Ramsay Hunt Syndrome – 1 with Polyneuritis Cranialis

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ABSTRACT

Ramsay Hunt Syndrome is a well known condition, caused by reactivation of herpes zoster infection in the geniculate ganglion and consists of severe lower motor neuron facial palsy along with a vesicular eruption in external auditory canal and sometimes in the oropharynx. Often the eighth cranial nerve is also affected. Here we report a case of 65 yrs. old man who presented with acuteinfranuclear facial palsy and typical herpes zoster rash over external ear canal along with eight, nineth and tenth cranial nerve palsy on same side which is a very rare entity.

INTRODUCTION :

Herpes Zoster infection is a viral infection caused by Varicella –Zoster virus (VZV), a double stranded DNA virus related to Herpes simplex group. It is characterised by painful rash with blisters in a limlited area on one side of body often in a stripe .The'zoster' is a Greek word meaning belt or girdle like. It is also known as' shingles' meaning cingulum or girdle in Latin.(1)

The initial infection withVaricella -Zoster Virus(VZV) causes acute, short lived illness - Chickenpox which generally occurs in children & young people. Once the episode of chickenpox has resolved the virusis not eliminated from body but can cause herpes zoster, often many years after initial infection. The virus becomes latent in nerve cell bodies & less commonly in non-neuronal satellite cells of dorsal roots, cranial nerves or autonomic ganglion without causing any symptoms. The virus can break out of nerve cell bodies many years after chickenpox infection and travels down nerve axons to cause virus infection of skin in the nerve region (dermatome). The virus may spread one or more ganglia along nerves of affected segments and infect corresponding dermatomes causing painful rash over the skin. Although the rash heals in 2 -4 weeks. Some patients experience residual pain for months to years after called as post- herpetic neuralgia (26%). Exactly how virus remains latent in nerve cell body and subsequently reactivates is not known. Ramsay Hunt Syndrome –1 (also called as Herpes Zoster Oticus or Geniculate neuralgia or Nervusintermedius neuralgia), is a distinct entity which is characterised by lower motor neuron facial palsyin association with typical herpetic rash in the external auditory canal, around the pinna,in mouth in sensory distribution of facial nerve and severe neuralgic pain. The other symptoms are hearing loss, vertigo,tinnitus & taste loss over the tongue.Ramsay Hunt Syndrome-2 , (DyssynergiaCerebellarisMyoclonica)is an autosomal recessive condition, characterised by progressive myoclonic epilepsy with ataxia secondary to mitochondrial encephalopathy.(2)

The risk factors are immunodeficient states secondary to cancer chemotherapy, radiation , diabetes, and HIV infection.The symptoms of meningo-encephalitis can occur in form of headache, neck pain but are uncommon. Sometimes very rarely other cranial nerves can also be affected producing palatal palsy due to involvement of nineth& tenth cranial nerve.(3).

Epidemiology : Throughout the world incidence of herpes zoster infection every year ranges from 1.2 to 3.4 cases per 1000 healthy indivijuals. It increases to 3.9to 11.8 per 1000 presons above the age of 65yrs. The syndrome was first described by James Ramsay

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Hunt in 1907. He defined it as VZV infection of head , neck that involves facial nerve. It accounts for 18%cases of all causes of unilateral facial palsies in adults. It is rare children below 6 yrs of age. It is thought to be the cause of 20% cases of Bell'sPalsy.The incidence of this syndrome among HIV + persons is not exactly known although it may occur at higher rate than generation population. Repeated attacks are rare & it is extremely rare to suffer from >3 recurrences.The infection does not spread frompreson to person. (4)However during the blister phase direct contact with rash can spread VZV to a non-immune person. Until rash develops crusts person is extremely contagious but not before blisters develop or during post-herpetic neuralgia phase. It causes chickenpox in people who have not had it previously. Unless immune system is compromised reactivation of virus does not occur.

The predisposing factors are: 1) Anyone who never had chickenpox 2) Newborn state 3) Pregnancy 4) Patients on immunosuppresants or 5) immunodeficient persons 6) Stress – physical and mental.

Classically in Ramsay Hunt Syndrome facial palsy is associated with vesicular rash of ear or mouth in 80% cases but rash might precede the onset of facial palsy. The onset of severe neuralgic pain usually precedes rash by several hours or days. Acute onset ipsilateral LMN facial palsy/paresis associated with vertigo and hearing loss occur next. Other features like tinnitus, otalgia, headache, gait ataxia, fever & cervical lymphadenopathy are also described. The facial weakness usually reaches to maximum severity by 1 week after onset of symptoms.The other cranial nerves may be involved rarely e.g. VIII, IX, X, V,VI in order of frequency.This infection gives rise to vesiculation& ulceration of external ear canal, ipsilateral anterior 2/3rd of tongue,and soft palate as well as ipsilateral VII neuropathy or radiculopathy or gangliopathy. The short and long term pain caused by herpes zoster virus comes from its widespread growth in infected nerve causing inflammation.(5)

CASE REPORT:

A 65 yrs. male patient, resident of Agargaon, Dist. Wardha, was admitted in our hospital with history of high grade fever of 10 days before admission which lasted for 4 days and development of painful eruptive skin lesions in right external ear &behind the pinna since 6 days before the admission. He developed deviation of angle of mouth to left, inability to close right eye as well as cough, dysphagia, nasal regurgitation , hoarseness of voice , tinnitus and Rt. sided headache, two days after that. There was no history of ear discharge, loss of consciousness, vomiting, neck pain, convulsions or weakness of any limbs. On general examination he was conscious & well oriented in time place &person.He had hoarseness of voice &dribbling of oral secretions from Right angle of mouth. There was pooling of secretions in oropharynx with choking sensation for which frequent suctioning was necessary.The past history,family history &personal history was not significant. Patient was mildly febrile. His pulse was 100/min, & blood pressure was 120/80 mm Hg. Patient was dyspneic for initial few days due to mild bilateral aspiration pneumonitis .Except mild pallor rest of the general examination was normal. No cervical lymphadenopathy or meningeal signs were present.

On examination of Rt. Ear – Erythematous vesicular eruptions were noted over Rt. External ear canal &behind the pinna. No vesicles were seen in oral cavity.(as shown in picture 3&4)

CNS exam. The examination of I, II, III, IV,V,& VI cranial nerves was normal. Pt. showed + ve consensual corneal response on Lt. side.The Fundus Exam. was normal.

Pt. had complete Rt. Infranuclear Facial Nerve (VII) palsy(as shown in picture 1&2). The taste sensations on Rt. half of tongue were absent. Pt also showed evidence of VIII CN involvement. The Rinne's Test was positive on Rt. Side and Weber's test was lateralised to Left side suggesting sensorineural deafness of Rt. Ear. He had Rt. Sided palatal palsy along with absent gag response.

The XI, XII cranial nerves were normal.

The rest of the neurological exam.including nutrition, tone, power, Tendon reflexes and plantars was normal. He did not have any involuntary movements.

Respiratory systemExam. showedbilateral scattered rhonci&crepts suggestive of pneumonitis.

The examination of cardiovascular system & abdomen was normal.

With the clinical history, neurological exam. & typical vesicular rash in ear canal and pinna the diagnosis of Ramsay Hunt Syndrome with Rt. Infranuclear facial palsy was obvious. The involvement of other cranial nerves i.e. VIII, IX,and X is a rare but well described feature in literature.To rule out the other causes of simultaneous CN involvement patient was evaluated in detail.

Routine Haemogram showed

Hb% - 10.6 gm%

TLC- 11,400/ cumm DLC- P-88%, L-9%, E-2%,

M-1%

Urine Exam – Normal. KFT- NAD LFT – NAD ECG – Normal

Blood Sugar – Fasting &postmealwere normal.

HIV status was Negative. X ray Chest - showed bilateral pneumonitis

MRI brain with contrast- shows age related mild cerebral atrophy . There is soft tissue intensity in Rt. mastoid air cellswhich is also seen extending in inner ear cavity. This on post contrast shows↖ ↖ enhancement with edematous VII &VIII CN complex suggestive of inflammatory focus consistent with clinical diagnosis of Ramsay Hunt Syndrome. (as shown in pictures 5&6 by arrows)

CSF studies were not done as clinically there was no evidence of meningitis.

Patient was put on broad-spectrum antibiotic in form of combination of Amoxycillin +Clavulanic acid (1.2 gm IV in BID doses for 5 days along with Antiviral drug Valaciclovir in doses of 1gm. TDS for 5 days.He was also given oral steroids as tab. Prednisolone 40mg. daily for 5 days & subsequently tapered off over next 2 weeks.Frequent suctioning of oropharynx was required and feeding was maintained on Ryles tube along with initial intravenous fluids support. He was given physiotherapy in form of nerve stimulation exercises (TENS) & neuromuscular exercises.

The patient showed gradual recovery of palatal palsy over next 3 weeks.But recovery of Rt. facial palsy did not occur.He is able to swallow liquids under supervision now & is given pad bandage and local antibiotic drops to prevent exposure keratitis.At present he is under our follow-up.

DISCUSSION: Herpes Zoster infection of geniculate ganglion producing LMN facial palsy is called as Ramsay Hunt Syndrome-1. It commonly occurs after the age of 60 yrs. Our patient is 65 yrs. old & has all classical features of Ramsay Hunt Syndrome along with rare manifestations of other CN involvement i.e. VIII, IX, X producing deafness & palatal palsy. This involvement of other CN is because of communicating branches of facial nerve with them. (5)It was reported by T. IriokaK,OhtaA,et al in2008 (6), Adachi M,(7)previously in 3 patients by H. Radhakrishnan et al in 2000. (8), by James Turner& Paul Geunes in 1997(9), by Sato K,Nakamura S et al in 1991 (10), by MathurSL,Bhandari R et al in 1985,(11) by AharonAviel, Gabriel Marshak in 1982(12) The immunocompromised conditions are associated with reactivation of latent VZV producing classical vesicular erythematous rash over skin in dermatomal distribution.

The confirmation of diagnosis can be done by a) VZV specific IgM antibodiesin blood. b) fluid from blisters can be tested for real time PCR which has 100% specificity &sensitivity.c) The MRI of brain with Gadolinium contrast is very important which shows enhancement of VII & VIII nerve complex and geniculate ganglion.If rash has appeared simple visual examination only is required since dermatomal distribution is very characteristic of herpes zoster infection.When rash is absent i.e. Zoster sine herpete, it is difficult to make clinical diagnosis. While it is not a life threatening condition shingles can be very painful - Post Herpetic Neuralgia is a well known complication. The other complications can be facial disfigurement , change of taste over tongue,damage to involved eye due to exposure keratitis, corneal ulceration &infection as well as cellulitis of external ear. Aberrant regeneration of facial nerve can produce 'crocodile tears' &hemifacialspasms.The spread of virus to meninges, brain & spinal cord can cause meningoencephalitis, convulsions and limb weakness secondary to myelitis.Although there is no reliable evidence that steroids,antiviral drugs or surgical decompression of facial canal make any difference but they are the only possible treatments available at present.Antiviral drugs&**steroids** if started within 72 hrs.of appearance of rash can reduce severity and duration of shingles. The treatment is started with **antiviral drugs** like Acyclovir (800mg PO , 5 times a

day) or Valaciclovir (1 gm PO TID) orFamciclovir (0.5gm TID) for 5-7 days and oral steroids. The narcotics,amitryptaline(25-75mg/d) , gabapentin(300-1200mg/d) are useful for **control of pain** .Topical Capsaicinointment,lignocaine patches or infiltration may be given for neuralgia.Care of involved eye is also important to prevent complications. There is a great role of **physiotherapy** in management of CN palsies in form of Transcutaneous Electrical Nerve Stimulation (TENS),facial neuromuscular exercises, automassaging, relaxation+coordination exercises.

The prevention of shingles is not possible if virus is already there in the body. VZV vaccine (Varivax) is given to all children in their primary immunization schedule between 12-18 months. It is also given to older kids &adults who never had chickenpox. This vaccine prevents chickenpox in most people and severity is very less if not able to prevent it. The FDA has also approved **Zostavax** injection to help prevent shingles & RHS like conditions in adults >60 yrs.of age. It is a live attenuated vaccine & given as single injection in upper arm without major side effects.

Prognosis: generally it is good. However hearing loss is often permanent. Facial weakness may improve completely if steroids & antiviral drugs are started early within 72 hrs.of onset of rash (75%).The poor prognostic factors for functional recovery are,age above 50 yrs. delay in initiation of treatment, complete facial palsy,lack of VII nerve excitability on electrophysiological studies.(13)

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Fig. 1

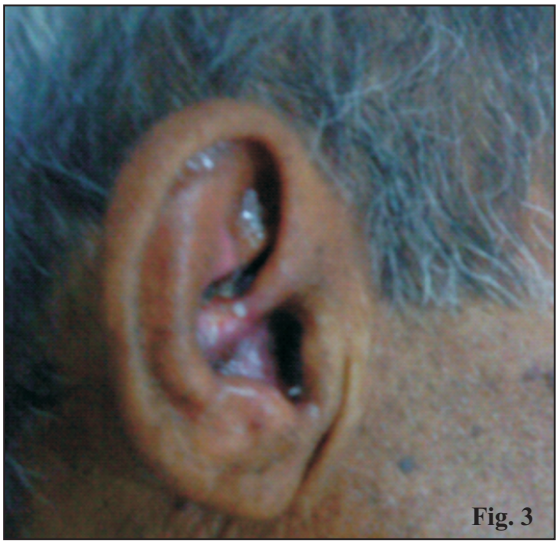

Fig. 3

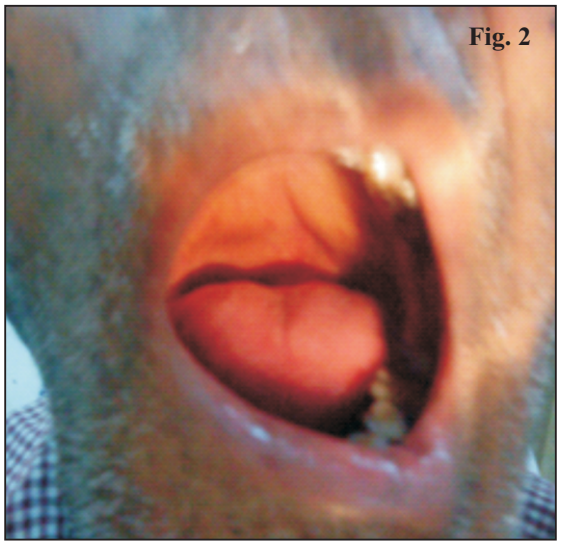

Fig. 2

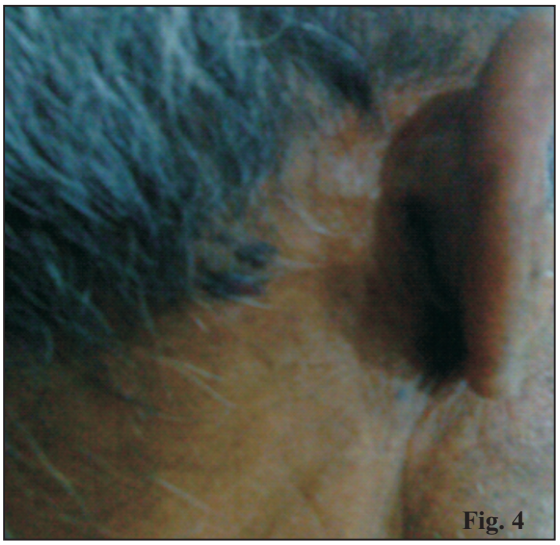

Fig. 4