Stroke Note

Cerebrovascular Diseases

Published online: December 13, 2011

Cerebrovasc Dis 2012;33:94–95
DOI: 10.1159/000332808

Observation of Indirect Neorevascularization after Leptomeningeal Biopsy in a 34-Year-Old Woman with Moyamoya Syndrome – Should Burr Holes Be Considered as an Alternative Revascularization Technique in Younger Adults with Moyamoya?

Jan C. Purrucker[a], *Stefan Rohde*[b], *Sibylle Stampfl*[b], *Peter A. Ringleb*[a]

Departments of [a]Neurology and [b]Neuroradiology, University Hospital of Heidelberg, Heidelberg, Germany

Case Report

A 34-year-old woman with right-sided hemiparesis and worsening of existing aphasia presented for reevaluation. When in summer 2009 the patient was admitted for the first time to our stroke unit, she suffered from posterior infarction on the left side. Three months later, she developed ischemia in the left semioval center. In 2001 and 2008, the patient had already suffered from basal ganglia infarction on the left side. Cerebral angiography demonstrated a stenosis of the left middle cerebral artery (MCA, M1 segment) and left anterior cerebral artery (ACA, A3 segment) with formation of a rete mirabile – consistent with either vasculitis or moyamoya syndrome. In October 2009, the patient underwent an open left frontoleptomeningeal biopsy: during craniotomy, a bone flap of 3 × 3 cm was removed and after biopsy fixed by two CranioFix®. Additionally, two drill holes for intraoperative dura sutures were made. Neither pathological examination, nor further laboratory diagnostics did show signs of vasculitis. After only 16 months of no further ischemic episode, the patient suffered from two new transient ischemic attacks (TIA). This time, cerebral angiography demonstrated a new stenosis of the right ACA, a progress of the left MCA stenosis and an increase of the cloudlike rete (fig. 1f), as well as the formation of anastomosis between the posterior and anterior pericallosal artery (Fischer anastomosis). Interestingly, we observed new collateralization with branches from both the left and right external carotid artery through the trepanation defect with anastomosis to left-sided peripheral MCA and ACA branches supplying blood to the left frontal cortex (fig. 1b). MRI scans showed reduced perfusion particularly in the left dorsal medial territory (fig. 1h), but improved perfusion compared to preoperative scans in the frontal MCA territory (fig. 1g).

Discussion

Moyamoya in general is a progressive disease and the neurological status at the time of treatment, more than age, predicts long-term outcome. Since medical treatment alone is shown not to be sufficient [1, 2], patients should be evaluated for surgical treatment at an early state. Revascularization approaches include several indirect techniques and the technically difficult superficial temporal artery to MCA bypass (recently reviewed in Parray et al. [1] and Baaj et al. [3]). Although we early evaluated our patient for a possible superficial temporal artery-MCA bypass, the patient showed a refusing attitude. Our findings of neorevascularization one and a half years after craniotomy with consequent enhancement of the frontal medial territory perfusion seems to be more than a welcomed side effect. Indirect revascularization by multiple burr holes is a technique used since the 1980s for children suffering from moyamoya disease [4]. In 1998, Kawaguchi et al. [5] published a small series of 3 children treated with multiple burr-hole operation, without recognizing new postoperative ischemic attacks despite progressive stenosis of the major cerebral arteries. Fourteen children with pseudomeningoceles as the main complication but lack of further ischemic attacks were treated by Sainte-Rose et al. [6]. A 10-year-old boy with frequent TIAs and radiological features of moyamoya disease was treated with 5 burr holes on each side but suffered from another TIA during a 6-month follow-up [7]. Another case of a 12-year-old girl with strong neorevascularization after placing 6 burr holes after recurrent right-hemispheric ischemia is reported [8]. There is only one study examining multiple burr-hole operation in adults with moyamoya disease. Ten patients (mean age 37.8 years; 2 of them suffering from intraventricular hemorrhage) were treated with 1–4 burr holes and showed neorevascularization in 41 of 43 burr holes. Preoperative symptoms improved in patients with both infarction and hemorrhage, and TIAs disappeared [9]. Our observation supports burr holes as a treatment option in early stages not only in children, but also in younger adults. Patients can undergo multiple burr-hole operation under local anesthesia, perioperative complications are rare and further complex treatment options are not excluded if they become necessary.

References

1 Parray T, Martin TW, Siddiqui S: Moyamoya disease: a review of the disease and anesthetic management. J Neurosurg Anesthesiol 2011;23:100–109.
2 Scott RM, Smith ER: Moyamoya disease and moyamoya syndrome. N Engl J Med 2009;360:1226–1237.
3 Baaj AA, Agazzi S, Sayed ZA, Toledo M, Spetzler RF, van Loveren H: Surgical management of moyamoya disease: a review. Neurosurg Focus 2009;26:E7.

KARGER

Fax +41 61 306 12 34
E-Mail karger@karger.ch
www.karger.com

1015–9770/12/0331–0094$38.00/0

Accessible online at:
www.karger.com/ced

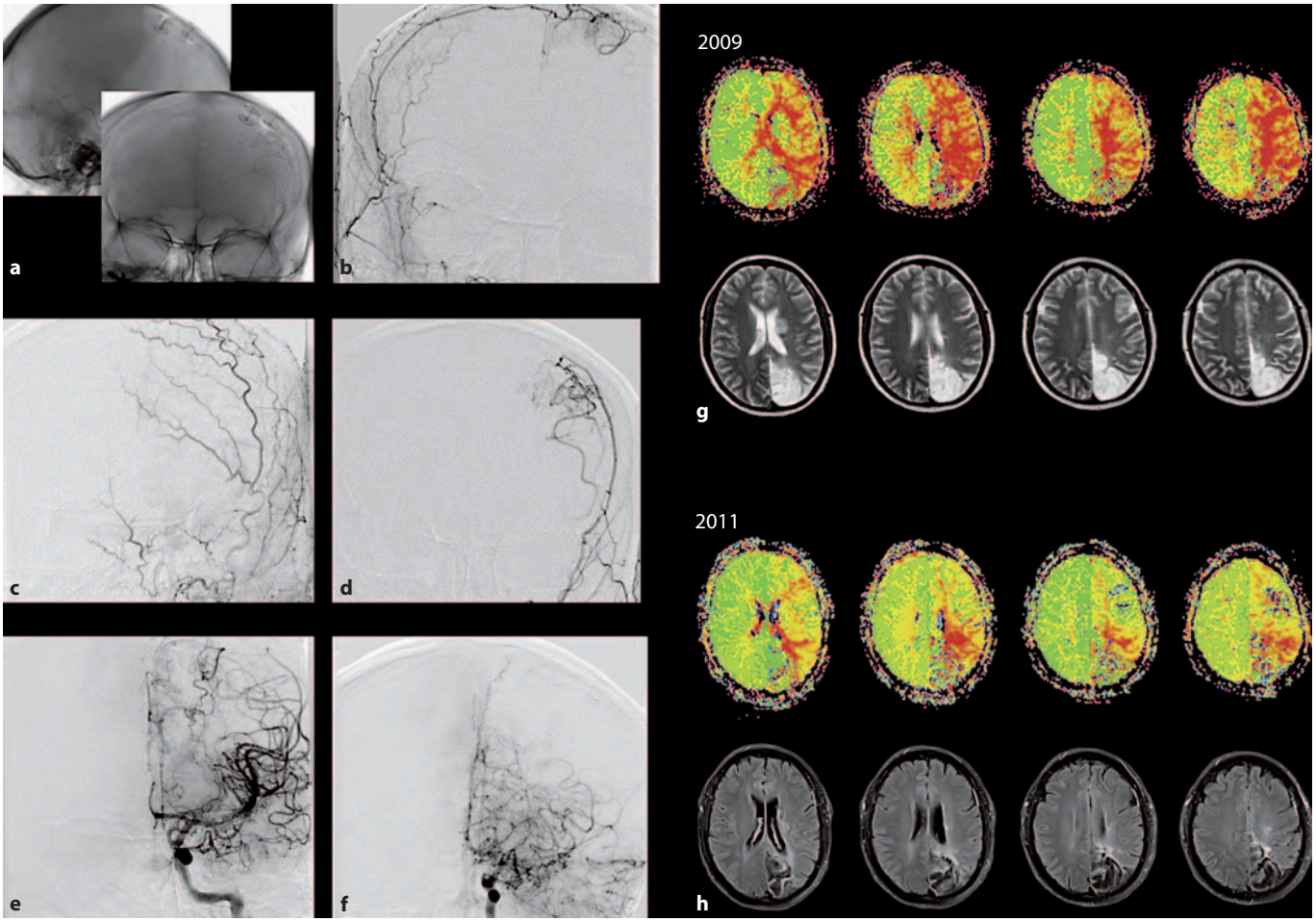

Fig. 1. a–f Digital subtraction angiography of the extra- and intracranial vasculature. The unsubtracted lateral and frontal views (**a**) demonstrate the left frontal position of the two CranioFix. **b** A run from the right external carotid artery (frontal view) shows transcranial/transdural anastomoses of the contralateral superficial temporal and the middle meningeal artery to peripheral MCA and ACA branches. **c**, **d** Frontal view of the left external carotid artery before (**c**, 2009) and after surgery (**d**, 2011) showing intensive neorevascularization at the site of the burr hole trepanation. **e**, **f** Cerebral angiography of the left internal carotid artery (**e**, 2009) in contrast to **f** (2011) showing the progressively rarefied branches of the MCA and the typical cloudlike rete of moyamoya. **g** Preoperative perfusion-weighted MRI scans (time-to-peak) taken 6 days before surgery show reduced perfusion in the whole left MCA territory. T_2-weighted sequences (lower row) show the old posterior infarction and subacute ischemia in the left semioval center. **h** Latest perfusion-weighted MRI scans (time-to-peak) demonstrate delayed perfusion in the frontoparietal MCA territory. Interestingly, the perfusion in the frontal MCA territory below the trepanation burr hole now shows a normal time-to-peak pattern (upper row). Fluid-attenuated inversion recovery sequences (lower row) show – besides the old infarctions – white matter lesions especially located in the dorsal MCA territory.

4 Endo M, Kawano N, Miyaska Y, Yada K: Cranial burr hole for revascularization in moyamoya disease. J Neurosurg 1989;71:180–185.
5 Kawaguchi T, Fujita S, Hosoda K, Shibata Y, Komatsu H, Tamaki N: Usefulness of multiple burr-hole operation for child moyamoya disease. Neurol Surg 1998;26:217–224.
6 Sainte-Rose C, Oliveira R, Puget S, Beni-Adani L, Boddaert N, Thorne J, Wray A, Zerah M, Bourgeois M: Multiple bur hole surgery for the treatment of moyamoya disease in children. J Neurosurg 2006;105:437–443.
7 Kapu R, Symss N, Cugati G, Pande A, Vasudevan C, Ramamurthi R, Lakshimpathi A: Multiple burr hole surgery as a treatment modality for pediatric moyamoya disease. J Pediatr Neurosci 2010;5:115.
8 Spetzler RF, Geber MS: Burr holes for moyamoya disease. Barrow Quarterly 2002;18:22–24.
9 Kawaguchi T, Fujita S, Hosoda K, Shose Y, Hamano S, Iwakura M, Tamaki N: Multiple burr-hole operation for adult moyamoya disease. J Neurosurg 1996;84:468–476.

Dr. Jan C. Purrucker
Department of Neurology, University Hospital of Heidelberg
Im Neuenheimer Feld 400, DE–69120 Heidelberg (Germany)
Tel. +49 622 156 37718, E-Mail jan.purrucker@med.uni-heidelberg.de