Commentary

Chorea in adults may be caused by distinct etiologies such as: neurodegenerative, structural, metabolic, pharmacological, or autoimmune.[1] Determining the cause of chorea and deciding on a cost-effective investigation process can be a challenge, due to the wide diversity of causes. Neurodegenerative diseases, such as Huntington's disease, its phenocopies (Huntington's disease like 1, 2, and 3, dentatorubro-pallidoluysian atrophy, and spinocerebellar ataxia type 17), and the neuroacanthocytosis syndromes, usually present with chronic generalized chorea in the context of other neurological symptoms.[2]

A subgroup of patients with chorea present with hemichorea, likely caused by acquired diseases or structural lesions within the basal ganglia.[2] Hemichorea is frequently associated with proximal, wide and violent movements of the limbs, namely hemiballism; thus, the term hemichorea-hemiballism is often applied. In patients with hemichorea, the time of onset and progression may be an important clue to investigation. Acute onset hemichorea is more likely related to ischemic stroke or intracranial hemorrhage involving the basal ganglia.[3]

Another important cause of acute hemichorea is hyperglycemia, mainly in the context of nonketotic hyperglycemia. The pathophysiology of this condition is not completely understood, but high signal in the contralateral putamen in T1-weighted images on magnetic resonance imaging (MRI) is a typical finding.[4] Whether this abnormality is related to an increased local cerebral blood flow, hyperviscosity, excitotoxicity, or inflammation remains unclear.

Expansive lesions within the basal ganglia are also well known causes of hemichorea, usually with a gradual or subacute onset. Among them, infectious granulomas and brain tumors need to be considered.[2] Infectious granulomas are often seen in HIV-positive patients; they can be caused by toxoplasmosis, tuberculosis, cryptococcus, neurosyphilis, and other opportunistic agents.[5] These infections can also occur in immunocompetent patients.

Neoplastic diseases can be related to chorea through distinct mechanisms: Brain expansive lesions and surrounding edema disrupting the striatopallidothalamocortical

motor circuitry, or paraneoplastic syndrome manifesting with chorea. In the former, focal limb chorea and contralateral hemichorea are expected,[2] whereas the latter usually manifests with generalized chorea.[6] Among brain tumors, primary central nervous system lymphoma, pituitary macroadenoma[7] and cavernous angioma[8] have already been associated with hemichorea.

Paraneoplastic chorea is a rare condition and its phenotype has not been completely elucidated, so far. For this reason, consideration of paraneoplastic etiology in patients with otherwise unexplained chorea is reasonable. A recent study showed that patients with paraneo-plastic chorea have a mean age at onset of 72 years, male predominance, and generalized moderate to severe chorea. Weight loss and peripheral neuropathy often go along with the movement disorder in these cases. Some onconeural antibodies are highly predictive of cancer, for instance CRMP-5 IgG, ANNA-1, ANNA-2, and amphiphysin.[9] In contrast, patients with autoimmune chorea (non-paraneoplastic) are younger (mean age 45) and female. They present with mild chorea associated with behavioral or cognitive symptoms. Coexisting autoimmune diseases including systemic lupus, antiphospholipid syndrome, and Sjogren are common in these patients.[9]

In this volume, the authors report a case of "*Hemichorea and dystonia due to frontal lobe meningioma*" in a 65-year-old female.[10] According to the description, she has presented with a combination of left hemichorea and dystonia, in the absence of hemiballism. Her symptoms developed along many years. The MRI showed a large meningioma in the right brain hemisphere compressing the right caudate, putamen, and deep white matter. Other causes of acquired chorea were excluded through laboratorial tests, however, a definite cause–effect mechanism could not be proven.[10]

Although the tumor has been implied in the abnormal movements and structural epilepsy in this case, a conservative approach was adopted.[10] For most cases of acquired hemichorea, treatment of underlying disease results in improvement of the involuntary movements. Moreover, it can spontaneously resolve after weeks of months (e.g. in cases of stroke). In patients with persistent chorea or untreatable diseases, dopamine blocker agents, such as antipsychotics, or monoamine depleting drugs, such as tetrabenazine, can be used.

In conclusion, a detailed medical history, and neurological exam can be important guides to the investigation of patients with chorea. Laboratorial tests, including glucose levels, auto-antibodies, and serology are often indicated. Neuroimaging is mandatory in focal and hemichoreic syndromes.

Camila Catherine Aquino

Movement Disorders Unit, Universidade Federal de Sao Paulo, Sao Paulo, Brazil

Address for correspondence:
Dr. Camila Catherine Aquino,
Discipline of Neurology, 715 Napoleao de Barros St, 2nd floor.
Vila Clementino. Sao Paulo - SP, Brazil. 04024-002.
E-mail: camilacathe@gmail.com

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