Ramsay-Hunt syndrome complicating osteonecrosis of edentulous maxilla and mandible: report of a rare case

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Abstract Review of literature revealed atleast 30 cases of post herpes zoster osteonecrosis of maxilla or mandible. To our knowledge this is a first reported case of Ramsay-Hunt syndrome with post herpetic neuralgia and post herpes zoster osteonecrosis of edentulous maxilla and mandible. We have briefly reviewed the pathophysiology and management of post herpes zoster osteonecrosis and post herpetic neuralgia.

Keywords Herpes zoster · Ramsay-Hunt syndrome · Osteonecrosis · Maxilla · Mandible · Post herpetic neuralgia

Introduction

Herpes zoster (shingles, zona) is an acute neurodermic viral infection of the dorsal root ganglia of the spinal cord or the extramedullary cranial nerve ganglia [1] or motor portion of the geniculate ganglion of the facial nerve. In cases where the facial nerve becomes infected, the clinical manifestations are hemifacial paralysis and xerostomia. This condition is termed the Ramsay Hunt syndrome [2]. Previous reports have shown that the condition is attributable to reactivation of latent varicella zoster virus in the dorsal root ganglia after an earlier attack of chicken pox [3]. Varicella–zoster virus reactivates with increasing age or immunosuppression of the infected person; however, the biologic mechanisms that underlie the transition from latency to active viral replication are unknown.

Perhaps most common complication of herpes zoster infection is post herpetic neuralgia [4], others include motor nerve palsy, optic neuropathy, blindness, encephalitis, and cutaneous calcinosis [5–8], there are few reports of bony and dental complications by herpes zoster infection, all of which were isolated in a single quadrant. These include devitalized teeth [9,10], abnormal development of permanent teeth [11], internal resorption [12], and spontaneous exfoliation of the

teeth with osteomyelitis of the alveolar bone [13–17].

A review of literature revealed atleast 30 cases of post herpes zoster osteonecrosis of either maxilla or mandible.

The simultaneous appearance of the Ramsay Hunt Syndrome has been discussed only twice previously [18,19].

We are reporting a case of Ramsay-Hunt syndrome with post herpetic neuralgia and post herpes zoster osteonecrosis of both edentulous maxilla and mandible in a 86-years-old male patient.

Case report

A 86-years-old male patient was referred by a private practitioner with complaints of pain during mastication in left maxillary and mandibular segment, difficulty in opening mouth and vertigo since 6 weeks, and decreased hearing from left ear and lower motor neuron facial palsy on the left side since 3 weeks. Patient was non-diabetic, non-smoker and does not abuse alcohol. Patient was on antihypertensive medication since 10 years and negative for syphilis, HIV and HBsAg. There was no history of irradiation, malignancy or trauma in the past. Six week back patient had severe pain and itching on the left side of the face following which vesicles appeared along the course of left maxillary and

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mandibular division of trigeminal nerve with ipsilateral ear involvement. Vesicles ruptured to leave raw areas that healed with hyperpigmentation initially and subsequently got depigmented. Intraorally ulcers were present on the left maxillary buccal sulcus and hemipalate. In addition, there was extensive necrosis of buccal and alveolar mucosa exposing the alveolar bone in the left mandibular region. He was diagnosed clinically as herpes zoster infection with facial nerve paralysis and treated with antiviral drugs for seven days and was referred to department of oral and maxillofacial surgery after 4 weeks for further management.

Extraoral examination revealed hypopigmented scar on the left side of the face not crossing the midline, extending from temporal to symphyseal region (Fig. 1). Blood crusted areas seen on left tragus. Low-grade lower motor neuron facial palsy was present on left side. Intraoral examination revealed edentulous upper and lower arches. Oral hygiene was poor with marked halitosis. There was extensive fibrosis of the labial and buccal mucosa with obliteration of the labial and buccal sulcus in the left maxillary and mandibular region. Depapillation and white scarring of the left half of the tongue not crossing the midline was present (Fig. 2). The alveolar bone was exposed in the mandibular arch from left central incisor to left retromolar

Fig. 1 Showing hypopigmented scar seen in relation to left hemiface along the course of maxillary and mandibular division of trigeminal nerve not crossing midline

Fig. 2 Showing white scarring of left anterior half to the tongue

Fig. 3 Showing completely edentulous mandibular arch with mucoperiosteal necrosis and exposure of yellowish necrotic mandibular left alveolar bone extending from midline to retromolar region with no evidence of pus discharge

Fig. 4 Showing isolated areas of necrosis with exposure of bone in the maxillary left alveolar ridge. Buccal mucosa on the left side is non-elastic

region and was yellowish in color (Fig. 3). In the maxillary arch alveolar bone was exposed in the left premolar and molar region (Fig. 4). There was no evidence of pus discharge. ENT consultation revealed sensorineural hearing loss, vertigo and secondary infection of left ear.

Orthopantomogram showed no sequestrum formation or separation of vital bone from non-vital bone (Fig. 5). In view of herpes zoster infection of left maxillary and mandibular division of trigeminal nerve with ear involvement and facial paralysis, a diagnosis of Ramsay Hunt Syndrome complicated by bimaxillary unilateral alveolar necrosis and post herpetic neuralgia was made. Hematologically patient was stable. Patient had undergone surgical trimming of exposed bone in upper and lower arch under local anesthesia and was started on Amoxicillin 500mg and Metronidazole 400mg thrice-daily postoperatively for one week. Cured

bone was sent for histopathological examination. The report described areas of necrotic bone with inflammatory cells with no evidence of fungal or bacterial colonies (Fig. 6).

Initial treatment resulted in wound dehiscence and treatment repeated after 3 weeks and same antibiotic course repeated. Wound healed satisfactorily.

Discussion

Herpes Zoster Infection (HZI) is predominantly a disease of the middle-aged and elderly. From 5–10 cases per 1000 persons are seen between the sixth and eighth decades of life [20], less than 5% of attack occurs in persons younger than 10 years of age [21,22]. The increasing frequency of the HZI with the age has been suggested to be due to the disappearance of zoster-neutralizing antibodies, which usually

persists for 40 years after the original attack of the chicken pox [21].

Approximately 8% of patient with HZI have an underlying disease such as leukemia, Hodgkin disease, myelomatosis, or carcinomatosis [20]. Attacks also may be precipitated by surgical stress, immunosuppressive therapy, physical trauma and radiation therapy [13,21,23,24]. In the case presented here there is no known predisposing factor.

The trunk (especially T3-L3) and the trigeminal area are most commonly affected [20,25]. Characteristically herpes zoster of the trigeminal nerve distribution manifest as a painful, vesicular eruption of the skin and mucosa innervated by the affected nerve. Oral vesicles usually appears after the skin manifestations. The vesicles rupture and coalesce, leaving mucosal erosions. Crusting and healing follow without subsequent scarring in most of the cases. A rare form without vesicular eruption also has been documented [26].

Hunt first described the syndrome of herpes zoster cephalicus, is a rare complication of shingles. The syndrome, caused by the spread of the virus to facial nerves, is characterized by a viral prodrome followed by intense ear pain, a rash around the ear, mouth, face, neck, tongue, larynx, or buccal mucosa and paralysis of facial nerves. The distribution of the vesicles depends on which sensory fibers are infected. Any of the nerve branches that communicates with the facial nerve may be involved, including cranial nerves V, VIII, IX, and X, and cervical nerve II through IV. In the mildest form, neurologic signs are absent, whereas in severe cases there may be accompanying sensorineural hearing loss, disturbed vestibular function, and even viral encephalitis.

Other symptoms may include loss of taste, dry mouth or eyes. The prognosis is good. However, in some cases, hearing loss may be permanent. Vertigo may last for days or weeks. Facial paralysis may be temporary or permanent.

The actual pathophysiology of osteonecrosis in HZI of the trigeminal nerve remains a contentious subject. Various hypothesis concerning the pathogenesis of the herpes zoster-bone necrosis include:

1. Local vasculitis caused by direct extension of the neural inflammatory process to the adjacent blood vessels. This eventually may cause an infarction of the trigeminal vessels that accompany the trigeminal nerves supplying the jaws [15,27].

2. A generalized infection of trigeminal nerves supplying the periosteum and periodontium is believed to cause avascular necrosis over a larger area [13,14,17]. Garty [16] et al., however claims that a generalized avascular necrosis is not likely, especially in the maxilla, because of its rich vascular supply.
3. Denervation of the bone, which seems unlikely to cause bone necrosis.
4. Systemic viral infection can cause injury to odontoblasts and cause degenerative tissue changes that result in pulp necrosis [28].

Aseptic osteonecrosis represents a stage in which portions of the bone are no longer viable. This phenomenon may be a result of inadequate vascularity of bone. The role of vascular alteration in the development of bone necrosis is further supported by the fact that osteonecrosis usually occurs in the patients with compromised vascularity because of ageing, irradiation, or chronic inflammation. Infection frequently intervenes and septic osteonecrosis, which represents active infection within bone, occurs. Further, it seems reasonable to assume that preexisting pulpal or periodontal inflammatory conditions have the potential to contribute to a greater probability of the tooth exfoliation and bone necrosis.

One of the earliest signs of osteonecrosis of the jaw bones associated with a HZI is spontaneous exfoliation of teeth prior to the overt signs of osteonecrosis. In non-viral osteonecrosis, however, dental pain usually heralds the exfoliation of teeth. This suggests that the periosteal blood flow is altered in the HZI, leading to necrosis of the periodontal ligament and exfoliation of the tooth prior to the development of alveolar necrosis [14]. Management of osteonecrosis consists of surgical trimming of the exposed bone, sequestrectomy and antibiotic coverage. Mintz [29] et al. reported the use of closed nasal vestibular drainage system for the ultimate control of maxillary bone viability.

Most common complication of zoster is Post Herpetic Neuralgia (PHN), which is pain that persists for more than six weeks after the development of rash. Post herpetic neuralgia develops slightly more frequently in women than in men, and it occurs after zoster in a trigeminal distribution [30]. Once post herpetic neuralgia disappears, it does not recur. PHN does not occur before 50 years of age [31]. PHN is believed to be due to a loss of inhibitory sensory function with a selective necrosis of larger trigeminal ganglion cells by zoster viruses [32].

Fig. 5 X-ray OPG does not show separation of vital bone from non-vital bone

Fig. 6 Photomicrograph showing necrotic bone with inflammatory cells with no evidence of fungal or bacterial colonies (10 x 40X hematoxylin and eosin stain)

Accordingly, a mild loss of large fiber fine-tactile senses is often measurable on routine neurosensory examination, especially notable in the loss of vibratory and two-point tactile sensibilities. The lesions effectively induce a partial deafferentation in the trigeminal-brain stem complex. There is other evidence that either the deafferentation itself or the viral lesions may stimulate an upregulation or activation of sympathetic nerve fibers in the lesioned trigeminal nerve distribution, bringing about a sympathetic mediated pain component. Chronic pain may emerge from acute herpes symptoms and is manifest in the trigeminal nerve distributions long after the crusted lesions have disappeared. Patients display allodynia pain in response to fine touch stimulation over the face. Bonica has taught that two-eight stellate ganglion blocks, given in the early weeks of zoster infection, may prevent the emergence of chronic neuralgia [33].

Comprehensive reviews of treatment for acute zoster and PHN have been published [34–37], although no protocol is universally accepted. The acute symptoms are managed aggressively with systemic steroids such as Dosepak cortisone (5 days course of 20 mg cortisol

equivalent per day) and an acyclovir medical regimen (200 to 400 mg four times daily for 7 days) [38]. Medical management of chronic PHN consists of topical or systemic therapies [38]. Topical capsaicin [39] preparations are available. Systemic therapies of choice are tricyclic antidepressants [40], anticonvulsant agents [41], alpha blocking agents [42], and systemic lidocaine [43].

More studies are needed to fully understand the pathophysiology of this condition for the purpose of prevention and effective management. There is also a need for early detection to prevent the complication that has been highlighted. Every case of herpes zoster should be closely followed for at least 6 months to enable early treatment.

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