Oculomotor Abnormalities in Large Cerebellopontine Angle Tumors: A Case Series of Bruns’ Nystagmus

Giada Bilotta1, Joost Stultiens2, Tomas Menovsky3,4, Yasin Temel5, Marc Lammers4,6, Olivier M. Vanderveken4,6, Raymond van de Berg2,7, Vincent Van Rompaey2,4,6

1Faculty of Medicine and Health Science, University of Antwerp, Antwerp, Belgium
2Department of Otorhinolaryngology & Head and Neck Surgery, School for Mental Health and Neuroscience, Faculty of Health Medicine and Life Sciences, Maastricht University Medical Center, Maastricht, The Netherlands
3Department of Neurosurgery, Antwerp University Hospital, Antwerp, Belgium
4Department of Translational Neurosciences, Faculty of Medicine and Health Science, University of Antwerp, Antwerp, Belgium
5Department of Neurosurgery, School for Mental Health and Neuroscience, Faculty of Health Medicine and Life Sciences, Maastricht University Medical Center, Maastricht, The Netherlands
6Department of Otorhinolaryngology and Head & Neck Surgery, Antwerp University Hospital, Antwerp, Belgium
7Faculty of Physics, Tomsk State Research University, Tomsk, Russia


ABSTRACT

Patients with large or giant large cerebellopontine angle lesions can present with a wide range of audiovestibular symptoms, including Bruns’ nystagmus. This is a rare variant of bidirectional nystagmus, characterized by a combination of slow, large-amplitude nystagmus when looking to the side of the lesion and rapid small-amplitude nystagmus when looking to the contralateral side. This phenomenon arises due to a unique situation in which 2 different neural circuits, specifically the floccular pathways and peripheral vestibular pathways, are simultaneously involved. The presence of Bruns’ nystagmus is a good indicator for large CPA lesions of at least 3 cm in diameter with compression and displacement of the cerebellum, comprising the flocculus, and/or the brainstem. A case series of 4 different cases illustrates that investigating the presence of such nystagmus could be incorporated into the common diagnostic work-up of CPA lesions, since it is of particular interest in localizing large CPA tumors requiring urgent imaging.

Keywords: Brainstem, cerebellum, neurilemmoma, pathologic nystagmus, vertigo

Introduction

Vestibular schwannomas (VS) are associated with a wide range of neurotologic symptoms. The most common presenting symptoms of VS are progressive unilateral or asymmetric hearing loss, tinnitus, ear fullness, imbalance, and vertigo.1-4 Cerebellar symptoms, including incoordination, ataxia, and disequilibrium, usually occur late in the course of tumor growth.4 Typical trigeminal nerve dysfunction related to VS results in reduced corneal reflex, facial hypoesthesia or paraesthesia, and weakness in the muscles of mastication. Facial nerve dysfunction can occur in large cerebellopontine angle (CPA) tumors leading to facial twiching and muscle weakness which should be graded according to the House–Brackmann (HB) system. Other cranial nerve deficits can occur with large tumors. Diplopia and lateral gaze can result from impairment of the abducens nerve. Glossopharyngeal and vagal nerve compression may cause hoarseness, dysphagia, and aspiration. Hypoglossal nerve involvement can lead to dysarthria.3,4 Contralateral weakness can be present if there is significant compression of the brainstem. Symptoms related to hydrocephalus, induced by obstruction of the fourth ventricle by large CPA tumors, include headache, nausea, vomiting, diplopia, papilledema, and changes in mental status.4

Clinical examination should include a complete ear–nose–throat examination, including micro-otoscopic evaluation, evaluation of soft palate movement to assess glossopharyngeal movement, and vocal fold evaluation by laryngoscopy to assess vagal nerve function. Neurological examination includes...
B-ENT 2022; 18(3): 204-207
Bilotta et al. Oculomotor Abnormalities in Large Cerebellopontine Angle Tumors

Functional evaluation of the cranial nerves and cerebellar function as it may demonstrate subtle signs which are indicative of the underlying pathology and its possible extent. Vestibular examination often reveals spontaneous and induced nystagmus by performing positional testing, head impulse test, and looking for either sound- or pressure-induced nystagmus. Bruns' nystagmus is a rare variant of bidirectional nystagmus, with a prevalence of 5%-12%, observed in patients with a large or large CPA lesion, caused by compression and displacement of the cerebellum and/or the brainstem. All patients with CPA tumors should receive air and bone conduction pure tone audiometry. Caloric testing is the most common vestibular function test described in relation to CPA lesions and in particular reference to VS. Contrast-enhanced magnetic resonance imaging (MRI) of the CPA is the golden standard tool for diagnosing VS, as it is capable of detecting small tumors. Auditory brainstem response and high-resolution computed tomography are complementary modalities, but they have a lower sensitivity for smaller tumors.

The objective of this case series is to describe 4 cases with Bruns' nystagmus to emphasize the usefulness of clinical examination in determining whether urgent imaging is warranted and how it can help in localizing large CPA lesions.

Case Presentation

Case 1
A 60-year-old man was referred with left-sided profound sensorineural hearing loss (SNHL), dizziness, gait unsteadiness, and diplopia. Left-sided hypogeusia, facial paraesthesia, and hemifacial spasm were reported with normal facial function. Micro-otoscopic examination was normal. Vestibular examination revealed a positive head impulse test on the left side and Bruns' nystagmus with slow large-amplitude nystagmus to the left and rapid small-amplitude nystagmus to the right (Figure 1). Audiometry identified a severe sensorineural hearing loss in the left ear. Electronystagmography demonstrated a reduced caloric response on the left side. The MRI scan identified a left-sided VS with a maximal diameter of 3.7 cm with a significant impact on the brainstem, cerebellum, and left cerebellar peduncle, causing hydrocephalus due to compression of the fourth ventricle (Figure 2). Retro-sigmoid subtotal tumor resection was performed. At 10-month follow-up, the gait was stable, Bruns’ nystagmus, dizziness, instability, and diplopia had disappeared, and facial nerve function had evolved from HB grade 4 2 days post-operatively to grade 1.

Case 2
A 30-year-old woman was referred with right-sided hearing loss and tinnitus, gait ataxia, bilateral facial dysesthesia, and...

Main Points
- Bruns' nystagmus is a rare variant of bidirectional nystagmus which can occur in “large” CPA lesions.
- Evaluation of nystagmus can guide the clinician toward localization of large cerebellopontine angle lesions.
- Urgent imaging is needed in patients presenting with Bruns’ nystagmus.

Figure 1. Video-oculography recordings during gaze to the right (A) and to the left (B).
visual impairment. Micro-otoscopic examination and facial nerve function were normal. Vestibular examination revealed a Bruns’ nystagmus with slow, large-amplitude nystagmus when looking to the right and rapid small-amplitude nystagmus when looking to the left, poor fixation-suppression, and saccadic smooth pursuit. Wide-based gait and abnormal tandem gait were observed. Romberg testing showed a tendency to fall to the right. Coordination testing with the finger-to-nose test and finger-to-finger test revealed dysmetria. Audiometry showed right-sided mild SNHL. Magnetic resonance imaging of the CPA identified a right-sided CPA lesion of 4 cm maximal diameter, resulting in the compression of the fourth ventricle without hydrocephalus. The CPA tumor was treated by translabyrinthine partial resection and postoperative radiotherapy. At 3-month follow-up, the tinnitus and facial dysesthesia persisted. Postoperative facial nerve paresis HB 2-3 was observed, which remained unchanged.

Case 3
A 65-year-old man was referred with a retro-auricular squamous cell carcinoma (SCC), presenting with subacute imbalance and right-sided hearing loss for several years. The SCC was removed surgically, but 1 week postoperatively, he complained of diplopia and progressive gait ataxia. Micro-otoscopic examination and facial nerve function were normal. Vestibular examination revealed skew deviation and right-sided head tilt. There was Bruns’ nystagmus with slow large-amplitude gaze-evoked nystagmus to the right and rapid small-amplitude nystagmus to the left. Furthermore, smooth pursuit was saccadic, and a right-sided positive head impulse test was observed. Wide-based gait was observed. Audiology revealed a rightsided profound SNHL. Magnetic resonance imaging revealed a large right-sided cystic VS, compressing the fourth ventricle, with signs of obstructive hydrocephalus. Retrosigmoid subtemporal tumor removal and postoperative radiotherapy were performed. Postoperatively, facial nerve paresis HB 3 improved to HB 2 after 1 week. The complaints of imbalance, diplopia, and ataxia during walking resolved almost completely 3 months after surgery.

Case 4
A 78-year-old female was referred because of severe dizziness 2 weeks after retrosigmoid resection of a right-sided petroclival meningioma. She complained of dizziness, imbalance, diplopia, and nausea, only improving when lying still with her eyes closed. She experienced diplopia during prolonged concentration. Pre-operatively, her meningeoma was diagnosed after consulting a neurologist with symptoms of imbalance and falls. Micro-otoscopic examination was normal. Vestibular examination revealed a Bruns’ nystagmus with slow large-amplitude nystagmus when looking to the right and rapid small-amplitude nystagmus when looking to the left, as well as gaze-evoked nystagmus to the upper side. There were saccadic eye movements as well and a positive head impulse test on both sides. Dix-Hallpike was normal, but the lateral roll evoked apogeotropic nystagmus on both sides. She was diagnosed with vestibular hypofunction due to a central component resulting from the tumor compression and a peripheral component due to the surgical resection. The patient reported improvement in her symptoms after 6 months of vestibular rehabilitation therapy.

Verbal informed consent was obtained from the patients who agreed to take part in the study.

Discussion
Bruns’ nystagmus is a rare variant of bidirectional nystagmus, which enables the localization of large CPA tumors. It occurs with compression and displacement of the cerebellum, comprising the flocculus, and/or the brainstem, and therefore requires urgent imaging. In the 19th century, neurologists like Ludwig Bruns relied on this kind of clinical sign to justify the undertaking of a surgical procedure with significant morbidity and mortality in his era. Rarely, it has also been reported in pontine stroke and cerebellar apoplexy. Bruns’ nystagmus is primarily a form of jerk nystagmus, characterized by a combination of a slow large-amplitude gaze-evoked nystagmus when looking to the side of the lesion and rapid small-amplitude nystagmus when looking to the contralateral side. In nystagmus, the slow component refers to deviation of the eyes away from focusing the target on the fovea. The saccadic rapid component is corrective in nature, refocusing the target back on the fovea. The slow components of the large-amplitude and small-amplitude nystagmus are different in nature and are attributed to different neural networks. The slow phase of the gaze-evoked large-amplitude
Oculomotor Abnormalities in Large Cerebellopontine Angle Tumors

Bilotta et al.

nystagmus is attributed to central vestibular dysfunction and decreases exponentially. Gaze-evoked nystagmus in all directions indicates compression and therefore disinhibition of the neuronal integrator which is responsible for maintaining eccentric gaze-holding, particularly the flocculus. Instead, predominately vertical gaze-evoked nystagmus occurs due to a disruption of the brainstem vestibular pathways to the interstitial nucleus of Cajal in the mesencephalon, which is the neuronal integrator of vertical (and torsional) eye movements. The small-amplitude nystagmus is attributed to the ipsilateral peripheral vestibular dysfunction. This peripheral vestibular nystagmus can be caused by direct compression of the tumor on the vestibular nerve, by neurotoxic substances released from the tumor or by the deterioration of the labyrinth, causing an imbalance in vestibular tone between the labyrinths or vestibular nerves.

Because of these specific conditions, Bruns’ nystagmus only appears with tumors of at least 3 cm in diameter. As a test for CPA tumor size greater than 3.5 cm, Bruns’ nystagmus has a sensitivity of 42% and a specificity of 98%. Thus, the presence of Bruns’ nystagmus is a good indicator of a large CPA tumor, but its absence does not preclude the presence of a large tumor. Due to the characteristics of the nystagmus, the large CPA tumor can be localized on the side where slow-large-amplitude nystagmus occurs when the patient looks in that direction. Investigating the presence of such nystagmus should be incorporated into the routine diagnostic work-up of the neurootology patient. If Bruns’ nystagmus is identified, urgent imaging of the CPA and brainstem should be advised.

Informed Consent: Verbal informed consent was obtained from the patients who agreed to take part in the study.

Peer-review: Externally peer-reviewed.


Declaration of Interests: The authors have no conflict of interest to declare.

Funding: The authors declared that this study has received no financial support.

References

1. Gréant E, de Heyning PV, Ihtijarevic B, Topsakal V, Menovsky T, Van Rompaey V. Sporadic vestibular schwannoma: correlation between tumour size, hearing levels, age and radiologic features in 384 patients. B-ENT. 2020;16(2):97–102. [CrossRef]

207