Clinical study of 35 patients with dysarthria-clumsy hand syndrome

A Arboix, Y Bell, L García-Eroles, J Massons, E Comes, M Balcells, C Targa

OBJECTIVES: Although dysarthria-clumsy hand syndrome (DCHS) is a well known and infrequent lacunar syndrome, there are few data regarding the spectrum of associated clinical characteristics, anatomical site of lesion, and aetiopathogenetic mechanisms. We report a clinical description of this subtype of lacunar stroke based on data collected from a prospective acute stroke registry.

METHODS: From 2500 acute stroke patients included in a hospital based prospective stroke registry over a 12-year period, 35 patients were identified as having DCHS.

RESULTS: DCHS accounted for 1.6% of all acute stroke patients (35/2110), 1.9% of acute ischaemic stroke (35/1840), and 6.1% of lacunar syndromes (35/570) admitted consecutively to a neurology department and included in the stroke registry over this period. The results supported the lacunar hypothesis in 94.3% of patients (n = 33). Atherothrombotic and cardioembolic infarction occurred in only one patient each (2.9%). No patient with DCHS had an intracerebral haemorrhage. Outcome was good (mortality in hospital 0%, symptom free at discharge 45.7%). After multivariate analysis, absence of limitation at discharge, limb weakness but not cerebellar-type ataxia, and internal capsule (40%), pons (17%), and corona radiata (8.6%) location were significantly associated with DCHS.

CONCLUSIONS: DCHS is a rare cerebrovascular syndrome, and supports the criteria of the lacunar hypothesis. The majority of patients in this study had internal capsule infarcts. The prognosis is good with striking similarity compared with other types of lacunar strokes. There are important differences between DCHS and non-lacunar strokes. Internal capsule and pons are the most frequent cerebral sites.

Dysarthria-clumsy hand syndrome (DCHS) is the most uncommon and poorly studied of the classic lacunar syndromes. Some authors have used the labels “dysarthria-clumsy hand” and “ataxic hemiparesis” interchangeably, not adhering to Fisher’s original descriptions, and thus confusing the clinical distinction. Additionally, DCHS is infrequently individualised in different prospective stroke registries and little is known regarding the frequency and natural history of this disorder. While the lacunar hypothesis is supported in most lacunar syndromes, it has not been tested in DCHS, probably because of its infrequent presentation, despite the fact that most of these cases are caused by a lacunar infarction, although cases secondary to intracerebral haemorrhage in pontine or cerebellar sites have been reported. The sites of lesions responsible for DCHS are not definitively established, although based on a study in six patients, it is assumed that pons is the main site; however, cases of DCHS have been also described in lacunar infarctions of the anterior limb of the internal capsule, genu of the internal capsule, or corona radiata. Therefore, a clinical study of 35 patients with DCHS collected from a prospective stroke registry was carried out, in order to assess: (a) the frequency of DCHS caused by different stroke subtypes and (b) differential demographic, clinical, neuroimaging, and outcome data of DCHS compared with patients with other lacunar syndromes and with those with non-lacunar stroke.

METHODS

Between January 1986 and December 1997, the data of 2500 stroke patients admitted consecutively to the Department of Neurology of Sagrat Cor (an acute care 350 bed hospital in Barcelona, Spain) were collected prospectively in a stroke registry. For the purpose of this study, patients with transient ischaemic attack (n = 328), subarachnoid haemorrhage (n = 35), and spontaneous subdural haematoma (n = 27) were excluded. The study population consisted of 2110 patients with acute ischaemic (n = 1840) or haemorrhagic (n = 270) stroke. Subtypes of stroke were classified according to the Cerebrovascular Study Group of the Spanish Society of Neurology, which is similar to the National Institute of Neurological Disorders and Stroke classification and has been used by our group in previous studies. Subtypes of stroke included 553 patients with atherothrombotic infarcts, 484 lacunar infarcts, 468 cardioembolic infarcts, 248 infarctions of undetermined origin, 87 infarctions of unusual aetiology, and 270 intracerebral haemorrhages. Definitions of cerebrovascular risk factors and lacunar syndromes (pure motor stroke, pure sensory stroke, sensorimotor stroke, ataxic hemiparesis, DCHS, and atypical lacunar syndromes) were those used in recent studies.

For the purpose of this hospital based prospective study, 570 consecutive patients with lacunar syndromes (secondary to lacunar, n = 484 or non-lacunar cerebral infarcts, n = 86) were collected. There were 277 patients with pure motor stroke, 99 with pure sensory stroke, 81 with sensorimotor stroke, 23 with ataxic hemiparesis, 35 with DCHS, and 55 with atypical lacunar syndromes. Atypical lacunar syndromes included isolated dysarthria (n = 32); hemichorea-hemibal-lismus (n = 5); isolated hemiataxia (n = 5); unilateral (n = 2) or bilateral (n = 3) paramedial thalamic infarct syndrome; pure motor hemiparesis with transient subcortical aphasia (n = 4); and pure motor hemiparesis with transient internuclear ophthalmoplegia (n = 4).

Patients selected as having DCHS (n = 35) met the following criteria: (a) dysarthria without dysphasia; (b) unilateral “central” facial weakness with ipsilateral
closeness appearing as a cerebellar-type ataxia (dysmetria, dysrhythmia, dysdiadochokinesia, gait ataxia), or with mild or no weakness; and (c) no sensory symptoms or signs. Other lacunar syndromes included all patients with lacunare stroke with the exception of DCHS (n = 535). Non-lacunar stroke included all patients whose clinical picture did not conform to the preceding subgroups (n = 1540).

All patients were admitted to the hospital within 48 hours of onset of symptoms. On admission, demographic characteristics, salient features of clinical history and neurological examination, results of routine laboratory tests, chest radiography, and twelve lead electrocardiography were recorded. In all patients, brain CT scans were performed within the first week of hospital admission. Patients with negative results had a second CT during their stay in hospital or were studied by MRI. Other investigations included angioMRI (51% of patients), echo Doppler of the supra-aortic trunks (43%), arteri digital subtraction angiography (8%), B mode echocardiography (40%), and lumbar puncture (4%).

Demographic variables included age and sex. All other findings were dichotomised as present v absent. Anamnestic findings comprised history of hypertension, diabetes, myocardial infarction or angina, rheumatic heart disease, congestive heart failure, atrial fibrillation, smoking (>20 cigarettes/day), alcohol misuse (>80 g/day), intermittent claudication, transient ischaemic attack, previous cerebral infarction, hyperlipidaemia, nephropathy, cirrhosis or chronic liver disease, obstructive pulmonary disease, asthma, diabetes, myocardial infarction, hyperlipidaemia, nephropathy, cirrhosis or chronic liver disease, obstructive pulmonary disease, asthma, diabetes, or hypertension (n = 93, age >85 years. Clinical variables were sudden onset of symptoms (minutes), headache, dizziness, seizures, nausea or vomiting, altered consciousness (drowsy, stuporous, comatose), limb weakness (hemiparesis or hemiplegia; Babinski’s sign not mandatory), sensory symptoms, hemianopia, aphasia or dysarthria, ataxia, and cranial nerve palsies. Neuroimaging variables comprised internal capsule, basal ganglia, cerebellum, mesencephalon, pons, middle cerebral artery, and basilar artery. Outcome variables were mortality in hospital, degree of clinical disability at discharge, cardiac events (acute myocardial infarction, heart failure, tachyarrhythmia), respiratory events (pneumonia, atelectasis, respiratory infection), urinary events, vascular events, and infectious complications.

**Statistical analysis**

Demographic characteristics, clinical events, and outcome of patients with DSCH were compared with those of patients with lacunar syndromes and patients non-lacunar stroke. Univariate and multivariate analysis were performed. In the univariate analyses, continuous variables were compared with Student’s t test and categorical variables with the $\chi^2$ test (with Yates’ correction when necessary). Statistical significance was set at $p<0.05$.

In the comparison of DCHS and other lacunar syndromes, variables related to DCHS in the univariate analysis plus age (used as a continuous variable with a constant odds ratio (OR) for each year) and sex were studied in a multiple linear regression model based on demographics, risk factors, clinical data, neuroimaging, and outcome variables, a total of eight variables. In the comparison of DCHS and non-lacunar strokes, two multiple linear regression models were set. The first predictive model, with 11 variables, was based on demographic, vascular risk factors, and clinical data. The second predictive model was based on demographic, risk factors, and clinical and neuroimaging data, and had 16 variables. In all cases, DCHS (coded as absent = 0, present = 1) was the dependent variable. The level of significance was set as 0.15, and the tolerance level at 0.0001. The maximum likelihood approach was used to estimate weights of the logistic parameters. OR and 95% confidence intervals (CI) were calculated from the beta coefficients and standard errors. The hypothesis that the logistic model adequately fitted the data was tested by means of the goodness of fit $\chi^2$ test. The SPSS-PC+ and the BMDP2 computer programs were used for statistical analyses.

**RESULTS**

The 35 patients with DCHS accounted for 1.6% of all cases of acute stroke, 1.9% of ischaemic stroke, and 6.1% of lacunar syndromes. Males made up 71% of patients, with a mean (SD) age of 71.3 (10.5) years. Hypertension and diabetes were the main cardiovascular risk factors in 62.9% and 34.3% of patients, respectively. Stunned onset of symptoms (minutes) was recorded in 35.6% of patients. In 71.5% of the cases (n = 25), neuroimaging studies confirmed the brain lesion sites; these were the internal capsule in 40% of cases (in the anterior limb, genu or near the genu), pons in 17.1% (paramedian rostral lesions), corona radiata in 8.6%, basal ganglia 2.9%, and thalamus in 2.9%.

With regard to stroke subtype, lacunar infarction was diagnosed in 33 patients and non-lacunar stroke in the remaining two (atherothrombotic infarction, n = 1; cardioembolic infarction, n = 1). No case of DCCH secondary to haemorrhagic stroke was recorded. Outcome of DCCH was good, with a 0% mortality rate in hospital and 45.7% of patients having no neurological disability at discharge. The mean length of hospital stay was 11.7 days. There were no significant differences in demographic data, risk factors, clinical variables, or outcome between 25 patients with DCCH and confirmed sites, and the remaining 10 patients in which the site of the lesion was not confirmed.

When the groups of DCCH and other lacunar syndromes (n = 535) were compared, limb weakness (but not cerebellar-type ataxia), speech disturbances, corona radiata and basilar artery involvement, and freedom from symptoms at discharge were significantly more frequent in DCCH group (table 1); only limb weakness and absence of limitation at discharge appeared to be significant variables related to DCCH after multivariate analysis (table 2). However, the comparison between the groups of DCCH and non-lacunar stroke (n = 1540) showed important differences, particularly in respect to the unfavourable outcome of non-lacunar stroke (higher mortality in hospital, longer hospital stay, and fewer days of clinical disability at discharge). The 35 patients with DCHS accounted for 1.6% of all cases of acute stroke, 1.9% of ischaemic stroke, and 6.1% of lacunar syndromes. Males made up 71% of patients, with a mean (SD) age of 71.3 (10.5) years. Hypertension and diabetes were the main cardiovascular risk factors in 62.9% and 34.3% of patients, respectively. Stunned onset of symptoms (minutes) was recorded in 35.6% of patients. In 71.5% of the cases (n = 25), neuroimaging studies confirmed the brain lesion sites; these were the internal capsule in 40% of cases (in the anterior limb, genu or near the genu), pons in 17.1% (paramedian rostral lesions), corona radiata in 8.6%, basal ganglia 2.9%, and thalamus in 2.9%.

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**DISCUSSION**

In this prospective hospital-based stroke registry, DCHS accounted for 6.1% of lacunar syndromes, 1.9% of ischaemic infarcts, and 1.6% of acute strokes. These findings are consistent with the North American Symptomatic Carotid Endarterectomy Trial, in which DCHS accounted for 4.6% of the 283 possible lacunar strokes and 7.1% of 210 probable lacunar strokes. In agreement with previous studies, DCCH is the most infrequent classic lacunar syndrome. In an investigation of 68 consecutive patients with sudden onset dysarthria due to a single infarction confirmed by MRI or CT, DCCH syndrome was observed in eight patients. To our knowledge, the present series of 35 patients with DCHS is the largest reported in the literature.

This study shows that DCHS was caused by a lacunar infarct in 94% of patients and by other stroke subtypes in 6%. Therefore, the lacunar hypothesis was justified in 94% of patients. A main finding of the study is that different topographies of lesions may cause DCHS, which is in contrast to current concepts.
Study of 35 patients with dysarthria-clumsy hand syndrome

<table>
<thead>
<tr>
<th>Table 1 Results of univariate analysis. Comparison of patients with dysarthria-clumsy hand syndrome (DCHS) with patients with other lacunar syndromes and with patients with non-lacunar stroke</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Outcome</strong></td>
</tr>
<tr>
<td>Total patients</td>
</tr>
<tr>
<td>Sex, male</td>
</tr>
<tr>
<td>Age, years, mean (SD)</td>
</tr>
<tr>
<td>Age&lt;85 years</td>
</tr>
<tr>
<td>Risk factors</td>
</tr>
<tr>
<td>Hypertension</td>
</tr>
<tr>
<td>Diabetes</td>
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<tr>
<td>Atrial fibrillation</td>
</tr>
</tbody>
</table>
| Motor fibre along the course of the pyramidal tract secondary to a small cerebral lacunar infarct that disrupts the corticospinal fibers independently of the sites of the lesion. An alternative or complementary explanation may be that although supratentorial small, deep infarcts can be seen on imaging in DCHS, there is a possibility that the clinical syndrome could be secondary to a tandem non-imaged lesion in the pons. Future studies using diffusion weighted imaging would allow more precise discrimination of the sites of lesions in DCHS with higher sensitivity and specificity.

From a clinical point of view, this study highlights the favourable outcome in DCHS and, as expected, the difference between DCHS and non-lacunar stroke. More interestingly, however, is the comparison between DCHS and other lacunar syndromes. Limb weakness but not cerebellar-type ataxia, speech disturbances, corona radiata and basilar artery involvement, and freedom from symptoms at discharge were significantly more frequent in patients with DCHS, but after multivariate analysis, only limb weakness and absence of functional disability at discharge were significant predictors of DCHS. Unilateral pyramidal signs in the form of mild weakness is a well known distinguishing feature of DCHS, being absent in the majority of atypical lacunar syndromes and in other lacunar syndromes, such as pure sensory stroke. In the literature, differentiation between ataxic hemiparesis and DCHS is not always clear, but in accordance with Fisher & Mohr & Marti-Vilalta, patients with obvious ataxia and definitive weakness were classified as having ataxic hemiparesis and were not included among

Clinical findings

<table>
<thead>
<tr>
<th>Subtype of stroke</th>
<th><strong>DCHS</strong></th>
<th><strong>Other lacunar syndromes</strong></th>
<th><strong>Non-lacunar stroke</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Lacunar infarct</td>
<td>33 (94.3)</td>
<td>403 (75.3)</td>
<td>&lt;0.005</td>
</tr>
<tr>
<td>Atherothrombotic infarction</td>
<td>1 (2.9)</td>
<td>34 (6.3)</td>
<td>518 (33.6)</td>
</tr>
<tr>
<td>Cardioembolic infarction</td>
<td>1 (2.9)</td>
<td>20 (3.7)</td>
<td>447 (29)</td>
</tr>
<tr>
<td>Infarction of unusual cause</td>
<td>0</td>
<td>40 (7.5)</td>
<td>85 (5.5)</td>
</tr>
<tr>
<td>Infarction of unknown cause</td>
<td>0</td>
<td>8 (1.5)</td>
<td>240 (15.6)</td>
</tr>
<tr>
<td>Intracerebral haemorrhage</td>
<td>0</td>
<td>20 (3.7)</td>
<td>250 (16.2)</td>
</tr>
<tr>
<td>Neuroimaging findings</td>
<td>Internal capsule</td>
<td>14 (40)</td>
<td>180 (33.6)</td>
</tr>
<tr>
<td>Thalamus</td>
<td>1 (2.9)</td>
<td>81 (15.1)</td>
<td>&lt;0.03</td>
</tr>
<tr>
<td>Basal ganglia</td>
<td>1 (2.9)</td>
<td>42 (7.8)</td>
<td>NS</td>
</tr>
<tr>
<td>Pons</td>
<td>6 (17.1)</td>
<td>37 (6.9)</td>
<td>&lt;0.04</td>
</tr>
<tr>
<td>Corona radiata</td>
<td>3 (8.6)</td>
<td>26 (4.8)</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Basilar artery involvement</td>
<td>6 (17.1)</td>
<td>37 (6.9)</td>
<td>&lt;0.04</td>
</tr>
<tr>
<td>Outcome</td>
<td>Symptom free at discharge</td>
<td>16 (45.7)</td>
<td>129 (24.2)</td>
</tr>
<tr>
<td>Severe disability at discharge</td>
<td>0</td>
<td>13 (2.4)</td>
<td>NS</td>
</tr>
<tr>
<td>Urinary complications</td>
<td>0</td>
<td>18 (3.3)</td>
<td>NS</td>
</tr>
<tr>
<td>Infectious complications</td>
<td>1 (2.9)</td>
<td>22 (4.1)</td>
<td>NS</td>
</tr>
<tr>
<td>Length of hospital stay, mean (SD)</td>
<td>11.71 (5.75)</td>
<td>12.64 (8.45)</td>
<td>20.55 (25.1)</td>
</tr>
<tr>
<td>Mortality in hospital</td>
<td>0</td>
<td>4 (0.7)</td>
<td>NS</td>
</tr>
</tbody>
</table>

*DCHS v other lacunar syndromes.
†DCHS v non-lacunar stroke.
Percentages in parentheses or otherwise stated.
patients with DCHS. DCHS presents characteristic and well-
differentiated clinical and semiological features, and the
absence of neurological disability in 46% of patients indicates
that DCHS is the lacunar syndrome with the most favorable
outcome.

In summary, DCHS is a rare cerebrovascular syndrome (6% of
lacunar strokes) and, in the majority of patients, is due to
small vessel disease as the stroke mechanism at capsular or
pontine sites. The majority of patients in our study had internal
capsule infarcts. The prognosis is good with striking
similarity to clinical features of other lacunar syndromes.
There are important differences between DCHS and non-
lacunar strokes in demographic and risk factors, clinical, sites
of lesion, and outcome.

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