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CORTICO-BASAL GANGLIONIC DEGENERATION

A CASE REPORT

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SUMMARY — The case of a Brazilian patient with cortico-basal ganglionic degeneration (CBGD) is presented. Since three years ago, a 71-year old male displays asymmetric ideomotor apraxia, gait apraxia, cortical sensory impairment, myoclonus, limp dystonia and rigidity. His mental status is spared. There is neither consanguinity nor similar cases in his family. The differential diagnosis of CBGD is discussed. A brief review of the literature is made stressing the clinical and pathological features of CBGD. This disease is poorly known and probably underdiagnosed. Its diagnosis can be safely made based on clinical grounds.

KEY WORDS: cortico-basal ganglionar degeneration, clinical diagnosis, management.

Degeneração ganglionar córtico-basal: registro de caso.

RESUMO — É apresentado o caso de um paciente brasileiro com degeneração córtico-ganglionar basal (DCGB). Há três anos um homem de 71 anos de idade apresenta-se com apraxia ideomotora, apraxia de marcha, déficit sensitivo cortical, rigidez, mioclonias e distonia apendicular. Todos estes sinais são assimétricos. Seu estado mental está preservado. Não há consangüinidade ou casos semelhantes na família. O diagnóstico diferencial desta nova entidade é discutido. Também é feita breve revisão da literatura, dando ênfase aos aspectos clínicos e patológicos da DCGB. Esta doença é mal conhecida e, possivelmente, pouco diagnosticada. O diagnóstico pode ser feito, com segurança, baseado em dados clínicos.

PALAVRAS-CHAVE: degeneração córtico ganglionar basal, diagnóstico clínico, conduta.

Watts et al.[20], in 1985, coined the term cortico-basal ganglionic degeneration (CBGD) to designate a disease characterized by involvement of basal ganglia and specific cerebral cortical regions. However, the clinical and pathological features of this entity was recognized in 1968 by Rebeiz and colleagues [16]. Since then, over twenty cases have been described [6]. We report on a Brazilian patient with CBGD.

CASE REPORT

MGB, 71 years-old, male, retired. At the age of 68, the patient noticed that his right leg was clumsy and stiff, leading to frequent falls. The gait difficulties kept increasing and by the age of 69 he was unable to walk without assistance. By this time, the right upper and the left lower limbs became also involved. From then on, he has been confined to a wheelchair. In the last two years his disease has been slowly but relentlessly progressive. On attempting to walk, his legs won't move and there is a tendency to fall backwards. It has become impossible for him to write and take food to the mouth with his right arm. He complains that the right side of his body does not obbey his mind. Additionally, abnormal movements have

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appeared in the right leg. He described them as involuntary spasms which produce flexion of the knee and the hip. These movements were particularly frequent at night, interfering with his sleep. There were no sphincter disturbances. Except for poliomyelitis in childhood, which resulted in a sequela in the left leg, and mansoni schistosomiasis previously treated, his past history was unremarkable. Consanguinity and similar cases in his family were denied.

At the time of examination, he was depressed but collaborative. His general examination was normal. He had a score of zero on the 6-item test of orientation-memory-concentration [9] and of 26 points on the minimental status examination [3]. In the latter he had problems with tasks requiring writing and drawing. The cranial nerves, including fundus oculi and the external ocular motility, were spared. The mandibular reflexes were exacerbated. The glabelar, snout, palmomental reflexes, and an intense bilateral grasp were observed. The alien limb sign was noted in the right arm, which also displayed a dystonic posture. Action myoclonus was present in both legs and reflex myoclonus was noted in the right side of the body, i.e., there was a clearcut dissociation between the performance of voluntary and automatic movements. The former, impaired; the latter, intact. Attempts to move the left leg evoked involuntary and similar movements in the right leg. Due to severe gait apraxia, the patient could not walk. The muscle strength was normal, except for the distal lower left limb muscles, which were atrophic and weak due to poliomyelitis. The deep reflexes were normal and the plantar responses were equivocal. The pressure, pain, and temperature sensation were spared. There were mild hypopallesthesia, severe hypographesthesia, and hypobaresthesia in the right limbs. The hand-face test was positive in the right.

Two cerebral computed tomographies, in the first and in the third year of the disease, were normal, as well as routine hematological and biochemical tests. A levodopa/carbidopa trial was ineffective in relieving patient's problems.

COMMENTS

Our patient shows features suggestive of cortical and basal ganglia injury. A distinctive characteristic of his disease is its asymmetry. The right side of the body was much more involved. The preservation of basic sensations with loss of cortical modalities indicates parietal lobe lesion [2]. Injuries to this region can cause apraxia, although it can also result from secondary motor areas involvement [2]. Focal action and reflex myoclonus are commonly from cortical origin [13]. The genesis of alien limb is not clear but it seems to express parietal lesion [6]. In spite of its pathogenesis still being disputed, dystonia is usually associated with basal ganglia lesions [8].

Several diseases can combine cortical and basal ganglionic features. Among them are Alzheimer's disease, Pick's disease, prions-related disorders, multiple system atrophy, progressive supranuclear palsy, and CBGD [6]. Obviously, the patient does not meet the diagnostic criteria of Alzheimer's disease [11]. The lack of dementia, defined by the patient's score on the 6-item test of orientation-concentration-memory [9] and on the minimental status examination [3], makes this diagnosis impossible. However, non-Alzheimer's disease forms of cerebral atrophy have been recently described. They include dementia of frontal-lobe type and lobar atrophy [12]. But both are characterized by absence of subcortical signs, and are thought to represent focal forms of Alzheimer's disease [7,12]. Diffuse Lewy body disease has been growingly recognized as cause of simultaneous cortical and subcortical injury [12]. However, its evolution is often characterized by dementia and dysautonomia [10]. The normal mental status rules out Pick's disease or prions-related disorders. The former usually displays severe temporal signs, and extrapyramidal involvement is not prominent [1]. Although Creutzfeldt Jakob disease is often accompanied by myoclonus, its cause is invariably rapid and with severe dementia [14]. In multiple system atrophy, cortical involvement is seldom found and its clinical expressions are quite different from the picture of our patient [15]. Progressive supranuclear palsy requires for its diagnosis the presence of down-gaze supranuclear ophthalmoplegia [5]. Other features of this disease, also absent in our patient, are dementia, pseudobulbar palsy, and axial dystonia.

CBGD is a disease characterized by asymmetric involvement of parietal lobes, expressed by loss of cortical sensation and apraxia [1,6,16-18]. The alien limb is an invariable finding in this disorder [6,18]. Extrapyramidal manifestations like myoclonus, dystonia, and rigidity are common findings of CBGD [4,6,17,18]. Its course is relentlessly progressive, slow, and exquisitely characterized by sparing of the mental status until the late stages of the disease [4,6,16,18]. Pathologically, there is cortical, striatal, and

nigral lesions consisting of neuronal dropout as well as features of swelling, pallor, and achromasia [6,16-18]. Another striking characteristics are the asymmetric distribution of the lesions and the presence of eosinophilic cytoplasmatic inclusions [4,6,16], which were termed corticobasal inclusions by Gibb et al.[4]. These authors stress that these inclusions are unique to CBGD and conclude that, in spite of neuropathological similarities to Pick's disease, the features of CBGD are distinct. Several drugs, like levodopa, bromocriptine, trihexyphenidyl, benztropine, ethopropazine, amantadine, propanolol, diazepam, clonazepam, valproic acid, baclofen, dantrolene sodium and piracetam have been used with little or no success in treating these patients [6,18]. Our patient is still alive. But comparing his clinical characteristics with the data found in the literature, we can reliably conclude that his diagnosis is CBGD. Several authors [6,17-19] emphasize that this disease can be readily diagnosed on the basis of clinical findings alone. CBGD is a poorly known and likely misdiagnosed disorder [19]. The increasing knowledge of its features will probably lead to recognition of cases.

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