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Opercular Syndrome Without Opercular Lesions: Foix-Chavany-Marie Syndrome in Progressive Supranuclear Motor System Degeneration

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Summary. A patient is described with slowly progressive supranuclear motor system degeneration (primary lateral sclerosis) characterized by pure bulbar spasticity for six years until a spastic tetraparesis developed. Clinically and electrophysiologically there was and still is no evidence of lower motor neuron involvement. Recently the patient presented with a syndrome of complete inability to move face and tongue voluntarily with preservation of the ability to move them "automatically", e.g. within gestures: automatic-voluntary motor dissociation. Loss of voluntary innervation of the facio-pharyngo-glosso-masticatory muscles with preservation of involuntary innervation are the features of the Foix-Chavany-Marie syndrome, which to date has most often been described in association with bilateral vascular lesions of the opercula or their cortigofugal projections.

Key words: Foix-Chavany-Marie syndrome – Opercular syndrome – Automatic-voluntary dissociation – Motor system degeneration – Primary lateral sclerosis

Introduction

The essential feature of amyotrophic lateral sclerosis (ALS) is the combination of lower motor neuron signs, i.e. wasting, atrophy, and fasciculations, and upper motor neuron signs, i.e. spasticity, enhanced reflexes, and pathological reflexes, without sensory findings (Mitsumoto et al. 1988; Munsat et al. 1988). The spectrum of motor system degeneration, however, includes pure lower motor neuron involvement, presenting as progressive muscle atrophy, as well as pure supranuclear motor degeneration, referred to as primary lateral sclerosis (PLS) (Beal and Richardson 1981; Russo 1982). The existence of a strictly supranuclear variant, PLS, has been debated, but in recent years several authors have provided evidence to support the nosological entity of PLS (Russo 1982; Sotaniemi and Myllyla 1985; Gastaut et al. 1988; Younger et al. 1988). PLS runs a less progressive course than classic ALS and affects preferentially the long descending corticospinal pathways, clinically apparent as a syndrome of slowly progressive spastic paraplegia or tetraplegia, less often including a pseudobulbar syndrome (Gastaut et al. 1988). This report describes a variant of supranuclear motor system degeneration that so far appears to be unique in the literature. It is characterized by a syndrome of pseudobulbar palsy reminiscent of the Foix-Chavany-Marie syndrome.

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Case report

In 1983 an insurance agent aged 52 years presented first to a neurologist because he felt that 1 year previously his speech had changed quite abruptly and had never returned to normal. He also had noticed a loss of quick tongue movements on playing a trumpet. CT did not show the expected vascular brain-stem lesion or other intracranial pathology. Doppler studies of the extracranial vessels, tensilon test for myasthenia, EEG, and lumbar CSF were all normal. On direct questioning in 1989, the patient and his wife recalled that as early as 1981 a mild inability to swallow saliva had been noticed but had never been of major concern. Already in 1984 the ENT department of the local university recommended logopaedic therapy because the patient felt severely handicapped in his job by the dysarthria. At the same time he underwent surgery for bilateral struma nodosa. In 1985 the patient was first seen at our department. Severe dysarthria, bilateral central facial weakness, palatal weakness, tongue stiffness and pathological laughing were observed, but there were no corticospinal signs and no sensory findings. CT and EEG were again normal, as were the sensory and brain-stem evoked potentials. The EMG of the deltoid and quadriceps muscles did not provide any evidence of lower motor neuron disease. A pathological orbicularis oculi reflex with delayed and reduced R2 component suggested the presence of a medullary process. For the first time a diagnosis of ALS was considered. In 1986 the patient was reassessed because of progressive dysarthria and dysphagia. Repeated CT and an EMG of the tongue were normal. In 1987 the patient had become anarthric, food intake was restricted to mashed food and liquids, and the pharyngeal collection of saliva was now of major concern to the patient. A severe supranuclear bulbar syndrome with affective disinhibition of pseudobulbar type was present, tendon reflexes were now brisk, and there was a suggestion of spasticity in the left leg. MRI and deltoid EMG were unremarkable. In 1989 the patient was readmitted because a progressive spastic tetraparesis with en-

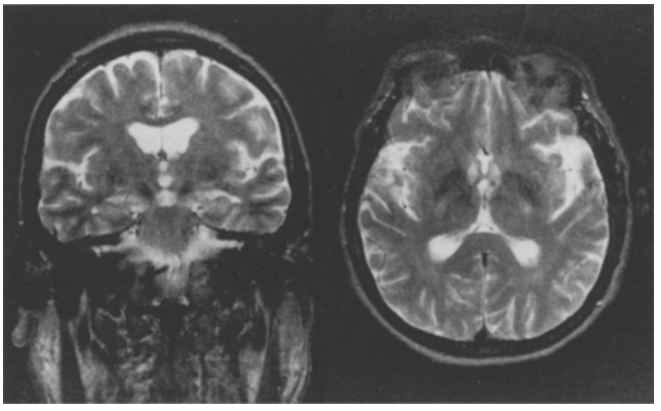

Fig. 1. The MRI scan, T_2-weighted (TR 2.0; TE 90), shows mild atrophy without circumscribed lesions

hanced reflexes and bilateral Babinski's signs had developed. Generalized, mild, symmetrical muscular atrophy was present to an extent attributable to disuse and immobilization. There were no fasciculations; bladder and bowel control were preserved; extraocular muscles were spared. Head and neck were often held in an abnormal posture for many minutes (oreiller psychique). The gag reflex was absent. We observed a striking discrepancy between the lack of voluntary innervation of the facial muscles on command, on the one hand, and the intact mimic involuntary innervation as well as the preservation, for example, of yawning, on the other. There was no sign of spasticity of the facio-pharyngo-glosso-masticatory muscles. The full range of ocular movements could only be elicited using strong visual stimuli, e.g. asking the patient to follow one's face rather than merely a finger. There was no gaze in any direction merely on an examiner's command, indicative of mild frontal lobe disorder and disinhibition. MRI with special attention given to the corticobulbar pathways and the opercula remained normal (Fig. 1); the EMG of the anterior tibial, gastrocnemius, and triceps brachii muscles showed no evidence of denervation.

Discussion

Pure supranuclear motor system degeneration, PLS, has been reported to progress much more slowly than ALS (Beal and Richardson 1981; Gastaut et al. 1988), matching the 8 years of clinical progression in our patient. Although long corticospinal tract degeneration seems to be the most prevalent form of supranuclear motor system degeneration (Hübbe and Mouritzen 1971; Ungar-Sargon et al. 1980; Russo 1982), many clinical descriptions of ALS date back to times where evoked responses, MRI, CT, and myelography were not readily available.

It is likely that lesions such as demyelinating disease, syringomyelia, lesions at the foramen magnum, spinal neoplasms and arteriovenous malformations may have been missed (Beal and Richardson 1981; Gibson 1983; Younger et al. 1988).

The supranuclear bulbar motor disturbance in our patient can be considered a form of the opercular syndrome of Foix-Chavany-Marie, which is characterized by dysarthria and a central facio-pharyngo-glosso-masticatory diplegia for voluntary innervation (Mariani et al. 1980). Involuntary muscle innervation is not or only mildly affected, producing the classical automatic-voluntary dissociation probably first described by Magnus in 1837 (Bruyn and Gathier 1969). The site of the lesion can be located to the frontal opercula or their corticofugal projections, respectively, areas which have occasionally been severely affected in ALS (Hudson 1981). All clinical cases with a Foix-Chavany-Marie syndrome (anterior operculum syndrome), except for two patients with astrocytoma, a case of meningoencephalitis (Bruyn and Gathier 1969), a case of viral encephalitis (Mao et al. 1989) and a related clinical syndrome due to cortical opercular maldevelopment recently described by Graff-Radford et al. (1986), have been traced to cerebrovascular disease (Mariani et al. 1980; Sandyk and Brennan 1983). Although MRI did not provide evidence of a circumscribed bilateral opercular atrophy, a predominant motor neuron loss affecting the operculobulbar projection has to be held responsible for the clinical picture of automatic-voluntary dissociation in our patient.

A related syndrome may have been present in a patient (case 4) described by Fisher (1977), who was reported to hold "the lips in a position of constant smiling" but also „moved the lips well in laughter".

Only a minority of the patients with Foix-Chavany-Marie syndrome have had a pure voluntary facio-pharyngo-glosso-masticatory diplegia without associated neurological signs, e.g. hemiparesis, aphasia, or other neuropsychological deficits. Mao et al. (1989) have emphasized the missing or decreased gag reflex as a typical sign of the Foix-Chavany-Maria syndrome, setting this entity apart from pseudobulbar palsy syndromes. The gag reflex could not be elicited in our patient.

The anatomical and functional basis of the syndrome has not been uniform in the documented cases; the extent of lesions in the area of the opercula was quite variable (Mariani et al. 1980), and the precise pathogenesis of the automatic-voluntary dissociation has remained obscure. Cases of PLS similar to our patient, where the bulbar syndrome was the initial manifestation and dominated the clinical picture for an extended period of time, have been described (Beal and Richardson 1981; Gastaut et al. 1988), notably the three female patients reported by Fisher (1977).

Another aspect of interest illustrated by this case is the association of motor system degeneration and thyroid disease which, according to some authors (Appel et al. 1986), cannot be accounted for just by chance alone. A convincing hypothesis explaining this apparent association, however, has not yet been put forward. MRI data on motor system degeneration are too scarce to be of diagnostic significance at this stage, although abnormalities have been reported (Goodin et al. 1988). Completely normal MRI (Fig. 1) in the face of a severe clinical syndrome compatible with PLS would probably support the clinical diagnosis (Gastaut et al. 1988).

References

Appel SH, Stockton-Appel V, Stewart SS, Kerman RH (1986) Amyotrophic lateral sclerosis. Associated clinical disorders and immunological evaluations. Arch Neurol 43:234–238

Beal MF, Richardson EP Jr (1981) Primary lateral sclerosis. A case report. Arch Neurol 38:630–633

Bruyn GW, Gathier JC (1969) The operculum syndrome. In: Vinken PJ, Bruyn GW (eds) Handbook of clinical neurology, vol 2. Localization in clinical neurology. North Holland, Amsterdam pp 776–783

Fisher CM (1977) Pure spastic paralysis of corticospinal origin. Can J Neurol Sci 4:251–258

Gastaut JL, Michel B, Figarella-Branger D, Somma-Mauvais H (1988) Chronic progressive spinobulbar spasticity. A rare form of primary lateral sclerosis. Arch Neurol 45:509–513

Gibson JM (1983) Primary lateral sclerosis. Arch Neurol 40:459–460

Goodin DS, Rowley HA, Olney RK (1988) Magnetic resonance imaging in amyotrophic lateral sclerosis. Ann Neurol 23:418–420

Graff-Radford NR, Bosch EP, Stears JC, Tranel D (1986) Developmental Foix-Chavany-Marie syndrome in identical twins. Ann Neurol 20:632–635

Hübbe P, Mouritzen DA (1973) Spastic paraplegia of unknown origin. Acta Neurol Scand 49:536–542

Hudson AJ (1981) Amyotrophic lateral sclerosis and its association with dementia, parkinsonism, and other neurological disorders: a review. Brain 104:217–247

Mao CC, Coull BM, Golper LAC, Rau MT (1989) Anterior operculum syndrome. Neurology 39:1169–1172

Mariani C, Spinnler H, Sterzi R, Vallar G (1980) Bilateral perisylvian softenings: bilateral anterior opercular syndrome (Foix-Chavany-Marie syndrome). J Neurol 223:269–284

Mitsumoto H, Hanson MR, Chad DA (1988) Amyotrophic lateral sclerosis. Recent advances in pathogenesis and therapeutic trials. Arch Neurol 45:189–202

Munsat TL, Andres PL, Finison L, Conlon T, Thibodeau L (1988) The natural history of motoneuron loss in amyotrophic lateral sclerosis. Neurology 38:409–413

Russo LS Jr (1982) Clinical and electrophysiological studies in primary lateral sclerosis. Arch Neurol 39:662–664

Sandyk R, Brennan MJW (1983) The operculum syndrome. J Comput Assist Tomogr 7:130–131

Sotaniemi KA, Myllyla VV (1985) Primary lateral sclerosis. A debated entity. Acta Neurol Scand 71:334–336

Ungar-Sargon JY, Lovelace RE, Brust JCM (1980) Spastic paraplegia-paraparesis. A reappraisal. J Neurol Sci 46:1–12

Younger DS, Chou S, Hays AP, Lange DJ, Emerson R, Brin M, Thompson H Jr, Rowland LP (1988) Primary lateral sclerosis. A clinical diagnosis reemerges. Arch Neurol 45:1304–1307