

Pseudobulbar Affect: Mysterious Disorder and Bizarre Behavior

Awad Magbri, MD, Eussera El-Magbri, MD, Mariam El-Magbri, BSC, MPH

Toledo Vascular Access Center, Toledo, OH.

elmagbri@hotmail.com

**Corresponding Author: AwadMagbri MD, Toledo Clinic Inc., Toledo, OH.*

Abstract

The author discussed a case of pseudobulbar affect following a traumatic brain injury with weakness on the right side of the body in a middle aged man. The case highlighted the under-treatment and under-diagnosis of PBA in various neurological disorders and how familiarities with diagnosis and treatment of PBA can make a dramatic changes in patients' lives. The author also discussed the pathophysiology and the treatment of the condition. Research into the pathophysiology of the disorder along with cognitive and drug treatment are desperately needed to understand this mysterious disorder.

Keywords: *Pseudobulbar affect, cerebellum, cognitive dysfunction, pathological laughter and crying, emotional lability, traumatic brain injury, depression.*

CASE HISTORY

The patient is 65 year-old man with past medical history of hypertension and type-2 diabetes both well controlled with medications. He was involved in motor vehicle accident about 18 months ago which resulted in traumatic brain injury (TBI) with weakness of the right side of the body. He had intensive physical therapy with remarkable recovery and almost resolution of the weakness of the right side of the body. However, his family and close friends noticed that the patient has bursts of uncontrollable crying and emotional lability over the last 6-8 months. He describes them as expression of his emotions in the form of bursts of exaggerated crying that does not match what he feels inside since the accident. He denies being depressed. The patient is highly educated and works as freelancing writers in politics and economy. He is multilingual mastering the faculty of speaking and writing in 4 languages. He is well respected in the community and among his associated and friends. He is able to cope with his daily activities with mild residual weakness of the right side of the body. The patient was assessed by his neurologist who diagnosed him as pseudo-bulbar affect (PBA) secondary to the TBI. He is being treated with dextromethorphan/quinidine (20/10mg) with notable improvement in his symptoms and self-confidence.

CASE DISCUSSION

Pseudobulbar affect is a syndrome characterized by uncontrolled bursts of crying or laughing which are considered to be inappropriate to the social situation. Therefore, there is dissociation between the patient's emotional expression and his or her emotional experience. Pseudobulbar affect has been described by many confusing terms like involuntary emotional expression disorders, emotional lability, emotional dysregulation, emotional incontinence, and pathological laughter and crying. Pseudobulbar affect may be encountered in 48% of patients with traumatic brain injury, 39% in Alzheimer's disease and dementia, 28% of patients with stroke, 46% of patients with multiple sclerosis, 50% in patients with amyotrophic lateral sclerosis (ALS), and up to 24% of patients with Parkinsonism (1-8). The impact of the disease is tremendous on the patients, family and the caregivers alike. This will result in restriction of social interaction, withdrawal, and a low quality of life (9). These patients have high degree of anxiety, depression, and poor social functioning (10).

Depression and anxiety are common in patients with PBA affecting up to 35% (11-13). Communications with these patients are difficult at times, and their use of motor functions like swallowing are sometimes

challenging. The recognition of this syndrome by the clinicians along with treatment familiarities are important to have positive impact on the patients' lives(14). The prevalence rate of this syndrome is difficult to estimate, but scoring criteria used for online instruments estimated the prevalence between 9.4-37.5% in various disease processes mentioned earlier (15). This means that PBA is a significant national health issue in the USA, occurring in greater numbers in individuals than those affected by Parkinson's disease, MS, or ALS.

The pathophysiology of PBA is not fully understood, but lack of voluntary control (disinhibition) over the emotions is probably the underlying mechanism. Neuroimaging and neurophysiological studies incriminate cerebellum as a major player in PBA (1,8). The faulty pathways from the motor cortex to the pons and cerebellum are thought to be the underlying culprit in this syndrome. In support of this hypothesis is that patients with cerebellar lesions may demonstrate emotional affect and emotional lability (16,17). One hypothesis is that cerebellum may play a modulating effect between emotions and social circumstances to keep them appropriate and concordant to each other (2,8). The input from the motor cortex, frontal, and temporal lobes passes through the brainstem and into the cerebellum to be modulated in accordance to social settings (8,18). The primary neurotransmitter in PBA is believed to be serotonin and glutamate. The balance between these neurotransmitters is thought to control the emotions according to the social condition.

The clinical features of PBA is characterized by episodes of emotions that are perceived by others as being unprovoked and disconnected from the social circumstances. Bursts of crying that seems to be uncontrolled by the patients is more common than laughing. The degree of emotional response by the patient is often striking. The periods of crying or laughing persists for a considerable period and unable to be suppressed by the patient (12,13,19,20).

The differential diagnosis of PBA should include depression, cerebellar disorders, bipolar disorders, other neurological and psychiatric disorders, and drug-induced effects. Many lability scales and scoring systems have been utilized for the diagnosis of PBA with variable sensitivity and specificities in different disease process associated with PBA. In practice PBA is under-diagnosed and under-treated disorder. This attitude should be improved if we want to change the lives of these patients.

Different medications have been used to treat the condition with variable success, this include; tri-cyclic antidepressants (TCAs), selective serotonin reductase inhibitor (SSRIs), recently dextromethorphan which inhibits the glutamatergic neurotransmitter via action on the N-methyl-D-aspartate receptors and α -1 receptors (21) have been used more frequently. Dextromethorphan is rapidly metabolized by the liver which limits the availability of the drug to the central nervous system where it should act. To decrease this first-pass metabolism of the drug in the liver the compound is administered with a small dose of quinidine sulfate (22), leading to higher and sustained plasma concentrations of dextromethorphan. In October 2010, the US Food and Drug Administration (FDA) approved Nuedexta (dextromethorphan/quinidine) for patients with MS or ALS who have PBA (23).

In conclusion: PBA is recognized more and more in patients with various neurological conditions. The recognition of this syndrome by the clinicians along with treatment familiarities are important to have positive impact on these patients' lives. Research into the pathophysiology of the disorder along with cognitive and drug treatment are desperately needed to understand this mysterious disorder.

REFERENCES

- [1] Parvizi J, Arciniegas DB, Bernardini GL, et al. Diagnosis and management of pathological laughter and crying. *Mayo Clin Proc.* 2006; 81(11): 1482-86
- [2] Parvizi J, Coburn KI, Shillcutt SD, Coffey CE, Mendez MF. Neuroanatomy of pathological laughing and crying: a report of the American Neuropsychiatric Association Committee on research. *J Neuropsychiatry Clin Neurosci.* 2009; 21(1): 75-87
- [3] Cummings JL, Arciniegas DB, Brooks BR, et al. Defining and diagnosing involuntary emotional expression disorder. *CNS Spectr.* 2006; 11(6): 1-7
- [4] Gallagher JP. Pathological laughter and crying in ALS: a search for their origin. *Acta Neurol Scand.* 1989; 80(2): 114-117
- [5] McCullagh S, Moore M, Gawel M, Feinstein A. Pathological laughing and crying in amyotrophic lateral sclerosis: an association with prefrontal cognitive dysfunction. *J Neurol Sci.* 1999; 169(12): 4348

Pseudobulbar Affect: Mysterious Disorder and Bizarre Behavior

- [6] Miller RG, Jackson CE, Kasarskis EJ, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive behavioral impairment (an evidence-based review): report of the quality standards subcommittee of the American Academy of Neurology. *Neurology*. 2009; 73(15): 1227-33
- [7] Strowd RE, Cartwright MS, Okun MS, Haq I, Siddigui MS. Pseudobulbar affect: prevalence and quality of life impact on movement disorders. *J Neurol*. 2010; 257(8): 1382-87
- [8] Miller A, Pratt H, Schiffer RB. Pseudobulbar affect: the spectrum of clinical presentations, etiologies and treatments. *Expert Rev Neurother*. 2011; 11(7): 1077-88
- [9] Colamonico J, Formella A, Bradley W. Pseudobulbar affect: burden of illness in the USA. *AdvTher*. 2012; 29(9): 775-98
- [10] Tateno A, Jorge RE, Robinson RG. Pathological laughing and crying following traumatic brain injury. *J Neuropsychiatry ClinNeurosci*. 2004; 16(4): 426-34
- [11] Calvert T, Knapp P, House A. Psychological associations with emotionalism after stroke. *J Neurol Neurosurg Psychiatry*. 1998; 65(6): 928-29
- [12] Miller U, Murai T, Bauer-Wittmund T, von Cramon DY. Paroxetine versus citalopram treatment of pathological crying after brain injury. *Brain Injury*. 1999; 13(10): 805-11
- [13] Moore SR, Gresham LS, Bromberg MB, Kasarkis EJ, Smith RA. A self-report measure of affective lability. *J NeurolNeurosurg Psychiatry*. 1997; 63(1): 89-93
- [14] Sacco S, Sara M, Pistoria F, Conson M, et al. Management of pathological laughter and crying in patients with locked-in syndrome: a report of 4 cases. *Arch Phys Med Rehabil*. 2008; 89(4): 775-78
- [15] Work SS, Colamonico JA, Bradley WG, Kaye RE. Pseudobulbar affect: an under-recognized and under-treated neurological disorder. *AdvTher*. 2011; 28(7): 586-601
- [16] Schmahmann JD, Sherman JC. The cerebellar cognitive affective syndrome. *Brain* 1998; 12(4): 561-579
- [17] Parvizi J, Joseph J, Press DZ, Schmahmann JD. Pathological laughter and crying in patients multiple system atrophy-cerebellar type. *MovDisord*. 2007; 22(6): 798-803
- [18] Haiman G, Pratt H, Miller A. Brain responses to verbal stimuli among multiple sclerosis patients with pseudobulbar affect. *J Neurol Sci*. 2008; 271(1-2): 137-147
- [19] House A, Dennis M, Molyneux A. et al. Emotionalism after stroke. *BMJ* 1989; 298(6679): 991-994
- [20] Rosen HJ, Cummings J. A real reason for patients with pseudobulbar affect to smile. *Ann Neurol*. 2007; 61(2): 92-96
- [21] Werling LL, Keller A, Frank JG, Nuwayhid SI. A comparison of the binding profiles of dextromethorphan, memantine, fluoxetine and amitriptyline: treatment of involuntary emotional expression disorder. *Exp Neurol*. 2007; 207(2): 248-257
- [22] Piore EP. Current concepts in the pharmacotherapy of pseudobulbar affect. *Drugs*. 2011; 71(9): 1193-1207
- [23] Choi-Kwon S, Han SW, Kwon SU, Kang DW, et al. Fluoxetine treatment in post-stroke depression, emotional incontinence, and anger proneness: a double-blind placebo-controlled study. *Stroke*. 2006; 37(1): 156-161.

Citation: Awad Magbri, MD, Eussera El-Magbri, MD, Mariam El-Magbri, BSC, MPH. *Pseudobulbar Affect: Mysterious Disorder and Bizarre Behavior. Archives of Psychiatry and Behavioral Sciences*. 2018; 1(1): 01-03.

Copyright: © 2018 Awad Magbri, MD, Eussera El-Magbri, MD, Mariam El-Magbri, BSC, MPH. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.