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Manuscript title:

CT perfusion imaging for childhood moyamoya disease before and after surgical revascularization

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Running title: CT perfusion imaging for moyamoya disease

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Summary

Moyamoya disease is a progressive occlusive disease of the circle of Willis with prominent collateral arterial formation. We report on a 12-year-old girl with moyamoya disease presenting with transient ischemic attacks (TIAs). Surgical indirect revascularization was performed. The patient did not suffer further TIAs at 12 month follow-up. Pre and postoperative cerebral perfusion were studied in quantitative single photon emission computerized tomography (SPECT) and CT perfusion imaging. CT perfusion imaging demonstrated postoperative increased cerebral blood flow as well as SPECT before and after revascularization. Furthermore, the area of decreased vascular reserve in SPECT with acetazolamide corresponded with areas of increased cerebral blood volume in CT perfusion imaging. CT perfusion imaging was equivalent to SPECT in accuracy, and superior in spatial resolution. CT perfusion imaging is likely to become more widely available as an easy-to-perform technique for assessing cerebral perfusion in a patient with moyamoya disease.

Key words: childhood; CT perfusion image; moyamoya disease; SPECT

Introduction

Moyamoya disease is a progressive vascular occlusion of the circle of Willis with abnormal net-like vessels comprising dilated perforating arteries (so called moyamoya vessels) [16]. As a result, transient ischemic attacks (TIAs) tend to occur in childhood. Surgical revascularization for childhood moyamoya disease can significantly decrease TIAs by improving regional cerebral blood flow (rCBF) [10, 12]. Positron emission tomography (PET), single photon emission computerized tomography (SPECT), xenon CT or, more recently, MR imaging have been used for the assessment of cerebral perfusion with moyamoya disease [5-7,13, 18], but to date there has been no report of the use of CT perfusion imaging for this type of evaluation and, further, no report of comparison of SPECT with CT perfusion imaging. We describe CT perfusion imaging in a case of childhood moyamoya disease.

Case report

A 12-year-old girl presented with TIAs on the right side resulting in loss of consciousness several times a month and a visit to our hospital. The MR image showed findings that suggested moyamoya disease. On admission, neurological examination showed no deficits. A conventional angiogram revealed the following (Fig. 1): The bilateral internal carotid artery (ICA) was occluded distal to the origin of the anterior choroidal artery; moyamoya vessels were around the base of the brain; a large proportion of the cerebral parenchyma was fed bilaterally by the posterior circulation system through leptomeningeal anastomose.

We studied CBF using N-isoprosyl-p-[^{123}I]-iodoamphetamine (IMP)-SPECT and CT perfusion imaging. The CBF values in the IMP-SPECT were quantified by

continuous arterial blood sampling method [9]. The rCBF values (ml/100g/min) in quantitative IMP-SPECT were as follows: right ACA (61.6), left ACA (64.1), right MCA (63.8), left MCA (61.6), right PCA (59.7) and left PCA (61.6) (Fig. 2A). Vascular reserve (%) was defined as a ratio of the difference between the acetazolamide-activated rCBF and resting rCBF to resting rCBF and were as follows: right ACA (13), left ACA (25), right MCA (31), left MCA (34), right PCA (45) and left PCA (49) (Fig. 2B).

A CT scan with a 16-detector row scan (LightSpeedQxi, General Electric Medical Systems) was used and an Advantage Workstation 4.1 (Perfusion 3, General Electric Medical Systems) was used for perfusion image analysis. The scanning protocol was as follows. Once we had selected the sections of interest, we obtained four contiguous 5-mm-thick slices per second in this area. Scanning was performed by continuous acquisition (cine) with 120 kVp and 100 mA for a duration of 40 sec with 1 rotation/sec, which allowed us to obtain 160 axial images 5 mm thick (40 images for 4 sections). We injected 40 ml of non-ionic contrast medium at an infusion rate of 5 ml/sec with a time delay of 5 sec from the beginning of the injection to the beginning of the image acquisition. The rCBF values (ml/100g/min) in preoperative CT perfusion images were follows: right ACA (64.6), left ACA (69.2), right MCA (65.7), left MCA (64.4), right PCA (62.9) and left PCA (63.2) (Fig. 3A). The regional cerebral blood volume (rCBV) values (ml/100g) in preoperative CT perfusion images images were follows: right ACA (4.04), left ACA (4.05), right MCA (3.74), left MCA (3.63), right PCA (3.09) and left PCA (3.17) (Fig. 3B). The mean transit time (MTT) map showed prolongation in bilateral ACA territories (Fig. 3C).

We diagnosed childhood moyamoya disease in stage 3, according to the definition of angiographical stages by Suzuki et al [16], and decided to perform surgical revascularization because of the presenting ischemic events and decreased vascular reserves. The encephalo-duro-arterio-synangiosis (EDAS) for the bilateral MCA territories and the burr hole method for bilateral ACA territories were performed without complications. After surgery, TIAs disappeared completely within one month.

Angiography 8 months after revealed marked revascularization bilaterally to the ACA and MCA territory through leptomeningeal anastomosis by burr hole method and EDAS. The rCBF values (ml/100g/min) in quantitative IMP-SPECT 10 months after were as follows: right ACA (64.2), left ACA (66.0), right MCA (64.4), left MCA (65.2), right PCA (56.8) and left PCA (63.4). The rCBF values (ml/100g/min) in CT perfusion images were follows: right ACA (71.3), left ACA (75.7), right MCA (74.8), left MCA (76.9), right PCA (65.1) and left PCA (70.5) (Fig. 4A). The rCBV values (ml/100g) in CT perfusion images images were follows: right ACA (3.31), left ACA (3.54), right MCA (3.36), left MCA (3.39), right PCA (2.66) and left PCA (2.88) (Fig. 4B). In CT perfusion images before and after surgery, rCBF increased as well as SPECT, and rCBV decreased in whole brain, especially in bilateral ACA territories with a decreased vascular reserve (Table 1). The MTT map showed improved delay in bilateral ACA territories (Fig. 4C). During the 12 month follow-up period, TIAs did not recur.

Discussion

Moyamoya disease is a rare, cerebral arterial angiopathy characterized by chronically progressive stenosis and occlusion at the supraclinoid portion of the ICAs, the proximal portions of the ACAs and MCAs [16]. There is a higher incidence of the disease among Japanese and Asian populations, and the etiology is unknown. Two separate age peaks of incidence have been recognized, the first at about 3 to 4 years and second at about 35 years. Children have a higher rate of cerebral ischemia resulting in transient ischemic events or strokes and intellectual deterioration [4]. Surgical direct and/or indirect revascularizations have been performed to encourage the development of natural collaterals, and may alter the course of progressive hemodynamic cerebral ischemia in patients with childhood moyamoya disease [5, 10, 18]. A direct bypass such as anastomosing a superficial temporal artery to a MCA may be a technically difficult procedure, especially in children, due to the small and fragile arterial walls of the MCA branch [17]. Indirect bypass such as EDAS is a technically simple procedure, and it allows for the formation of sufficient collaterals [12, 14]. Understanding cerebral hemodynamics is thought to be essential in determining the indicators for vascular reconstruction in patients with moyamoya disease [18]. Identification of changes in cerebral perfusion may be more important than angiographic findings both for predicting the risk of stroke and assessing the success of treatment [13].

The cerebral perfusion in patients with moyamoya disease has been studied with PET, SPECT, xenon CT and MR imaging [5-7, 13, 18]. In PET of childhood moyamoya disease, Ikezaki et al. [6] observed an increase of rCBV and regional oxygen extraction fraction were compensating for reduced rCBF. This phenomenon has been termed *misery perfusion* [1]. Ikezaki et al. [6] suggested that clinical

symptoms might improve by increasing the vascular reserve in the absence of improved rCBF after revascularization. In IMP-SPECT with acetazolamide, Hoshi et al. [5] reported the low rCBF and vascular reserve in childhood moyamoya disease. Using ^{99m}Tc-hexamethylpropyleneamine oxime SPECT, Touho et al. [18] reported that a reduced cerebral perfusion and improved vascular reserve after revascularization, and that those regions with hemodynamic compromise could be determined by measuring regional cerebral perfusion and vascular reserve. An increased rCBV represented dilatation of the vascular beds and was coincident with low reactivity to acetazolamide in IMP-SPECT. In the ischemic area, the vessels of the leptomeningeal anastomosis are thought to dilate maximally and the collateral vascular beds to enlarge in order to compensate for the reduced CBF, and hence there is no further dilatation in response to acetazolamide [19]. Using xenon CT to assess childhood moyamoya disease, McAuley et al. [13] reported that the findings of improved rCBF as revealed by xenon CT correlated better with an improved clinical outcome than did the angiographic findings. They proposed that xenon CT might permit assessment of stroke risk in children with moyamoya disease and predict surgical outcomes earlier than angiography. Kim et al. [7] compared perfusion MRI with SPECT in childhood moyamoya disease. They concluded that perfusion MRI might be a valuable tool for characterizing and monitoring ischemia, and that perfusion MRI had a potential role comparable to SPECT in the evaluation of moyamoya disease. Various imagings, including quantitative PET, SPECT and xenon CT or semiquantitative perfusion MR imaging, have used to study cerebral perfusion in childhood moyamoya disease. However, to the best of our knowledge,

there is no reported case using CT perfusion imaging as a tool for evaluating the rCBF in moyamoya disease.

CT perfusion imaging has been used recently for the quantitative measurement of the CBF based on improved helical scanning, multi-detector CT, and advances in the software used to analyse the data. CT perfusion data involves only the sequential acquisition of cerebral CT sections achieved on an axial mode during less invasive intravenous administration of iodinated contrast material. The data is easily analyzed by available software, which produces a color-map of CBF, CBV, and MTT. The CBF map from xenon CT has proved to be quantitative and accurate, however the imaging necessitates excellent collaboration from the patient [2, 11, 20]. Wintermark et al. [20] reported a good correlation between CT perfusion imaging of rCBF and xenon CT. Although the validity of the quantitative CBF values from CT perfusion imaging should be evaluated by more studies, the utility of CT perfusion imaging has been already reported in the evaluation of patients with acute stroke, recently with chronic cerebrovascular insufficiency [3, 8, 15].

In the CBF calculations with CT perfusion imaging, the choice of a reference artery is critical. Artery of anterior circulation such as ACA or MCA usually is selected as reference artery automatically. However, in a patient with moyamoya disease, ACAs and/or MCAs might be occluded. Therefore, we emphasize that it is important to select basilar artery or P1 segment of PCA as reference artery manually.

In the present case, after surgical revascularization, TIAs disappeared completely. With CT perfusion imaging, we also described the increase in rCBF before and after revascularization, as reported in another method previously [5-7, 13, 18]. Preoperative rCBF values in CT perfusion imaging were about equivalent to those in quantitative

IMP-SPECT. However, postoperative rCBF values in CT perfusion imaging were higher than those in quantitative IMP-SPECT. This error may be due to marked neovascularization of the cortex after the operation as perfusion CT derived CBF values in those ROIs including large blood vessels are more highly variable than those not including large blood vessels [3]. The areas of decreased vascular reserve in IMP-SPECT with acetazolamide coincided with areas of increased CBV in CT perfusion imaging. If the methodology of rCBF and rCBV derivation with CT perfusion imaging is standardized, CT perfusion imaging may offer a minimally invasive alternative technique for assessing brain perfusion which is easy to perform and accurate [15, 20].

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Figure legends

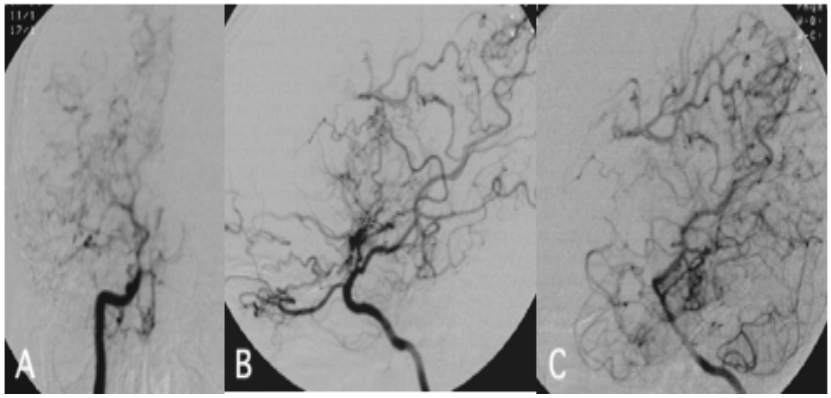

Fig. 1

(A) The right carotid angiogram of anteroposterior projections demonstrated an occlusion in distal to the origin of the anterior choroidal artery.

(B) The left carotid angiogram of lateral projections revealed an occlusion distal to the origin of the anterior choroidal artery. Moyamoya vessels were seen around the base of the brain.

(C) Vertebral angiogram of lateral projections showed that a large part of the cerebral parenchyma was fed by the posterior circulation system through leptomeningeal anastomosis.

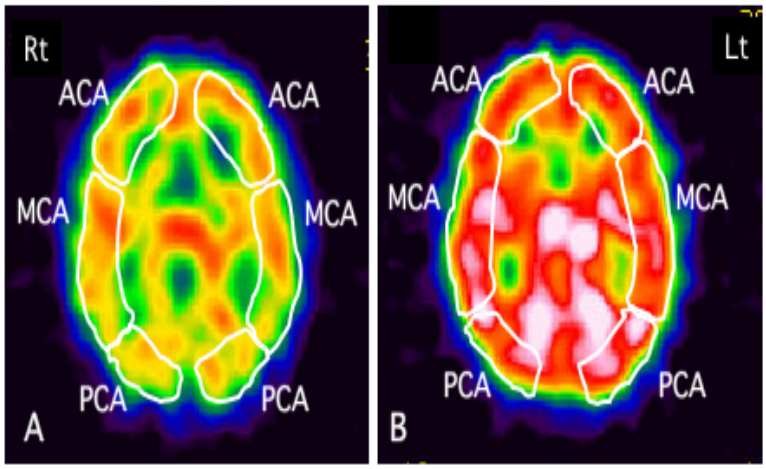

Fig. 2

(A) IMP-SPECT image before acetazolamide showed CBF values (ml/100g/min) as follows: right ACA (61.6), left ACA (64.1), right MCA (63.8), left MCA (61.6), right PCA (59.7) and left PCA (61.6).

(B) IMP-SPECT image after acetazolamide showed CBF values (ml/100g/min) as follows: right ACA (70.2), left ACA (80.6), right MCA (84.0), left MCA (83.0), right PCA (87.1) and left PCA (91.8). Vascular reserve (%) were as follows: right ACA (13), left ACA (25), right MCA (31), left MCA (34), right PCA (45) and left PCA (49). The vascular reserve decreased in anterior circulation, especially in territories of bilateral ACAs.

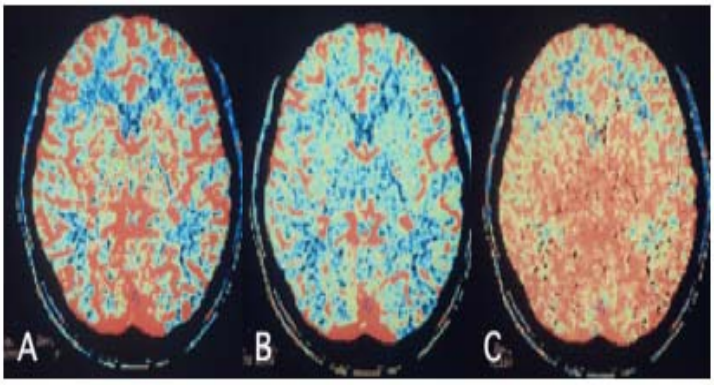

Fig. 3

(A) The CBF map in CT perfusion images showed CBF values (ml/100g/min) as follows: right ACA (64.6), left ACA (69.2), right MCA (65.7), left MCA (64.4), right PCA (62.9) and left PCA (63.2).

(B) The CBV map in CT perfusion images showed CBV values (ml/100g) as follows: right ACA (4.04), left ACA (4.05), right MCA (3.74), left MCA (3.63), right PCA (3.09) and left PCA (3.17).

(C) The MTT map showed prolongation in bilateral ACA territories.

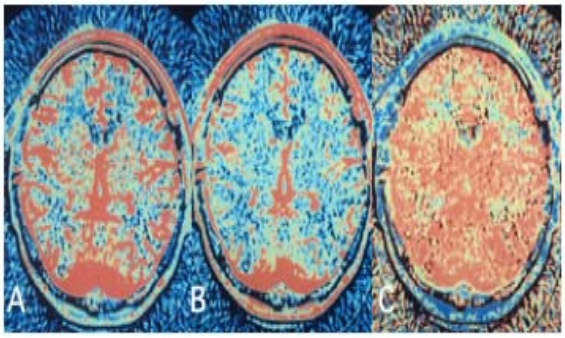

Fig. 4

(A) The CBF map in CT perfusion images showed CBF values (ml/100g/min) as follows: right ACA (71.3), left ACA (75.7), right MCA (74.8), left MCA (76.9), right PCA (65.1) and left PCA (70.5).

(B) The CBV map in CT perfusion images showed CBV values (ml/100g) as follows: right ACA (3.31), left ACA (3.54), right MCA (3.36), left MCA (3.39), right PCA (2.66) and left PCA (2.88).

(C) The MTT map showed improvement of delay in bilateral ACA territories.

Table 1. *Relation among CBF, CBV and VR in SPECT and CT perfus*

	Before surgery				A
Vascular territory	CBF in SPECT	VR in SPECT	CBF in CT-P	CBV in CT-P	CBF in SPECT
Rt ACA	61.6	13	64.6	4.04	64.2
Lt ACA	64.1	25	69.2	4.05	66.0
Rt MCA	63.8	31	65.7	3.74	64.4
Lt MCA	61.6	34	64.4	3.63	65.2
Rt PCA	59.7	45	62.9	3.09	56.8
Lt PCA	61.6	49	63.2	3.17	63.4
Mean±SD	62.0±1.6	32±13	65.0±2.2	3.62±0.41	63.3±3.32