Impairment of visuospatial function in idiopathic spasmodic torticollis

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Abstract We studied visuospatial function in 15 patients with idiopathic spasmodic torticollis (ST) and 15 age- and sex-matched controls. All subjects underwent a battery of visuospatial tests, assessing different functional components of spatial ability. The performance of ST patients on tasks of spatial perception did not significantly differ from that of normal subjects, but patients performed significantly worse on spatial tasks requiring mental manipulation of personal space. This distinct pattern of visuospatial impairment may result from basal ganglia dysfunction.

Key words Torticollis • Visuospatial function • Basal ganglia

Introduction

It is now widely accepted that the basal ganglia are not only involved in motor control but may also contribute to cognitive processes. This concept has been supported by neuropsychological studies in non-demented patients with Parkinson disease (PD) and in patients with early Huntington’s disease (HD), who were shown to have significant difficulty in performing tasks on spatial perception and orientation [4–7, 9, 15, 17, 20, 27, 28, 30, 31]. However, the neural substrates for these cognitive deficits are still controversial, mainly because the pathological process may spread beyond the basal ganglia and involve other parts of the brain in these disorders. Therefore, studies of cognitive function in patients with other basal ganglia syndromes seem to be warranted. Though its pathological and biochemical mechanisms are still unknown, torticollis, like other forms of dystonia, is thought to arise from basal ganglia dysfunction. Up to now, only a limited number of studies on cognitive function in patients with dystonia have been published [16, 22], potentially owing to the fact that motor abnormalities represent the hallmark of the syndrome whereas intellectual impairment does not occur, either in early or in advanced stages of disease. Pathology in idiopathic dystonia is restricted to the basal ganglia during the course of disease, with cortical areas remaining unaffected. For this reason, patients with primary dystonia may represent an appropriate study group for further investigation of the potential role of basal ganglia in cognitive processing. Against this background we decided to study visuospatial ability in patients with primary cervical dystonia.

Subjects and methods:

Subjects

Fifteen patients with spasmodic torticollis (ST), attending the outpatient department of the Hamburg Neurological University Hospital, and 15 age- and sex-matched controls were enrolled in this study. All patients underwent careful neurological examinations,
and personal history was thoroughly reviewed in order to obtain detailed information on duration and clinical course of disease, previous treatment, concomitant diseases and family history. Routine diagnostic procedures in all patients included radiography of the cervical spine, polygraphic needle-EMG of neck muscles and computed tomography of the brain. Apart from the torticollis, none of the patients had evidence of other neurological or psychiatric disease. Patients who had undergone neurosurgical therapy (including thalamotomy) for their torticollis or revealed any clinical features suggesting secondary dystonia were excluded from the study. Six patients were receiving centrally acting drugs including trihexyphenidyl (n = 3; 2–6 mg/day), amitriptyline (n = 2; 25–50 mg/day), flunitrazepam (n = 1, 0.5 mg/day), diazepam (n = 1; 5 mg/day) or triapid (n = 1; 300 mg/day). All patients were tested before therapy with botulinum toxin. Characteristics of the patient population are given in Table 1. Eleven healthy volunteers and 4 inpatients with polyneuropathy (n = 3) or benign progressive muscular dystrophy (n = 1) served as a control group (mean age: 50.4 years; range: 30–72). None of the control subjects had evidence or history of CNS disease.

Methods

All subjects underwent a series of preliminary tests to assess verbal IQ (Mehrfachwahl-Wortschatztest) and memory function (block tapping test, recurring-digits test). We selected six visuospatial tests, measuring various functional components of visuospatial ability. The tests are briefly summarized below. The entire battery of tests was administered in one session, and no time limit was set on each test.

Preliminary tests

Mehrfachwahl-Wortschatztest. In Germany, this test is commonly used for estimation of premorbid intelligence and can be regarded as functionally equivalent to the National Adult Reading Test. Subjects are asked to recognize the only meaningful word in a sequence of four otherwise meaningless words. The test consists of 37 four-word sequences of increasing complexity.

Hebb's recurring-digits test [19]. The subject is asked to repeat orally presented digit lists, each of them one digit longer than his/her immediate memory span. A total of 24 sets of digits are read to the subject, one set being repeated every third trial. This test is considered to be a highly sensitive test of left hippocampal function [25].

Table 1 Demographics and clinical signs of patients with spasmodic torticollis (ST)

| Gender:     | 8 female; 7 male |
| Mean age (range): | 50.9 years (23–69) |
| Mean duration of disease (range): | 8.5 years (1–31) |

Symptoms:

Rotational ST.: n = 3
- Laterocollis: n = 1
- Laterocollis + rotational ST.: n = 6
- Retrocollis + rotational ST.: n = 5
- Head tremor: n = 2

Mean Tsui-Scorea (range) 8.3 (2–13)

a See [39]

Corsi’s block tapping test [14]. The test consists of nine black cubes fastened in random order to a board. The examiner taps the blocks in a prearranged sequence, and the subject must attempt to copy this tapping pattern. After determining the subject’s span for immediate recall, he/she is tested on 24 series of tapping sequences that are one more than his/her attention span. One sequence is repeated every third trial. The test is considered to be a sensitive test of right hippocampal function [25].

Visuospatial tests

Judgement of the visual vertical (Aubert experiment) [2]. The subject sits upright in a chair fitted with a head-rest, in a completely dark room. By rotating two knobs he/she is instructed to align to the gravitational vertical a motor-driven, 15-cm luminous line, which is placed 180 cm in front of the subject. Patients are tested in three different positions: with the head maintained as straight as possible, with the head tilted 30° to the right and with the head tilted 30° to the left. A small luminous tape is attached to the subject’s forehead, and a semicircular, luminous scale is placed behind him/her, which enables the examiner to observe the subject’s head position in spite of complete darkness. Each sequence consists of 6 estimations, giving a total of 24 trials. After each positioning the examiner reads the deviation.

Benton’s line-orientation test [3]. The test consists of a card showing an array of lines numbered from 1 to 11, each separated by an angle of 18°. Subjects are presented 30 stimulus cards consisting of two lines, each of which represents one half of a response-choice line. The task is to indicate the lines of the response-choice card having the same angles as the two stimulus lines.

Route-walking test. The test consists of five maps originally used by Semmes et al. [35]. Nine dots on each represent nine circles laid out on the floor of a room in a 270 × 270 cm square. One wall of the room is designated as “North”, and “North” is also marked on the maps. Holding the map in front of him/her, the subject is required to walk the designated path without turning the map. The total score is based on the number of correct turns (maximum possible score = 35).

Personal-orientation test [35]. Subjects are successively shown five diagrams with a dorsal and ventral representation of a human figure on which parts of the body are numbered. Subjects are asked to touch their body according to the numbered schematic diagrams. The maximum possible score of correctly indicated body parts is 35.

Ratcliff’s mental re-orientation test [32]. Twenty cards, showing the Little Man figure in different positions are successively presented to the subject. Each of the four positions is shown five times: in half the cases the black disc is on the figure’s right, in half on the left. The subject’s task is to state whether the black disc is on the figure’s right or left side.

Standardized road-map of direction sense. With the road map lying in a fixed position in front of them, subjects have to describe the right or left turns which would need to be made while following the route designated on the map.

Statistics

Statistical analyses were carried out using the computerized statistical program SPSS-PC (Version 4.0). A two-tailed t-test for independent samples was used to assess the effect of medication on test performance within the patient group. Group means for patients and controls on tests of verbal IQ and memory function were compared by t-tests. Analysis of co-variance (ANCOVA) with verbal