Neurological Case Reports

Case Report
Published: 22 Jul, 2020

A Case of Cervical Dystonia with Pathological Laughter

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Abstract

Dystonia is a movement disorder whose main feature is a sustained or intermittent muscle contraction causing abnormal, often repetitive, movements. Cervical Dystonia (CD) is a type of focal dystonia affecting cervical muscles leading to abnormal postures and movements of the head, neck and shoulders. Pathological laughter and crying is a condition characterized by uncontrollable episodes of laughter and crying. It occurs without any apparent triggering stimulus or in response to a stimulus which had not resulted in cry or laughter prior to the onset of the condition. It is characterized as a disorder of emotional expression rather than disorder of feelings. Here we are presenting the case report of a patient with pathological laughter and cervical dystonia without any other neurological complications.

Keywords: Cervical dystonia, pathological laughter; ET; EL

Introduction

Cervical Dystonia is associated with many co morbid conditions. The lifetime risk for patients with cervical dystonia meeting diagnostic criteria for depression is 15% for anxiety disorders is 26.4%. The prevalence of pathological laughter and crying in individual diagnostic categories was: 4.7% (18/387) of idiopathic Parkinson's disease (PD), 2.7% (2/74) of primary dystonia, 3.1% (2/65) of Essential Tremor (ET) and 7.8% (8/108) of patients with other forms of Parkinsonism. Dystonia is a movement disorder whose main feature is a sustained or intermittent muscle contraction causing abnormal, often repetitive, movements [1]. Cervical Dystonia (CD) is a type of focal dystonia affecting cervical muscles leading to abnormal postures and movements of the head, neck and shoulders [2,3]. Dystonic movements are twisting, and tremulous. It is characterized as a disorder of emotional expression rather than disorder of feelings [4]. The lifetime risk for patients with cervical Dystonia meeting diagnostic criteria for depression is 15% for anxiety disorders is 26.4% [5]. The prevalence of pathological laughter and crying in individual diagnostic categories was: 4.7% (18/387) of idiopathic Parkinson's disease (PD), 2.7% (2/74) of primary dystonia, 3.1% (2/65) of Essential Tremor (ET) and 7.8% (8/108) of patients with other forms of Parkinsonism [6]. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation [7]. The term Emotional Liability (EL) was first described by Pierre-Marie in 1892 [8]. The term Pathological Crying and Laughing (PCL) was used by Wilson in the year 1924, in which he categorized his unifying theory of the disorder [9]. "Pathological crying and laughing," is used interchangeably with "pseudobulbar affect," "emotional lability," and "involuntary emotional expression disorder". It can be differentiated from mood disorders in which laughter or crying is associated with periods of happiness or sadness and from regular laughter and crying which is concurrent with the triggering stimulus [4]. Pathological laughter and crying is associated with common in several neurological conditions including, in order of prevalence: Motor Neuron Disease (MND), Traumatic Brain Injury (TBI), Multiple Sclerosis (MS), stroke, Multiple System Atrophy-Cerebellar type (MSA-C), Alzheimer's disease (AD) and Parkinson's disease (PD) [10-15]. Pathological laughter in a case of cervical dystonia without any degenerating features in the brain is a relatively rare occurrence and hence we report a case.

Case Presentation

A 70 year old female presents to the OPD with complaints of dull aching pain in the nape of the neck and shoulders for 3 years with movement of the head towards the left. She was from a rural area and her occupation is carrying wooden logs on her neck for long distances. Involuntary uncontrollable low pitch laughter for last 8 months. She assumes a sensory trick for getting relieved from the pain and abnormal head pulling by touching her right cheek with her arm. Involuntary sudden outbursts of laughter increased in frequency in the last 3 months. It was elicited by non-specific, trivial and neutral stimuli (such as enquiry about her health) lasted for a few minutes until

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Received Date: *08 Jun 2020*
Accepted Date: *17 Jul 2020*
Published Date: *22 Jul 2020*

Citation:
Vishnupriya V. A Case of Cervical Dystonia with Pathological Laughter. Neurol Case Rep. 2020; 3(1): 1016.

Figure 1: Case of dystonia.

she gained some control. It was not associated with any emotionally provoking situations. Her personal and social behavior was entirely appropriate. She did not have any co-morbid medical illness, no family history of psychiatric or neurological illness and no past history of any psychiatric illness. On examination, left sided sternocleidomastoid is taut and she moves her chin downwards frequently and supports her right cheek with her right arm to mitigate the pain (Geste Antagonist). Systemic examination Findings revealed 60º head rotation to the left and left shoulder elevation and a taut Sternocleidomastoid (Figure 1). Toronto Western Spasmodic Torticollis Rating Scale score was 42. Fundus examination was normal. The following psychological assessment scales were applied. University of Florida Modified PBA Screening Question test was positive. CNS-LS score was 13. MMSE score was 20. Beck Depression Inventory and the Beck Anxiety Inventory score dint showed significant scores for depression and anxiety. Base line investigations done were normal. Vitamin B12 levels were normal and thyroid function test revealed a normal T3 and T4 levels , but TSH was 8.8 ng/ml. EEG done during laughter was normal. CT Brain showed Ill defined hypo dense areas seen involving bilateral periventricular white matter suggestive of small vessel disease and MRI Brain confirmed Small vessel disease. She was started on Selective Serotonin Reuptake Inhibitor tablet sertraline 50 mg, tablet clonazepam 0.5mg, tablet baclofen 20 mg. Neurologist opinion was sort and advised tablet trihexyphenidyl 6 mg divided doses, Tab. Aspirin/Atorvastatin 75/10 mg. Botulinum toxin in the subsequent visits if dystonic movements persisted. After one week, frequency of her laughter episodes reduced. CNS-LS scores were 8. After 1 month of discharge, she came for follow up, laughter episodes were there only occasionally and pain in the neck reduced dystonic movements reduced in intensity.

Discussion

Cervical dystonia also known as spasmodic torticollis is the most common type of focal dystonia. It usually begins between the ages of 30 and 50 years, often with initial neck stiffness and restricted head mobility. Neck and shoulder pain occur in 75 percent of cases. Sensory tricks (Geste Antagonist) such as lightly touching the face or chin reduce the severity of symptoms in most patients. The differential diagnosis includes essential head tremor, Tardive dystonia in which retrocollis is common, anterocollis caused by cervical myopathy or multiple system atrophy, and secondary torticollis associated with neck injury, atlantoaxial dislocation, cervical disk disease, spinal-cord neoplasm, or soft-tissue infections of the neck [16]. Our case report is unique, as fixed dystonia with pathological laughter has few reports till date. On examination there was no focal neurological sign. It was a focal, static, isolated with onset at late adulthood. There was no history of psychiatric treatment. Overuse Cervical Dystonia (CD) should be suspected when typical symptoms and signs of CD develop in the context of chronic repetitive use or overuse of cervical muscles, especially where exacerbating tasks involve asymmetric postures. Cervical Dystonia (CD) has rarely been associated with chronic overuse, with very few case reports [17]. Absence of neurodegenerative changes was again a fascinating point in this patient as pseudobulbar effect is commonly associated with neurodegenerative disorders like motor neuron disease or multiple sclerosis. Anatomical correlates between cervical dystonia and pathological laughter is the involvement of basal ganglia and cerebellum. Cerebellum plays a vital role in modulating emotional responses and keeps them appropriate to the social situation and to the patient's mood based on input from the cerebral cortex. Disruption of corticopontine-cerebellar circuits results in impairment of this cerebellar modulation, causing pathological laughter. Motor cortex including the frontal and temporal cortices sends inputs to brainstem which is modulated by the cerebellum. The motor input is mediated by inhibitory input from the somatosensory cortex. Reduced inhibitory input results in disinhibition of the cerebellum, leading to pathological laughter [18]. Studies prove that patients with cervical dystonia had an abnormal cerebellar cortical connectivity at rest and the plasticity of this cerebellar pathway is also altered [19]. Even in the absence of overt cerebellar signs, cerebellar dysfunction was identified in most patients with cervical Dystonia [20]. Though we are unable to establish any basal ganglia or cerebellum function in our patient through MRI, still higher imaging techniques like fMRI can be done to establish a neurobiological correlate.

References

1. TT Warner. Dystonia: Clinical Features, Diagnosis and Treatment. 2010;1-13.
2. Chan J, Brin M, Fahn S. Idiopaqthic cervical dystonia: Clinical characteristics. Mov Disord. 1991;6(2):119-26.
3. Dauer WT, Green P, Fahn S. Current concepts on clinical features, aetiology and management of idiopathic cervical dystonia. Brain. 1998;121(4):547-60.
4. Parvizi J, Anderson SW, Martin CO, Damasio H, Damasio AR. Pathological laughter and crying-A link to cerebellum. Brain. 2001;124(9):1708-19.
5. Ozel-Kizil ET, Akbostanci MC, Ozguven HD, Atbasoglu EC. Secondary social anxiety in hyperkinesias. Mov Disord. 2008;23(5):641-45.
6. Siddiqui MS, Fernandez HH, Garvan CW, Kirsch-Darrow L, Bowers D, Rodriguez RL, et al. Inappropriate crying and laughing in Parkinson disease and movement disorders. World J Biolog Psychiatry. 2009;10(3):234-40.
7. Albanese A, Bhatia KP, Bressman SB, DeLong MR, Fahn S, Fung SCV, et al. The Phenomenology and classification of dystonia: A consensus update. Mov Disord. 2013;28(7):863-73.
8. Marie P, Lecons Sur Les Maladies De La Moelle. Paris: G Masson. 1892.
9. Wilson SAK. Original papers. Some Probl Neurol. 1924;299-333.
10. Engelman W, Hammond FM, Malec JF. Diagnosing pseudo bulbar affect in traumatic brain injury. Neuropsychiatr Dis Treat. 2014;10:1903-10.
11. Patel N, Combs H, York M, Phan C, Jimenez-Shahed J. Pseudobulbar affect correlates with mood symptoms in Parkinsonian disorders but not amyotrophic lateral sclerosis. J Neuropsychiatry Clin Neurosci. 2018;30:214-9.
12. Parvizi J, Schiffer R. Exaggerated crying and tremor with a cerebellar cyst. J Neuropsychiatry Clin Neurosci. 2007;19:187-90.
13. Vidovic V, Rovazdi MC, Kraml O, Kes VB. Pseudobulbar affect in multiple

sclerosis patients. Acta Clin Croat. 2015;54(2):159-63.

14. Brooks BR, Crumpacker D, Fellus J, Kantor D, Kaye RE. PRISM: A novel research tool to assess the prevalence of pseudobulbar affect symptoms across neurological conditions. PLoS One. 2013;8(8):72232.

15. Phuong L, Garg S, Duda JE, Stern MB, Weintraub D. Involuntary emotional expression disorder (IEED) in Parkinson's disease. Parkinson Relat Disord. 2009;15:511-5.

16. Tarsy D, Simon DK. Current concepts Dystonia. N Engl J Med. 2006;355:818-29.

17. Hogg E, Tagliati M. Overuse Cervical Dystonia. A case report and literature review. Tremor Other Hyperkinet Mov (NY). 2016;6:413.

18. Ahmed A, Simmons Z. Pseudobulbar affect: Prevalence and management. Ther Clin Risk Manag. 2013;9:483-89.

19. Porcacchia P, Álvarez de Toledo P, Rodríguez-Baena A, Martín-Rodríguez JF, Palomar FJ, Vargas-González L, et al. Abnormal cerebellar connectivity and plasticity in isolated cervical Dystonia. 2019;14(1):211367.

20. Philip P, Lungu O, Bares M. Dystonia and the cerebellum: A new field of interest in movement disorders? Clin Neurophysiol. 2013;124(7):1269-76.