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LETTERS TO THE EDITOR
CASE REPORT

Idiopathic basal ganglia calcification as a differential diagnosis of parkinsonism: A case report in an older adult

Dear Editor,

We present the case of a 76-year-old man who was admitted to the emergency room of Dario Fernandez General Hospital, ISSSTE (Mexico City, Mexico) with slurred speech of a few hours of onset. His relevant background included hypertension since he was 68 years-of-age without current treatment, and a posterior cervical laminectomy because of medullar injury when he was 72 years-of-age.

During the physical examination, motor aphasia was found. His gait was slow, with short steps and reduced arm swing; his trunk bent forward and there was slight flexion of the neck. Increased muscle tone in the pelvic limbs and muscle stretch reflexes were also found. There were no signs of cerebellar involvement, and the plantar extensor reflex was present. In order to assess parkinsonism, a Unified Parkinson's Disease Rating Scale (UPDRS) was carried out, resulting in a score of 17 points. No behavioral or psychiatric symptoms were found.

Because of these findings, a cranial tomographic scan was carried out (Fig. 1). Symmetrical and bilateral nodular calcifications in the basal ganglia, and the cerebellar dentate nuclei were reported, without evidence of ischemic or hemorrhagic stroke.

In order to discard secondary sources of calcinosis; laboratory investigations were carried out: prostate-specific antigen 1.27 ng/mL, parathyroid hormone 71 pg/mL, calcium 8.8 mg/dL, phosphorus 3.30 mg/dL, magnesium 2.03 mg/dL, alkaline phosphatase 74 IU/L and uric acid 4.3 mg/dL. Consensus among the faculty staff concluded that the final diagnosis was transient ischemic attack and idiopathic basal ganglia calcification (IBGC). The patient was prescribed treatment and appointed for follow-up in 3 months.

This entity has a frequency, in the general population, of less than 0.5%, and some authors considered it a rare disease.[1] In 1930, Karl Theodor Fahr described an 81-year-old patient with a long history of dementia, decubitus ulcer and "immobility without paralyses". Post-mortem examination of the brain showed brain calcifications localized in the semiovale centrum and the striatum.[2]

There are a number of names given for this disease including Fahr's disease bilateral striopallidodentate calcinosis or IBGC (which is codified in the International Classification of Diseases as G23.8), among others; however, the most common name today is IBGC. The main characteristic is bilateral symmetrical calcification of the basal ganglia, thalamus, cerebellum, dentate nucleus and brain hemispheres,[3] in the absence of other metabolic causes of calcinosis[1] – such as hypoparathyroidism.[4] Although still considered idiopathic, there is some evidence of an association with genetic abnormalities; in particular with an autosomic dominant transmission.[5,6]

The clinical characteristics of this condition are circumscribed to movement (mainly parkinsonism), but

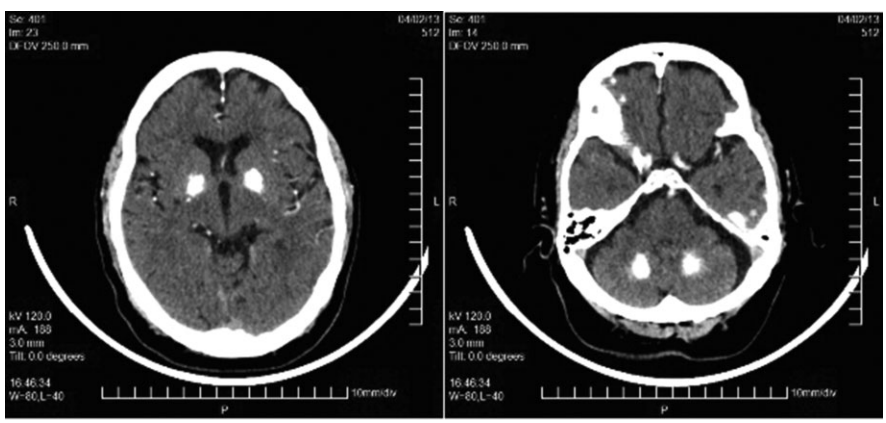

Figure 1 Computed tomography scan of the brain, tranversal vision showing the calcifications in the basal ganglia and in the cerebellar dentate nuclei.

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several reports have described cognitive impairment and cerebellar manifestations.[3]

Diagnosis depends on the combination of clinical manifestations, neuroimaging and discarding secondary sources of calcinosis. Regarding brain calcinosis, it has been reported with the following dimensions: basal ganglia 1.39 ± 0.28 cm^3, thalamus 0.26 ± 0.06 cm^3, dentate nucleus 1.02 ± 0.34 cm^3 and centrum semiovale 0.64 ± 0.22 cm^3.[2]

Calcifications representative of IBGC are also presented in another entity that shares clinical signs found in the present patient (parkinsonism); diffuse neurobrillary tangles with calcification (DNTC), a tauopathy characterized by slowly progressive presenile dementia and neuropsychiatric manifestations.[7,8] Even though it is a differential diagnosis for the case presented here, there are some clues that point to IBGC rather than DNTC. It is more frequent in the Asian population, almost all reported cases have either cognitive or neuropsychiatric manifestations, and images in DNTC have a shorter diameter and are slightly heterogenous.[8] Nevertheless, because of the frequency of dementia in older adults, this entity should always be considered when finding bilateral calcifications in brain images of older adults; and follow up is entitled in order to recognize any new symptomatology.

We consider the dissemination of information about rare diseases in the elderly population important not only because such publications in this age group are scarce, but also because in the specific case of IBGC this should be considered as a differential diagnosis of geriatric syndromes, such as cognitive impairment (Alzheimer's disease) and gait/movement disorders (Parkinson's disease, sarcopenia, frailty).

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RESEARCH STUDIES

Central benzodiazepine receptor imaging in Alzheimer's disease with cerebrovascular disease

Dear Editor,

Alzheimer's disease (AD) with cerebrovascular disease (CVD) is a common cause of dementia in the elderly.[1] However, the clinical distinction between AD with CVD and mixed Alzheimer's/vascular dementia (MD) can be difficult in clinical practice. In our previous single photon emission computed tomography (SPECT) study, regional benzodiazepine receptor (rBZR) reduction was found predominantly in the frontal lobe of vascular dementia (VaD) and MD compared with AD.[2] In the present study, we examined rBZR and regional cerebral blood flow (rCBF) using SPECT in AD with CVD and MD patients.

Eight AD with CVD patients and seven MD patients underwent SPECT studies with N-isopropyl-p-[^{123}I]-iodoamphetamine and ^{123}I-iomazenil (IMZ) to measure rCBF and rBZR. Six MD patients in a 2012 study[2] were included in the MD group. AD with CVD was defined as a typical AD associated with clinical and radiological evidence of CVD in the deep gray and white matter.[3] MD was defined as possible AD with clinical and imaging features indicative of subcortical VaD according to the criteria for MD in the 4th edition of the Diagnostic and Statistical Manual of Mental Disorders.[4] The measurement of rCBF and rBZR using SPECT is described elsewhere in detail.[2] To assess rCBF and rBZR changes, the ratio of each value in several brain regions

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