cluster headache (12.41%), paresthesia (l0.33%), inability to eat or speech (6.40%), and constant unilateral pain (59.39%) (Table 1). In main neurological disorders an the second selected group, 174 males and 212 females with the age range of 22-68 years, comprising 30.11% of all patients, showed dysphagia (19.95%), seizure (12.18%), ocular disturbances (17.10%), weakness of facial and jaw muscles (15.80%) and pain and burning sensation (34.97%) (table 2). In neoplastic and tumoral neurological lesions as the third group, 126 males and 117 females with the age range of 6 months- 74 years, comprising 18.93% of all patients, showed multiple nodules or neuromas (34.16%), central bony lesions (5.77%), tumors (11.11%), papules (17.28%) and single nodules (31.68%) (table 3).

In congenital anomalies of neurological diseases as the fourth group, 60 males and 63 females with the age range of 6-55 years, comprising 9.48% of all patients, showed angioma (15.45%), hemangioma (20.32%), fibroma (8.14%), nodules (32.52%), and neurofibroma (23.57%) (table 4).

Description of details of oral manifestations of these neurological diseases was not the purpose of this article. This study has tried to show oral manifestations of related neurological disorders for two main purposes: First, if a dentist observes such oral manifestations in a patient with no known other related etiological factors, he or she could be reminded of these neurological disorders as the causative agents. Second, if dentists will be confronted with patients with known cases of neurological diseases, they will know about these oral manifestations for better diagnosis of them in relation to these patients.

## Discussion

Orofacial manifestations of neurological diseases have direct relation with dentistry, therefore, dentists should be familiarized with those common diseases between neurology and dentistry [1, 18]. The overlap of these two specialties has not received enough attention but in daily clinic, recognizing of them is very important and unfamiliarity with them may be troublesome [3]. That is why in this study, out of many neurological disorders, 34 diseases with

**Table 2** Orofacial Manifestations in Main Neurological Disorders *

| Signs and symptoms | Location | Age range | % in this group | % in all groups | Male | | Female | | Total number of patients |
|---|---|---|---|---|---|---|---|---|---|
| | | | | | No. | % | No. | % | |
| Dysphagia | Oral cavity | 37- 48 | 19.95 | 6.01 | 31 | 8.03 | 46 | 11.92 | 77 |
| Seizure | Face | 27- 53 | 12.18 | 3.67 | 19 | 4.92 | 28 | 7.25 | 47 |
| Ocular disturbances | Eye | 22- 46 | 17.10 | 5.15 | 39 | 10.11 | 27 | 6.99 | 66 |
| Weakness of facial and jaw muscles | Face | 25- 51 | 15.80 | 4.76 | 26 | 6.74 | 35 | 9.07 | 61 |
| Pain and burning sensation | Face | 26- 68 | 43.97 | 10.52 | 59 | 15.28 | 76 | 19.69 | 135 |
| Total | --- | 22- 68 | 100 | 30.11 | 174 | 45.08 | 212 | 54.92 | 386 |

*** Name of the diseases in this group:** Epilepsy (grandmal), multiple sclerosis, Parkinson's disease, Myasthenia Gravis, Guillain- Barr syndrome, Huntington Chorea, Amylotrophic lateral sclerosis, Riley- Day syndrome, orolingual paresthesia, temporal arteritis

orofacial manifestations clinically subdivided to four major categories, have been evaluated for these manifestations. Worldwide research in this area in a broad- sense is not too many and most of them are case reports [14, 16-17, 19]. In Iran, this article with respect to its volume of data, happens to be the first kind of its own and results obtained from this research, seem to be a good source to increase the knowledge of dentists and even other physicians regarding orofacial manifestations of related neurological diseases.

Cranial nerves V, VII, IX, X and XII disorders as a category, are important for a dentist because all sensory and motor functions in the orofacial region are under the control of these nerves [3, 18].

G.C.S Cousin describes development of Bellś palsy as a progressive lpselateral facial weakness and hyperaccusis in a- 38- year old man after injection of local anesthesia [19]. Rabinovich A., et al describes trigeminal neuralgia in a- 52- year- old woman with intense left-sided facial pain [20].

Karunananthan et.al reported an unusual case of bilateral Frey syndrome in a woman having facial flushing upon food mastication [21].

In main neurological disorders category, Gayatri NA., et.al, reported progressive bulbar dysfunction in a-6-year- old boy with epilepsy [22].

Daisy Chemaly et.al reported numbness of left side of face in a-38-year old woman with multiple sclerosis [23]. Durham TM et al described the use of a bruxism splint to reduce orofacial pain in a patient with parkinsonś disease [24].

In neoplastic and tumoral neurological lesions category, Shih- Chieh Chuang et al showed neurilemmoma of tongue in a -21 -year old man [25].

Shah SS., et al, showed intraosseous traumatic neuroma of the maxilla after excision of giant cell granuloma [26]. Halachmi S et al, described trigeminal neurinoma in maxillary sinus of a patient with Numb chin syndrome [27]. Ribas M et al, described multiple impacted teeth with adenomatous lesion in mandible of a patient with Gardnerś syndrome [28].

In congenital anomalies category, Freitas TM, et al, showed gingival hemangiomatosus lesion restricted to the epsi lateral maxilla and mandible in a- 34- year old man with Sturge- Weber syndrome [29]. Lopez J, described fibroma in lower lip and both cheekś mucosa in a patient with tuberous sclerosis [30]. Minic S, et al, reported peg like teeth, delayed eruption of teeth, partial anodontia and gothic palate in 25 patients with incontinentia pigmenti [31].

Steven L., et al, described marked mandibular

**Table 3** Orofacial Manifestations of Neoplastic and Tumoral Neurological lesions *

| Clinical appearance | Location | Age range | % in this group | % in all groups | Male | | Female | | Total number of patients |
|---|---|---|---|---|---|---|---|---|---|
| | | | | | No. | % | No. | % | |
| Multiple nodules (neuromas) | Face | 21- 45 | 34.16 | 6.47 | 46 | 18.93 | 37 | 15.23 | 83 |
| Central bony lesions | Mandible | 26-53 | 5.77 | 1.09 | 8 | 3.29 | 6 | 2.47 | 14 |
| Tumor | Maxilla | 6 months- 32 years | 11.11 | 2.10 | 16 | 6.58 | 11 | 4.53 | 27 |
| Papules | Face | 52-74 | 17.28 | 3.27 | 24 | 9.88 | 18 | 7.41 | 42 |
| Single nodules | Lip and tongue | 30- 64 | 31.68 | 6.00 | 32 | 13.17 | 45 | 18.51 | 77 |
| Total | --- | 6 months- 74 years | 100 | 18.93 | 126 | 51.85 | 117 | 48.15 | 243 |

*** Name of the diseases in this group:** Multiple Endocrine Neoplasia (MEN llb), Neurilemmoma, traumatic neuroma, neurofibrosarcoma, melanotic neuroectodermal tumor of infancy, palisaded encapsulated neuroma of oral mucosa (PEN), neuroendocrine carcinoma, olfactory neuroblastoma

**Table 4** Orofacial Manifestations of Congenital Anomalies of Neurological Diseases *

| Clinical appearance | Location | Age range | % in this group | % in all groups | Male | | female | | Total number of patients |
|---|---|---|---|---|---|---|---|---|---|
| | | | | | No. | % | No. | % | |
| Angioma | Gingiva | 16-55 | 15.45 | 1.33 | 8 | 6.51 | 11 | 8.94 | 19 |
| Hemangioma | Oral cavity | 11-49 | 20.32 | 1.96 | 11 | 8.94 | 14 | 11.38 | 25 |
| Fibroma | Buccal mucosa | 6-14 | 8.14 | 0.79 | 6 | 4.88 | 4 | 3.25 | 10 |
| Nodules | Oral cavity | 12-39 | 32.52 | 3.13 | 22 | 17.89 | 18 | 14.63 | 40 |
| Neurofibroma | Face | 16-44 | 23.57 | 2.27 | 13 | 10.57 | 16 | 13.01 | 29 |
| Total | --- | 6-55 | 100 | 9.48 | 60 | 48.79 | 63 | 51.21 | 123 |

*** Name of the diseases in this group:** Sturge- Weber syndrome, tuberous sclerosis, Apert syndrome, VonRecklinghausen neurofibromatosis1 (NFI), incontinentia pigmenti, Crouzon syndrome.

prognathism with maxillary retrusion and hypoplastic molar processes in a family with Crouzon syndrome [32]. All above mentioned name researches and many other ones being done in different countries, reported one or two orofacial manifestations of only one neurological disease mostly as case- reports. The unique difference of this study with other ones is that this article with its collective data obtained in fourteen years, describes different orofacial manifestations of related neurological diseases in a broad spectrum. This present study which is at least the first kind of its own in Iran, familiarizes dentists and even other physicians with these manifestations, enabling them to have better diagnosis upon treating these manifestations in such patients.

## Conclusion

Oral manifestations of some of the neurological diseases show a logical and understandable connection of these diseases with dentistry. According to the data of this fourteen years study, at least 34 different neurological disorders classified into four major categories based on neural-oral manifestations manifested oral signs and symptoms and clinical lesions.

The data obtained in this study, showed cranial nerve disorders had most of oral signs and symptoms followed by main neurological disorders, neoplastic and tumoral lesions, and congenital anomalies of neurological diseases respectively. So, to familiarize dentists and even other physicians with theses manifestations is important for them to have a better recognition, diagnosis and correct decision upon treating these manifestations in such patients.

## References

[1] Bricker SL, Langlaris RP, Craig S. Oral Diagnosis, Oral Medicine and treatment Planning. 2nd ed., BC Decker Inc: Hamilton, Ontario; 2002. p. 325-360.

[2] Friedman MH. Atypical facial pain: the consistency of ipsilateral. Maxillary area tenderness and elevated temperature. J Am Dent Assoc 1995; 126: 855-860.

[3] Fauci AS, Braunwald E, Isselbacher KJ, Wilson JD, Martin JB, Kasper DL, et al. Harrison's Principles of Internal Medicine. 14th ed., Mc Graw-Hill companies, Inc: United States of America; 1998. p. 2307-2451.

[4] Smirnioto poulos JG. The phakomatosis tuberous sclerosis complex. A JNR 1992; 13: 732-737.

[5] Langmore SE, Lehman ME. Physiologic deficits in the orofacial system underlying dysarthria in amyotrophic lateral sclerosis. J Speech Hear Res 1994; 37: 28-37.

[6] Neville BW, Damm DD, Allen CM, Bouquout JE. Oral & Maxillofacial Pathology. 2nd ed., Philadelphia: W.B.Saunders Co.; 2002. p. 40-41.

[7] Abell J. Tumors of the peripheral nervous system. Human pathol.1985; 1: 530.

[8] Shafer WG, Hine MK, Levy BM. Text book of Oral Pathology. 4th ed., Philadelphia: W.B. Saunders Company; 1983. p. 854-877.

[9] Greenberg MS, Glick M, Burket's Oral Medicine Diagnosis and Treatment. 10th ed., BC Decker Inc. Hamilton, Ontario; 2003. p. 592-604.

[10] Carney JA. Psammomatous melanotic schwannoma. A distinctive, heritable tumor with special associations, including cardiac myxoma and the Cushing syndrome. Am J Surg Pathol 1990; 14: 206-222.

[11] Wood NK, Goaz PW. Differential Diagnosis of Oral and Maxillofacial lesions. 5th ed., Mosby-Year book, Inc.: United States of America; 1997. p. 329.

[12] Rushton B. Glossopharyngeal neuralgia. Arch Neurol 1982; 38: 201.

[13] Little JW, Falace DA, Miller CS, Rhodus NL. Dental Management of the medically compromised Patient. 6th ed., Mosby, Inc.: United States of America; 2002. p. 417-438.

[14] Jacobs L, Kaba S, Pullicino P. The lesion causing

continuous facial myokymia in multiple sclerosis. Arch Neurol 1994; 51: 1115-1119.

[15] Forrest K, Weismer G. Dynamic aspects of lower lip movement in parkinsonian and neurologically normal geriatric speakers' production of stress. J Speech Hear Res 1995; 38: 260-272.

[16] Kini PG. Juvenile myasthenia gravis with predominant facial weakness in a 7-year-old boy. Int J Pediatr Otorhinolaryngol 1995; 32: 167-169.

[17] De Assis JL, Marchiori PE, Scaff M. Atrophy of the tongue with persistent articulation disorder in myasthenia gravis: report of 10 patients. Auris Nasus Larynx 1994; 21: 215-218.

[18] Amanat D, Yassami S. Orofacial manifestations of neurological diseases (dissertation). Shiraz University of Medical Sciences: School of Dentistry; May 1997. p. 35-56.

[19] Cousin GC. Facial nerve palsy following intra-oral surgery performed with local anaesthesia. J R Coll Surg Edinb 2000; 45: 330-333.

[20] Rabinovich A, Fang J, Scrivani S. Diagnosis and Management of Trigeminal Neuralgia. Columbia Dental Review 2000; 21: 4-7.

[21] Karunananthan CG, Kim HL, Kim JH. An unusual case of bilateral auriculotemporal syndrome presenting to an allergist. Ann Allergy Asthma Immunol 2002; 89: 104-105.

[22] Gayatri NA, Hughes MI, Clarke MA, Martland TR. Epilepsy with reversible bulbar dysfunction. Dev Med Child Neurol 2002; 44: 770-772.

[23] Chemaly D, Lefrançois A, Pérusse R. Oral and maxillofacial manifestations of multiple sclerosis. J Can Dent Assoc 2000; 66: 600-605.

[24] Durham TM, Hodges ED, Henry MJ, Geasland J, Straub P. Management of orofacial manifestations of Parkinson's disease with splint therapy: a case report. Spec Care Dentist 1993; 13: 155-158.

[25] Shih-Chieh Chuang. Neurilemmoma of Tongue-A Case Report. Tzu Chi Med J 2000; 12: 73-76.

[26] Shah SS, Ghannoum J, Carness A, Freedman PD. Intra-osseous traumatic neuroma of the maxilla after excision of giant cell granuloma: a case report. J Oral Maxillofac Surg 2004; 62: 1161-1164.

[27] Halachmi S, Madeb R, Madjar S, Wald M, River Y, Nativ O. Numb chin syndrome as the presenting sympt-om of metastatic prostate carcinoma. Urology 2000; 55: 286-289.

[28] de Oliveira Ribas M, Martins WD, de Sousa MH, de Aguiar Koubik AC, Avila LF, Zanferrari FL, et al. Oral and maxillofacial manifestations of familial adenomat-ous polyposis (Gardner's syndrome): a report of two cases. J Contemp Dent Pract 2009; 10: 82-90.

[29] Freitas TM. Sturge-Weber syndrome: case report with oral manifestations. Odontalagia Clin Cientif Recife Maio/ago 2004; 3: 143-146.

[30] López-López J, Rodríguez-de-Rivera-Campillo E, Marques-Soares MS, Finestres-Zubeldia F, Chimenos-Küstner E, Roselló-Llabrés X. Tuberous sclerosis and its oral manifestations. A clinical case. Med Oral 2004; 9: 216-223.

[31] Minić S, Novotny GE, Trpinac D, Obradović M. Clinical features of incontinentia pigmenti with emphasis on oral and dental abnormalities. Clin Oral Investig 2006; 10: 343-347.

[32] Steven L. Dentofacial features of a family with Crou-zon syndrome. Case reports. Australian Dental Journal 1997; 42: 11-17.