Case Report

Clinical Presentation and Short-Term Prognosis of Faciobrachial Dystonia

Hossain R[1], Raknuzzaman M[2], Mia MS[3], Miah MZ[4], Khan MZR[5]

Conflict of Interest: None
Received: 15.08.2023
Accepted: 01.09.2023
www.banglajol.info/index.php/JSSMC

Abstract: *Wide range of disorders fall into the intersection between Psychiatry and Neurology. Movement Disorders namely dystonia, chorea, tics, tremors and myoclonus often raise dilemma between coarse brain pathology and functional disorder. Faciobrachial Dystonia due to autoimmune encephalitis presents with dystonic movements of face and arm; memory impairment and psychiatric symptoms which may create uncertainty between the diagnosis of neurologic disorder and psychogenic movement disorder.*

This is a case of a middle-aged lady who presented in the chamber of a psychiatrist with irrelevant speech, jerky movement of right side of face and hand and forgetfulness of recent events. MMSE score at the presentation were 20. Her symptoms were not responsive to conventional anti-epileptic, anxiolytic or sedative-hypnotics. Thus, she was advised an MRI on the basis of which she was referred to a neurologist. On the basis of clinical presentation, physical and mental state examination and laboratory investigation, the lady has been diagnosed as a case of Faciobrachial Dystonia due to anti LGI 1 autoimmune encephalitis. She was given Methyl Prednisolone 1 gm daily for 5 days followed by oral methyl Prednisolone and rapid tapering within 1 month. Following high dose steroid treatment, the frequency of her dystonic movements improved robustly.

Psychiatrists are often the first point of contact for the patients with movement disorder. Knowledge and skills for early diagnosis and timely referral makes the care more comprehensive for the patients. Moreover, they can collaborate with neurologists and other healthcare professionals to provide integrated, multidisciplinary care that addresses both the physical and mental health needs of the patients.

Key Words:
Faciobrachial Dystonia, Dystonia, Movement Disorder, Psychiatry, Neurology

[J Shaheed Suhrawardy Med Coll 2023; 15(2): 49-51]
DOI: https://doi.org/10.3329/jssmc.v15i2.81867

Authors:

1. Redwana Hossain, Assistant Professor, Department of Psychiatry, Shaheed Suhrawardy Medical College Hospital,Dhaka
2. Md Raknuzzaman , Junior Consultant (Neurology), National Institute of Neuroscience,Dhaka
3. Md. Sajib Mia , Registrar, Department of Psychiatry, Mymensingh Medical College Hospital,Mymensing
4. Mohammed Zubayer Miah , Assistant Professor, Department of Psychiatry, Shaheed Suhrawardy Medical College,Dhaka
5. Muhammad Zillur Rahman Khan , Associate Professor, Department of Psychiatry, Shaheed Suhrawardy Medical College,Dhaka

Correspondence:

Redwana Hossain, Assistant Professor, Department of Psychiatry, Shaheed Suhrawardy Medical College
Email address: hossainredwana@gmail.com

Introduction

Intersection between neurologic and psychiatric disorder has been increasingly studied in recent years. Psychiatrists and neurologists often encounter the cases with movement disorders. Tremor, tics, chorea, myoclonus and dystonia are most prevalent movement disorders presented to neurologists. On the other hand, catatonia and psychotropic induced movement disorders often present to psychiatrists. Thus, the diagnostic dilemma often oscillates from coarse brain pathology to functional disorder. Genetics, infection, toxins, metabolic disorder, vascular event, brain injury or adverse effect of prescribed medication may predispose or precipitate movement disorder. Autoimmune encephalitis is an infectious disorder of brain that often presents with overlapping features of psychiatric and neurological symptoms. New classifications of autoimmune encephalitis based on mechanism-related autoantibodies herald a new era which bridges neurology, psychiatry, immunology and synaptic biology[1].

Among autoimmune encephalitis, N-methyl-d-aspartate receptor (NMDAR) and anti-leucine-rich glioma-inactivated 1 protein (LGI1) are most common auto antibodies directed against cell surface proteins. Anti LGI1 autoim-

mune encephalitis often presents with clinical picture that may mimic to psychogenic movement disorder. These group of autoantibody related encephalitis are relatively treatment sensitive[2]. Early detection of such seropositive autoimmune encephalitis assures enhanced patient outcomes and minimizes disability. Thus, the aim of this case study is to present a case of Facio brachial dystonia who has been diagnosed as a case of psychogenic movement disorder for long before confirmation of the neurologic diagnosis.

Case Presentation

Mrs. F, a 39 years old Muslim married woman from Dhaka visited to the chamber of a psychiatrist (the first author) with the complaints of forgetfulness, stereotyped movement of right side of head, face and arm and repeated utterance of "I feel uneasy inside head". Her symptoms started with headache on June 2022 which was gradually increasing. Approximately 3-4 months after the onset of headache, she has developed jerky movement of right side of face, right forearm and hand which was also increasing. Initially the movement occurs 5-6 times a day. At this stage Mrs. F went to an internist who advised her EEG and CT scan of brain, the report of which were unremarkable. The physician prescribed her conventional antiepileptic (valproate) with the suspicion of possible seizure. However, the dystonia getting worse day by day. After 2 months of onset, the dystonia becomes persistent throughout the day. Moreover, her family members state that she has been losing attention and forgetful of basic things (asking her unmarried daughter "where is your husband?"). Considering this as an irrelevant speech, family members of Mrs. F brought her to the chamber of the psychiatrist. On query it has been revealed that she has been also suffering from insomnia from the beginning.

On mental state examination, tremor of right side of face and arm was observed; mood was anxious; speech was evolving around her headache; thought, perception, judgment and insight were intact. All other general and neurologic examination revealed no abnormality. Abnormal head, face and arm movement associated with headache and forgetfulness, not responding to conventional antiepileptic drug raises suspicion of the neurological origin. Thus, she has been advised Mini Mental State Examination (MMSE) and MRI of brain (Fig 1 and 2).

MMSE score was 20 on the first day. MRI of brain was suggestive of chronic toxic/ hypoxic-ischemic encephalitis and mild degenerative cerebral cortical atrophy with temporo- parietal predominance. Based on the clinical presentation, psychometric evaluation and MRI report, the patient has been referred to a neurologist (2nd author). Neurologist conducted the general physical examination and neurological examination but nothing significant was found except tremor and dystonia. Her laboratory examination including liver function, renal function, serum copper and ceruloplasmin tests, were within normal range. On the basis of clinical presentation, physical and mental state examination and laboratory investigation, Mrs. F has been diagnosed as a case of Faciobrachial Dystonia most likely due to autoimmune encephalitis. She was given Methyl Prednisolone 1 gm daily for 5 days followed by oral methyl Prednisolone and rapid tapering within 1 month. Following high dose steroid treatment, the frequency of her dystonic movements improved robustly.

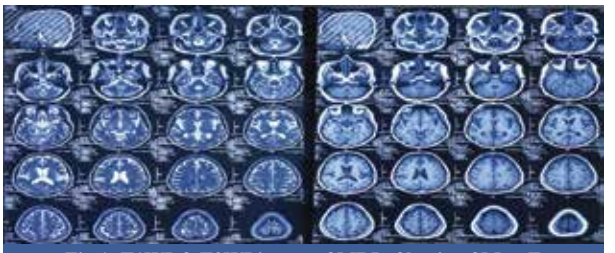

Fig 1: T1WI & T2WI image of MRI of brain of Mrs. F

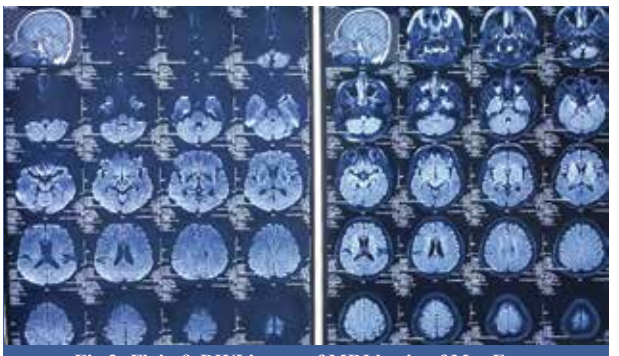

Fig 2: Flair & DWI image of MRI brain of Mrs.F

During her follow up visit after one month, Mrs F shows no dystonic movement, memory function improves (MMSE 25) and sleep has been restored.

Discussion

Movement disorders due to neurological abnormality often does not exhibit any hard sign. Structural brain abnormalities are one of the important causes of movement disorder. Etiologically, there is disconnection between Limbic cortex and motor cortex which lead to dystonia[3]. On the contrary, psychogenic movement disorders are not related to any structural changes. The precise

mechanism behind psychogenic movement disorder is not clearly known and it is postulated that stressor causes disconnection between brain and body and the person become unaware of his voluntary movement. Presenting features of both types of movement disorder often raises clinical dilemma between psychogenic and neurologic functional disorder.

Faciobrachial dystonia is an immunotherapy responsive, brief and focal type of movement disorder often preceding the cognitive impairment4. Though exact mechanism of Faciobrachial Dystonia (FBD) is unknown, from neuroimaging studies, it is evident that FBD seizure arises from motor cortex and basal ganglia. Cognitive impairments were the most prevalent related condition, with seizures follow a clinical characteristic[2].

Unusual clinical presentation and initially normal laboratory reports often mislabel Faciobrachial dystonia as functional or psychiatric origin[5]. Autoimmune antibody testing and response to immunotherapy are important markers for diagnosing autoimmune encephalitis. Pleocytosis brain abnormalities on T2-weighted and fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging (MRI) confined to medial temporal lobes are another finding commonly detected in autoimmune encephalitis[6].

Weishuai (2018) suggests FBD seizure occur early and have a high incidence in LGI1 antibody encephalitis and upon definitive diagnosis, immunotherapy be initiated immediately[7]. As autoimmune antibody panel testing is time consuming and expensive, thus clinical evaluation and MRI findings remain the earliest convenient diagnostic tool. Irani (2012), investigated the response of immunotherapy in FBD and observed excellent short-term outcome of dystonia4. As Mrs. F has been suffering from movement disorder and cognitive impairment for long 8 months, so based on her presentation only MRI of brain has been advised. As the MRI is very much suggestive of autoimmune encephalopathy, so treatment begun immediately. Rapid resolution of symptoms confirms the diagnosis of Faciobrachial Dystonia due to autoimmune encephalitis.

One important limitation of the study is unavailability of the autoimmune encephalitis panel as patient party refused to do the test. However, as other clinical parameter and imaging study is very much supportive of the diagnosis, so the treatment initiated which ultimately alleviate patient's suffering.

Conclusion

Psychiatrists plays a vital role in the comprehensive care of patients with movement disorders. When patients present at early stage of movement disorder and clinical examination and baseline investigation reveal no abnormality, psychiatric disorders become first possibility. Psychiatrists should be cautious to deal such cases and do not hesitate to do extensive evaluation.

Early interdisciplinary referrals ensure proper management and lessens the suffering of patients. Psychiatrists possess the expertise to assess and manage the emotional, cognitive, and behavioral aspects associated with these conditions. Moreover, they can collaborate with neurologists and other healthcare professionals to provide integrated, multidisciplinary care that addresses both the physical and mental health needs of the patients

Reference

1. Zandi MS. Defining and treating leucine-rich glioma inactivated 1 antibody associated autoimmunity. Brain. 2013 Oct;136(Pt 10):2933-5. doi: 10.1093/brain/awt256. PMID: 24072484.
2. Siriratnam P, McArthur L, Chen Z, Kempster P and Monif M (2023) Movement disorders in cell surface antibody mediated autoimmune encephalitis: a meta-analysis. Front. Neurol. 14:1225523. doi: 10.3389/fneur.2023.1225523
3. Cummins G, Zandi M, Barker RA. Movement disorders and psychiatry: Five new things. Neurol Clin Pract. 2015 Apr;5(2):143-149. doi: 10.1212/CPJ.0000000000000113. PMID: 29443187; PMCID: PMC5764446.
4. Irani SR, Stagg CJ, Schott JM, Rosenthal CR, Schneider SA, Pettingill P, Pettingill R, Waters P, Thomas A, Voets NL, Cardoso MJ, Cash DM, Manning EN, Lang B, Smith SJ, Vincent A, Johnson MR. Faciobrachial dystonic seizures: the influence of immunotherapy on seizure control and prevention of cognitive impairment in a broadening phenotype. Brain. 2013 Oct;136(Pt 10):3151-62. doi:10.1093/brain/awt212. Epub 2013 Sep 6. PMID: 24014519.
5. Flanagan EP, Kotsenas AL, Britton JW, McKeon A, Watson RE, Klein CJ, Boeve BF, Lowe V, Ahlskog JE, Shin C, Boes CJ, Crum BA, Laughlin RS, Pittock SJ. Basal ganglia T1 hyperintensity in LGI1-autoantibody faciobrachial dystonic seizures. Neurol Neuroimmunol Neuroinflamm. 2015 Oct 1;2(6):e161. doi: 10.1212/NXI.0000000000000161. PMID: 26468474; PMCID: PMC4592539.
6. Simabukuro MM, Nóbrega PR, Pitombeira M, Cavalcante WCP, Grativvol RS, Pinto LF, Castro LHM, Nitrini R. The importance of recognizing faciobrachial dystonic seizures in rapidly progressive dementias. Dement Neuropsychol. 2016 Oct-Dec;10(4):351-357. doi: 10.1590/s1980-5764-2016dn1004016. PMID: 29213481; PMCID: PMC5619277.
7. Li, Weishuai & Wu, Si & Meng, Qingping & Zhang, Xiaotian & Guo, Yang & Cong, Lin & Cong, Shuyan & Zheng, Dongming. (2018). Clinical characteristics and short-term prognosis of LGI1 antibody encephalitis: A retrospective case study. BMC Neurology. 18. 10.1186/s12883-018-1099-z.