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A Review on Some Associated Disorders with Chorea Based on a Neurobiochemical Perspective

Saberi B*

Medical Research, Iran

***Corresponding author:** Behzad Saberi, MD, Medical Research, Esfahan, Iran, Tel: 989131263487; Email: sab64b@yahoo.com

Review Article

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Abstract

Chorea which is characterized by random, brief, irregular and arrhythmic movements which would start from a body part and go to another one, in most of the cases has association with various secondary features like parakinesis, athetosis, ballism, partial suppressibility, motor impersistence, gait and deep tendon reflex abnormalities. The movements in chorea have low amplitude in general and all of the muscles can be engaged in such unpredictable movements. There are some disorders which have association with chorea. This brief review tries to point to some disorders which have association with chorea from a Neurobiochemical perspective.

Keywords: Chorea; Associated Disorders; Differential Diagnosis; Inherited; Acquired; Sporadic; Neurobiochemical Perspective

Abbreviations: HDL1 & HDL2: Huntington's Disease-Like One and Two; MELAS: Mitochondrial Encephalopathy with Lactic Acidosis and Stroke-Like Episodes; PLAN: Phospholipase A2G6-Associated Neurodegeneration; PKAN: Pantothenate-Kinase-Associated Neurodegeneration; vCJD: variant Creutzfeldt-Jakob Disease; AHD: Acquired Hepatocerebral Degeneration; CBD: Corticobasal Degeneration.

Chorea which is characterized by random, brief, irregular and arrhythmic movements has association with some disorders. Having knowledge about the differential diagnosis of disorders which have association with chorea is of importance both in clinical and basic settings. In a general classification of disorders which have association with chorea, two main categories can be defined as "inherited" and "acquired" or "sporadic".

Inherited disorders can be sub classified into X-linked, mitochondrial, autosomal dominant and autosomal recessive disorders. Acquired or sporadic disorders can be sub classified into metabolic, neoplastic, infectious, vascular, endocrine, immune-mediated, drug-induced and miscellaneous ones [1-3]. Leigh's syndrome which causes seizures, developmental delays, cardiac issues and difficulties in breathing and mitochondrial encephalopathy with lactic acidosis and stroke-like episodes or MELAS are among the mitochondrial disorders. Lesch–Nyhan syndrome which is caused by hypoxanthine-guanine phosphoribosyltransferase enzyme deficiency which occurs by HPRT1 gene mutations, McLeod syndrome which is caused by XK gene mutations and Lubag syndrome or X-linked dystonia Parkinsonism, are among the X-linked disorders.

Dentatorubropallidoluysian atrophy, Benign hereditary chorea, Huntington's disease, Neuroferritinopathy, Huntington's disease-like one and two (HDL1 and HDL2), Glucose transporter type one or GLUT1 deficiency and types one, two, three and seventeen of Spin cerebellar ataxia are among the autosomal dominant disorders.

Ataxia-telangiectasia, Wilson's disease, Friedreich's ataxia, Neuroacanthocytosis, Huntington's disease-like three (HDL3), Ataxia with oculomotor apraxia, Phospholipase A2G6-associated neurodegeneration or PLAN, Pantothenate-kinase-associated neurodegeneration or PKAN which was formerly called Hallervorden–Spatz syndrome, Aceruloplasminemia, Propionic acidemia or Propionic aciduria or Propionyl-CoA carboxylase (PCC) deficiency and Glutaric aciduria are among the autosomal recessive disorders. Among the autosomal recessive disorders, the PLAN, PKAN and Aceruloplasminemia are among the Neurodegeneration with brain iron accumulation or NBIA disorders [1,3-5]. Lymphoma, Toxoplasmosis, Human immunodeficiency virus encephalopathy, Mycoplasma encephalitis, viral encephalitis, Cysticercosis, Neurosyphilis and variant Creutzfeldt-Jakob disease or vCJD are among the infectious disorders. Vascular malformations haemorrhage or infarction in subcortical regions and basal ganglia and Moyamoya are among the vascular disorders.

Acquired or Non-Wilsonian hepatocerebral degeneration or AHD, hyperglycaemia and hypoglycaemia, deficiency in vitamin B12 and disturbances in the electrolytes are among the metabolic disorders. Central nervous system lymphoma, Brain tumours and Paraneoplastic syndromes are among the neoplastic disorders. Hypoparathyroidism, Hyperparathyroidism, Chorea gravidarum which is a movement disorder which rarely may be developed during pregnancy and Hyperthyroidism are among the endocrine disorders. Antiphospholipid antibody syndrome, Sydenham's or Rheumatic chorea which occurs after being infected with specific type of bacteria which would be Streptococcus group A, Central nervous system vasculitis, Paraneoplastic syndromes, Systemic collagen vascular disorders and Systemic lupus erythematosus or SLE are among the Immune-mediated disorders.

Multiple sclerosis, Hypoxic-Ischemic encephalopathy, Polycythaemia Vera which is an uncommon myeloproliferative neoplasm in which excessive production of red blood cells would be done by the bone marrow, Corticobasal degeneration or CBD, Postpump chorea or PPC which would be choreoathetoid movements development within two weeks following cardiopulmonary bypass, Alzheimer's disease and Toxins are among the miscellaneous disorders.

Neuroleptic antipsychotics, Amphetamines, Oral contraceptives, Methadone, Lithium, Levodopa, Dopamine Agonists, Valproate, Ethanol withdrawal, Gabapentin, Prochlorperazine, Cocaine, Metoclopramide, Amphetamine, Carbamazepine, Lamotrigine and Phenytoin are among the drug-induced causes for development of the chorea [6-8].

Conclusion

It is important for clinical neuroscientists and neurobiochemists to have knowledge about the disorders which can accompanied by chorea. Having such knowledge is of importance to consider the disorders which may present themselves with chorea with or without other signs and symptoms and approach the affected patients with such disorders more accurately and with more precision at the bedside.

Having this knowledge is also of importance for basic scientists to study about chorea, relevant pathologies and pathophysiology with more precision in basic study settings.

The author recommends studying other references in neuroscience, neurobiology and neurochemistry to gain more knowledge about the introduced pathologies in this study which discussing more about them in more detail is beyond the scope of this short review.

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