The procerus sign was described by Romano and Colosimo (1) in two patients with progressive supranuclear palsy (PSP), being defined as the appearance of vertical wrinkles in the glabellar region and the bridge of the nose due to dystonic contractions of the procerus muscle, associated with a combination of reduced blinking, spasticity, lid retraction, and gaze palsy. This occurs because the procerus muscle originates in the nasal bone and inserts in the skin in the center of the forehead between the eyebrows (1).

Recently, two letters (2, 3) have been published suggesting that the name “procerus sign” is erroneous, because the vertical wrinkles in the forehead are not formed by contractions of the procerus muscle, but rather by those of the corrugator supercilius muscle. The procerus muscle acts by drawing down the medial angle of the eyebrows, producing the transverse wrinkles over the bridge of the nose. The reply from Romano and Colosimo (4) was as follows: the peculiar contractions of the upper facial muscles in PSP patients are complex and not easily explained by simple contraction of the procerus muscle, where the corrugator supercilius and orbicularis oculi muscles are also involved. They stated that their aim was not to detail the complex anatomy of the muscles involved in the human facial expression, but simply to draw attention to a distinctive semiological aspect of PSP using the procerus sign, thus with no need to withdraw the name of the procerus sign (4).

Are the peculiar facial expressions with vertical wrinkles in the forehead associated with “astonished”, “worried” or “reptile-like” features, specific to PSP? Warabi et al (5) have recently reported similar features in a patient with clinically diagnosed corticobasal degeneration (CBD).

The patient was a 66-year-old woman who showed progressive neurologic deteriorations since the age of 62, including left dominant bilateral limb-kinetic apraxia, cortical sensory disturbance, agraphia, dystonia of the left upper limb, rigidospasticity and hyperreflexia. Additionally, a mask-like face with prominent vertical wrinkles in the glabellar region was noted. Brain MRI revealed asymmetric cortical atrophy, more severe on the right, without marked atrophy of the mid-brain. The clinical features of this patient were compatible with those of the classical form of CBD (6, 7). The ordinary clinical criteria for the classical form of CBD had approximately 100% specificity for the diagnosis of pathological CBD, while sensitivity was less than 50% (8). CBD shows a wide spectrum of clinical symptoms, and there is a group of patients with early aphasia, dementia, or behavioral disorders without laterality in motor disturbances, which Litvan et al (9) called atypical CBD. In the latter cases, the diagnosis of CBD is difficult clinically, although the concentrations of tau protein in CSF could be a clinical biological marker of CBD (10).

In my collection of photos of the faces of patients with clinical PSP or CBD (5 PSP and 7 CBD patients) who were admitted to our neurologic ward, one PSP patient and two CBD patients showed the procerus sign (unpublished data). The procerus sign was marked in one CBD patient and mild in one PSP patient and in another CBD patient. Therefore, the procerus sign may not be uncommon in both diseases with differing severity in each patient. Of course, the vertical wrinkles in the forehead associated with blepharospasm are common in Meige’s syndrome. Blepharospasm with increased blinking seen in Meige’s syndrome excludes the procerus sign (1). The vertical wrinkles in the forehead are sometimes present in healthy people, some of whom ask for cosmetic treatment using a local injection of botulinum A toxin (11). It should be taken into consideration as to what degree of vertical wrinkling in the forehead is abnormal.

PSP and CBD share common pathology, especially the existence of neurofibrillary tangles in the substantia nigra and glial tangles (coiled bodies and argyrophilic threads) in the cerebral cortex (12). Moreover, 4-repeat tau isoforms are common in the brains of both CBD and PSP patients (13). Clinically, CBD is sometimes misdiagnosed as PSP. Morimatsu and Negoro (14) surveyed clinically diagnosed (probable) and pathologically confirmed (definite) CBD patients in 29 neurologic institutions in Japan in 2001, where 151 clinical cases and 13 autopsy proven cases were collected. Clinical diagnosis of the 13 autopsy cases was CBD in 8 (62%), PSP in 3 (23%) and other disorders in the remaining 2, thus demonstrating clinical differentiation between CBD and PSP is occasionally difficult. Dystonia appears in both diseases, which in the early stage is marked in the limbs in CBD and in the axial muscles in PSP, later becoming generalized in both. The site of lesions responsible for dystonia is still unestablished; the basal ganglia, caudate, lentiform nucleus, thalamus or cerebral cortex has been suggested to be responsible in various kinds of
dystonia (15). It is possible that the procerus sign as a focal dystonia appears in both diseases.

The following problems remain to be solved: 1) the name of this sign; the procerus sign or the corrugator supercilii sign, 2) a diagnostic significance whether it appears in PSP and CBD alone, and finally 3) the responsible site of lesion for this focal dystonia.

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References