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Review Article

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Prototypical Variability Meets Phenomenology in Movement Disorders : A Bench to Bed Approach in Clinical Parkinsonism

Massod Vadiee BS, MD. PhD*

Queen Mary University of London, United kingdom Jagiellonian University Medical College, International Department

***Corresponding Author**
Massod Vadiee BS,MD.PhD, Queen Mary University of London, United kingdom Jagiellonian University Medical College, International Department, United states.

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Abstract

The movement disorders are chronic neurodegenerative group of diseases such as; Parkinson's disease, Huntington's disease, tremor, and dystonia which are increasingly becoming more prevalent. The establishing of their classification, accurate diagnosis and clinical management can be a difficult and lengthy process in part due to their complex natural history and heterogeneous presentations. Unfortunately, the under recognition and under diagnosed movement disorders are also common partly as a result of the clinicians inadequate understanding of disease phenomenology and prototypical variability and challenges related inability to differentiate functional and secondary movement disorders from organic movement disorders. In addition to challenges of clinicians knowledge and attitudes and the lack of coordination and obstacles in the dynamics of care pathways that link the primary care services with the higher specialists care can significant have adverse impacts on of quality of the chronic neurological care delivery needed among the vulnerable patient population. A unique group of conditions that are commonly encountered by the community healthcare providers are the secondary movement disorders or the mimickers of movement disorders that include the drug-induced and the systemic or metabolically-induced movement disorders which present both as a diagnostic and management challenge and opportunity for the general practitioners and community neurologist alike.

Keywords: Parkinson's Disease, Parkinsonism, Dystonia, Tremors, Movement Disorders; Chronic Neurological Care, Secondary Movement Disorders; Drug and Metabolically -Induced Movement Disorders, Clinical Encounters

1. Introduction

The management of patients with movement disorders require an integrated and responsive service that can provide accurate diagnosis,appropriate treatment, a sustained long term care and if needed timely referral. With increasing aging population and the inadequate access to chronic neurological care, novel care delivery approaches are needed. The principal component of any quality healthcare system is the timely access to a patient-centered care and this may vary considerably depending on the type of care sought, patient populations profile the geography. The failure to accurately diagnose neurological conditions may result in delayed treatment and patient's unnecessary loss of daily function and productivity. The complex interface of care pathways between patient's point of contact with the primary care settings and the complementary care services including neuropsychiatry, neuroradiology, palliative care, physiotherapy and rehabilitation are also either not optimized or unable to meet patient's needs and expectations.

The availability of a sustained chronic care among patients Parkinson's, tremors and dystonia and other with movement disorders is fundamental and currently, limited data is available on the level of integration and coordination that is embedded in our care pathways and in particular between rural community practitioners and the specialist care. Surprisingly, in comparison with other medical disciplines and specialties, the specialized neurological l care particularly at the community level appears to be least integrated into the overall healthcare delivery system. With respect to the movement disorders there is an urgent need for innovative approaching delivery and service utilization to address the current gaps and ensure truly patient-centered outcomes. For majority patients living with movement disorders such as Parkinson's, Dystonia, Tremors and other forms of chronic neurological conditions the access to primary care for initial clinical assessment and referral to specialists essential.

Moreover the clinicians interpretations of patient chief complaints, history, clinical examination and the ability to distinguish and differentiate functional and secondary movement disorders from organic in movement disorders requires a considerable clinical knowledge and diagnostics acumen. This transition from primary to secondary care pathway services can be a costly and stressful experience for patients, while the outcomes can vary with prac-

titioner's knowledge, attitudes, and the availability of coordinated care between primary and tertiary specialized centers. For a favorable management of movement disorders, a focused initial assessment of patient's early clinical presentations by the primary care team and timely referral for further investigations can have a major impact on disease progression and eventual outcome. Currently, no clear policy exists on the preferred pathway of care and the extent by which the primary care sector should be involved in management of patients with movement disorders and chronic neurological conditions.

2. Key Points

Establishing classification and understanding phenomenology are crucial in arriving at a diagnosis of movement disorders. The phenomenology is determined from the specific combination of the dominant movement disorder(akinetic–rigid and hyperkinetic) and presence of additional neurological or non-neurological abnormalities.

• A systematic approach is recommended when approaching patients with movement disorders. The drug and metabolically – induced movement disorders as mimickers that are commonly encountered in community healthcare setting and provide an opportunity for the skilled general practitioners' to evaluate before proceeding to seek second opinion and specialist referrals.

• To ensure optimal clinical outcome and a patient-centered care patients we should foster clinicians enhanced knowledge and attitudes and scale up coordinated care pathways that link the primary and the higher specialist care levels into the overall ecology of long term chronic neurology services

3. Method

We decided to use a mini-review to search of literature in order to uncover the novel causal factors that exist between the classification sand diagnostics aspect of chronic neurological care. In particular,with regards to the movement disorders we were also interested in the exhibiting literature on topics related to clinician's knowledge and attitudes, the neurological are integration into primary care pathway. We intended to explore and describe the challenges related to possible neurological service gaps experienced by patients when navigating between general practitioners and specialist neurologist services in the community healthcare setting. We performed a pub med search of published literature from 2015 to 2015. The key search terms included;neurological care, movement disorders, primary care, neurology specialist referral practitioners' knowledge and attitude. We also performed a second literature search for publications that had addressed the non-neurological secondary movement disorders such as; systemic and metabolic-induced and or drug-induced movement disorders commonly encountered by community neurologist and general practitioner in primary healthcare centers

4. Challenges of Classifications and Understanding the Phenomenology

Movement disorders (i.e.; Parkinson's disease, tremor,Huntington's disease dystonia, monoclonal, tics, etc) are a related group of neurological conditions that previously referred to extraterrestrial disorders' and anatomically originate from basal ganglia, although clinical phenomenology plays a more important role than anatomic location in classification and diagnostic schemes. These abnormal movements maybe voluntary or involuntary and categorized as either hyperkinetic or hypo kinetic voluntary and automatic movements that are not unrelated to weakness or plasticity. The movement disorders furthermore can be categorized as either primary, or secondary depending on the underlying cause, with primary movement disorders the abnormal movement considered as the primary manifestation of the disorder, while in secondary movement disorders, a broader systemic, structural, metabolic, toxic or inherited factor might be the etiological culprit [1,2].

Currently biological investigations on animal models have shown insights into the pathophysiology of movement disorders and understating of the natural history of movement disorders and other neurodegenerativeconditions. Currently, the clinical examinations based on our understanding the phenomenology of the abnormal movement are the pillars of our diagnostic approach, and the therapeutic management remain largely symptomatic with select cases the radio-surgery and deep brain stimulation have shown some promising results [2-4].The epidemiological studies show that a fifth of all movement disorders were diagnosed to be probably drug-induced, and Parkinsonism and increasing trend in clinical burden indicates need for increased resource allocation, education and training meet the increased demand for movement disorders specialist as community neurology practitioners in resource limited community healthcare settings [5-7].

The Parkinson's disease and other common movement disorders are usually neurodegenerative progressive,disabling conditions and associated significant economical burden and reduction of patients daily activity and quality of life. Anesthesiology are not exclusively motor but also non motor features(i.e.; seborrhea) with psychiatric sequel which are stigmatizing impairing patients emotional well-being and can exert substantial economic burden on patient and caregivers [8-10]. The general classification of movement disorders is a complex and diagnostic task is often compounded by its clinical phenotype variability. Nonetheless, attention to pattern recognition such as; akinetic–rigid,or hyperkinetic movements or presence or absence of jerky character will often lead the clinician to diagnosis of movement disorders.

Although the complex and mixed-patterns movement disorders require a sophisticated understanding the phenomenology of the clinical syndrome, and additional systematic search for presence of dominant abnormal movement in conjunction with neurological or non-neurological presentations are required in order to arrive at an accurate diagnosis [11,12]. WITH the clinical heterogeneity phenotype diversity that is commonly observed among patients with movement disorders the diagnostic value of a detailed systematic approach is quite justifiable. The search for phenomenology of the movement disorders is grounded in an age appropriate focused clinical examination and relevant information on quality of disease onset, progression, and possibility of existing drug toxicity [12,13].

The classification and categorizing movement disorder patient scan be cumbersome task using current diagnostic criteria without additional euro-pathological and psychiatric investigations. Therefore, detailed examination and lengthier clinical investigations are often needed in order to accurately differentiate between patients who might represent an atypical presentations of movement disorder or those who indicate a population that is concurrently unrecognized or under-diagnosed [14,15]. A large cohort study of patients with Parkinsonism showed that 74.7%, were drug-induced Parkinsonism followed by vascular Parkinsonism other rare sporadic, genetic,infectious etiologies [16-18]. Therefore in the absence of a biologically meaningful biomarkers or reliable gold standard test it is of paramount importance to anticipate the prodromal phase of movement disorders either with motor, no-motor or cognitive symptoms among patients with atypical variant of Parkinson's disease and ensure early accurate diagnosis [17-20]

A number of studies illustrate that with regards to the management of movement disorders, adhering to diagnostic criteria and timely recognition of prodromal clinical signs and symptom scan potentially yield moderate to high predictive power of the likelihood for an earlier diagnosis, adequate neuro-cognitive support and timely referral and long term care planning [21-25]. A coordinated service delivery service link between the primary care physicians and higher specialist neurologists would allow systematic and comprehensive evaluation of patients that could investigate memory, cognitive, perceptual-motor as well as balance and gait. This requires a reform in the manner by which [24-26].The new data and technology driven neuro-radiological diagnostic sand artificial inelegance have inspired new pathways of delivering chronic neurological care and also opportunities for novel practice of neurology as a discipline [27-29]. Additional challenges continue to exist with respect to the traditional medical education curriculum and its failure to adequately train and sensitize a more knowledgeable generation of residents and community based neurology practitioners that can manage patients with movement disorders and other long term neurological conditions [30-33].

5. Movement Disorders Mimickers: Drug Induced and Neuro-Metabolic Movement Disorders

A wide range of conditions, both neurological and non-neurological can mimic various movement disorders, therefore itis vital for the clinicians to systematically approach patients who present with one or more types of movement disorder. Among the diverse movement disorders, the secondary dystonia, and overstatements are a good illustrations of pathological and genetic heterogeneity associated with secondary movement disorders and may etiologically be associated with exogenous processes including focal and diffuse neuronal damage, impaired systemic metabolism, regulations noxious substances, and toxic effects of therapeutics drugs that invariably can lead to a pathological process known as oligonucleotide repeats processes [34,45]. The gerontological nature of movement disorders like pseudo-dystonia psychogenic dystonia require a broader knowledge of natural history of these conditions and a clinical precision in establishing the differential diagnosis to successfully recognize and diagnose different clinical scenarios [34-36].

The current research on phenomenology and clinical presentations of movement disorders (Parkinson's disease, Dystonia,tremors; etc) indicate that they commonly precede with nonspecific symptom presentations that include; tremor, memory and cognitive decline depression fatigue, dizziness, urinary dysfunction, seborrhea,constipation and gait and balance abnormalities.. Therefore robust clinical knowledge and a judicial anticipations of nonspecific presentations early in the course of the disease by the primary care physicians is essential for earlier diagnosis and referrals [37-40].While in Parkinson's disease and other movement disorders the core etiologies are vascular infections, and or space-occupying lesions, with secondary movement disorders the underlying cause maybe psychogenic and drug induced which often missed or under–recognized by the primacy providers [39,40].

The psychogenic movement disorders as mimickers of organic movement disorders present with normal laboratory and imaging tests and routinely exhibit bizarre gaits, tremor, dystonia, paroxysmal and jerkiness that could involve face, neck, trunk or limbs the diagnosis is based on paroxysmal nature and variability of tremor direction, and swaying gait and balance without falling [41-43]. The dug-induced movement disorders are commonly the result of dopamine receptor-blocking agents (Levodopa neuroleptics,-calcium channel blockers, illicit drugs) causing dyskinesias dystonia,hyperkinetic or hypokinetic movement disorders. With the aging populations and higher rate of comorbidity and pharmacotherapy potential risk of drug-induced movement disorders among the elderly requires for a careful risk assessment and a comprehensive review of current medications by primary care physicians should be of considerations [44-46].

Another group of therapeutics that precipitate drug-induced movement disorders are anti psychotics,anti epileptics, antimicrobial, arrhythmic, gastrointestinal drugs that are causative agents for Parkinsonism and the extra pyramidal side effects such as tremor, chorea-ballistics, dystonia, tar dive dyskinesia,myoclonus, and tics [46-48]. The drug-induced movement disorders are among the most commonly encountered and at the same time under-recognized conditions by the primary practitioners, with the younger patients typically presenting with acute presentations and elderly exhibiting tardive or sub-clinical Parkinsonism. In the absence of a definitive laboratory and imaging tests for movement disorders,the clinicians awareness of clinical presentation, knowledge of risk populations and the ability to distinguish the drug-induced Parkinsonism from Parkinson's disease remains a challenge particularly in the communities with limited access to higher levels of care pathways and [49-52].

6. The Hepatic and Metabolically-Induced -Movement Disorders

Movement disorders may also be under recognized when arising in the context of systemic disease and metabolic disorders. Abnormal movements may be the initial manifestation of a systemic disease,with the pro-inflammatory mediators and microbial act-

ing as triggers that accelerating metabolic and biochemical dysregulationsin the central nervous system [53,54,66]. Therefore ascertaining movement phenomenology with acute or sub-acute presentations and unexplained prodromal clinical and psychiatric symptoms might suggest that a systemic process such as metabolic, endocrine infectious, autoimmune diseases might be the underlying cause of the movement disorders [54-56]. The movement disorders arising in the context of a broad range of metabolic disorders or inborn errors of metabolism present with acute and sub-acute ataxia, hyperkinetic,hypokineticr or rigid movement syndrome.

Among the systemic metabolic syndromes, the cardiovascular and the diabetes mellitusType 2 disease are known as risk factors for secondary movement disorder as these metabolic syndromes compromise neuronal function due to hyperglycemia and pro-oxidative states undermine neuronal protection which lead to unique movement disorders [57-59]. The current research evidence increasingly point to the brain-gut macrobiotic axis and the role that the gut innate immune system might have as a result of up-regulation of inflammatory cascades and enteric neurological cells that hinder the neuronal protection in genesis of the motor and cognitive symptoms among patients with Parkinson and other progressive movement disorders [60, 61].

The dystonia are among the movement disorders that frequently can be the manifestation of metabolically conditions that clinically present with paroxysmal onset factorial movements involvement with associated neurological and extra-neurological features that can prompt the clinician to consider a more diligent history and particular attention to phonotypical presentation of the particular movement disorder [62-64]. The underlying physiological cause of metabolically induced movement disorders broadly associated with hepatic syndromes role in causing mitochondrial a acidemias, purine-creatine metabolism and lipid storage disuse that lead to permeability of the blood-brain barrier noninflammatory induction of circulating cytokines encountered in hepatic encephalopathy, myelopathy, and cirrhosis-relatedparkinsonism [65-67].

Finally, in the Wilson disease as an inherited metabolic diseases causing excess upper buildup can cause a distinct category of neurodegenerative abnormal movement disorders that in clinically may present with chorea, dystonia, myoclonus, tremor,and Parkinsonism in children, while on the other hand the inborn abnormal iron deposition also with genetic etiology can present withextrapyramidal movement disorder without intellectual disability disorders [68-71]. A timely recognition and characterization of these secondary movement disorders in the context of age of onset and clinical presentation is essential and requires addressing the underlying metabolically disorder [72-73].

7. Conclusion

The movement disorders are a group of chronic neurodegenerativeconditions that increasingly are becoming more prevalent and continue to be a considerable economical burden on healthcare system. The establishing of classification and an accurate diagnosis of movement disorders can be a difficult process in part due to complex and heterogeneous presentations and its natural history. There are indications that among undeserved populations such as elderly with less access to specialized neurological care in general he chronic neurological conditions and specifically the movement disorders areunrecognized or under-diagnosed. The complex interface of care pathways often lack integration and the an optimal management,treatment and continuous and coordinated care for patients with movement disorders remains a challenging task as it require snot only a well trained knowledgeable and sensitized primary care team but also a cross disciplinary care pathway that is capable of providing timely evaluation and specialist referral. A unique group of conditions that are neurological and non-neurological that mimic various movement disorders and are either the drug or metabolically-induced movement disorders are commonly encountered in community healthcare setting, providing both a challenge and an opportunity for general practitioners' to evaluate and potentially manage before referrals. More research is needed to explore the current extent and availability of coordinated care that exists between primary and higher specialist care for a sustainable care aimed at patient with movement disorders. Moreover, an innovative approaches vital for novel approaches into the organization and integration of community neurology services and into the overall ecology of the healthcare system.

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