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 and/or CT ischaemic changes, and a severe hemiplegia with altered level of consciousness and/or haemorrhagic transformation involving more than 50% 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 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 unilateral or bilateral mydriasis (9/10), Cheynes-Stokes hypventilation (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) v 65 (19), p=0.48, Wilcoxon test) and proxy (49 (17) v 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. The patient’s 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>
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<tr>
<th>Patients/age (years)/sex</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>
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<td>100</td>
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</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.
assessment of physical domains recovery was lower (39/100) than the patient assessment. In addition, the disability measured by the mRS showed that 6/8 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 partici- cation. As expected, patients with aphasia had a lower rate of recovery for communica- tion than patients without, though the difference did not reach statistical signifi- cance, presumably because of the small numbers. In the same way, the global percentage of recovery was lower, but not significantly, in patients with aphasia than in patients without. Interestingly, the proxy’s assessment of psychosocial recovery, though lower, was close to the patient’s assessment.

In conclusion, this study shows that after craniectomy for malignant MCA infarctions, even though the perception of physical aspects of disability is high, that of psycho- social impairment is lower. Open surgery for craniectomy for malignant MCA infarction indicate that surgery decreases death rates. However, randomised trials are needed, taking into account not only death and depen- dency but also quality of life.

Acknowledgements

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K Vahedi, J Benoist, A Kurtz
Service de Neurologie, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France

J Mateo
Département d’Anesthésie-Réanimation, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France

A Blanquet
Service de Neurochirurgie, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France

M Rossignol
Département d’Anesthésie-Réanimation, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France

P Amarenco
Service de Neurologie, Hôpital Bichat, AP-HP, Paris, France

A Yelínk
Service de Médecine Physique et de Réadaptation, Hôpital Lariboisière-Fernand Widal, AP-HP, 200 rue du Faubourg, Saint-Denis, 75010 Paris, France

E Vicaut
Unité de Recherche Clinique, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France

D Payen
Département d’Anesthésie-Réanimation, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France

M G Bousseau
Service de Neurologie, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France

Correspondence to: Dr Katayoun Vahedi, Service de Neurologie, Hôpital Lariboisière, AP-HP, 2 rue A. Paré, 75010 Paris, France; katayoun.vahedi@bichat.ap-hp.fr

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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. However, the precise mechanism 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 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 medica- tions, including oral contraceptives, before admission. The patient had no intracranial hypertension; her fundi showed no papillo- edema, and intracranial pressure measured by lumbar puncture was normal (4.3 mmHg).

Brain computed tomogram (CT) showed a high density lesion in the left tempo-occipital lobe (fig 1A). Magnetic resonance venogram (MRV) on the first hospitalised day showed a filling defect in the 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 %), and D-dimer (330 μg/ml). Other major factors related to coagulation and fibrinolysis, including antithrombin III (112 %), fibrinogen (330 mg/ml), plasmine 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 war- farin potassium. A relative fibrinolytic enhancement following the strict anticoagu- lation may have caused recanalisation of the lateral sinus, which was confirmed by the following brain 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. In this patient, 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-adosterol on scintigram. These findings were consistent with Cushing’s syndrome. After the laparo- scopic left adrenalectomy, the patient received replacement therapy with hydrocorti- solone for approximately 1 year. Plasma levels of factor VIII and VWF decreased gradually to the normal level (130 % and 140 %, respec- tively), 1 year after adrenalectomy.

Discussion

We report the first case of cerebral sinus thrombosis associated with Cushing’s syn- drome. Thromboembolic complications are well known to occur in patients with hypercortisolism.1 Most are deep vein throm- boses 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.2 As well as blood group, sex, age, inflammation, and endothe- lial dysfunction,2 hypercortisolism is reported to be an important determinant factor for plasma levels of VWF.3 Huang et al4 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 con- cordant increase of factor VIII and VWF in plasma is generally observed.4 As a result, studies show that high plasma level of factor VIII (especially over 150 %) is an independent risk factor for venous thromboembolism,5 including cerebral sinus thrombosis.6 In our patient, 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 syn- drome. 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.7 In our patient, considerably elevated factor VIII and PAI-1 levels were observed before craniectomy for malignant MCA infarction. However, high but significant elevation of PAI-1 in this case may also have contributed to the thrombus formation. However, factor V Leiden, a common coagulation abnormality in Western populations, may not have partici- pated in thrombus formation in our case, because it is considered that the mutation is not present in the Japanese population.7
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. The 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 antibodies, cortisol, thyroid hormones, vitamin B12, and folic acid, immunoglobulin G, and lipoprotein electrophoresis, and cerebrospinal fluid examination, was normal.

The electrophysiological examination revealed findings compatible with anterior horn involvement in both patients. Specifically, the electromyogram showed chronic active denervation of distal muscles, with large amplitude motor unit potentials and presence of polyphasic potentials and spontaneous activity (fibrillations and positive waves), more pronounced in the lower limbs. Nerve conduction studies showed normal motor and sensory conduction velocities, with normal amplitudes, latencies, and F waves. No conduction blocks were recorded. Electrophysiological investigation of both parents was normal. Both parents refused consent for muscle biopsy.

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 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. Additionally, 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 neuro-pathy, and facial dysmorphism. There is also a report of familial congenital cataracts and Dandy-Walker anomaly with lissencephaly.

References

We were not able to locate in the literature any reports of distal SMA in combination with any of the currently 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. Future investigation of similar cases should include genetic studies relevant to all three main features of the disorder.

M Panas, K Spengos, G Tsivgoulis, N Kalfakis, C Sfagos, D Vassiliopoulos
Department of Neurology, Eginition Hospital, University of Athens, Greece

N Markomichelakis
Department of Ophthalmology, Georgiaio Gennimatas General Hospital, Athens, Greece

Correspondence to: Dr M Panas, Eginition Hospital, Neurogenetics Unit, Department of Neurology, University of Athens, 74 Vas. Sofias Avenue, Athens 11528, Greece; mpanas@med.uoa.gr
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References


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.

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


This pocket sized book consists of a comprehensive 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 categories (eg, SUNDCT syndrome, gelastic epilepsy) throughout 936 pages. The entries are organised in a structured way and usually include information on pathophysiology, clinical features, investigations and diagnosis, differential diagnosis, and treatment and prognosis. Some literature is quoted and extensive cross references 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

Craniocervical junction and the pharynx

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 four 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 it 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 anatomical 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 additions 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 Neurofunctional 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 Youssry
Quality of life after decompressive craniectomy for malignant middle cerebral artery infarction

K Vahedi, L Benoist, A Kurtz, J Mateo, A Blanquet, M Rossignol, P Amarenco, A Yelnik, E Vicaut, D Payen and M G Bousser

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