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Leptomeningeal contrast enhancement in moyamoya: its potential role in postoperative assessment of circulation through the bypass

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Abstract Leptomeningeal contrast enhancement (LMCE) is one of the MRI features of moyamoya. Its clinical significance, however, is not elucidated. Our purpose was to characterise LMCE on MRI and to evaluate its role in the assessment of circulation through a surgically established bypass in moyamoya. We studied 16 patients with idiopathic moyamoya (seven males, nine females, includingt four children, aged 7 to 54 years, mean 24 years) who underwent T1-weighted MRI before and after intravenous contrast medium. The presence of LMCE, its intensity and anatomical distribution, catheter angiographic findings, and relation of LMCE to the bypass surgery were assessed. More LMCE was seen in the cerebrum in most patients with moyamoya than in normal controls. LMCE in the brain stem and cerebellum was minimal, similar to that seen in the controls. LMCE was less prominent following surgery than before operation or in patients who did not undergo surgery. In three patients examined both before and after operation LMCE became less prominent following bypass surgery. As LMCE becomes less prominent after "effective" bypass surgery, this may be used for evaluation of effectiveness of surgery in moyamoya.

Key words Moyamoya · Surgery, bypass · Contrast enhancement, leptomeningeal · Magnetic resonance imaging

Introduction

Classic MRI features of moyamoya include new or old infarcts or haemorrhage, brain atrophy and ventricular dilatation; lack of flow void in the distal internal carotid artery and/or the proximal portions of the anterior and middle cerebral arteries; and fine flow void in the dilated perforating arteries at the base of the brain (moyamoya vessels) [1, 2, 3, 4, 5, 6]. In addition to these features, Ohta et al. [7] reported leptomeningeal enhancement by gadolinium diethylenetriamine-pentaacetic acid (Gd-DTPA) in children with moyamoya, which they called the "ivy sign" because the appearances resembled creeping ivy on a stone. We have evaluated this leptomeningeal contrast enhancement (LMCE) on MRI in children and adults with idiopathic moyamoya and discuss its clinical significance in the assessment of blood flow through a surgically established bypass.

Materials and methods

We studied 16 patients with idiopathic moyamoya: seven males and nine females, of whom four were aged less than 17 years old, aged 7 to 54 years, mean 24 years. The diagnosis of moyamoya was established by clinical manifestation and conventional catheter angiography in all patients. The initial symptoms were ischaemia (transient ischaemic attack or cerebral infarct) in all cases except for patient 2 (who had a small haemorrhage) and patient 12 (who was asymptomatic). Ten patients had undergone bypass surgery, three were awaiting surgery or were not operated upon, and three were examined both pre- and postoperatively. Bypass

surgery was either superficial temporal artery–middle cerebral artery (STA-MCA) anastomosis or enceplalo-duro-arterial synangiosis (EDAS). STA-MCA anastomosis was performed on both sides in four patients and EDAS on both sides in nine. Patient 6 underwent MRI preoperatively and following both unilateral and bilateral EDAS. Patients 4–6 developed occasional transient ischaemic attacks, and therefore subsequently underwent bypass surgery. The other 13 patients (three unoperated and 10 postoperative) were neurologically stable without ischaemic or haemorrhagic episodes at least for 1 year prior to the MRI.

All patients underwent axial T1-weighted spin-echo MRI before and after Gd-DTPA 0.1 mmol/kg body weight at 1.0 Tesla. Imaging parameters were: repetition time 560, echo time 15 ms, two excitations, slice thickness 6 mm, matrix 256 × 256, field of view 230 mm. The patients did not move between the pre- and postinjection imaging. Three normal volunteers without cerebrovascular disease (all men, aged 31, 55 and 60 years) underwent similar imaging.

Four-vessel catheter cerebral angiograms were performed within 12 days of the MRI in patients 3–8. In the other 10 patients, the interval between MRI and angiography ranged from 6 months and 4 years 3 months, mean 1 year 9 months; no neurological events occurred in the interim. Antegrade internal carotid artery flow to the territories of the anterior and middle cerebral arteries and flow from the external carotid system were assessed separately and rated as none, minimal, moderate and marked (scored 0–3). "Marked" was defined as supply to more than half the territory of the anterior and middle cerebral arteries in the frontal, temporal and parietal lobes, while "minimal" indicated supply to less than a quarter of these areas; "moderate" was intermediate. Collateral flow from the posterior cerebral arteries was not assessed because antegrade internal carotid flow and collateral flow from the external carotid system usually shows a reciprocal relationship, unless cerebral infarcts occur.

Subtraction images (unenhanced images subtracted from corresponding contrast-enhanced images) of patients and normal subjects were created by a computer program, to facilitate detection of subtle signal changes after contrast medium infusion. LMCE was judged as : none, minimal, moderate and marked (scored 0–3). "Minimal" was defined as invisible on T1-weighted, but visible on subtracted images, while "marked" was prominent on unsubtracted images; "moderate" was again intermediate.

Patients 3,5 and 6, (aged 19, 24 and 54 years) underwent examinations to investigate the time-course of LMCE. Patients 3 and 6 were awaiting surgery and patient 5 had already had bypass surgery. These patients underwent imaging immediately, 10, 20 and 30 min after injection of Gd-DTPA at the standard dose. Subtraction images could not be derived from the images at 10, 20 or 30 min, because of motion artefacts.

Two neurosurgeons (MK, MN) jointly reviewed the unenhanced and contrast enhanced T1-weighted and subtracted images and catheter angiography films retrospectively, without blinding. By consensus, the degree of LMCE, its anatomical distribution and the extent of blood supply on the catheter angiograms were interpreted.

Results

In the three normal subjects, contrast enhanced T1-weighted images showed enhancement of the choroid plexus, pituitary gland, relatively large cortical veins and venous sinuses, but no LMCE was apparent on unsubtracted images. Subtraction images disclosed minimal symmetrical LMCE over the cerebrum, brain stem and cerebellum, but LMCE was less apparent over the cerebellum due to flow artefacts caused by the adjacent large venous sinuses, because phase-encoding was horizontal (Fig. 1).

Results in the patients are shown in Table 1. Moderate or marked LMCE was observed over the cerebrum in most of the patients (Figs. 2a, 3a). However, little or no LMCE was seen in regions of brain atrophy caused by old cerebral infarcts. LMCE was minimal over the brain stem or cerebellum (Fig. 3f). In patients 4–6, examined both pre- and postoperatively, LMCE became less apparent after bypass surgery (Fig. 2b). In patient 6, left and right EDAS were performed separately with an interval of 9 months. At 1 month after the left EDAS, LMCE did not change (Fig. 3i), but it was less prominent on only the operated side 7 months postoperatively (Fig. 3j). After bilateral EDAS, LMCE become less prominent bilaterally (Fig. 3k).

The average scores for pre- and postoperative LMCE in the frontal, temporal, parietal and occipital lobes and the entire hemisphere were 2.3, 2.6, 2.4, 2.3, 2.5 and 1.8, 1.7, 1.3, 1.4, 1.6, respectively. If the data of Patient 12 are excluded (because there was almost no collateral flow from the external carotid system in this patient), the postoperative scores become 1,7, 1.6, 1.2, 1.4 and 1.5, respectively. There was significant reduction in the LMCE after "effective" bypass surgery ($P < 0.05$); LMCE in the cerebrum was usually less prominent in the presence of surgically established anastomoses or spontaneously established transdural anastomoses.

In patients 3–8, who underwent catheter angiography (four pre- and five postoperatively) within 12 days of MRI, the average scores for angiographic collaterals from the external carotid system had a negative correlation with the degree of LMCE ($r = -0.6$). LMCE was less apparent in the patients with adequate surgically established and spontaneous transdural anastomoses than in those without. It was thus generally less prominent postoperatively than in patients who had not undergone surgery. The exception was patient 12, who had almost no collateral flow through anastomoses; even following surgery, marked LMCE was observed over the cerebrum.

In the patients who underwent sequential imaging, LMCE was observed on unsubtracted images at all stages after injection) (Fig. 3a–d). LMCE over the cerebrum was, however, most prominent on images obtained soon after injection (Fig. 3a). LMCE rapidly decreased in intensity, but was still faintly visible 30 min after contrast medium administration (Fig. 3d). There was no delayed parenchymal enhancement in these patients.

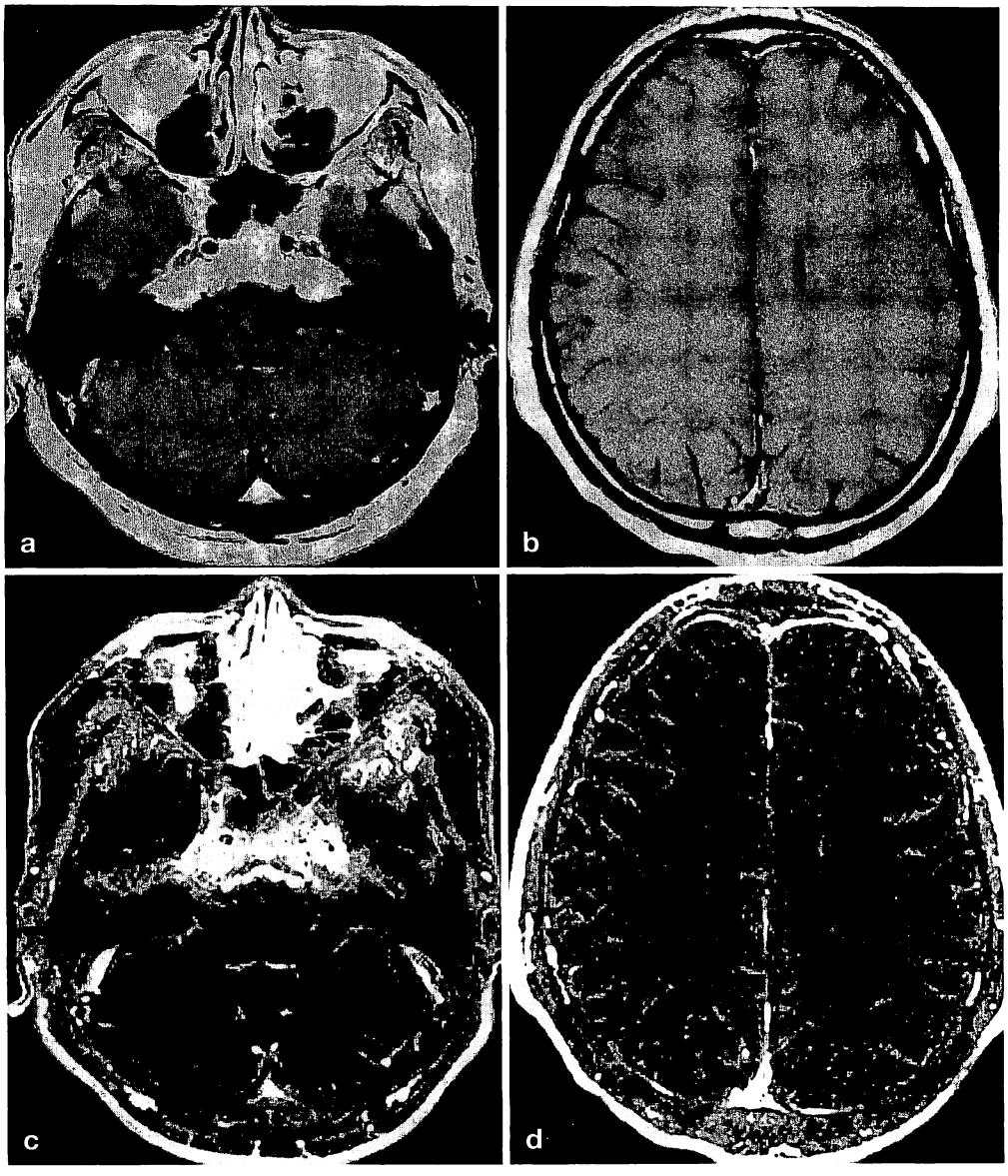

Fig. 1 a–d A 31-year-old normal subject. Contrast-enhanced T1-weighted (**a, b**) and subtracted images (**c, d**). Only the latter reveal minimal leptomeningeal contrast enhancement (LMCE) over the cerebrum. LMCE in the cerebellum is less apparent due to flow artefacts

Discussion

The classic MRI features of moyamoya have been listed above. MRI is less sensitive to leptomeningeal and transdural anastomoses than to flow void in moyamoya vessels [4, 5].

MR angiography (MRA) features are stenosis or occlusion of the distal internal carotid and/or proximal anterior and middle cerebral arteries and fine vascular structures at the base of the brain. MRA is less sensitive to advanced moyamoya, especially in adults because the moyamoya vessels are less apparent than in children [6].

In normal subjects, LMCE was observed only on subtracted images. As the intensity of LMCE decreased rapidly and it was observed only in the leptomeninges, this enhancement is presumably caused by T1-shortening of intravascular protons in the leptomeninges.

Demaerel et al. [8] observed marked meningeal enhancement on MRI in a child with moyamoya. Ohta et al. [7] noted that LMCE occurred in children with moyamoya and that its intensity decreased after bypass surgery. Maeda and Tsuchida [9] observed LMCE on fluid-attenuated inversion-recovery images in a 15-year-old girl. They did not, however, describe it in normal subjects or adults with moyamoya, or discuss its frequency in moyamoya, its occurrence on the brain stem and cerebellum.

We observed that LMCE was: minimal over the brain stem, cerebrum and cerebellum in normal subjects; minimal over the brain stem and cerebellum in patients with

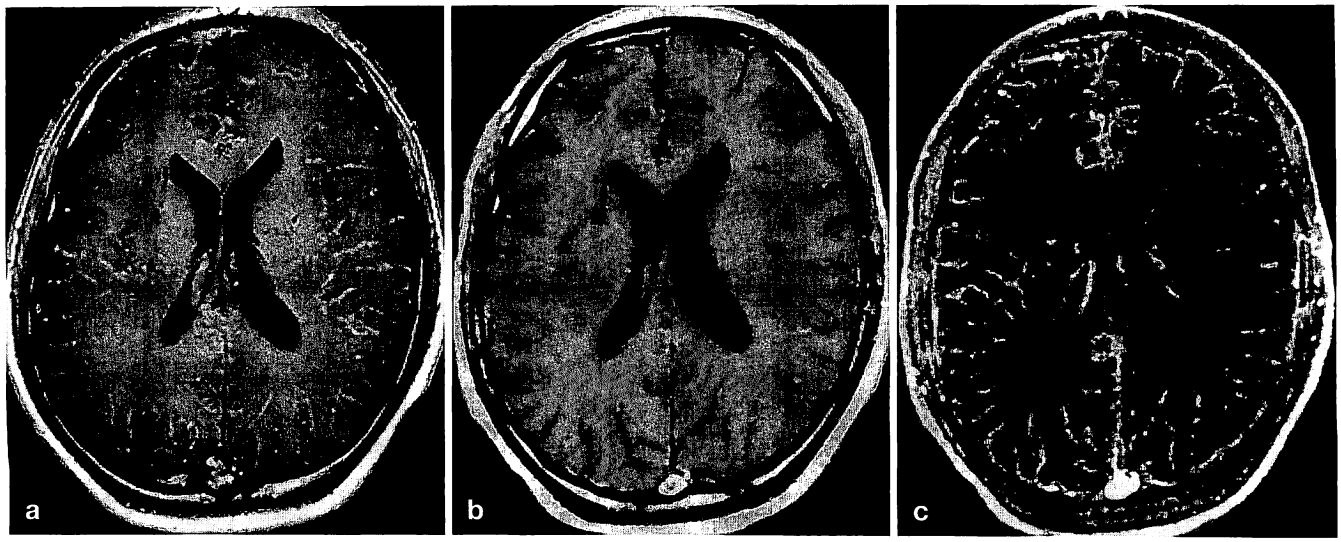

Fig. 2 a–c Patient 5 developed a transient ischaemic attack with alternate hemiparesis several times when singing loudly during the 2 years preceding preoperative MRI. Superficial temporal -middle cerebral artery anastomoses were performed bilaterally. MRI was repeated 1 year after surgery. LMCE over the cerebrum is marked preoperatively (**a**) but after bypass surgery (**b**) it becomes moderate, while a subtracted image shows it more clearly (**c**)

moyamoya; increased over the cerebrum in most children and adults with moyamoya; minimal or absent in regions of brain atrophy due to old infarcts; reduced over the cerebrum by spontaneous transdural anastomoses; most prominent in patients with moyamoya soon after administration of contrast medium, fading rapidly thereafter; less prominent after effective bypass surgery; marked in a patient with inadequate surgical anastomosis from the external carotid system; reduced ispilaterally by unilateral bypass surgery; and unchanged 1 month, but reduced 7 months after EDAS.

Ohta et al. [7] postulate that LMCE is attributable to fine arterial neovascularisation over the leptomeninges, which is observed over the cortical surface during bypass surgery. Maeda and Tsuchida [9] suggested that it represents slow, retrograde flow in engorged pial vessels via leptomeningeal anastomosis. LMCE was, however, detected even in the normal subjects. We believe that LMCE in these individuals is an enhancement of the slow blood flow in the normal vessels of the leptomeninges, and that in the patients with moyamoya there is enhancement of the slow, stagnant flow in the markedly dilated arteries of the leptomeninges. We postulate that this vascular enhancement is similar to the "intravascular enhancement sign" in acute cortical infarction [10, 11, 12], in which arterial "flow void" due to high-velocity signal losses and spin-dephasing effects decreases, with consequent high intensity from the vessels, from the mixture of blood and contrast medium. Slow antegrade flow is caused by progressive occlusive change in the distal internal carotid arteries and slow retrograde flow by leptomeningeal collaterals from the posterior cerebral arteries. The slow, stagnant arterial flow is observed as prolonged cerebral circulation on conventional cerebral angiograms. Bypass surgery usually results in reduction in the circulation time of the pial blood flow from the external carotid system. The negative correlation between the scores for angiographic collaterals from the external carotid system and LMCE ($r = -0.6$) and the significant reduction in LMCE after "effective" bypass surgery ($P < 0.05$) may support this speculation. Reduction in LMCE after bypass surgery was slightly more prominent in the parieto-occipital than the frontotemporal region. This may be attributable to the location of the surgical anastomoses and pre-existing leptomeningeal collaterals from the posterior cerebral arteries. Since the anastomoses are usually posterior

Fig. 3 a–m Patient 6 developed transient weakness and occasionally involuntary movements of the right arm during the year preceding the MRI examination. **a-d** T1-weighted images immediately, 10, 20 and 30 min after contrast medium: the intensity of LMCE decreases with time, but it is faintly visible even in **d**. Subtracted images immediately after injection (**e, f**) show LMCE to be marked over the cerebrum but minimal LMCE over the cerebellum. **g, h** Preoperative right internal and external carotid angiography shows poor blood flow. The dural branch of the right ascending pharyngeal artery (arrow) supplys the contralateral occipital lobe, where LMCE is not prominent. **i** LMCE is not decreased 1 month after a left encephalo-duro-arterial synangiosis (EDAS), but is reduced 7 months after surgery (**j**) especially in the parietal region; on the right it remains prominent. **k** It has decreased bilaterally 20 months after the left and 11 months after a right EDAS. **l,m** Postoperative right internal and external carotid angiography shows increased collateral flow from the external carotid system, correlating with decreased LMCE

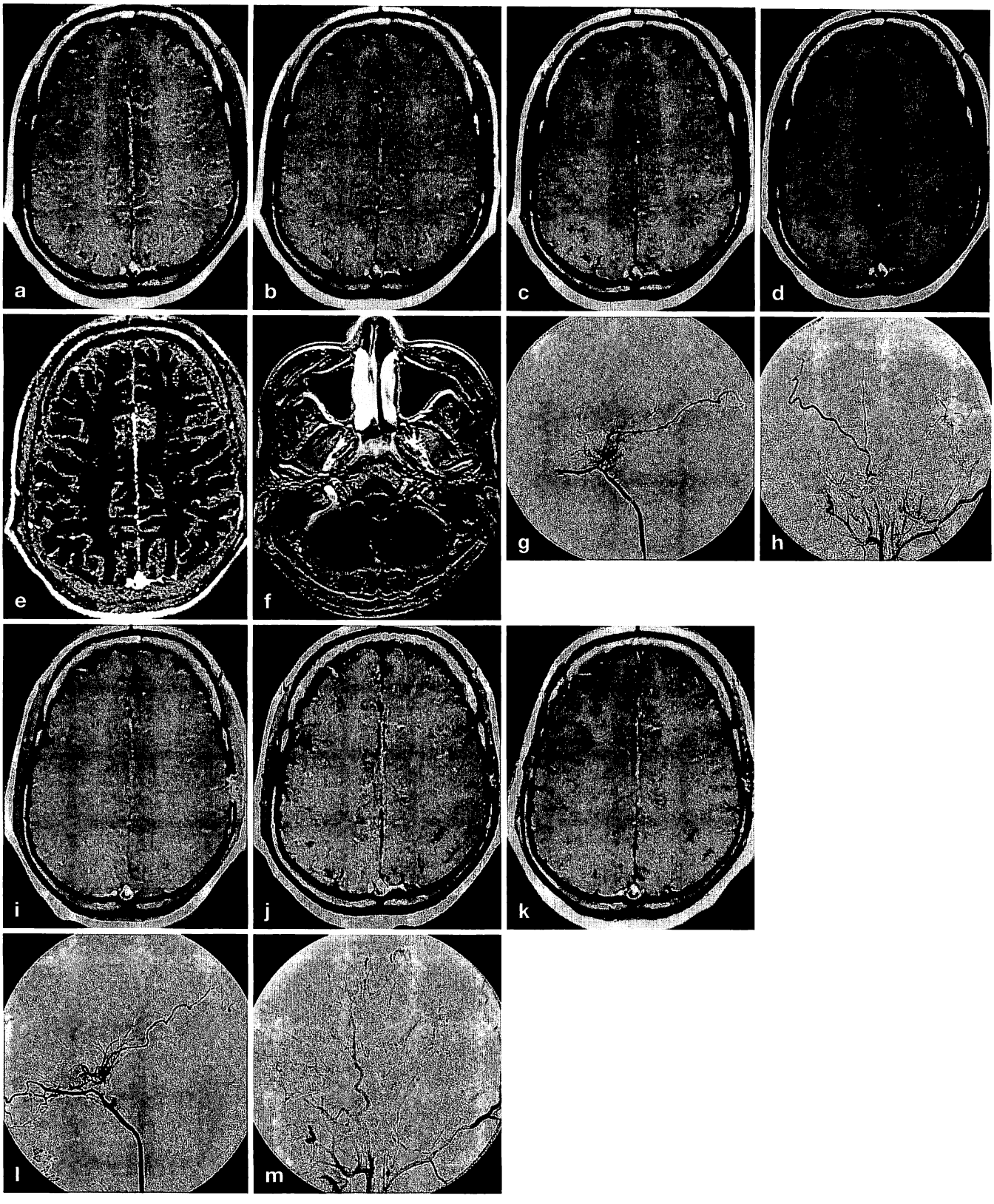
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Table 1 Clinical data on patients (*B* bilateral, *EC* external carotid artery system, *EDAS* encephalo-duro-arterial synangiosis, *F* female, *Fr* frontal lobe, *IC* internal carotid artery system, *L* left, *m* month, *M* male, *O* occipital lobe, *P* parietal lobe, *R* right, *STA-MCA* superficial temporal artery-middle cerebral artery anastomosis, *T* temporal lobe, *y* year)[a, b]

Patient	Age (years), sex	Bypass surgery	Time from surgery to MRI	Time from initial symptom to MRI	Leptomeningeal cointrast enhancement score									Angiographic score				Remarks
					R				L				Average	R		L		
					Fr	T	P	O	Fr	T	P	O		IC	EC	IC	EC	
1	21, M	-	..	8 y	3	3	3	2	2	2	2	2	2.4	2	2	3	1	
2	44, M	-	..	3 y	2	2	1	1	2	2	1	1	1.5	3	0	3	0	Small putamimal haemorrhage
3	54, F	-	..	2 y	2	3	3	3	2	2	3	3	2.6	2	1	1	1	
4	7, F	-	..	3 y	3	2	1	2	3	3	3	2	2.4	2	0	2	0	Right parietal atrophy, left parietal infarct
4	7, F	B-STA-MCA	6 m	3.5 y	2	2	1	2	2	2	1	2	1.8	1	1	3	3	Bilateral parietal atrophy
5	24, M	-	..	3 y	3	3	3	3	3	3	3	3	3	1	1	1	1	
5	25, M	B-STA-MCA	1 y	4 y	2	2	2	2	2	2	2	2	2	1	2	1	3	
6	19, M	-	..	1 y	3	3	3	3	3	3	3	2	2.9	1	1	1	1	Left occipital spontaneous transdural anastomosis
6	19, M	L-EDAS	L 1 m	13 m	3	3	3	3	3	3	3	2	2.9	..	..	..	..	
6	20, M	L-EDAS	L 7 m	19 m	2	3	3	3	3	2	1	1	2.3	1	1	1	2	
6	20, M	B-EDAS	L1 y R 3 m	2 y	2	2	3	3	3	2	1	1	2.1	..	..	..	..	R-EDAS added
6	21, M	B-EDAS	L 20 m, R 11 m	2 y 8 m	1	1	1	2	2	1	1	1	1.3	1	3	1	3	
7	13, F	B-EDAS	8 y	10 y	1	1	1	1	2	2	2	2	1.5	1	2	2	2	Right hemisphere atrophy
8	13, F	B-EDAS	6y	7 y	1	2	1	1	1	2	1	1	1.3	3	2	2	1	
9	15, F	B-STA-MCA	2 y	8 y	2	2	2	2	1	1	1	1	1.5	2	2	1	1	Left hemisphere atrophy
10	19, M	B-EDAS	14 y	14 y	3	2	2	1	3	2	2	1	2	2	2	2	2	
11	21, F	B-EDAS	15 y	15 y	1	1	0	1	2	1	1	1	1	1	2	1	2	Right hemisphere atrophy
12	21, F	B-EDAS	5 y	5 y	3	3	3	2	3	3	3	2	2.8	3	0	3	1	Asymptomatic; poor collateral through bypass
13	23, M	B-EDAS	15 y	16 y	2	1	1	1	1	1	0	1	1	1	3	2	2	Bilateral parietal atrophy (left > right)
14	26, M	B-EDAS	13 y	16 y	2	2	2	2	2	2	1	1	1.8	1	3	2	2	
15	29, F	B-EDAS	10 y	13 y	2	2	1	1	2	2	1	1	1.5	1	3	1	2	
16	35, F	B-STA-MCA	2 y	2 y	1	1	1	2	1	1	1	1	1.1	1	2	1	3	Bilateral hemisphere atrophy

[a] Degree of LMCE is rated as 0–3: none (0), minimal (1), moderate (2) and marked (3).

[b] Degree of vascularisation to the territory of the middle cerebral artery from the internal and external carotid system is rated as 0–3: none (0), minimal (1), moderate (2) and marked (3).

frontal, the anterior frontal and temporal regions are distant from them, and from collaterals from the posterior cerebral arteries, giving slightly less reduction in LMCE in these regions. LMCE does not increase over the brain stem and cerebellum probably because the posterior cranial fossa circulation is essentially normal and leptomeningeal collaterals do not develop in these regions in moyamoya. We therefore believe that LMCE may be used to assess the effectiveness of bypass surgery, which diminishes the prolonged cerebral circulation time in moyamoya.

Differential diagnoses of LMCE in moyamoya should include diseases which show leptomeningeal contrast enhancement: meningeal carcinomatosis [13], meningitis [13, 14, 15], angiitis [16], encephalitis [15, 17], idiopathic cerebellitis [15], and venous sinus thrombosis [15, 18]. Clinical manifestations may easily differentiate moyamoya from these other conditions. LMCE in moyamoya is observed essentially over both cerebral hemispheres, while in these other diseases it tends to be localised. MRA is also useful for differential diagnosis. Postoperative enhancement of surgical margins and dura mater [19, 20] should also be considered. In bypass surgery for moyamoya, only dural enhancement occurs, which can be differentiated from LMCE because it is confined to the dura mater, predominantly on the side of the operation. Postoperative dural enhancement is prominent within a month of surgery, and may last several years although its intensity decreases [19, 20].

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