Quality of life after decompressive craniectomy for malignant middle cerebral artery infarction

Malignant middle cerebral artery (MCA) infarction is a devastating condition leading to early death in nearly 80% of cases due to the rapid rise of intracranial pressure despite maximum medical management of the ischaemic brain oedema. Decompressive craniectomy (DC) has been proposed to prevent brain herniation in malignant MCA infarction, but it remains controversial in the absence of randomised controlled trials and because of the fear of a severe residual disability after surgery. We present herein the results of a quality of life assessment using patient and proxy versions of the Stroke Impact Scale (SIS) in eight patients 12–30 months after craniectomy for malignant MCA infarcts.

Methods

Between March 1999 and November 2000, all consecutive patients with malignant MCA infarction were treated by DC and durotomy at Lariboisière Hospital if they were younger than 55 years of age, had a complete MCA infarct as defined by complete MCA territory CT ischaemic changes, and a severe haemorrhagic transformation involving more than 50% of the MCA territory, and significant contralateral ischaemia.

Results

Ten patients were included (eight men and two women, mean (SD) age 41 (12) years, range 15–54). The mean (SD) NIH score scale at admission was 21 (3), range 16–25. Five patients had a left sided stroke with severe aphasia. The mean time between stroke onset and surgery was 65 (68) h, range 12–252. One patient had a late DC because of recurrent MCA infarct at day 9 after the first stroke. All patients had signs of temporal brain herniation before surgery including uni- or bilateral mydriasis (9/10), Cheyne-Stokes hypoventilation (8/10), or decerebration (6/10). The mean (SD) duration of hospitalisation in the intensive care unit was 22 (20) days, range 3–58. Two patients died, one from a cerebral abscess and the other from a large epidural hematoma.

All living patients (8/10) were followed for a mean (SD) duration of 21 (20) months, range 12–30. All were managed in a specialised stroke rehabilitation unit with a mean (SD) hospital stay of 12 (11) months, range 4–24, after which they returned home with either home rehabilitation facility or day hospital care. At the end of follow up, 7/8 patients had an mRS ≤ 4 (table 1). The mean (SD) NIH score scale was 13 (4), range 8–18.

The two youngest patients had the best scores on disability (mRS = 2) and were fully independent for the activities of daily living (BI ≥ 90) (table 1).

The 64 SIS items could be measured in all patients except patient 7 who had severe aphasia (table 1). The proxy version of the SIS was administered to a close relative (five spouses, two parents) or an employed caregiver (one). The mean (SD) patient assessment of global perception of stroke recovery was 59 (16). The score was lower, but not significantly so, in patients with aphasia compared to patients without, both in patient (55 (15) vs 65 (19), p = 0.48, Wilcoxon test) and proxy (49 (17) vs 57 (18), p = 0.45, Wilcoxon test) versions of the measurement.

The combined mean (SD) physical domain recovery was 48 (16) when assessed by patients and 39 (16) when assessed by proxies. The lowest scaling success rate was for hand function and the highest for emotion domain recovery. However, during the follow up, two patients had a major depressive episode. In addition, one spouse attempted suicide (patient 8). As expected, patients with aphasia had a lower mean (SD) rate of recovery for communication (50 (37)) than those without (91 (14)), although the difference was not statistically significant (p = 0.21, Wilcoxon test). No patient returned to his or her prior employment, although one patient, the youngest (patient 3), returned to school.

Discussion

This study shows that the SIS measurement is applicable to patients with malignant MCA infarction 12–30 months after craniectomy. Patients’ assessment of the physical aspects of disability at 12–30 months post stroke was high (all physical domains mean recovery of 48/100). Interestingly, the proxy

Table 1

<table>
<thead>
<tr>
<th>Patients/age (years)/sex</th>
<th>SIS version</th>
<th>Stren-</th>
<th>Hand</th>
<th>Mobili-</th>
<th>ADL/</th>
<th>Physical</th>
<th>Emot-</th>
<th>Mem-</th>
<th>Commu-</th>
<th>Partic-</th>
<th>% of</th>
<th>Stroke</th>
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<tbody>
<tr>
<td>mRS-B*</td>
<td>gth</td>
<td>function</td>
<td>ty</td>
<td>ADL/ IAD†</td>
<td>combined score‡</td>
<td>Emotion</td>
<td>Memory</td>
<td>Communication</td>
<td>Participation</td>
<td>recovery</td>
<td>recovery (VAS)</td>
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<tr>
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<td>98</td>
<td>85</td>
<td>61</td>
<td>100</td>
<td>95</td>
<td>210</td>
<td>7</td>
<td>75</td>
<td>75</td>
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<td>55</td>
<td>0</td>
<td>100</td>
<td>93</td>
<td>62</td>
<td>96</td>
<td>87</td>
<td>210</td>
<td>7</td>
<td>74</td>
<td>70</td>
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<td>0</td>
<td>60</td>
<td>6</td>
<td>24</td>
<td>64</td>
<td>87</td>
<td>210</td>
<td>7</td>
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<td>42</td>
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<td>100</td>
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<td>75</td>
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<td>57</td>
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<td>67</td>
<td>48</td>
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*Modified Rankin Scale-Barthel Index; †fatvicty of daily living/instrumental activities of daily living; ‡combined physical score calculated from the strength, hand function, and mobility domain scores.

ND, not done; VAS, visual analogue scale.
assessment of psychosocial recovery was lower (39/100) than the patient assessment. In addition, the disability measured by the mRS showed that 68 living patients had an mRS=3, which may indicate a poor outcome. It may be that in patients with malignant MCA infarction, the patient version of the SIS overestimates the physical recovery because of cognitive dysfunction including unilateral neglect, anosognosia, or aphasia.

One main concern in malignant MCA infarction is the psychosocial impact of stroke. In our study, the percentage of recovery was good for emotion and memory but moderate for communication and participation. As expected, patients with aphasia had a lower rate of recovery for communication than patients without, though the difference did not reach statistical significance, presumably because of the small numbers. In the same way, the global participation is lower in patients with aphasia than in patients without, though the proxy’s overestimates the physical recovery because of other factors.

Cerebral sinus thrombosis in a patient with Cushing’s syndrome

It is well known that hypercortisolism induced by Cushing’s disease or syndrome, or by administration of glucocorticoids, causes thromboembolic complications.1 However, the precise mechanisms underlying the hypercortisolism induced hypercoagulable state still remain unknown. Here we describe a case of cerebral lateral sinus thrombosis with Cushing’s syndrome. Glucocorticoid induced overproduction of factor VIII and von Willebrand factor (VWF) may have contributed to the development of the cerebral thrombosis in this patient.

Case report

A mildly obese 30 year old woman was admitted to our hospital because of headache and nausea. She was not taking any medications, including oral contraceptives, before admission. The patient had no intracranial hypertension; her fundi showed no papilloedema, and intracranial pressure measured by lumbar puncture was normal (4.3 mmHg). Brain computed tomography (CT) showed a high density lesion in the left temporopontal occipital lobe (fig 1B). Magnetic resonance venogram (MRV) on the first hospitalised day showed a filling defect in the left lateral sinus (fig 1B). These findings were consistent with cerebral lateral sinus thrombosis.

Laboratory data showed elevation of factor VIII (183 %), one stage clotting assay; normal range 60–150 %), VWF (275 %; normal range 60–150 %), thrombin–antithrombin III complex (15.5 ng/ml), plasminogen activator inhibitor-1 (PAI-1) (123 %), and D-dimer (2.1 µg/ml). Other major factors related to coagulopathy and fibrinolysis, including antithrombin III (112 %), fibrinogen (330 µmol/l), plasminogen (117 %), plasminogen–α2- plasmin inhibitor complex (0.9 µg/ml), protein C (87 %), protein S (95 %), were within normal limits. Markers of acute phase reaction such as C reactive protein and erythrocyte sedimentation rate were not elevated. Neither antiphospholipid antibodies nor antinuclear antibodies were detected. The patient was treated with intravenous heparin and subsequent oral administration of warfarin potassium. A relative fibrinolytic enhancement following the strict anticoagulation may have caused recanalisation of the lateral sinus, which was confirmed by the following MRV (fig 1C). The patient’s symptoms disappeared completely.

During the extensive examination of thrombotic causes, we suspected the presence of hypercortisolism because of the presence of central obesity and moon facies. As a result, we found a left adrenal tumour, which was accompanied by hypercortisolism (210 µg/l) with suppressed adrenocorticotropic hormone (3 µg/ml). The left adrenal mass showed a high uptake of 131I-iodo-cholesterol on scintigram. These findings were consistent with Cushing’s syndrome. After the laparoscopic left adrenalectomy, the patient received replacement therapy with hydrocortisone for approximately 1 year. Plasma levels of factor VIII and VWF decreased gradually to the normal level (130 % and 140 %, respectively), 1 year after adrenalectomy.

Discussion

We report the first case of cerebral sinus thrombosis associated with Cushing’s syndrome. Thromboembolic complications are well known to occur in the patients with hypercortisolism.1 Most are deep vein thromboses and pulmonary thromboembolism. However, there are no reports so far to show association with cerebral sinus thrombosis and Cushing’s syndrome.

A few reports suggest that factor VIII and VWF may have roles in the development of thromboembolic complications associated with hypercortisolism.1 As well as blood group, sex, age, inflammation, and endothelial dysfunction,2 hypercortisolism may have enhanced VWF release3 and secretion to the circulation. Because VWF is generally observed.

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1182 PostScript
Spinal muscular atrophy, Dandy-Walker complex, and cataracts in two siblings: a new entity?

Lower motor neurone involvement is the main feature of several neurological disorders, including the various forms of spinal muscular atrophy (SMA). A distinct form of SMA is characterised by predominantly distal weakness and atrophy of the limbs. Various combinations of SMA with neural and extraneural defects, mainly pontocerebellar hypoplasia, have also been reported.

We report a combination of distal SMA with Dandy-Walker complex and anterior polar cataracts in two brothers. The patients were aged 25 and 23 years. Their parents, who originated from the same area of Greece, were unrelated and asymptomatic. Since the age of 10 years, both brothers presented with progressively deteriorating symmetrical distal muscle weakness and atrophy of the lower limbs, which affected mainly the anterior tibialis and peroneal muscles and, to a lesser degree, the gastrocnemius, resulting in an almost “stoke-like” appearance of the legs. Bilateral anterior polar cataracts had been diagnosed in both patients at the age of 9–11 months. Additional findings of the neurological examination in both patients were slight muscle strength reduction in both hands and forearms and decreased tendon reflexes in the upper and lower limbs, while the Achilles’ tendon reflexes could not be elicited. No sensory, plantar responses were normal. No sensory, cerebellar, or cognitive impairment was found. Dysmorphic features were not observed. The general physical examination was normal in both patients.

Extensive haematological, biochemical, and immunological investigation of both patients, including levels of creatine kinase, prolactin, hexosaminidase A, anti-GM1 and antisialidase antibodies, cortisol, thyroid hormones, vitamin B12 and folic acid, immunoglobulin, and androgen receptor genes was negative.

Magnetic resonance imaging revealed the presence of Dandy-Walker complex in both patients. There was enlargement of the cisterna magna, with slight hypoplasia of the vermis and slight elevation of the tentorium (fig 1). No supratentorial or brainstem abnormalities were observed. The magnetic resonance imaging of the spine was normal in both brothers, as were visual and brainstem evoked responses. Ophthalmological examination confirmed the presence of anterior polar cataracts in 15 both patients.

The karyotype was normal in both patients. Molecular genetic analysis for mutations in the survival motor neurone (SMN; exon 7 and 8 deletions), neuronal apoptosis inhibitory protein (NAIP; exon 5 and 6 deletions), and androgen receptor genes was negative.

Discussion

Our patients were two brothers with almost identical clinical and laboratory findings. One of the main features was the involvement of the anterior horn cells, which was compatible with distal SMA, according to published criteria. Additional features were Dandy-Walker complex and bilateral anterior polar cataracts.

There are several reports of SMA with additional features (SMA plus), among them pontocerebellar hypoplasia. These cases, however, are characterised by proximal muscles involvement, early presentation with profound floppiness at birth, mental retardation, and cerebellar signs. There are also rare reports of recessive distal SMA with additional features: diaphragmatic paralysis or pyramidal signs. None of these cases of proximal or distal SMA plus has been linked to chromosome 5q.

Familial cases of Dandy-Walker complex are not uncommon; however, the combination of the disorder with SMA seems to be quite unusual. The aforementioned severe cases of pontocerebellar hypoplasia in SMA plus clearly constitute a different nosological entity.

The coexistence of early onset cataracts with neuromuscular disorders is also unusual. Apart from the well known occurrence of cataracts in myotonic dystrophy, there are some reports of cataracts in combination with spastic paraparesis, spinocerebellar degeneration or neuropathy, and facial dysmorphism. There is also a report of familial congenital cataracts and Dandy-Walker anomaly with lissencephaly.

In conclusion, physicians should be aware that Cushing’s syndrome is a possible cause of cerebral sinus thrombosis. Plasma levels of factor VII and VWF may play an important role in the hypercoagulable state in hypercortisolism.

References


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We were not able to locate in the literature any reports of distal SMA in combination with any form of Dandy-Walker variant and/or congenital anomalies. We were not able to locate a genetic defect, a not altogether unreasonable task, in view of the available information on the genetics of the main features of our patients, contiguous gene syndrome, pleiotropy, and/or contiguous gene syndrome. The fact that the patients were first-degree relatives and presented with identical phenotypes is a strong indication that the disorder is genetically determined. With the available information on the genetics of the main features of our patients, contiguous gene syndrome appears unlikely. We were not able to locate a genetic defect, a not altogether unexpected result, as most recessive distal SMA families remain to be genetically determined.

Future investigation of similar cases should include genetic studies relevant to all three main features of the disorder.

**References**