Procerus Sign in Corticobasal Degeneration

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In a recent description, Romano and Colosimo (1) named, for the first time, a typical facial expression seen in a progressive supranuclear palsy (PSP) patient "the procerus sign". This sign may be due to a focal dystonia of the procerus muscles, as well as to facial bradykinesia, and it presents vertical wrinkles in the glabella region. This facial expression has been described as "astonished," "worried," or "reptile-like" expression in PSP.

We also have had a case of clinically probable corticobasal degeneration (CBD) with the procerus sign. The relationship between PSP and CBD has yet to be clarified (2, 3), but this sign may be important in the association of these diseases clinically. Therefore, we give a summary of our case. This is the first reported case of procerus sign in CBD.

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The patient is a 66-year-old woman who experienced left hand clumsiness at age 62. Parkinson's disease was suspected. She was treated with L-DOPA, but it was ineffective. At age 64, a neurological examination detected left dominant bilateral limb-kinetic apraxia and cortical sensory disturbance that included agraphesthesias, agraphia, dystonia of the left upper limb, rigospastica, and hyperreflexia. There was no gaze palsy. Brain MRI showed asymmetric cortical atrophy, which was more severe on the right (Fig. 1A). Despite this, her midbrain was normal. CBD was diagnosed. The disease has gradually worsened, and now at age 66, paretic dysarthria and gait disturbance have appeared, and her limb dystonia has intensified. She shows masked face and the procerus sign occurs without concomitant blepharospasm (Fig. 1B). Brain MRI reveals cortical atrophy progression. But her midbrain is normal in size and shape for her age (Fig. 1C).

CBD is characterized clinically by ideomotor apraxia, limb dystonia and asymmetric akinetic-rigid syndrome (3). Descri-
tions of facial expressions in CBD are few except for nonspecific signs of parkinsonism: masked face, oily face, Myerson phenomenon, and forced mouth opening reaction (4). On the other hand, Jean Martin Charcot commented on facial bradykinesia and contracted forehead muscles in atypical Parkinson’s disease in the nineteenth century (5). Drawings from his original lesson showed frightened expressions in these patients. This facial expression, however, has not been well characterized.

We agree with Romano and Colosimo (1) that “the procerus sign” should be considered a newly identified neurological sign. The present patient’s procerus muscle showed contracting and mounding as a focal dystonia, similar to “the procerus sign”. We believe that our patient’s facial expression reflects focal dystonia because it appeared with the progression of limb dystonia, and it is also apparent when she blinks or smiles. The etiology of her expression may be the same as that of “the procerus sign” in PSP. Electromyography (EMG) should provide useful information about what this sign represents.

PSP and CBD have characteristics in common neuropathologically and genetically, e.g., the tau haplotype (2). Both diseases are neuropathologically characterized by selective 4-repeat tau deposition. Houlden et al (2) described two extended tau haplotypes (designated H1 and H2) that cover the entire tau gene. Tau haplotype and genotype were analyzed in PSP and CBD cases, and the frequency of H1 and H1/H1 was significantly increased in both diseases. This suggests that they have the same etiology. Little, however, is known about the clinical relationship of these diseases, except for nonspecific parkinsonism (3). Moreover, CBD is considered a heterogeneous disorder presenting parkinsonism and dementia. For neurologists, “the procerus sign” should prove a significant clinical tool for showing the association of CBD and PSP.

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