LETTER TO THE EDITOR

To The Editor

Vertebrobasilar Hypoplasia Associated with Loss of Consciousness Induced by Laughter

Keywords: Vertebrobasilar, Hypoplasia, Syncope, Laugh

Syncope is a common clinical problem characterized as a non-traumatic transient loss of consciousness due to cerebral hypoperfusion with a rapid onset, short duration, and spontaneous complete recovery.[1] Its pathophysiology relies on a final common pathway, that is, the fall in systemic arterial pressure to levels lower than those tolerated by autoregulation, but it can be distinguished into three groups: neurally mediated syncope, syncope due to orthostatic hypotension, and cardiac syncope.[1]

Laughter-induced or gelastic (from the greek word *gelos,* laughter) syncope is a rare subtype of situational syncope[1] which is classified as a low-risk syncope and that seems to be physiologically kindred to other vasovagal syncopes resulting from increased intrathoracic or intra-abdominal pressure that reduces venous return and, therefore, cerebral perfusion.[2-4]

Here, we report the case of a middle age man with vertebrobasilar hypoplasia presenting episodes of syncope after vigorous laughter.

A 43-year-old man sought for health assistance because of recurrent loss of consciousness induced by intense laughter since adolescence. The events lasted less than 1 minute and were accompanied with full regain of consciousness. The patient reported that they were frequent and occasionally associated with stereotyped movements that, by the description given, resemble post-syncopal seizures. He denied dizziness, vertigo, sleep attacks, sleep paralysis, and hypnagogic hallucinations. He also reported that his brother has similar symptoms. Review of systems was notable for hypertension, anxiety disorder, and obstructive sleep apnea (OSA), for which the patient was in irregular use of continuous positive airway pressure therapy. Concerning lifestyle habits, he is sedentary and drinks alcohol almost daily. On physical examination, there were no remarkable findings except for a body mass index of 37.6 kg/m^2. Approximately 3 years before the patient's evaluation, he was submitted to Sleeve gastrectomy; however, there was no consistent weight loss. Furthermore, the patient had been taking amitriptyline, hydrochlorothiazide, losartan, and amlodipine.

Medical investigation revealed normal results of transthoracic echocardiogram, tilt table test, Holter and ambulatory blood pressure monitoring. A polysomnogram showed severe OSA with an apnea/hypopnea index of 75.6 events/hour and he presented oxygen desaturation below 90% in almost one quarter of his sleep duration.

Further investigation with cranial magnetic resonance imaging (MRI) and video electroencephalogram was unremarkable. Cervical MRI showed mild upper cervical canal stenosis with no cerebellar tonsils herniation. Study of cervical and intracranial blood vessels with magnetic resonance angiography (MRA) demonstrated hypoplastic vertebral V4 segment and basilar artery associated with fetal posterior cerebral artery (Figure 1). Supplemental screening with transcranial Doppler ultrasound (TDU) and carotid/vertebral duplex scan showed considerable reduction in the diastolic component with an increase of systolic peak speed and abnormal high resistance index for matching age during forced Valsalva maneuver (Figure 2). TDU did not demonstrate any flow reduction after neck rotation. The exam, yet, did not trigger any symptoms.

Laughter is an emotional reaction which generates spasmodic contraction of thoracic muscles and the diaphragm in response to humorous stimuli that activates several neuronal pathways involving the hypothalamus, temporal cortex, basal ganglia, pontine reticular formation, periaqueductal grey matter, and the cerebellum.[5,6] Gelastic syncope is a subtype of syncope and it is thought to be neurally mediated.[3,4] The prognosis of the disease depends on the underlying etiology[3], although, fortunately, it has majoritarily a benign prognosis.[6] Vertebrobasilar hypoplasia, such as in our case, has not been reported as the etiology of laughter induced elsewhere.

The current most accepted hypothesis is the Valsalva-like mechanism that triggers neurally mediated reflexes, just like in coughing preceding vasovagal syncope, since both have sustained state of repetitive bursts of progressive and forced expiration.[7]

In normal conditions, there is compensation for these changes on account of cerebral vascular autoregulation mediated by increased sympathetic tone; however, in patients with neurally mediated syncope, the compensatory mechanism fails to prevent hypotension and, thus, diminished cerebral perfusion, which leads to the loss of consciousness. The underlying cause of this disrupted autoregulation comes from inappropriate overstimulation of the parasympathetic system by the left ventricle wall.[3]

Several clinical scenarios could yield this pattern, but the main differential diagnoses are epilepsy, bradycardia or arrhythmias, hypoglycemia, vertebrobasilar insufficiency, and cataplexia. Thus, it is recommended to evaluate the cardiovascular and neurological systems, and family history in order to recognize any risk factors for those diseases. Depending on the findings taken from history and the physical exam, supplementary diagnostic tests, such as laboratorial or imaging studies, should be considered.[3] In our case, we proceeded with cerebral vascular study while only another report[8] has done so; therefore, we recommend that this approach should be used when further clarification of the etiology is needed.

It is important to state that we cannot fully exclude that our patient's finding on MRA is coincidental, since the finding of vertebrobasilar hypoplasia is common in normal individuals. We, however, believe that his vertebral hypoplasia can be contributing with the mechanism underlying gelastic syncope by heightening his autoregulatory impairment.

The treatment consists in behavioral therapy by the avoidance of trigger events, beta-blockers, or alpha-1-agonists.[4] Our patient is currently in use of propranolol and has not presented other syncope episodes.

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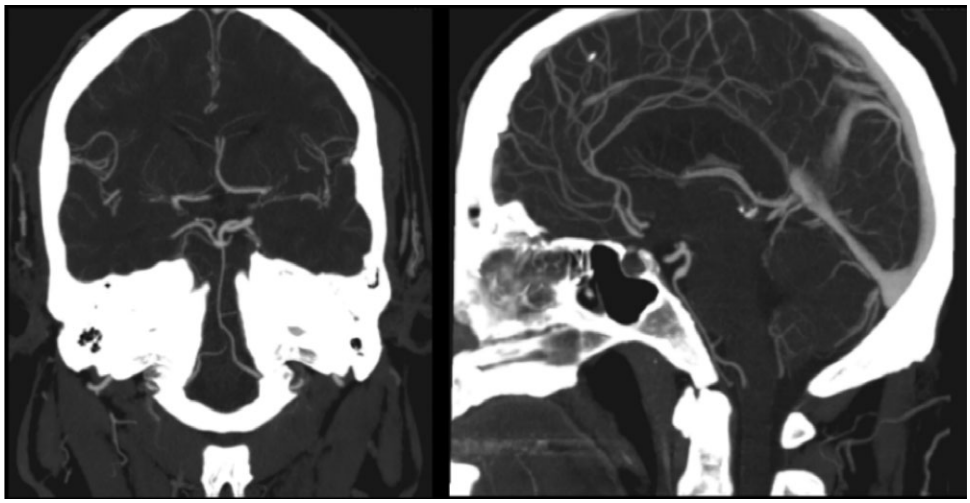

Figure 1: *CT angiography. CT angiography demonstrates diffuse narrowing of the vertebrobasilar arteries, without focal stenosis or other abnormalities, suggesting presumed hypoplasia.*

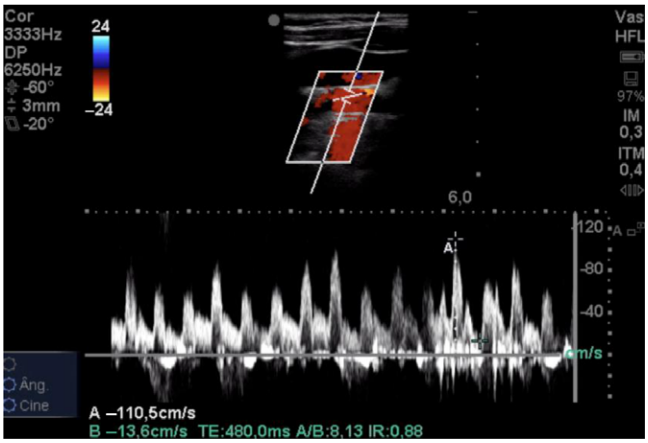

Figure 2: *Vertebral duplex ultrasound. Vertebral duplex scan of the right V1 vertebral showing the hemodynamic changes during the maneuver (variation of the systolic peak, abnormal resistance index, and color mode artifacts due to Valsalva/induced laughing).*

Conflict of Interest

The authors report no disclosures relevant to the manuscript.

Statement of Authorship

E.G.B designed and conceptualized study and drafted the manuscript for intellectual content. D.A.D and A.F.L had a major role in the acquisition of data. S.C.M had a major role in the acquisition of data and revised the manuscript for intellectual content.

Esther Grangeiro Barreto
University of Fortaleza (UNIFOR), Health Science Center, Fortaleza, Ceará, Brazil

Daniel Aguiar Dias
Department of Radiology, Hospital Universitário Walter Cantídio, Fortaleza, Ceará, Brazil

Adson Freitas de Lucena
Department of Neurology, Hospital Geral de Fortaleza, Fortaleza, Ceará, Brazil

Samir Câmara Magalhães
University of Fortaleza (UNIFOR), Health Science Center, Fortaleza, Ceará, Brazil

Correspondence to: Esther Grangeiro Barreto, University of Fortaleza (UNIFOR), Health Science Center, Fortaleza, Ceará, Brazil. Email: esthergbarreto@edu.unifor.br

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