

Focal Dystonia and Speech Impairment Responding to Anticholinergic Therapy

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Focal dystonia of the right hand and speech impairment as a result of a circumscribed left-sided anterior thalamic lesion is reported in a 9-year-old girl with borderline normal psychomotor development. Both focal dystonia and speech impairment improved during anticholinergic treatment and worsened after drug withdrawal.

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Introduction

The site of the lesion responsible for symptomatic dystonia has been localized to the contralateral caudate nucleus, putamen, globus pallidus or thalamus, or a combination of these structures [1]. The most useful pharmacologic approach in dystonia involves anticholinergic drugs demonstrating a better response in children as compared to adults [2]. Thalamic lesions can account for speech and/or language disturbances also [3], for which no specific therapy is available other than speech therapy.

Case Report

A 9-year-old white girl is presented with borderline normal psychomotor development with partial defects consisting of a speech and language impairment and nonprogressive focal action dystonia of the right hand. Family history of neurologic disease was negative. She was born spontaneously after 41 weeks of normal gestation; birth weight was 3,500 gm. There was no history of ante-, peri-, or postnatal complications, especially no signs of birth asphyxia or neonatal jaundice. From

the age of 1 year her parents noticed the function of her right hand to become backward. Although her psychomotor development was borderline normal, a more pronounced impairment of speech and language became gradually apparent.

Neurologic examination revealed focal action dystonia with ulnar deviation and swan-neck position of hand and fingers without atrophy. No sensory disturbances were found in this medically healthy girl. Radiographic examination of the cervical spine and chest, and extensive neurophysiologic investigation (EEG, EMG, SSEP) revealed no central or peripheral lesion. Whereas cerebral CT was negative, MRI revealed a cyst with surrounding gliosis at the site of the left anterior thalamic region characteristic for a lacunar infarct (Fig 1). Extensive laboratory examination did not reveal known etiologic causes such as clotting cascade disturbances, metabolic and (para)infectious disorders. Studies of thrombocytes, white and red blood cell counts, sickle cell, erythrocyte sedimentation rate, glucose, lipid profile, liver enzymes, urea and creatinine, lactate, creatine kinase, bilirubin, copper, ceruloplasmin, thyroid function, homocysteine, vitamin B₁₂, folic acid, anti-DNA, protein C and S, lupus coagulants, antithrombin-III, and syphilis were all normal. By exclusion the thalamic infarction was believed to be of perinatal origin.

Methods

On first referral (session 1) an extensive neuropsychologic, linguistic, and motor test battery was employed [4-7]. Tests of speech and manual motor function were repeated 4 times, twice during anticholinergic therapy (0.4 mg trihexyphenidyl twice daily) at time points 3 and 5 months (sessions 2 and 3, respectively), and twice after drug withdrawal, at time point 7 months (session 4: drug holiday of 1 week) and at time point 10 months (session 5: drug holiday of 14 weeks) (Table 1).

Results

Below-average scores on tests for intelligence were found (IQ = 78, verbal IQ = 78, performance IQ = 82 [4]; mental processing composite (problem solving skill; composite of sequential and simultaneous processing) = 78, sequential processing (serial or temporal order problem solving skill) = 78, simultaneous processing (gestalt-like or spatial problem solving skill) = 81 [5]; visuomotor integration = 81 (standard score) [6]). Although subtest profiles were not harmonious, no factors with respect to information processing emerged from these tests.

A discrepancy could be demonstrated between the receptive language skills (auditory word discrimination, receptive vocabulary), on which average percentile scores of 40 were obtained, and the expressive language skills (morphology, expressive vocabulary, sentence construction) with percentile scores of approximately 11. Speech remained slightly unfluent with perseveration but no signs of thalamic tachylalia.

Maximum repetition rates of syllables (repeat as fast as possible the sequences "papapa . . .," "tatata . . .," "kakaka . . .") [7] were initially slower than control children (session 1). After 3 and 5 months of medication (sessions 2 and 3), rapid repetitive monosyllabic rates had increased to normal reference values, but without medi-

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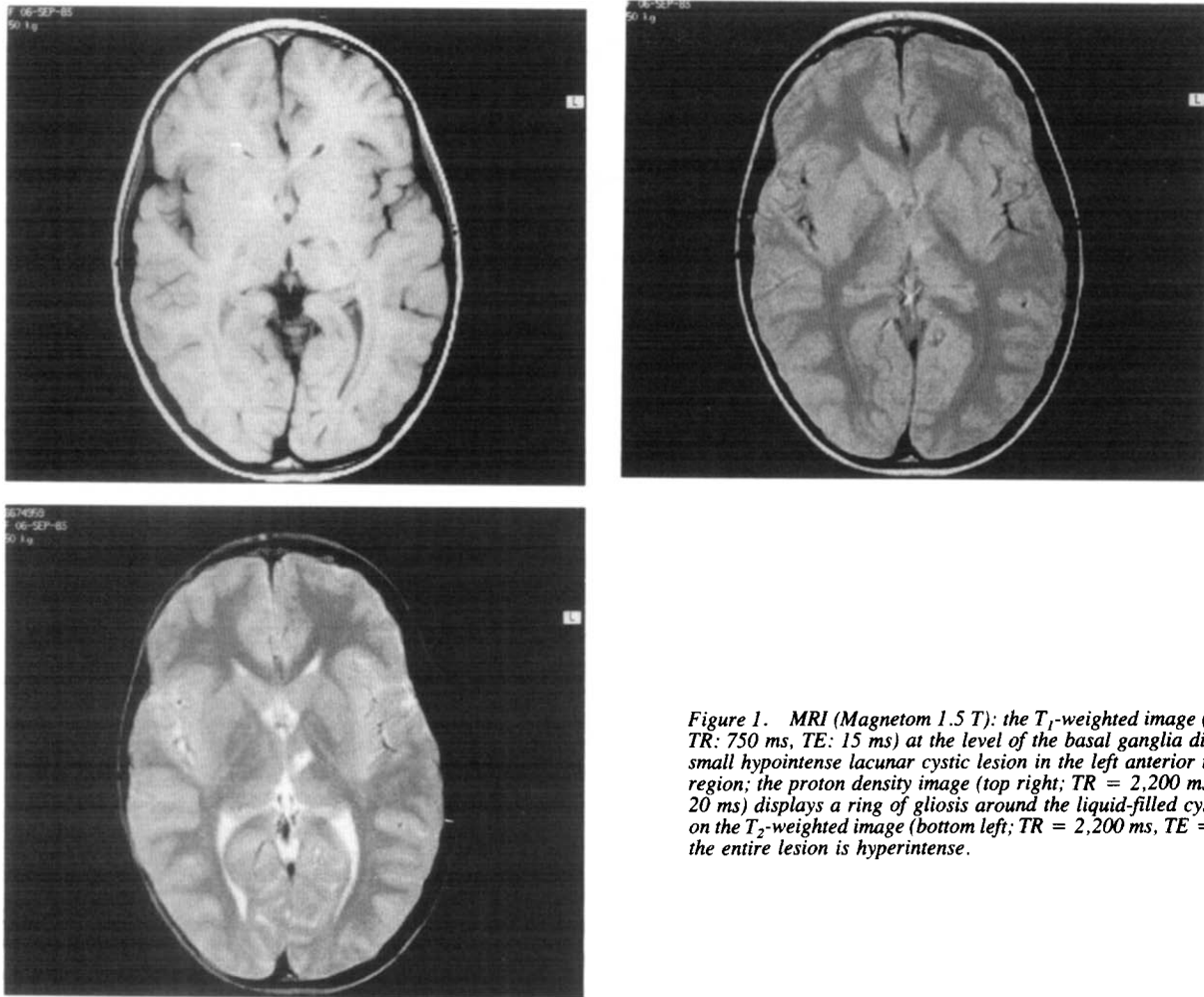


Figure 1. MRI (Magnetom 1.5 T): the T_1 -weighted image (top left; TR: 750 ms, TE: 15 ms) at the level of the basal ganglia displays a small hypointense lacunar cystic lesion in the left anterior thalamic region; the proton density image (top right; TR = 2,200 ms, TE = 20 ms) displays a ring of gliosis around the liquid-filled cyst, while on the T_2 -weighted image (bottom left; TR = 2,200 ms, TE = 80 ms) the entire lesion is hyperintense.

cation (sessions 4 and 5) syllable rates decreased again. The factor Sessions was significant in a nonparametric analysis of variance (Kruskal-Wallis: $\chi^2 = 12.6$; degrees of freedom = 4; $P = .0136$). The rapid alternating multisyllabic rate displayed less decline than the monosyllabic rate with values still being within the control range, although the pattern over time was similar. Apparently, the

production of rapid repetitive movements more than rapid alternating movements seems to be affected.

Maximum sound prolongation (sustain sounds “aa . . . ,” “ff . . . ,” “ss . . . ,” and “zz . . . ” for as long as possible) [7] was slightly reduced at all 5 sessions, but within the control range and without any beneficial effect of medication.

Table 1. Speech function tests

Session		1	2	3	4	5	C (n = 24; range)
Time point (months)		0	3	5	7	10	
Trihexyphenidyl medication (weeks)		—	13	22	—	—	
Drug holiday (weeks)		—	—	—	1	14	
Monosyllabic sequences (n/s) [7] (rapid repetitive movements)	/papa. . ./	3.08	5.15	5.41	4.33	4.51	5.18 (4.20-6.41)
	/tata. . ./	3.85	4.61	5.52	4.31	4.00	5.15 (3.92-6.17)
	/kaka. . ./	3.57	4.63	5.00	4.59	4.40	4.74 (3.61-6.13)
Multisyllabic sequence (n/s) [7] (rapid alternating movements)	/pataka. . ./	4.41	5.46	6.29	5.65	4.52	5.00 (3.77-7.14)

Abbreviations

C = Control

(n/s) = Number of sequences per second at session 1 (pretreatment), sessions 2 and 3 (trihexyphenidyl regimen), and sessions 4 and 5 (after 1 week and 14 weeks of drug withdrawal).

The articulation tests did not change during anticholinergic therapy; they revealed no dyspraxia, but a delayed articulation development according to her below-average IQ.

On the finger tapping task a huge difference was found between the normal left (3.6 taps/s) and the affected right hand (1.8 taps/s). Although the pattern over sessions was erratic, there was a tendency for the right hand to improve slightly during medication (2.2 taps/s), but not to the normal left-handed control level, and to impair again after drug withdrawal (1.7 taps/s and 0.7 taps/s). No left-right differences in reaction times (0.45-0.49 s) were detected on the 5 sessions: apparently, the production of rapid repetitive movements more than rapid movements per se (initiation time) is affected in the right hand, a result that reveals a striking similarity to the rapid repetitive and rapid alternating syllabic rates.

The dystonia of the right hand was noteworthy although it subjectively improved as independently reported by the patient herself, her parents, teachers, and caregivers: fine motoric hand movements could be carried out much more easily resulting in a more appropriate execution of activities of daily living and consequently in an overall enhanced well-being.

Discussion

In adults, dysphasia due to infarction of the anterior thalamic region is characterized by hypophonia, a reduced output, verbal paraphasias, moderate impairment of comprehension, and preserved repetition [8]. In contrast, our young patient did not meet this dysphasia profile although she too had an anterior thalamic infarction. This discrepancy between young and adult thalamic dysphasia is probably due to damage of a developing neuronal circuitry rather than damage of a matured brain with minor plasticity capacity.

In conclusion, this case report is clinically relevant in several respects:

(1) It is the first report in which both the focal dystonia of the right hand and the linguistic impairment can be attributed to the same circumscribed left-sided anterior thalamic lesion;

(2) Improvement of both manual motor function and speech impairment occurred after administering the anticholinergic drug trihexyphenidyl, without side effects. This clinical benefit was largely lost after the drug was withdrawn, and a learning effect seems therefore rather unlikely;

(3) Focal dystonia and developmental defects at motor level, speech, and probably at expressive language level occurred simultaneously, although the latter still can be attributed to the borderline normal psychomotor development;

(4) MR to CT neuroimaging techniques are preferred in detecting small cerebral lesions in focal dystonia.

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