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Brown-Vialetto-Van laere syndrome & auditory neuropathy spectrum disorder: A case study

Nithin AK and Sagar Jose

Abstract
The Brown-Vialetto-Van Laere syndrome (BVVL) may be a rare neurological disease characterised by progressive pontobulbar palsy related to sensorineural hearing impairment. Cardinal cases are reportable in mere over one hundred years. The feminine to male quantitative relation is roughly 3:1[1]. The syndrome most often presents with sensorineural hearing impairment that is sometimes progressive and severe.
Method: The patient conferred during this case study had undergone multiple tests tympanometry, acoustic reflex, pure tone audiometry, transient evoked otoacoustic emissions and auditory brain-stem response were used and continual 3 times throughout six years.
Results: This study shows the clinical symptoms pertinent to auditory Neuropathy spectrum disorder (ANSD) for a 15-year-old woman with Brown-Vialetto-Van Laere Syndrome (BVVL). The results are followed for 6 years between 2016 and 2022. Within the 1st session, the results discovered bilaterally normal transient evoked otoacoustic emissions, absent acoustic reflexes (ipsilateral and contralateral), delicate to moderate low tone loss within the sound audiometry, no auditory brain stem responses at high input intensities and totally different polarities and gift tube-shaped structure micro phonic part in single polarities. Within the second session, sound audiometry showed slight to gentle high tone loss but the opposite tests incontestable their same results. Within the final session, the latter results were repeated.
Conclusion: During this comprehensive follow up study, the patient with BVVL incontestable clinical symptoms of ANSD within which sensory system showed an impairment within the sense modality temporal synchronization and encoding.

Keywords: Brown-Vialetto-Van Laere syndrome, auditory neuropathy spectrum disorder, auditory brainstem response, pure tone audiometry

Introduction
The patient conferred during this case study was a 10-year-old girl on the primary admission referred together with her parents from a Child specialist in 2016 to assess auditory function and judge auditory responses. She was the sole kid of her oldsters and was born term in hospital. There have been no risk factors together with drive, baby medical care unit hospitalization, cytomegalovirus and no alternative issues related to development of central system throughout 1st six years of life. In contrast, after this period of your time and thanks to the incidence of issues in her face look and behaviour, medical specialty examination showed that she developed bilateral cranial polyneuropathy, bilateral descensus and esotropia, inability to laugh, blow and suck. Moreover, the patient was unable to stick out her tongue. Instinctive reflex was weak, however visual and sense modality functions appeared to be normal [1]. Initial diagnostic tests together with brain resonance imaging, diagnostic technique, and nerve conductivity speed, and neurotransmitter antibody studies were conducted to produce additional detail info concerning the progressive un-wellness. At the first time, differential diagnoses enclosed myasthenia gravis, Brown-Vialetto-Van Laere Syndrome (BVVL), polyneuritis cranialis, neurosarcoidosis, and malignant infiltration of the nerve roots. When four months, no abnormalities were found on repeat brain magnetic resonance imaging. However, electromyography and nerve conductivity speed showed nerve fibre injury of bilateral seventh nerves. More significantly, the auditory brainstem response (ABR) revealed no responses at high intensities showing an involvement of eight nerves bilaterally. Additionally, EMG demonstrated neurogenic changes within the tongue and facial muscles [1]. The results of the clinical and electrophysiological studies were indicative polyneuritis cranialis and therefore the kid received medications as well as intravenous immune serum globulin within the initial 2 years when of sickness and daily heptoliv in yet.

Despite the mentioned treatment, she experienced worsening of within the following years. Because of this condition, genetic study was performed and confirmed BVVL. It is indicated that the prevalence of this syndrome is extremely low and solely cardinal patients in only over a century. Half all cases are sporadic [1]. The bulk of familial cases demonstrate chromosome recessive inheritance, though chromosome dominant or sex chromosome inheritance has been instructed in a very few families. The female to male quantitative relation is about 3:1 in reported cases [1]. Half-dozen years later in 2021, the patient showed covert face, no ABR in each ears, bilateral descensus, reduced gag reflex and blurred speech. The patient suffered from perception difficulties of speech and hearing in strident things. Also, she suffered from depression because of her disabilities. In the present study, the patient was evaluated by complete auditory tests throughout six years to reveal her auditory issues objectively and behaviourally.

Method

The ethical approval was obtained from the Institutional ethical committee prior to the conductivity of the study similarly an informed consent was signed from the participant explaining all the procedures concerned during this study.

In the present study, a whole case history centred on hearing issues was taken from the kid and her parents within the 1st session of auditory system analysis. The collected key info provided an honest profile of child's issues in hearing perception of speech sounds in noisy situations like faculty lecture rooms. In order to gauge her sensory system, auditory take a look at battery together with tympanometry, acoustic reflex, pure tone audiometry, transient-evoked otoacoustic emissions (TEOAEs) and click-evoked ABR was used. In the 1st session back in 2016, otoscopic review showed normal condition of the external ear canal and tympanic membrane.

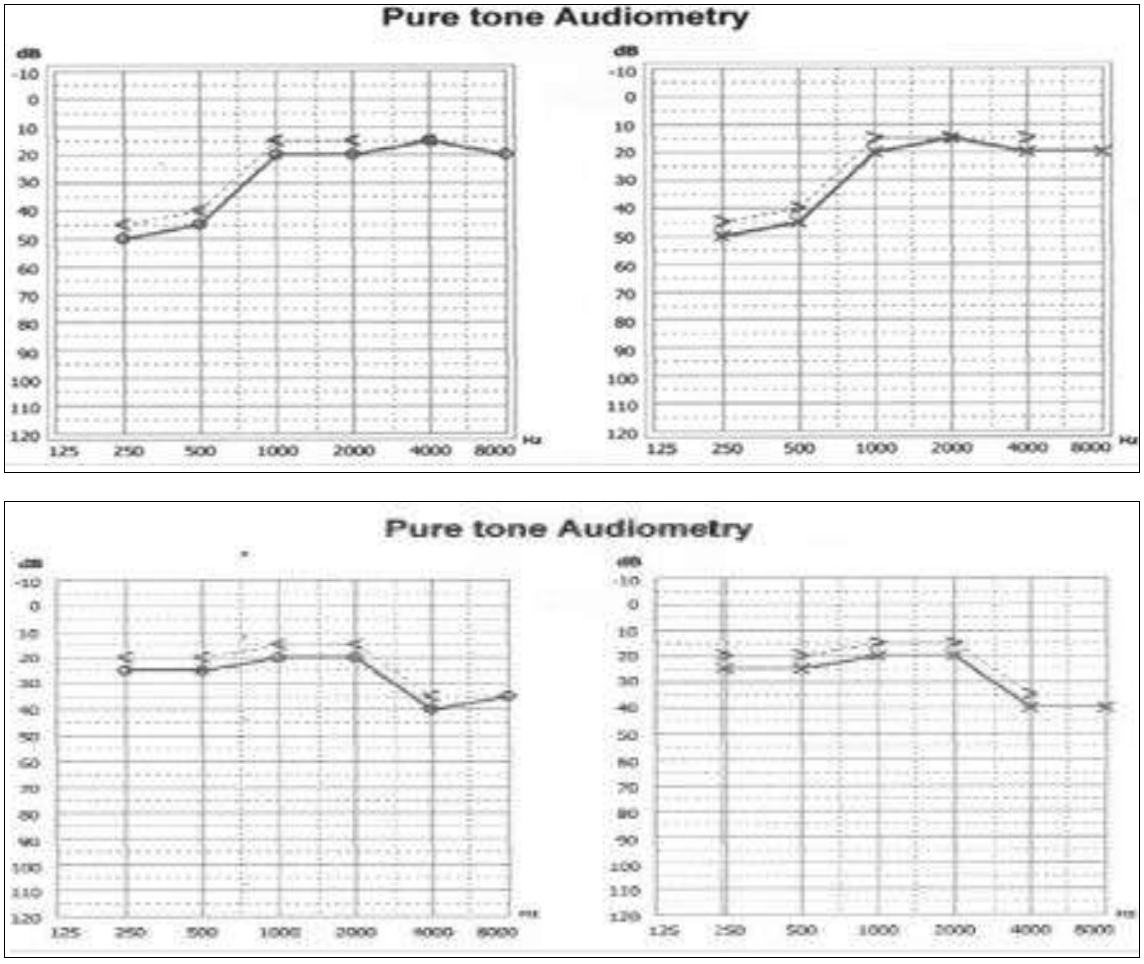

Fig 1: The initial and second session PTA up and down respectively

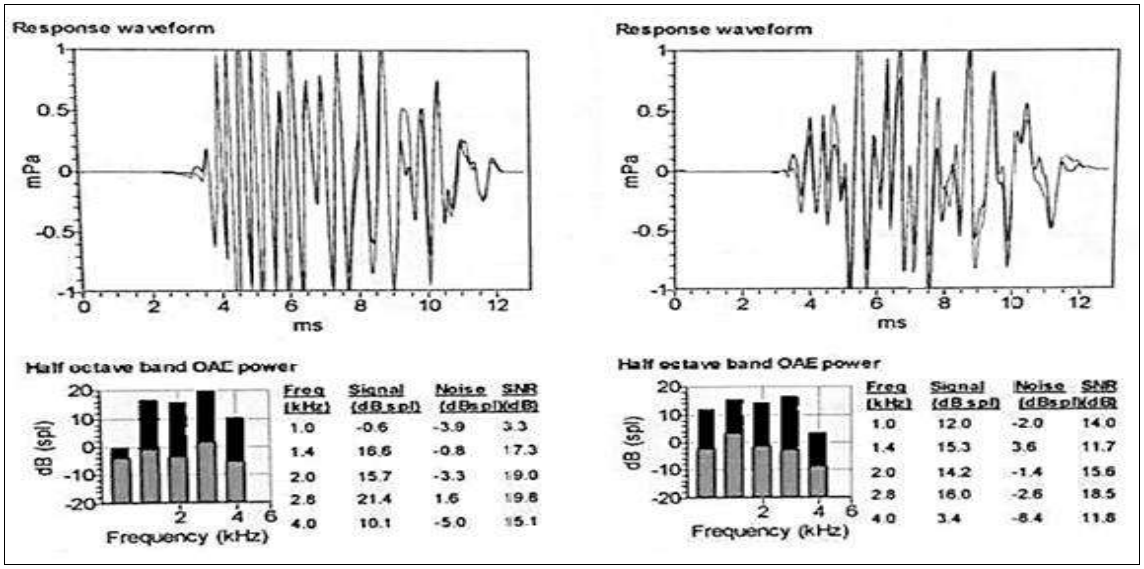

Fig 2: Transient-evoked otoacoustic emissions in both ears. The amplitude of responses was identified by dark colour

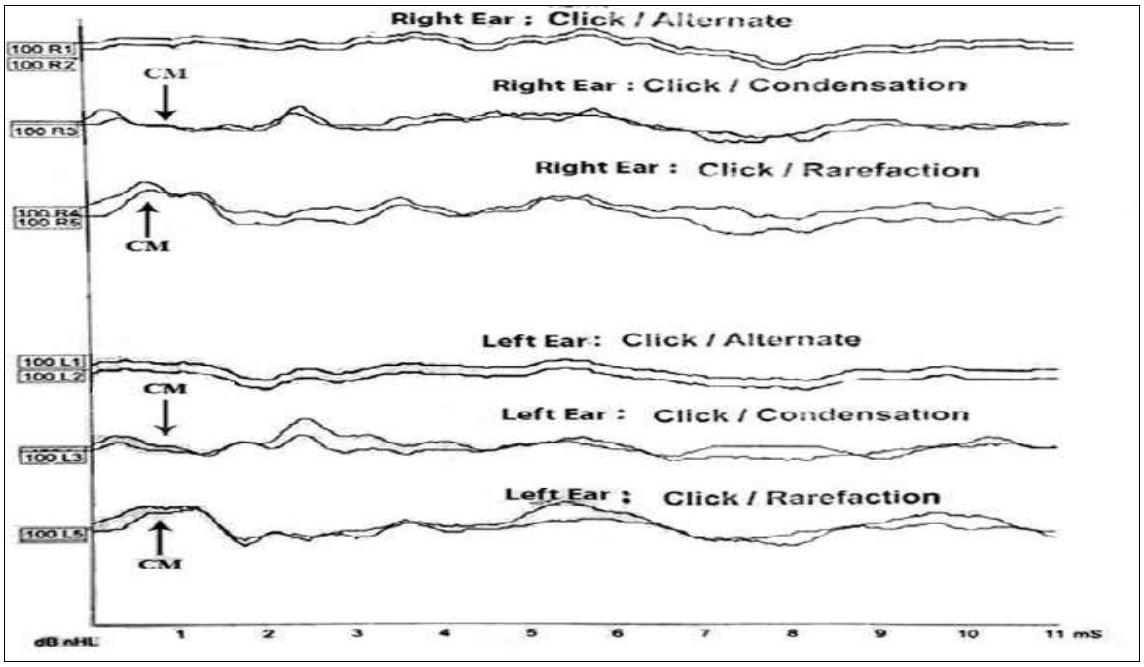

Fig 3: No auditory brainstem responses (waves I, III and V) at high stimulus intensity and different polarities in both ears. Cochlear micro phonic component could be distinguished in single polarities.

Admittance audiometry disclosed normal tympanograms (type 'A') and absent ipsilateral and contralateral acoustic reflexes bilaterally. Pure tone audiometry indicated bilaterally delicate to moderate low tone loss with excellent score (100%) in word recognition test in an acoustic chamber. 3 years later (second session), the audiometric results disclosed clinically normal thresholds in low frequencies however slight to delicate sensory-neural hearing impairment in high frequencies (Figure1). TEOAE demonstrated good signal to noise ratios in different frequencies indicating traditional perform of outer hair cells in each ears (Figure 2). The click-evoked ABR which can be conducted objectively to assess auditory nervous system in each ears illustrated no responses (waves I, III and V) at high stimulant intensity (100 decibel nHL) and completely different polarities (rarefaction, condensation and alternation). However, the results showed bilaterally cochlear micro phonic potential for rarefaction and condensation polarities (Figure 3). The other assessments showed the same. Finally, in final session, all of the tests were repeated and the results confirmed the latter ones.

Discussion

Brown-Vialetto-Van Laere Syndrome could be a rare progressive neurological disorder with worsening of symptoms in the following years of life [1]. The clear symptoms are progressive pontobulbar palsy related to bilateral ptosis, reduced reflex, bilateral disablement and tongue atrophy [2]. The age of onset might be from infancy up to as late as thirty years recent [1]. During this study, auditory test battery as well as behavioural, physical and electrophysiological modality tests was utilised to demonstrate function of auditory system in a child with BVVL. Interestingly, tone audiometry showed delicate to moderate low tone loss bilaterally within the 1st session modified to normal thresholds within the low frequency and slight to delicate high tone loss within the second session. The other significant result was no auditory brain-stem

responses bilaterally at high stimulation intensity and totally different polarities in 3 sessions. Against this, TEOAEs and cochlear micro phonic responses were present and repeated during this case. Overall, these attention-grabbing results showed auditory neuropathy spectrum disorder (ANSD) indicating possible problems in auditory temporal encryption [3, 4]. It is believed that sound transduction and speech perception need to a high level of synchronised activity. Disturbance to cranial nerve synchronizing interferes with the temporal coding of sound that is that the most significant consider speech perception [5]. It's according that almost all of the patients with auditory neuropathy/auditory dys-synchrony show hearing issues and report difficulties in hearing and auditory perception in blatant situations [6, 7]. In this study, the child with BVVL showed clear perception difficulties thanks to ANSD. supported the results, it is probable that the sensory system will show totally different behaviours to totally different acoustic stimuli; pure tones will produce traditional or nearly normal responses however transient stimuli like click will show no responses in cases with BVVL. This finding might be associated with abnormal synchronization in patients with ANSD [6, 7]. In fact, many patients with this disorder could report misunderstanding in modality environments with abrupt acoustic changes (especially in time) e.g. things with competing stimuli and educational school rooms. To survey and monitor the severity of central modality process involvement in these cases, a lot of behavioural (speech in noise test) and electrophysiological modality assessments (auditory evoked potentials using complicated modality stimuli including speech signals) ought to be conducted.

Conclusion

To sum up, it's claimed that this study is one amongst the first reports of watching auditory responses in patients with Brown-Vialetto-Van Laere Syndrome throughout a 6-year period once initial identification. The results of this study stressed on differing types of sensory system responses to totally different stimuli because of auditory neuropathy spectrum disorder.

Ethical considerations

The ethical approval was obtained from the Institutional ethical committee before the conductivity of the study similarly an informed consent was signed from the participant explaining all the procedures concerned during this study.

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Authors' contributions

N.A.K: The study design, data collection, analysis and interpretation of results, drafting the manuscript.
S.J: data collection, analysis and interpretation of results

Conflict of interest

The author does not have any financial or other interests relating to the study.

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