CASE REPORT

Nerve afflictions of maxillofacial region: a report of two cases

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Neurological disorders and conditions affecting the maxillofacial region result in disabilities that affect an individual's functioning. Sensory or motor disturbances of the nerves may be caused by trauma, infections, pressure effect or infiltration by tumours or other health conditions. Two rare cases of nerve afflictions are described here with their typical clinical features. The first case had an involvement of maxillary, mandibular and ophthalmic divisions of the trigeminal nerve (sensory) due to herpes zoster infection in a very young patient and the second case had a unilateral isolated hypoglossal nerve palsy (motor) secondary to infiltration of the nerve by carcinoma of pyriform fossa.

BACKGROUND

Neurological apparatus of the maxillofacial region is unique in many respects as they perform the vital processes of feeding, sensory perception, respiratory activity and external communication by means of facial expression and speech.¹ As the maxillofacial region is the most vulnerable to various pathologies and injuries, it is not surprising that the maxillofacial neurological apparatus is a prime target for characteristic pathology involving head and neck regions. We presented two rare cases of nerve afflictions affecting the maxillofacial region presenting with oral manifestations.

CASE PRESENTATION**Case 1**

A teenage boy reported to our department with pain for the past 10 days in his upper right posterior teeth. He had developed fever 2 weeks prior to the visit after which he had developed vesicular eruptions on the right side of his face and multiple painful ulcerations in the corresponding cheek mucosa. He had no history of any topical application, insect bite, drug intake or contact with persons with chicken pox. He had no lesions in other parts of the body.

Extraorally there was presence of a diffused swelling over the right side of face leading to closure of right eye. Erythema and crusted lesions were present all over the right side of the face including the infra-orbital region, cheek, upper lip, and right side of the nose and the tip of the nose (figure 1). Right submandibular lymph nodes were enlarged and tender. An examination of the oral cavity showed multiple irregular shallow ulcerations involving the right upper labial mucosa and hard palate which stopped abruptly at midline (figure 2). Ulcers were also seen on the mandibular attached gingiva, posterior buccal

mucosa and buccal vestibule on the right side. The ulcers were covered by white necrotic slough. Ophthalmological examination revealed ocular inflammation, slight visual loss and debilitating pain with crusted lesions in the eyes.

Diagnosis

Hence a clinical diagnosis of herpes zoster involving maxillary, mandibular and ophthalmic division of the right trigeminal nerve (V1, V2 and V3) was made.

Investigations

Complete blood count and ELISA for HIV helped to rule out most of the conditions causing immune suppression; however, no specific test was performed to rule out hereditary immunodeficiency conditions.

Treatment and follow-up

The patient was treated with the tablet acyclovir 400 mg five times a day for 7 days. Supportive treatment was also given and ophthalmology consultation was sought for eye lesions. On telephonic conversation with the patient after 1 month, he reported to have completely recovered, but could not come for a follow-up appointment.

Figure 1 Herpes zoster infection involving ophthalmic and maxillary division of trigeminal nerve. With crusted lesions on the malar region of the face on the right side, involving the tip of the nose, right upper lip and the swollen right eyes.

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Figure 2 Multiple irregular shallow ulcerations over right side of upper labial mucosa and hard palate which stopped abruptly at midline.

Case 2

A male patient in his early 40s, a known case of carcinoma of left pyriform fossa with left-sided neck swelling (figure 3) was referred to our department from oncology, for oral prophylaxis prior to radiation therapy. He had been operated for the same 2 years ago but it had recurred. Intraoral examination revealed an unusual deviation of the tongue to the left side on protrusion and the left half of the tongue appeared shrunken (figure 4).

Diagnosis and follow-up

This was attributed to left hypoglossal nerve palsy (HNP) secondary to infiltration of the nerve by carcinoma of the left pyriform fossa. On completion of radiotherapy for the malignancy, the patient could protrude his tongue without any deviation.

Figure 3 Carcinoma of left pyriform fossa with swelling on the left side of the neck.

DISCUSSION

Major cranial nerves innervating the maxillofacial region are V—trigeminal nerve, VII—facial nerve, IX—glossopharyngeal nerve and XII—hypoglossal nerve. Nerve afflictions of maxillofacial region may be due to direct injury to the nerve, narrowing of the skull base foramina, viral lesion of nerve ganglion, compression or infiltration of the nerve by tumours, nerve involvement in systemic diseases and various intrinsic pathologies of the nerve. The motor nerve involvement of the maxillofacial region results in either paralysis or spasm and the sensory nerve involvement manifests as pain of the involved site. The pain produced may be symptomatic having a definitive organic origin or it may be idiopathic without any organic origin.

The most common viral lesion affecting the nerve ganglion is the herpes zoster infection (HZI) (Shingles). Herpes zoster is an acute neurodermic viral infection of the dorsal root ganglia of the spinal cord or the extramedullary cranial nerve ganglia.² HZI occurs during the lifetime of about 10–20% individuals.³ HZI affects middle-aged and elderly individuals. The prevalence of attack increases with age. From 5 to 10 cases/1000 persons

Figure 4 Hypoglossal nerve palsy demonstrated clearly on protrusion of his tongue, it deviated to left side with atrophy of tongue seen on left side.

are seen between the sixth and eighth decades of life.⁴ Fewer than 5% of attacks occur in persons <10 years of age.^{5 6} Approximately 8% of patients with HZI have an underlying disease, such as HIV, diabetes mellitus, leukaemia, Hodgkin disease or other malignancies.⁴ Attacks may also be precipitated by surgical stress, immunosuppressive therapy, physical trauma or radiation therapy.⁵ Characteristically, the condition presents as a painful unilateral vesicular rash, usually restricted to the distribution of a sensory nerve. The thoracolumbar trunk (especially T3–L3) is most commonly affected.⁶ HZI may affect cranial nerves, more commonly the trigeminal nerve (18.5%–22% of total cases).⁶ It typically manifests as a vesicular rash in a unilateral dermatomal distribution associated with pain. The ophthalmic division of the trigeminal nerve is commonly affected. The skin manifestations of *herpes zoster ophthalmicus* strictly obey the midline with involvement of one or more branches of the ophthalmic division of the trigeminal nerve, namely the supraorbital, lacrimal and nasociliary branches. Because the nasociliary branch innervates the globe, the most serious ocular involvement develops if this branch is affected. Classically, involvement of the tip of the nose (Hutchinson's sign) has been thought to be a clinical predictor of ocular involvement which was clearly demonstrated in our patient. Oral manifestations appear when the second or third division is affected. A rare form without vesicular eruption also has been documented.⁷ As in our first case, there was involvement of all three divisions of the trigeminal nerve, complete blood count and ELISA for HIV helped to rule out most of the conditions causing immune suppression; however, no specific test was performed to rule out hereditary immunodeficiency conditions. Also the patient was 16 years old, it is very rare for a HZI to occur in this age group. This disease has three phases—prodrome, acute and chronic. The prodrome phase is usually associated with intense pain, sensitive teeth and otitis media. In the acute phase clusters of vesicles on the erythematous base appear along the path of the affected nerve and terminate at midline. The vesicles gradually become pustular and break open to form ulcers. In the chronic phase the lesions heal but the neuralgic pain continues for a long time (postherpetic neuralgia). Without treatment, symptoms usually resolve over several weeks to a month. However, up to 20% of patients may experience a prolonged and debilitating sequelae, chiefly postherpetic neuralgia. Other complications include motor nerve palsy, optic neuropathy, blindness, encephalitis and cutaneous calcinosis.⁸

Head and neck tumours such as orbital tumours, paranasal sinus tumours, rhabdomyosarcomas, esthesioneuroblastomas, infratemporal fossa tumours and those involving the skull base can result in the compression or infiltration of the cranial nerves situated in that area. One such instance has manifested in our second case, where due to the pressure effect of the tumour in the pyramidal fossa there resulted in a same-sided HNP in the patient. The hypoglossal nerve is a pure motor nerve, which supplies the somatic musculature of the tongue. It arises as a series of rootlets that issue from the medulla between the pyramid and the inferior olivary complex. The nerve leaves the skull through the hypoglossal foramen and innervates the genioglossus muscle, which acts to protrude the tongue; the styloglossus, which retracts and elevates its root; and the hypoglossus, which causes the upper surface to become convex. Cranial nerve palsies are seen in around 13–25% of patients with nasopharyngeal carcinoma at diagnosis.⁹ HNP (cranial nerve XII) is uncommon; damage usually produces contraction of the genioglossus muscle on the affected side resulting in the deviation of

the tongue. The tongue curves slightly to the healthy side at rest, but on protrusion it deviates to the affected side. The denervated side becomes wrinkled and atrophied, and fasciculations and fibrillations can be observed. By pushing the tongue against the cheek, the degree of one-sided weakness can be judged. HNP may be accompanied by other cranial nerve palsies (typically IX, X and XI). The hypoglossal nerve is liable to damage from lesions at the medulla oblongata, cisternal space, skull base, nasopharyngeal space, parapharyngeal space, carotid space and lingual levels. Approximately 49% of all HNPs are caused by malignant tumours.¹⁰ Other causes may include trauma, infarctions, multiple sclerosis, Guillain-Barré syndrome and infections.¹¹ Nuclear/infranuclear lesion results in the deviation of the tongue towards the side of the lesion and atrophy of the tongue musculature and fasciculation. While the supranuclear lesion results in deviation of the tongue away from the lesion with no fasciculation and atrophy of the tongue due to bilateral innervations.^{12 13} Hence, the second case reported is an infranuclear lesion. Moreover, unilateral isolated HNP is either extremely rare due to its complex course and proximity to other cranial nerves and vessels or under-reported.^{10 14} It also represents a diagnostic challenge in everyday clinical practice due to its diverse aetiologies.^{10 14} Hence, it would be worthwhile to report our case with unilateral isolated HNP on the left side secondary to carcinoma of the pyriform fossa.

Learning points

- Knowledge of the anatomy of cranial nerves helps to examine and recognise these nerve afflictions for a proper diagnosis and treatment plan.
- The diagnosis of maxillofacial neurological problems depends on an orderly process of (1) interpreting the symptoms and signs of altered neurophysiology, (2) determining the anatomical localisation of the disease process, (3) understanding the basic pathological processes that exist and (4) when possible, identifying the aetiological or precipitating factors.
- Treatment of these problems is a challenge to practitioners of many disciplines, and effective action may depend on a team approach. However, the dentist is in a position to make a strong contribution to this field because of his extensive knowledge of the maxillofacial symptomatology, anatomy, physiology and pathology.

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