CASE REPORT

RAMSAY HUNT SYNDROME—A RARE CAUSE OF FACIAL AND PALATAL PALSY

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ABSTRACT: Ramsay-Hunt Syndrome (RHS) is a rare entity in children produced by reactivation of varicella-zoster virus (VZV) in the geniculate ganglia and is characterized by peripheral facial palsy, skin eruption in the auricular canal and palatal palsy, with an incidence of 16.7% of children (1). We report a 12 years-old boy with Ramsay-Hunt Syndrome, who had complete resolution of the peripheral facial palsy with acyclovir. Since it is uncommon in children a high degree of suspicion should be kept in children with unilateral Facial Nerve Palsy, erythema, vesicles and/or auricular pain.

CASE REPORT: A 12 yrs old male child, third by order of birth, born of third degree consanguineous marriage brought by parents with history suggestive of chicken pox 25 days back

He had fever for 3 days, 15 days prior to development of deviation of mouth to left side for 4 days with history of unable to close right eye since last 3 days.

He was apparently all right 25 days back; to start with he had fever with rash of vesicular with central umbilication suggestive of chicken pox, distributed over face, body and extremities. And the rash was crusted and healed completely on 9th day of the illness. He started having pain in right ear. There was history of redness in right external auditory canal. After 4 days of above complaints, he developed deviation of the mouth to left side with drooling of saliva. He was unable to close his right eye, voluntarily and during sleep. There was history suggestive of nasal regurgitation and nasal twang to his voice.

Also associated with difficulty in deglutition since 3 days. He was immunized as per National immunization schedule. On examination he was conscious, co- operative, thin built. There was absence of naso-labial fold on right side, with deviation of angle of mouth to the left and drooling of saliva from the right side. There was absence of wrinkling over right side of forehead on frowning, suggestive of lower motor neuron facial palsy. His gag reflex was weak. No signs of cerebellar involvement. Clinical diagnosis of Ramsay Hunt Syndrome was made. He improved clinically with Tablet Acyclovir for 7 days and Tablet Prednisone in tapering doses over 10 days along with physiotherapy.

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DISCUSSION: RHS is defined as an acute peripheral facial neuropathy associated with erythematous rash of the skin of the ear canal, auricle (herpes zoster oticus) and/or mucous membrane of the oropharynx.

RHS is also known as geniculate neuralgia or nervous intermedius neuralgia. RHS can also occur in the absence of a skin rash, condition known as **zoster sine herpete** (2). This infection gives rise to vesiculation and ulceration of the external ear and ipsilateral anterior two thirds of the tongue and soft palate, as well as ipsilateral facial neuropathy (in Cranial Nerve VII), radiculoneuropathy or geniculate gangliopathy. VZV infection or reactivation involving the geniculate ganglion of CN VII within the temporal bone is the main pathophysiological mechanism of Ramsay Hunt Syndrome. RHS is estimated to account for 16% of all cases of unilateral facial palsies in children, and 18% of facial palsies in adults. Ramsay Hunt Syndrome is rare in children younger than 6 years (3).

CLINICAL FEATURES: Patients usually present with paroxysmal pain deep within the ear. The pain often radiates outward into the pinna of the ear and may be associated with a more constant, diffuse and dull background pain. The onset of pain usually precedes the rash by several hours and even days. Classical Ramsay Hunt Syndrome can be associated with vesicular rash of the ear or mouth (as many as 80% of cases), the rash might precede the onset of facial paresis/ palsy (involvement of the seventh cranial nerve, ipsilateral lower motor neuron facial paresis/palsy, vertigo and ipsilateral hearing loss, tinnitus, otalgia, headaches, dysarthria, gait ataxia, fever, cervical adenopathy, facial weakness usually reaches maximum severity by 1 week after the onset of symptoms. Other cranial neuropathies might be present and may involve cranial nerves VIII, IX, X, V, and VI. Ipsilateral hearing loss has been reported in as many as 505 of cases.

PHYSICAL FINDINGS: The primary physical findings in classical Ramsay Hunt Syndrome include peripheral facial nerve paresis with associated rash or herpetic blisters in the distribution of the nervus intermedius(4).

DIAGNOSIS: The diagnosis of RHS is usually made without difficulty when the clinical characteristics are present. If necessary, varicella zoster virus (VZV) may be isolated from the vesical fluid (5).

TREATMENT: Corticosteroids and oral Acyclovir are commonly used in the treatment of Ramsay Hunt Syndrome. A recent study concluded that controlled release oxycodone was safe and well tolerated in patients experiencing acute pain due to herpes zoster (6). Vestibular suppressants may be helpful if vestibular symptoms are severe. In Bell's palsy care must be taken to prevent corneal irritation and injury.

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