Aphemia as a first symptom of multiple sclerosis

Sir: Speech disorders are common in the course of multiple sclerosis, but aphasia is rare.1–3 We describe a patient with aphemia, that is, an articulatory disorder caused by a left hemisphere lesion, as a first symptom of multiple sclerosis.

A 29 year old right handed female medical student complained of speech problems and of a loss of strength in the right hand, which had developed insidiously in 5 days. The speech difficulties mainly consisted of an altered inflection. Writing was unimpaired, but less skillfully performed. She had no other symptoms and her medical history was uneventful. Apart from contraceptives no medication was taken. Neurological examination disclosed a minimal weakness of the right arm and leg. Cranial nerves, coordination, sensory testing, and reflexes were normal, with both plantar responses flexor. There were no signs of buccofacial apraxia. There was no dysphagia, increase of facial reflexes or lability of emotional expression. Her speech was monotonous with a loss of intonation and emotional expression. Neuropsychological examination did not show any language disorder. Conversational speech was fluent with a normal syntax and vocabulary, without paraphasias. Comprehension of spoken and written language was excellent. Confrontation naming, series speech, reading aloud, writing, and repetition of spoken language were entirely normal. No ideational or ideomotor apraxia was present. Visual spatial skills, tactile perception and memory functions were unimpaired. There was no attentional or planning disturbance.

The speech difficulty was characterised by hesitation in starting a sentence and in pronouncing words starting with a consonant. She did not simplify certain phonemes, but rather gave the impression of stuttering although there was no actual repetition of phonemes. The speech difficulty was not constant for a certain word or sound, but changed during conversation. Inflection was radically altered. She could not sing a scale or simple songs because of the dys prosody, but humming was normal. The patient was definitely concerned about her symptoms.

Extensive blood examination disclosed no abnormality. There was no serological evidence of viral or treponemal infection. A contrast enhanced CT scan of the brain on admission was normal. Cerebrospinal fluid (CSF) contained 73 erythrocytes, 16 lymphocytes, and 2 monocytes per mm3. Total protein was 0.35 g/l with an IgG index of 1.58 (normal less than 0.60) and a normal albumin ratio. Electrophoresis showed oligoclonal bands confirming intrathecal IgG synthesis EEG and evoked potential studies (visual, sensory and auditory) were normal. Within a few days the weakness disappeared and in 2 weeks there was a resolution of the speech disorder. A second CT scan now showed a hypodense area with central contrast enhancement in the frontal white matter of the left hemisphere (figa). She was discharged in good condition and resumed her studies without signs of recurrent neurological deficit during a follow-up of one year. After 6 months magnetic resonance imaging (MRI) showed multiple para- ventricular lesions (figb). Spontaneous recovery, CSF findings, and lack of any other explanation were suggestive of multiple sclerosis. In combination with the MRI findings 6 months later the criteria for laboratory supported definite multiple sclerosis were fulfilled.4–5

Abnormal speech with normal propositional language can be subdivided into three groups according to the kind of articulatory disorder and the accompanying neurological symptoms and signs: dysarthria, aprosodia, and aphemia.1 In dysarthria the articulation disorder is basically a simplification of articulation with articulatory distortions.6 In the aprosodies the affective components of speech are impaired after right hemisphere lesions.7 The third group, called aphemia or apraxia of speech comprises articulation disorders which have caused much debate. Different authors have stressed aphasic, dysarthric, or apraxic aspects, and named this group of articulation disorders accordingly.8 Basically aphemia concerns involvement of a purely expressive aspect of spoken language, and as such it is difficult to distinguish from motor aphasia.9 Propositional language however is entirely normal. There has been discussion about the interpretation of phonetic errors as aphasic or apraxic.10 The inconsistency of the articulatory problems is used as a distinction from dysarthria and to stress the apraxic aspect.6 A frequent concomitant finding is oral or facial apraxia.11 The symptoms of our patient fit in with the picture of aphemia. A left-sided lesion of the posterior part of the inferior frontal lobe and especially the deep white matter of the anterior limb of the internal capsule is the most consistent pathological finding. The aetiology is nearly always vascular or traumatic.6 In our patient a lesion became visible on the CT scan which extended into the area of the
putamen and the anterior limb of the internal capsule. The latter localisation could be responsible for the aphemia. To our knowledge aphemia has not been described previously as a first symptom of multiple sclerosis.

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References

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Limb girdle type muscular dystrophy associated with a Wolff-Parkinson-White syndrome

Sir: Cardiac involvement varies greatly with different types of muscular dystrophy. In general the so called dystrophic heart disease is common in Duchenne’s muscular dystrophy (50–80%), infrequent in limb girdle type muscular dystrophy and occasional in all other types of muscular dystrophy. The most important clinical features of myocardial disease in muscular dystrophies are tachycardias, arrhythmias, congestive heart failure and sudden death. Various publications emphasise the even more frequent appearance of an altered ECG.1 5 The Wolff-Parkinson-White syndrome has been described in patients with cardiomyopathy2 6 10 and in three cases with Duchenne’s muscular dystrophy.7 8 9 To our knowledge the syndrome has not been reported in association with limb girdle type muscular dystrophy.

A 40 year old male patient was admitted because of weakness and atrophy of shoulder and pelvic girdle muscles. He had a history of 12 years with lumbosacral pain and slow deterioration of muscular function which incapacitated the patient in his daily activities. Examination revealed normal vital signs, a weight of 45 kg and a height of 168 cm. Cardiologic examination was normal. The patient had severe asymmetrical proximal muscular atrophy of both upper and lower limbs. Distal, facial and abdominal muscles were not affected, and there were no skeletal deformities. Biceps, triceps and knee reflexes were diminished. The patient had a lordotic gait, difficulty in climbing stairs, combing his hair and Gower’s sign was positive. The rest of the neurological examination was normal. Laboratory data revealed a CK of 500 U (normal up to 170 U) with a muscle fraction of 1·1%, LDH, SGOT and all other standard laboratory tests were normal. Chest and abdominal radiographs were normal. The resting ECG showed a sinus bradycardia of 49 beats per minute and an axis of −20°. The QRS complex measured 0·15 ms, the PR interval 0·1 ms and the PQ interval 0·26 ms. Positive delta waves were seen in leads I, aVL and V4-V6 and a negative delta wave was observed in VI compatible with a type B Wolff-Parkinson-White syndrome (fig). The ECG also revealed sinus bradycardia which was reversed by exercise and thought to be physiological. An M-mode, two dimensional and dynamic echocardiogram did not reveal abnormalities and all standard measurements were normal. The EMG showed fibrillations of low amplitude and polyphasic motor deltoid, biceps, quadriceps and anterior tibial muscles. Neuroconduction was normal. The muscular biopsy specimen showed muscle fibre necrosis, wide variations in fibre size, increased connective tissue and internal muscle fibre nuclei, being compatible with muscular dystrophy. We have obtained detailed information of 38 family members. One brother of the patient was found to have the same progressive muscular process but without cardiac involvement. He was 9 years older than our patient and his disease had progressed further. A muscle biopsy showed the same myopathic features.

Limb girdle type muscular dystrophy is an autosomal recessive disorder characterised by insidious onset in early adulthood and

Letters

Fig Electrocardiogram demonstrating a prolonged QRS complex shortened QT and PJ times and delta waves.
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