Introduction

Ramsay Hunt syndrome (RHS) is first described by James Ramsay Hunt in 1907 [1]. It is caused by the reactivation of latent varicella-zoster virus (VZV) that has remained dormant within sensory root ganglia (commonly the geniculate ganglion) of the sensory branch of the facial nerve. Involvement of sensory branch of the geniculate ganglion of facial nerve leads Herpes zoster (HZ) oticus which is also known as RHS. Individuals with decreased cell-mediated immunity resulting from carcinoma, radiation therapy, chemotherapy or human immunodeficiency virus (HIV) infection are at greater risk for reactivation of latent VZV. Physical stress and emotional stress often are cited as precipitating factors. Incidence and clinical severity increases when host immunity is compromised. However it is uncommon to see Herpes zoster oticus in healthy individuals. In this report, we describe the clinical course of a patient who presented with Herpes zoster oticus in the absence of a known immunosuppressive condition. A brief review of literature on this topic is also presented.

Case Report

A previously healthy 65-year old man presented with a 3-days history of vesicular lesions with erythematous base on neck. It was followed by the spread of vesicular eruption to involve external auditory meatus of right side, and pinna over next 2-days. These are fluid filled vesicular eruptions which burst and break down on 2nd-3rd day with subsequent scab formation. There is no facial weakness. On examination, there was no lower motor neuron facial palsy. A neurologic examination revealed no weakness in the marginal mandibular branch of the left facial nerve. There were painful adherent crusts and scabs in left conchae and external auditory meatus (Fig. 1), associated with unclear hearing of left ear. The oral cavity and oropharynx were normal. Ocular examination demonstrated no nystagmus and normal conjunctiva. He has given history of stressful life events (sudden 3 deaths in family) in past 4 months. Review of systems was negative. The patient is not able to tell history of chickenpox during childhood or any recent exposure to it. There was no past history of diabetes, cardiac or pulmonary disease or lymphoma. The patient has not been on immunosuppressive or other medications. There is history of Psoriasis 2 years back showing on and off symptoms but on regular treatment. Patient is alcoholic, habit of tobacco chewing and occasional Pan chewing. On examination, the patient was afebrile (37.4°C). He had vesicles but not pustules, with crusting and swelling, in the distribution of the cervical division of VII (facial) cranial nerve. The left pinna is swollen, tender and red. There was no tympanic membrane involvement. Vesicles and scabs in various stages were also present lateral side of neck, in front of sternum and external ear (Fig. 1) and behind Right pinna (Fig. 2) and there was no lymphadenopathy.
Pulmonary, cardiovascular and abdominal examinations were normal. Complete blood count Hb- 13 gm%, BSL® - 81.5 mg% peripheral smear, routine biochemistry, liver function tests, and chest x-ray were normal. CD4 and CD8 lymphocyte counts were 1.34 × 109/L and 0.61 × 109/L respectively (CD4:CD8 ratio = 2.2). Serology for Human Immunodeficiency Virus (HIV), Hepatitis A, Hepatitis B, and Hepatitis C were negative and RPR was non-reactive.

Skin biopsy from fresh vesicle on the cheek and external ear showed ulceration with acute inflammation, necrosis and intranuclear inclusions within epithelial cells. The direct fluorescence antigen staining from skin lesions was positive for VZV. The patient was started on oral Acyclovir 800 mg every 8 hours. In the next 5 days, the eruption of new vesicles ceased, the rash started to resolve and his clinical status improved.

Figure 1. Vesicles below right pinna and in front of sternum

Figure 2. Vesicles behind right pinna

Discussion
Herpes zoster, also called shingles is the consequence of reactivation of latent VZV from the dorsal root ganglia. It is characterized by unilateral vesicular eruptions within a dermatome. Ramsay Hunt syndrome is a painful rash around the ear that occurs when the varicella zoster virus infects a nerve in the head. In Ramsay Hunt syndrome, the virus is believed to infect the facial nerve near the inner ear. This leads to irritation and swelling of the nerve. Herpes Zoster is seen a disease of older people (> 60 years old). The severity and incidence of the disease is depend on the cellular immunity of the patient [2]. This RHS type 2 is shingles of the geniculate ganglion, means the herpes zoster virus lies dormant in various nerve cells in the body and in check by the patient’s immune system. During an illness that suppresses the immune system, the virus is reactivated and travels to the end of the nerve cell, where it causes the symptoms described above [1]. This complication of zoster has been described in immunocompromised persons (HIV, cancer, patients on immunosuppressive therapy) and reported to be as common as 10%-40% [3,4]. However, Herpes zoster oticus in otherwise healthy persons who are not on immunosuppressive therapy and have no underlying cancer is rare. In our search we could identify less than ten cases with disseminated zoster in otherwise healthy persons [5,6]. Our patient presented with characteristic skin findings of herpes zoster oticus. The skin biopsy findings supported the clinical diagnosis of VZV. In our patient, significant age related depression in cellular immunity with history of psoriasis, alcoholic could have contributed to the dissemination of herpes zoster. Elderly patients should be recognized as a group in whom the risk of HZ oticus is higher than the average immunocompetent host. Patient of VZV without any immunosupression are at risk of infection of visceral organs, particularly lungs, liver and brain. Other complications include corneal ulceration and post herpetic neuralgia [3]. Therefore, identification and aggressive treatment of disseminated herpes zoster infection in elderly immunocompetent hosts is important. The treatment of choice is intravenous Acyclovir 10 mg/kg every 8 hours for 5–7 days. As patient reported earlier without complications, we preferred oral acyclovir for 5 days along with adjuvant drugs.
REFERENCES