Eur Arch Psychiatry Clin Neurosci (1992) 242:93–95

European Archives of Psychiatry and Clinical Neuroscience

Automatic-Voluntary Dissociation: an Unusual Facial Paresis in a Patient with Probable Multiple Sclerosis

Frank Eblen, Michael Weller, and Johannes Dichgans

Neurologische Universitätsklinik, Hoppe-Seyler-Str. 3, W-7400 Tübingen, Federal Republic of Germany

Received February 5, 1992

Summary. A patient with multiple sclerosis is described who presented with a unilateral loss of voluntary function of his lower face muscles. However, in an emotional situation, there was strong involuntary innervation of these muscles: automatic-voluntary dissociation. The subcortical afferents to the facial motor nucleus are discussed. It is hypothesized that cortical disinhibition of midbrain nuclei underlies the accentuated involuntary innervation.

Key words: Supranuclear – Facial expression – Voluntary-emotional-limbic-dissociation

Introduction

In an earlier issue of this journal, we described a patient with supranuclear motor system degeneration who presented with a unique syndrome of bilateral automatic-voluntary dissociation of facial muscle innervation (Weller et al. 1990). We have now had the opportunity to study an isolated unilateral dissociating facial paresis in a young male with probable multiple sclerosis.

Case Report

The 25-year-old male was admitted with a left facial paresis of 1 week's duration. He had been healthy until 12 months earlier when he noted a "flutter" of facial muscles in the left perioral region. This symptom subsided after 2 months of treatment with carbamazepine. There was no history of fever, visual loss, double vision, sensory loss, weakness of limbs, or tick bites. On examination of his cranial nerves, there was weakness of his left facial muscles. The left palpebral fissure was wider. Voluntary eye closure was weaker on the left side but complete. The corneal and blink reflexes were symmetric. When the patient was asked to frown, his frontal muscles were paretic on the left side; however, when he looked upwards, he was able to frown. When asked to show his teeth, the patient could not raise the corner of his mouth, nor could be purse his lips. On the other hand, when enjoying a joke or smiling involuntarily, he showed good innervation of the left side that preceded that on the right (Fig. 1). Taste was preserved, there was no impairment of lacrimation, no hearing or visual loss, no hyperacusis, and no weakness of jaw or pharyngeal muscles. A slight right-sided cerebellar syndrome with hemiataxia, bidirectional gaze-evoked nystagmus and a disturbance of smooth pursuit and optokinetic nystagmus were noted. Deep tendon reflexes were increased on the left, the plantar reflexes were flexor. The sensorium was intact. The patient appeared psychologically unremarkable. Lumbar puncture revealed clear fluid at a normal opening pressure with 3 cells/μl and autochthonous oligoclonal IgG production. No antibodies against *Borrelia sp.* were detectable in CSF or serum. Magnetic resonance imaging (MRI) with T2-weighted images revealed multiple large periventricular plaques and a small hyperintense lesion in the left midbrain close to the red nucleus (Fig. 2). Visual and somatosensory evoked potentials of the tibial nerve were normal. Four weeks after

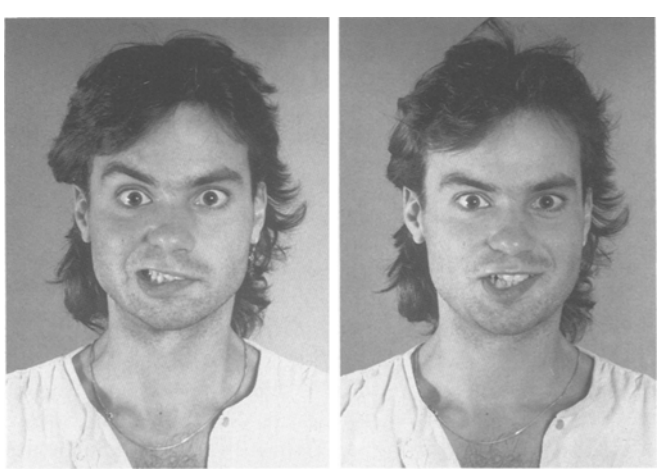

Fig. 1. Left: Left-sided weakness predominantly of the lower face at attempted voluntary movement. Right: Note stronger innervation of the upper lip at involuntary smile on the affected left side compared to the right side

Correspondence to: J. Dichgans

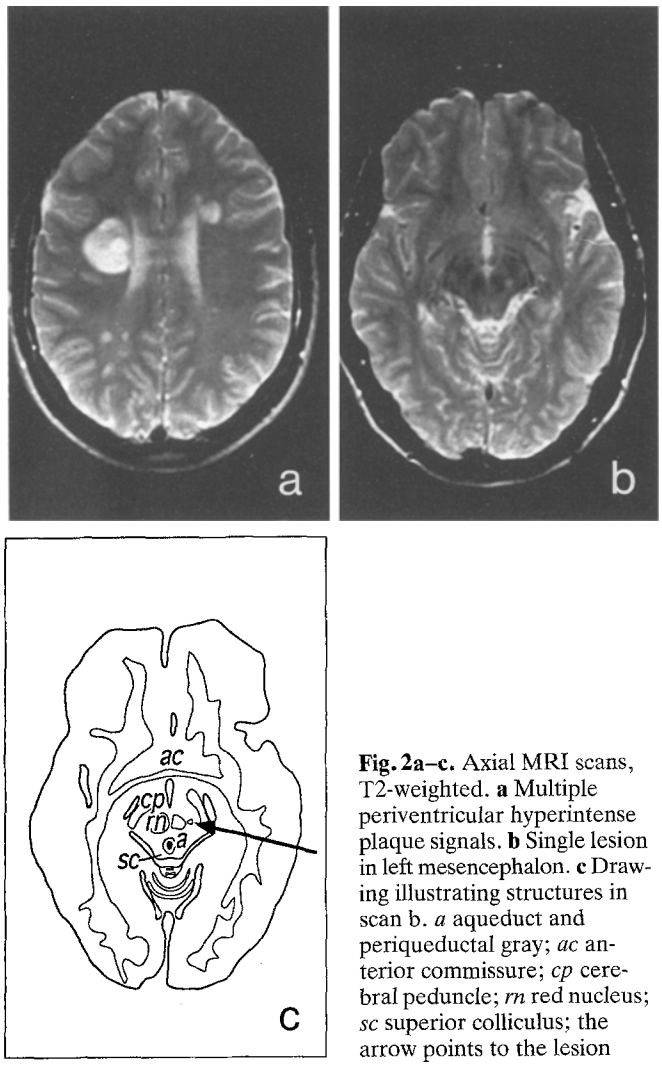

Fig. 2a–c. Axial MRI scans, T2-weighted. **a** Multiple periventricular hyperintense plaque signals. **b** Single lesion in left mesencephalon. **c** Drawing illustrating structures in scan b. *a* aqueduct and periqueductal gray; *ac* anterior commissure; *cp* cerebral peduncle; *rn* red nucleus; *sc* superior colliculus; the arrow points to the lesion

onset, the symptoms persisted. Electromyography performed of the left orbicularis oculi muslce showed no denervation; the evoked muscle action potentials after direct stimulation of the facial nerve were of normal amplitude. The central latencies of the blink reflex were within normal limits.

Discussion

The patient presented with a pattern of facial paresis consistent with a supranuclear lesion; the predominant involvement of the lower face, absence of Bell's sign, preservation of blink and corneal reflexes as well as intact secretory and sensory functions in the area of the seventh cranial nerve all pointed to a central etiology. Indeed, a diagnosis of multiple sclerosis was highly probable in the presence of a discrete cerebellar syndrome, a typical CSF pattern and MRI showing multiple white matter lesions (Poser et al. 1983). While the central character of the facial paresis was not in doubt, the degree of preserved involuntary facial muscle innervation was unusual. Automato-voluntary dissociation has not been described in multiple sclerosis and its anatomical basis is still far from clear.

Automato-voluntary dissociation can be observed at the cortical level with bilateral lesions of the frontal operculum. In this so-called anterior opercular syndrome, the upper and lower face tend to be equally affected ("pseudo-peripheral palsy"), and there is also bilateral loss of voluntary function of other cranial nerves (Foix et al. 1926; Bruyn and Gathier 1969). Steiner (1979) showed in normal and anencephalic newborns that the integrity of the corticobulbar tract is not required for reflectory smile. Other clinical evidence for separate pathways of voluntary and emotional facial innervation comes from patients with peripheral facial palsy who had accessorio-facial nerve anastomoses performed to reinnervate the face. While these patients learned to move their facial muscles without moving their shoulder, emotional expression was irreversibly lost (Kahn 1966).

Stimulation and lesion studies in animals suggest that the midbrain central gray is the lowest level of integration for emotional behaviour (Brown 1967; LeDoux 1987). The central gray blends dorsally with the deep layers of the superior colliculus and comprises the dorsal raphe nuclei ventrally. Like the parabrachial nucleus, it is part of the mesencephalic reticular formation. Studies in cats show that each of these nuclei projects to the lateral subnucleus of the facial motor nuclear complex which innervates perioral muscles (Courville 1966). The projections from the parabrachial and dorsal raphe nuclei are predominantly ipsilateral whereas the dorsal central gray projects mostly contralaterally (Takeuchi 1979; Holstege et al. 1984; Cowin and Holstege 1992). The dorsal raphe nucleus, central gray, and parabrachial nucleus, for their part, receive limbic afferents from the central nucleus of the amygdala and the lateral hypothalamus (Krettek and Price 1978; Swanson 1976; Saper et al. 1979) by way of the medial forebrain bundle and the dorsal longitudinal fascicle. Furthermore, the ventral pallidum and the substantia nigra pars compacta which have been viewed as a part of a limbic circuit through the basal ganglia (Alexander et al. 1990) also project directly and indirectly to the reticular formation (Beckstead at al. 1979; Graybiel and Ragsdale 1979; Haber et al. 1985). Unvoluntary facial innervation is likely to be conveyed through these multisynaptic pathways although this has not been demonstrated in humans.

Several questions arise. Could the lesion adjacent to the red nucleus (Fig. 2b, c) be responsible for automato-voluntary dissociation? This seems unlikely given that the red nucleus has not been implicated in limbic pathways and projects predominantly to the contralateral facial nucleus (Tageuchi et al. 1979). We are unable to determine the lesion responsible for the patient's symptoms with certainty given the extent of the demyelinating process. We would like to speculate that disruption of right-hemispheric corticomesencephalic fibres caused a disinhibition of limbic input to the facial motor nucleus, thus leading to a stronger emotional innervation on the left side (Monrad-Krohn 1924). Why then is automato-voluntary dissociation not routinely seen in capsular hemiparesis? While the true incidence of automato-voluntary

dissociation in patients with a hemispheric lesion has not been established and is likely to be underreported, it is possible that extensive lesions will affect not only the corticobulbar tract but also other multisynaptic descending pathways.

Acknowledgement. We thank Dr. Klott and colleagues, Stuttgart, for providing the MRI scans.

References

Alexander GE, Crutcher MD, DeLong MR (1990) Basal ganglia-thalamocortical circuits: Parallel substrates for motor, oculomotor, "prefrontal" and "limbic" functions. Progr Brain Res 85:119–146

Beckstead RM, Domesick VB, Nauta WJH (1979) Efferent connections of the substantia nigra and ventral tegmental area in the rat. Brain Res 175:191–217

Brown JW (1967) Physiology and phylogenesis of emotional expression. Brain Res 5:1–14

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

Courville J (1966) Rubrobulbar fibers to the facial nucleus and the lateral reticular nucleus (nucleus of the lateral funiculus). An experimental study in the cat with silver impregnation methods. Brain Res 1:317–337

Cowin RJ, Holstege G (1992) Dorsal mesencephalic projections to pons, medulla and spinal cord in the cat. Limbic and non-limbic components. J Comp Neurol 319:336–359

Foix C, Chavany JA, Marie P (1926) Diplégie facio-linguo-masticatrice d'origine cortico-sous-corticale sans paralysie des membres. Rev Neurol (Paris) 33:214–219

Graybiel AM, Ragsdale CW, Jr (1979) Fiber connections of the basal ganglia. In: Cuénod M, Kreutzberg GW, Bloom FE (eds) Development and Chemical Specificity of Neurons. Elsevier, Amsterdam, pp 239–283

Holstege G, Tan J, Van Ham J, Bos A (1984) Mesencephalic projections to the facial nucleus in the cat. An autoradiographic tracing study. Brain Res 311:7–22

Haber SN, Groenewegen HJ, Grove EA, Nauta WJH (1985) Efferent connections of the ventral pallidum: evidence of a dual striato-pallidofugal pathway. J Comp Neurol 235:322–335

Kahn EA (1966) On facial expression. Clin Neurosurg 12:9–22

Krettek JE, Price JL (1978) Amygdaloid projections to subcortical structures within the basal forebrain and brainstem in the rat and cat. J Comp Neurol 178:225–254

LeDoux JE (1987) Emotion. In: Mountcastle VB, Plum F (eds) Handbook of Physiology, Sect 1, vol V. American Physiological Society, Bethesda, pp 419–459

Monrad-Krohn GH (1924) On the dissociation of voluntary and emotional innervation in facial paresis of central origin. Brain 47:22–35

Poser CM, Paty DW, Scheinberg L, McDonald WI, Davis FA, Ebers CG, Johnson KP, Sibley WA, Silberberg DH, Tourtelotte WW (1983) New diagnostic criteria for multiple sclerosis; guidelines for research protocols. Ann Neurol 13:227–231

Saper CB, Swanson LW, Cowan WM (1979) An autoradiographic study of the efferent connections of the lateral hypothalamic area in the rat. J Comp Neurol 183:689–706

Swanson LW (1976) An autoradiographic study of the efferent connections of the preoptic area in the rat. J Comp Neurol 167:227–256

Steiner JE (1979) Facial expressions in response to taste and smell stimulation. Adv Child Dev Behav 13:257–295

Takeuchi Y, Nakano K, Uemura M, Matsuda K, Matushima R, Mizuno N (1979) Mesencephalic and pontine afferent fiber system to the facial nucleus in the cat: a study using the horseradish peroxidase and silver impregnation techniques. Exp Neurol 66:330–342

Weller M, Poremba M, Dichgans J (1990) Opercular syndrome without opercular lesions: Foix-Marie-Chavany syndrome in progressive supranuclear motor system degeneration. Eur Arch Psychiatry Neurol Sci 239:370–372