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 hemiplegia with altered level of consciousness with further neurological deterioration due to brain oedema, and if a close family member gave informed consent. Exclusion criteria were: prestroke moderate to severe disability defined by a modified Rankin scale (mRS) 2–5, haemorrhagic transformation involving more than 90% of the MCA territory, and significant contralateral ischaemia.

Disability was assessed using the mRS and the Barthel Index (BI), and quality of life using the French version of the SIS 2.0. The SIS comprises eight domains, four physical domains (including strength, hand function, mobility, and activity of daily living/instrumental activities of daily living) and four psychosocial domains (including emotion, communication, memory, and social participation) and includes the patient’s global assessment of percentage of recovery. The scores of each domain range from 0 to 100, with 100 being the best.

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 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 12–232. One patient had a late DC because of recurrent MCA infarct at day 9 after the first stroke. All patients had signs of temporal herniation before surgery including uni- or bilateral mydriasis (9/10), Cheynes-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 (21) 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. Our patient 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: Domain scores of the SIS questionnaire filled in by seven living patients and eight proxies 12–30 months after decompressive craniectomy

<table>
<thead>
<tr>
<th>Patients/age (sex)</th>
<th>mRS-BI*</th>
<th>SIS version</th>
<th>Strength</th>
<th>Hand function</th>
<th>Mobility</th>
<th>ADL/IADL†</th>
<th>Physical combined score‡</th>
<th>Emotion</th>
<th>Memory</th>
<th>Communication</th>
<th>Participation</th>
<th>% of recovery</th>
<th>Stroke recovery (VAS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/23/M 2–95</td>
<td>Patient</td>
<td>60</td>
<td>0</td>
<td>98</td>
<td>85</td>
<td>61</td>
<td>100</td>
<td>95</td>
<td>100</td>
<td>61</td>
<td>75</td>
<td>75</td>
<td>75 (10)</td>
</tr>
<tr>
<td>2/49/F 5–15</td>
<td>Proxy</td>
<td>55</td>
<td>0</td>
<td>100</td>
<td>93</td>
<td>62</td>
<td>96</td>
<td>87</td>
<td>100</td>
<td>61</td>
<td>74</td>
<td>70</td>
<td>70 (10)</td>
</tr>
<tr>
<td>3/15/M 2–90</td>
<td>Patient</td>
<td>31</td>
<td>0</td>
<td>60</td>
<td>6</td>
<td>24</td>
<td>64</td>
<td>87</td>
<td>75</td>
<td>42</td>
<td>45</td>
<td>40</td>
<td>40 (8)</td>
</tr>
<tr>
<td>4/45/M 4–85</td>
<td>Proxy</td>
<td>31</td>
<td>0</td>
<td>60</td>
<td>2</td>
<td>20</td>
<td>69</td>
<td>84</td>
<td>79</td>
<td>28</td>
<td>39</td>
<td>30</td>
<td>30 (5)</td>
</tr>
<tr>
<td>5/47/F 4–35</td>
<td>Patient</td>
<td>55</td>
<td>0</td>
<td>60</td>
<td>47</td>
<td>40</td>
<td>53</td>
<td>60</td>
<td>40</td>
<td>27</td>
<td>47</td>
<td>40</td>
<td>40 (5)</td>
</tr>
<tr>
<td>6/54/F 4–35</td>
<td>Proxy</td>
<td>55</td>
<td>0</td>
<td>44</td>
<td>55</td>
<td>46</td>
<td>84</td>
<td>60</td>
<td>43</td>
<td>69</td>
<td>58</td>
<td>50</td>
<td>50 (5)</td>
</tr>
<tr>
<td>7/46/M 4–60</td>
<td>Patient</td>
<td>40</td>
<td>0</td>
<td>36</td>
<td>52</td>
<td>32</td>
<td>84</td>
<td>57</td>
<td>37</td>
<td>67</td>
<td>48</td>
<td>60</td>
<td>60 (5)</td>
</tr>
</tbody>
</table>

*Modified Rankin Scale-Barthel Index; tactivity 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.

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competing interests: none declared

References
2 Brown MM. Surgical decompression of patients with large middle cerebral artery infarcts is ineffective: a meta-analysis. Stroke 2003; 34: 2305–6

Cerebral sinus thrombosis in a patient with Cushing's syndrome
It is well known that hypercortisolism induced by Cushing's disease and syndrome, or by administration of glucocorticoids, causes thromboembolic complications. 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 overproduction and von Willebrand factor (VWF) may have contributed to the development of the cerebral sinus 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 medication, 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 (14.3 mmHg). Brain computed tomography (CT) showed a high density lesion in the left temporo-occipital lobe (fig 1A). 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 60–160%), VWF (275% of normal range 60–170%), 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 coagulation and fibrinolysis, including antithrombin III (112%), fibrinogen (330 µg/ml), plasminogen (117%), plasminogen activator inhibitor complex (0.9 µg/ml), protein C (87%), and 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 multiple factors. We found a left adrenal tumour, which was accompanied by hypercortisolism (210 µg/dl) with suppressed adrenocorticotropic hormone (3 µg/ml). The left adrenal mass showed a high uptake of 131I-adsterol 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 patients with Cushing's syndrome. Most are deep vein thromboses and pulmonary thromboembolisms. 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. As well as blood group, sex, age, inflammation, and endothelial dysfunction, hypercortisolism is reported to be an important determinant factor for plasma levels of VWF. Huang et al showed that dexamethasone stimulated VWF release from cultured human endothelial cells. Factor VIII is mainly synthesised in the liver and secreted to the circulation. Because VWF protects factor VIII from proteases, a concordant increase of factor VIII and VWF in plasma is generally observed. Results in this study show that high plasma level of factor VIII (especially over 150%) is an independent risk factor for venous thromboembolism, including cerebral sinus thrombosis. In this case, considerable elevation of factor VIII and VWF was observed specifically before removal of the adrenal tumor. Thus, hypercortisolism may have enhanced VWF release from endothelial cells to increase factor VIII, thereby causing a hypercoagulable state. The present case also suggests that measurement of factor VIII and VWF may be useful to decide if anticoagulation therapy can be ceased after successful adrenalectomy in Cushing's syndrome. However, because hypercortisolism does not always cause hypercoagulable state, some genetic factors, such as polymorphism of steroid receptor, may determine whether glucocorticoids increase plasma levels of factor VIII and VWF.

It is also reported that PAI-1 is often elevated and may cause thromboembolic complications by lowering fibrinolytic activity and by increasing fibrin formation. In this case, a high uptake of 131I adsterol on scintigram may have contributed to the thrombus formation. However, factor V Leiden, a common coagulation abnormality in Western populations, may not have participated in thrombus formation in our case, because it is considered that the mutation is not present in the Japanese population.
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 neutral 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 neurologic 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, 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, α-galactosidase, and anti-syphilis antibodies, cortisol, thyroid hormones, vitamin B12 and folic acid, immunoglobulin, and androgen receptor genes was negative. The aforementioned severe cases of pontocerebellar hypoplasia in SMA plus clearly constitute a different nosological entity.

There are also rare reports of 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 flaccid paralysis at birth, mental retardation, and cerebellar signs. There are also rare reports of recessive distal SMA with additional features: diaphragmatic paralysis or pyramidal signs.

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 both patients.

The karyotype was normal in both patients. Molecular genetic analysis for mutations in the survival motor neuron (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 flaccid paralysis 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.

References

4. Nakayama H, Setoyama H, Kurosawa Y, et al. 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.
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tal cataracts.
form of Dandy-Walker variant and/or congeni-
reports of distal SMA in combination with any
We were not able to locate in the literature any
Figure 1 Brain magnetic resonance imaging of the older sibling. T1 and T2 weighted sagittal
images, showing enlargement of the cisterna magna and slight elevation of the tentorium.

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 congeni-
tal defects.

In summary, our cases represent an unusual
combination in distal SMA. This combination
does not seem to fit in any of the already
described syndromes and could be the result of
pleiotropy, contiguous gene syndrome, or
chance. 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.3
Future investigation of similar cases should
include genetic studies relevant to all three
main features of the disorder.

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Competing interests: none declared

References
1 Delonge P, Timmerman V, van Broeckhoven C. Second workshop of the European CMT
2 Zerres K, Rudnik-Schoneborn S. 93rd ENMC international workshop: non-Sq spinal muscular
atrophy (SMA) – clinical picture. 6–8 April 2001, Naarden, the Netherlands. Neuromusc

Cranial neuroimaging and clinical
neuroanatomy

Edited by Hans-Joachim Kretschmann,
Wolfgang Weinrich. Published by Thieme,
2003, €199 (hardback), pp 451. ISBN 3-13-
672603-0

Already a well established reference, this
third edition of Cranial Neuroimaging and Clinical Neuroanatomy was significantly updated,
thereby offering a more comprehensive
approach to neuroanatomy than did any of the previous editions.

This book is divided into 10 chapters and an atlas. The introduction gives an overview
of the scope of this book but also provides very useful information on “basics” such as
the various imaging planes, their historical
evolution, and their definitions. The next
chapter then briefly describes the various
neuroimaging techniques illustrating the
useful approaches to the imaging of the
various areas of the head and neck.

This is then followed by the atlas, which is
really at the heart of this book, as is emphasised
by the subtitle. The atlas is subdivided into four
sections—one section for each of the three
planes and a fourth section for the posterior
fossa. This edition retains in all of these
sections the excellent line drawings of previous
editions, which were obtained from gross
anatomic slices. These are now complemented
by new, large sized, state of the art T1 and T2
weighted MR images in all three planes as well as
CT images in the axial plane. These images increase significantly the practical utility of this
atlas, which is enhanced by the fact that each of
the four sections has a coloured margin of its
own.

The topography of the neurocranium, the
cranio-cervical junction, and the pharynx are
succinctly discussed in the following two
chapters covering among others the skull, the
CSF spaces, the vascular territories and the
subdivisions of the brain. Of particular
interest are the newly introduced three
dimensional reconstructions of the vascular
tree, as well as the detailed schemes of the
vascular supply of the posterior fossa.

The well illustrated chapter of the Neuro-
fuctional Systems relates the anatomy with
the physiology thereby underlining the clinical
utility of this book. This is finally followed by
the last chapter offering a succinct overview on
neurotransmitters and neuromodulators.

In summary, this an excellent companion
for students and medical trainees that will
help them both in their initial as well as in
their more advanced stages in getting a better
command of the complex but very seductive
world of neuroanatomy.

T A Yousry

The A–Z of neurological practice. A
guide to clinical neurology

Edited by Roger A Baker, Neil Scolding,
Dominic Rowe, Andrew J Larner. Published by
Cambridge University Press, 2004, £45.00

This pocket sized book consists of a comprehen-
sive series of entries from A to Z, each one
describing a specific aspect of neurology. The
authors provide overviews of major disease
groups (eg, headache, epilepsy) as well as more
detailed descriptions of specific disease catego-
ries (eg, SUNCT syndrome, gelastic epilepsy)
throughout 936 pages. The entries are organ-
ised in a structured way and usually include
information on pathophysiology, clinical fea-
tures, investigations and diagnosis, differential
diagnosis, and treatment and prognosis. Some
literature is quoted and extensive cross refer-
ces to other entries are provided.

This is a very useful reference book for
everyone who works in clinical neurology or
related areas. It can also be used by general
physicians who need some fast and succinct
information on neurological issues. For obvious
reasons this book cannot replace a textbook.
The overviews of the major disease groups
provide only the basic information, and the
entries are of limited value for differential
diagnosis and therapy. The main advantage of
this “guide to clinical neurology” is that it
provides relevant and up-to-date information
on each neurological topic in a readable and
accessible manner. This is of particular interest
if the treating neurologist or generalist is
confronted by one of the numerous rare
neurological disorders and/or syndromes. This
goal is also achieved by the myriad of entries
and cross references. In summary, we can
recommend this reference book as a useful
supplement to the traditional textbooks in the
neurologist’s bookshelf.

J C Möller, W H Oertel

BOOK REVIEWS