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# Illustrative Teaching Case

Section Editors: Daniel Strbian, MD, PhD, and Sophia Sundararajan, MD, PhD

## Malignant Hemispheric Infarction Diagnosis and Management by Hemicraniectomy

Lauri Soinne, MD, PhD; Sophia Sundararajan, MD, PhD; Daniel Strbian MD, PhD

### Case Description

A previously healthy, right-handed 43-year-old man was found lying on the floor in his apartment. His relatives had been unable to contact him for more than a day. On admission, he was somnolent but attempted to obey commands and had a right-sided hemiplegia, dense sensory loss, severe word-finding difficulty, and anisocoria with miosis and ptosis in left eye (National Institutes of Health Stroke Scale, 21). Emergent computerized tomographic scan revealed an extensive left-hemispheric infarction (Figure [A]). Computerized tomographic angiography showed a left carotid dissection. The neurosurgeon recommended medical management; hypertonic saline was initiated. Fifteen hours after admission, his level of consciousness had deteriorated (Glasgow coma score, 10), his left pupil was enlarged and fixed, and increased midline shift was visible on computerized tomography (Figure [B]). The patient was referred to decompressive hemicraniectomy (DHC), which was performed 20 hours after admission. By 72 hours, the patient was more alert and his left pupil was reactive (still enlarged). Because of ventilatory problems, he required intermediate care for subsequent 2 weeks. After tracheostomy removal 18 days after admission, the patient was alert but hemiplegic and hemianopic, obeying commands, and able to communicate with single words and short phrases. He was discharged to a rehabilitation hospital 10 days later and thereafter to a supported residential home. Six years later, he was able to walk with a cane, go shopping unaided, communicate reasonably, and read short sentences with difficulty. His right arm remained plegic, and the left arm has restricted movement because of a fracture at the time of stroke. Therefore, he needs help with dressing and grooming but not every day (modified Rankin scale, 3).

### Discussion

The case typifies a malignant middle cerebral artery infarction, which usually results from an internal carotid or middle cerebral artery stem occlusion. Middle cerebral artery infarction may be accompanied with anterior cerebral artery infarction (especially in internal carotid occlusions) but also with posterior cerebral artery infarction (internal carotid occlusion

in patients with prominent posterior communicating artery or as a result of transtentorial compression during the expansive stage, like in our patient). Such concomitant involvement is often fatal within the first week if managed conservatively (transtentorial herniation). The downward spiral of symptoms is marked with increasing drowsiness, reduced level of consciousness, pupillary enlargement, and coma. Approximately 1 of 20 strokes has a malignant course, which occurs more commonly in younger adults with less brain atrophy (less ability to accommodate edema). Life-saving treatment by DHC has been available for >25 years. Pooled major trials show a marked effect in reducing mortality and the most severe dependency in patients aged <60 years.<sup>1</sup> The number-needed-to-treat is striking: in patients aged <60 years, number-needed-to-treat is 2 in reduction of mortality and 4 in reduction of severe disability (modified Rankin scale, 0–3). A trial in patients aged >60 years (a protocol similar to one of the major trials) showed increased survival without complete dependency, although their outcome in general was worse compared with younger patients.<sup>2</sup> In the elderly, the number-needed-to-treat for survival without the most severe form of disability (modified Rankin scale, 0–4) was 5. However, it is notable that patients surviving because of DHC were invariably left with moderate to severe disability (modified Rankin scale, 4–5), which does not strictly support the notion of improved functional outcome.<sup>2</sup>

### Diagnosis and Timing

Early recognition of the malignant course is vital. Intuitively, it seems that secondary damage could be minimized with faster action. Some data indicate that an ultraearly intervention could bring extra benefit in reducing mortality but other not.<sup>3,4</sup> Although the proper timing may not be considered strictly evidence based, the available data support an early intervention rather than an expectant approach. Prediction tool for malignant development is needed, including potentially baseline stroke severity, level of consciousness (Glasgow coma score or National Institutes of Health Stroke Scale item 1a), and the need for mechanical ventilator support. Imaging parameters depicting the early infarct size (exceeding 50% of

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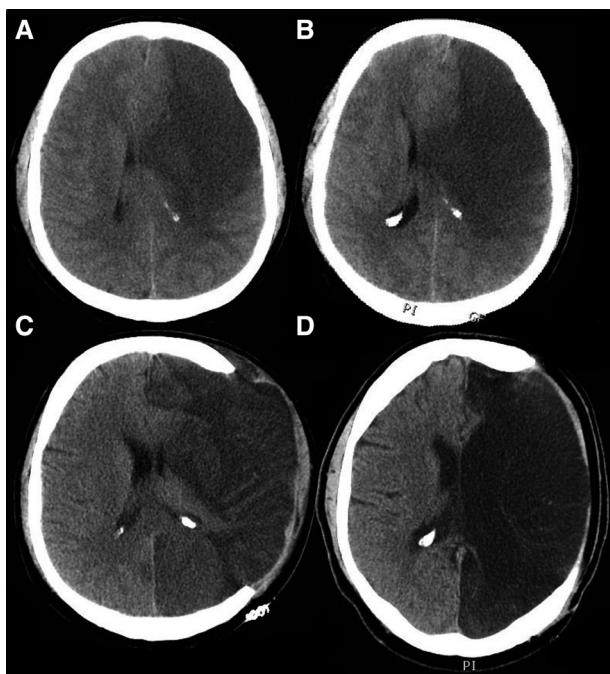
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**Figure.** **A**, An admission computerized tomographic scan showing an expansive left middle cerebral artery (MCA) infarction (midline shift 10 mm). **B**, Extended midline shift 15 hours later (subfalcine herniation). **C**, Follow-up scan a week later reveals a secondary infarction in the posterior cerebral artery territory in addition to the extensive anterior and MCA infarctions.

**D**, Years after, the bony defect has been replaced with an artificial lambeau.

the middle cerebral artery territory or perfusion deficits  $>2/3$  of the area) are central predictors of herniation. MRI is more sensitive than computerized tomography at defining infarction size early.<sup>5</sup> A diffusion-weighted lesion size exceeding 82 mL has proved a specific early predictor, although lacking sensitivity.<sup>5</sup> The sensitivity and overall predictive value of early imaging can be enhanced by clinical follow-up data (24 hours) or severity of ischemia.<sup>6,7</sup>

### Patient Selection

DHC is a highly effective, life-saving, evidence-based treatment, however, many physicians have concerns about quality of life: do we produce further suffering as a by-product of overzealous treatment? Reviewing subjective quality-of-life data is encouraging. Despite certain degree of disability, the majority of patients/caregivers would undergo DHC again (even patients with more severely restricted functionality). Physicians must avoid the pitfall of being overly pessimistic in the hyperacute stage in our assumptions about lost functions. Cognitive outcome may be better than expected on the basis of physical handicap.<sup>8,9</sup> The quality-adjusted life years are gained at a high cost, estimated to be  $\approx 60\,000$  € per quality-adjusted life years gained for lifetime in long-term modeling.<sup>10</sup> Without an early aggressive approach, a miserable outcome becomes a self-fulfilling prophecy of the worst kind. The trials do not support the view that favoring nondominant hemisphere strokes would be associated with better outcomes.<sup>4,9</sup> This previously adopted hypothesis may reflect the tendency to emphasize the importance of linguistic disability at the expense of the

cognitive sequelae in nondominant hemisphere strokes, as well as our limited understanding of what the patients consider acceptable outcomes. The pitfall of overly mechanistic view on cerebral localization is also exemplified by our case, in which the degree of recovery of linguistic functions exceeds all expectations for a right-handed individual with an extensive lesion. This suggests the possibility of a less conventional representation of eloquent areas in this particular case. Clearly DHC can be life saving and should be considered in patients with malignant infarction. More data are warranted for a better case-by-case management. Importantly, current data allow consideration of the biological age of patients as opposed to strict age limits for DHC.

### TAKE-HOME POINTS

- Malignant middle cerebral artery infarction can be predicted by neuroradiological and clinical examination.
- Decompressive hemicraniectomy is effective for malignant middle cerebral artery infarction, with dramatic reduction in mortality and improved functional outcome, although many patients remain with permanent disability (eg, shift from modified Rankin scale 5 to modified Rankin scale 3–4) and require costly support.
- Current data indicate that also patients aged  $>60$  years may benefit; however, each case should be considered individually because the patients are invariably left with moderate to severe disability.
- Timely surgery is recommended to minimize further injury because of cerebral edema and herniation.

### Disclosures

None.

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