International Journal of Research and Review
www.ijrrjournal.com E-ISSN: 2349-9788; P-ISSN: 2454-2237

Case Study

Speech and Language Profile for Acute Disseminated Encephalomyelitis (ADEM) Post Varicella Zoster Infection: A Case Study

Ambar Gupta[1], Perumal Santhanam Divya[2], Mathew Nishanth[1], Mary Vishali Prabu[3], Sujitha Satchithanandham[3]

[1]Postgraduate Student, [2]Clinical Supervisor, [3]Undergraduate Student,
MERF Institute of Speech and Hearing (P) Ltd, Chennai, India

Corresponding Author: Ambar Gupta

ABSTRACT

Acute Disseminated Encephalomyelitis (ADEM) Post Varicella Zoster Infection is a rare inflammatory demyelinating disease of the (CNS) and it is elucidated as a scattered focal or multifocal (disseminated) inflammation of brain and/or spinal cord. A 25 year old unmarried female had a compliant of headache, vomiting, photophobia and common cold for two days and she was diagnosed to have post varicella ADEM, sepsis - urosepsis and status epileptics. She underwent tracheostomy to restore respiration. Dysarthric component was present. This case study focuses on speech and language evaluation and management for ADEM.

Key words: ADEM, Speech, language, tracheostomy

INTRODUCTION

Chickenpox or herpes zoster is a contagious disease caused by varicella zoster virus. Chickenpox follows initial exposure to the virus and is typically a relative mild, self-limited childhood illness with a characteristic of exanthema, but can become disseminated in immuno-compromised children. Infection are usually self limiting but complication may occur like pneumonia, encephalitis, or secondary phylogenic skin infections Central Nervous System (CNS) complications rate varies from 0.1 to 0.7% in several series. [1] Complications include encephalitis and cerebellar ataxia but rare complication like Acute Transverse Myelitis (ATM), aseptic meningitis, Guillain-Barre Syndrome, Meningoencephalitis, optic neuritis and stroke have also been described. [11] Acute disseminated encephalomyelitis (ADEM) is one of the rare CNS complications of varicella zoster infection. ADEM is a rare inflammatory demyelinating disease of the (CNS) and is elucidated as a scattered focal or multifocal (disseminated) inflammation of brain and/or spinal cord. [2] Approximately 70% of patients with ADEM reported a precipitating event, e.g. viral or bacterial infections or vaccination.[2] Predominantly viral infection such as chickenpox, measles, rubella, mumps, influenza, Epstein Barre virus, Human Immunodeficiency Virus (HIV) and Mycoplasma leads to ADEM followed by vaccination or spontaneously. Classically ADEM is a monophagic diseases but it may have a recurring course. [1] ADEM due to varicella infection is much lower in comparison with ADEM comparing other viral infections. Disease specific reported incidence rates for varicella are 1 per 1000.

[2] Cerebellar ataxia may also be one of the signs following varicella-zoster infection, 50% of post varicella encephalitis may present with cerebellar ataxia. [2]

Dysarthria is a collective group of speech sound disorders resulting from a disturbance in the muscular control over the speech mechanism due to damage of central and peripheral nervous system.

Speech abnormality domains relevant to dysarthria are:

- Articulation may be slow with imprecise consonants
- Instability of pitch and loudness of voice
- Decreased time for phonation expiratory pressure
- Longer and frequent pauses with deficient control of loudness. The modern classification of dysarthria still rests largely on Darley's classical study of 1969 (Darley et al 1969). [3]

Dysphagia is defined subjectively as a sensation of delay in transit of liquid or solid bolus during the oropharyngeal or esophageal stages of swallowing (oropharyngeal and esophageal dysphagia respectively). This could be distinct from the objective measurement of dysphagia because various mechanisms of esophageal sensory functions may account for the sensation of dysphagia with apparent delay in bolus transit. [4]

CASE HISTORY

A 25 year old unmarried female had a complaint of headache, vomiting, photophobia and common cold lasting for two days. The onset of the problem was reported to be sudden and she was diagnosed as having post varicella ADEM, sepsis - urosepsis and status epilepticus. She reported a history of sinusitis and chickenpox. Due to respiratory distress the patient underwent tracheostomy (fenestrated cuffed tube) to protect the air way along with the support of ventilator. She was treated with high dosage steroids and intravenous immunoglobulin initially yielding mild improvement.

Medical Investigation Findings:

Table: 1 shows the Magnetic Resonance Imaging (MRI) findings

Date	Findings
6/05/2019:	Multiple patches T2 hyperintensive in bilateral cerebral white matter, varicella leuconchylitis / ADEM.
11/05/2019:	Confluent T2W/ FLAIR hyperintensivity in bilateral cerebral region, mild brain pons and bilateral middle cerebellar peduncles with pateolus area of diffusion restriction. Possible differentials include varicella leukoencephalitis / ADEM.

SPEECH AND LANGUAGE EVALUATION

After two weeks of admission speech and language evaluation was carried out.

During the visit the client was alert and oriented. On informal language assessment, her auditory comprehension for complex sentences was observed to be good. She communicates her needs nonverbally through eye pointing, eye blinks, and mutual eye gaze. Breath support was inadequate. Voice parameters could not be assessed as the cuff of the tracheostomy tube was inflated during the base line assessment. Food intake was through Nasogastric (NG) tube.

Table: 2 shows the Oral Peripheral Mechanism Examination

ARTICULATORS	STRUCTURE	FUNCTION
Face	• Drooping • Eyelid closure (completely)	• Rising of eyebrows - Present • Closing of eyes - Present
Lips	• No deviation	• Retraction/Puckering/ Rounding/ Pursing- Inadequate • Lip strength against resistance- Reduced • Range of motion- Reduced • Lip closure - inadequate • IOBP: Reduced
Tongue	• No deviation	• Tongue protrusion, retraction, lateralization, elevation : Inadequate
Teeth	• Normal	• Biting/ Chewing: Present (Reduced range of motion)
Jaw	• No deviation	• Jaw movement: Normal

Table: 3 showing the Cranial nerve examination:

Cranial Nerve	Function	Indicate
V Trigeminal	**SENSORY:** Face, cheeks, lips, jaw, forehead, eyes, eyebrows, nose (pain,temperature, touch, proprioception) Interior/exterior jaw and TMJ. Sensation to superficial and deep structures of face, mucous membrane of upper mouth, palate and tongue. **MOTOR:** Mastication, jaw lateralization and closure. Assists with: a) Upward/anterior movement of larynx b) Backward movement of tongue to soft palate c)Palatal elevation (tenses soft palate) posterior pharyngeal wall constriction	Inadequate
VII Facial	Taste -Salivation(submandibular and sublingual glands) - Lacrimation - Mandibular depression - Contributes to hyoid Elevation	Inadequate
IX Glossopharyngeal	-Elevation of the larynx and pharynx - Taste/sensation posterior 1/3 3of tongue and sensation to tonsils, soft palate, upper pharynx - Sensory portion of pharyngeal gag - Salivation (parotid gland)	Inadequate
X Vagus	**SENSORY:** To palate, pharynx, larynx, trachea, lungs, epiglottis. Taste receptors in posterior oral cavity Recurrent Laryngeal Nerve (RLN): sensation below true vocal cords. Superior Laryngeal Nerve (SLN): posterior tongue and larynx above true vocal cords. **MOTOR:** - Elevation/depression of soft palate. - Elevation of posterior tongue - Elevation/closure of larynx. Lowering of larynx after swallow.	Inadequate

Goals

- To establish regulated and modified breathing pattern by using breathing exercises.
- To establish muscle strength using oral motor exercises (active muscle exercise, muscle stretching, passive exercise).
- To increase the awareness of the oral mechanism through sensory stimulation.
- To normalize oral tactile sensitivity.
- To improve the precision of volitional movement of oral structure, differentiation of oral movements and feeding skill.
- To improve oro pharyngeal phase of swallowing using Head extension, flexion and lateralization techniques and by implying swallowing maneuvers such as effortful swallow and Mendelsohn maneuvers.

POST THERAPY ASSESSMENT:

Tracheostomy and NG tube was removed at the fourth visit to the speech language pathologist. Oro motor functions of Jaw, Lip, Tongue were adequate for both vegetative and speech skills. Strength and range of motion of the articulators were observed to be improved compared to baseline. Post therapy swallowing evaluation was done. On swallowing assessment, semi-solid and solid foods were administered and the following was observed: Laryngeal elevation was adequate, No delay in oral transient time, Aspiration absent, No residue present.

Table: 4 showing Respiratory system for Pre therapy and Post therapy assessment:

	Pre therapy assessment	Post therapy assessment
Breathing pattern for speech and non- speech activity	Could not be assessed	Thoracic breathing
Respiratory support for speech	Could not be assessed	Adequate.
Respiratory support for phona tion	Could not be assessed	Adequate

Table: 5 showing Voice Profile for Pre therapy and Post Therapy assessment :

	Pre therapy assessment	Post therapy assessment
Pitch	Could not be assessed	Appropriate
Loudness	Could not be assessed	Reduced
Quality	Could not be assessed	Moderate weak voice
Endurance	Could not be assessed	Poor

DISCUSSION

Herpes zoster or shingles is a painful rash eruption caused by the varicella zoster virus (VZV). VZV usually persists symptomless in the dorsal root ganglia of

any person who has had chickenpox. Reactivating from its dormant state was observed in about 25% of people resulting in traveling of virus along the sensory nerve fibers and causing vesicular lesions in the dermatome supplied by that nerve. Herpes zoster is more common in people with diminished cell mediated immunity. [5]

Neurological complications of varicella zoster reactivation are vasculopathy, meningoencephalitis, cerebelitis, post-hepatic neuralgia, myelopathy and ocular diseases. [6] ADEM is one of the rare neurological complications. ADEM in its more severe form is commonly referred as acute haemorrhagic leucoencephalitis (AHLE) representing non-infective central nervous system inflammatory diseases. [9] ADEM signs include ataxia, tremors, dysarthria, hemiparesis, cranial nerve palsy, optic neuritis and convulsions which indicates significant involvement of brain parenchyma, optic nerve and spinal cord. [7]

ADEM is often misdiagnosed as MS (Multiple sclerosis). Discrimination between ADEM and the first presentation of MS has important prognostic and therapeutic implications. Patients with ADEM generally recover completely, whereas those with MS may have recurrent relapses or progressive deterioration over time. No clinical feature is exclusive to either condition, but some characteristics are more commonly seen in one than the other. ADEM is more typically a monophasic illness, whereas the course of MS is usually multiphasic and characterized by relapses and remissions. [8]

A diagnosis of Varicella ADEM is usually made with a combination of clinical features and radiological findings, but this can be supplemented with detection of VZV

in Cerebrospinal Fluid and /or blood with the help of serological and Polymerase Chain Reaction (PCR). White matter lesions on MRI are suggestive of ADEM though not specific for condition.

Numerous reports have established that dysarthria often occurs with dysphagia. It is inferred that these simultaneous occurrences depend on speech and swallowing sharing many peripheral morphological structures, that is, oral, pharyngeal, and laryngeal structures, although they differ significantly in their neural mechanisms. Based on the clinical observation that speech and swallowing disorders frequently co-exist and that speech and swallowing involve much the same structures, it has been suggested that dysarthria and dysphagia can be improved concurrently. Dysphagia and dysarthria are quite common, especially in patients with brainstem stroke.

Concomitant dysphagia is more common in patients with flaccid, spastic and mixed dysarthria. Furthermore, dysphagia encompasses a broad range of severity levels with many individuals being severely impaired in these categories. On the other hand, concomitant dysphagia is less frequent in patients with ataxic, hypokinetic and unilateral upper motor neuron dysarthria, and severe swallowing problems seldom coexist. These results indicate that bilateral impairment of upper or lower motor neurons commonly results in severe dysphagia, while unilateral upper motor neuron, extrapyramidal system and the cerebellar system impairment rarely produce severe dysphagia. [10]

The patient's rate of speech was reduced to <60 words / min (80-120 words /min). Mean Length of Utterance (MLU) was one-two word per sentence. Inconsistent articulatory errors were observed to be present. Prosody aspects such as stress, rhythm, and intonation were observed to be inappropriate. No concerns regarding language and swallowing domains. Formal assessment for dysarthria was carried out using Frenchay's Dysarthria Assessment (FDA) which reveals Mild Dysarthria. Speech intelligibility was observed to be improved and her speech was understandable; however the patient feels that her speech is unclear. In addition she had rashes in the pinna, crusted eruptions on concha and in external auditory canal, henceforth adding to the high risk

category for Herpes Zoster Oticus (HZO). She was advised to have regular follow-up with ENT and high intensive care along with rehabilitation services which will enhance her recovery. Beside from all the medical care provided, her prognosis in speech and communication was better with the rehabilitative services given by Speech Language Pathologist and along with her family support.

REFERENCES

1. Sinha A, Kar SS, Purkait TP, Pandit UK. Acute disseminated encephalomyelitis in chicken pox. National Journal of Medical Research. 2016;6:103-4.
2. Murthy JM. Acute disseminated encephalomyelitis. Neurology India. 2002 Jul 1;50(3):238.
3. K K Jain MD *Originally released April 3, 2007; last updated March 26, 2019; expires March 26, 2022* http://www.medlink.com/article/dysarthria
4. Remes-Troche JM. The hypersensitive esophagus: pathophysiology, evaluation, and treatment options. Current gastroenterology reports. 2010 Oct 1;12(5): 417-26.
5. Wareham DW, Breuer J. Herpes zoster. *BMJ*. 2007; 334(7605):1211–1215.
6. Nagel MA, Gilden D. Neurological complications of VZV reactivation. Current opinion in neurology. 2014 Jun;27(3):356.
7. Stonehouse M, Gupte G, Wassmer E, Whitehouse WP. Acute disseminated encephalomyelitis: recognition in the hands of general paediatricians. Archives of disease in childhood. 2003 Feb 1;88(2):122-4.
8. Hynson JL, Kornberg AJ, Coleman LT, Shield L, Harvey AS, Kean MJ. Clinical and neuroradiologic features of acute disseminated encephalomyelitis in children. Neurology. 2001 May 22;56(10):1308-12.
9. Chaudhuri A, Kennedy PG. Diagnosis and treatment of viral encephalitis. Postgraduate medical journal. 2002 Oct 1;78(924):575-83.
10. Nishio M, Niimi S. Relationship between speech and swallowing disorders in patients with neuromuscular disease. Folia phoniatrica et logopaedica. 2004;56(5):291-304.
11. Dhar J, Kumar L, Ish P, Anuradha S. Acute Disseminated Encephalomyelitis: A Rare Complication of Varicella Zoster Infection in an Adult. Journal of Clinical & Diagnostic Research. 2018 Feb 1;12(2).

How to cite this article: Gupta A, Divya PS, Nishanth M et.al. Speech and language profile for acute disseminated encephalomyelitis (ADEM) post varicella zoster infection: a case study. International Journal of Research and Review. 2019; 6(7):59-63.

******