Clinical note

The role of brain perfusion SPECT in moyamoya disease

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A B S T R A C T

Moyamoya disease (MMD) is a cerebrovascular disorder involving stenosis of brain vessels. Brain perfusion SPECT in MMD demonstrates impaired perfusion in ischemic areas. We present a 6-year-old boy with MMD. The patient had numbness on the right arm and simultaneous electroencephalography changes while studying arms up on the table. To differentiate seizure and ischemic symptoms, brain perfusion studies were obtained when the patient was asymptomatic (SPECT-A) and during the symptoms-EEG changes (SPECT-B). SPECT-A showed perfusion defect on the right frontal cortex, hyperperfusion on the right parieto-occipital region and slightly increased perfusion on the left parietal cortex. SPECT-B displayed significant hyperperfusion on the left parietal cortex; hyperperfusion on the right parietal, right temporal, right parieto-occipital and left frontal cortex. Additionally, brain perfusion SPECT of the child’s younger brother diagnosed with MMD showed decreased regional cerebral perfusion. Physiopathological mechanisms of our patient’s SPECT findings and indications of brain perfusion SPECT in MMD were also discussed.

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El papel del SPECT cerebral de perfusión en la enfermedad de moyamoya

R E S U M E N

La enfermedad de moyamoya (EdM) es una alteración cerebrovascular que implica la estenosis de los vasos cerebrales. La SPECT de perfusión cerebral en la EdM demuestra alteración de la perfusión en las zonas isquémicas. Presentamos el caso de un niño de 6 años con EdM que presentaba adormecimiento en el brazo derecho y alteraciones en el EEG cuando estudiaba con los brazos sobre la mesa. Para diferenciar entre crisis epilépticas y síntomas isquémicos se efectuaron estudios cuando el paciente estaba asintomático (SPECT-A) y durante los síntomas con cambios en el EEG (SPECT-B). La SPECT-A mostró defecto de perfusión frontal derecho, hipoperfusión parieto-occipital derecha y leve incremento de la perfusión en la corteza parietal izquierda. La SPECT-B mostró hiperperfusión significativa en la corteza parietal izquierda e hipoperfusión en el parietal, temporal y región parieto-occipital derechos y corteza frontal izquierda. Además, la SPECT de perfusión cerebral de un hermano menor del niño diagnosticado de EdM mostró disminución de la perfusión cerebral regional. Se discuten los mecanismos fisiopatológicos de estos hallazgos y las indicaciones de la SPECT de perfusión cerebral en la EdM.

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Introduction

Moyamoya disease (MMD) is a rare cerebrovascular disorder involving progressive bilateral stenosis of the distal internal carotid arteries, major vessels of the circle of Willis and proximal portions of its principal branches. The disease was named as MMD due to the irregular perforating vascular networks, called moyamoya vessels.1

MMD usually presents with ischemic symptoms during childhood. Less common symptoms in children are hemorrhagic, epileptic and others.2 On children with ischemic symptoms, brain perfusion SPECT may be used to evaluate the regional cerebral blood flow in MMD and acetazolamide challenge can show blood flow reserve of the brain.3

Both focal and generalized seizures have been associated with MMD and are likely related to hypoperfusion.4 Seizures, which present early in the course of MMD, are usually associated with ischemia. Seizures also arise as a complication of surgery. Ictal brain perfusion SPECT can be recommended in selected cases to identify seizure focus.

This report indicates the value of brain perfusion SPECT in differentiation of ischemic symptoms and seizures during the course of MMD. Additional functional information which can be obtained from brain perfusion SPECT in MMD are also presented and listed along with the description of our patients.

Clinical case

A 6-year-old boy, who presented with weakness in left arm 5 years ago, had the diagnosis of MMD after initial diagnostic workup. Acute ischemic lesion was described on right frontal lobe on MRI. A brain perfusion SPECT, obtained with 99mTc-ethylene cysteine dimer (ECD) 2 years ago, revealed no perfusion on the right frontal lobe which was thought to be due to an infarct during the chronic ischemic course of MMD (Fig. 1). Perfusion of the other
cortical, bilateral subcortical brain regions and bilateral cerebellum was normal.

In order to induce the formation of blood vessels and increase the perfusion of the brain, the patient underwent a multiple burr-hole surgery involving the frontal, parietal and occipital areas bilaterally, 8 months ago. Although the child’s symptoms were mostly improved following the surgery, he was admitted to the hospital with numbness on the right arm and sometimes weakness of the left leg accompanied. The family realized that his symptoms were always associated with a specific studying position, when the boy studied arms up on the table for more than 10 min. EEG monitoring results showed slow delta waves during this studying position which was suspicious for seizure. To differentiate between ischemic attacks and seizure in our patient, two brain perfusion SPECT studies, 1 month apart, were obtained; $^{99m}$Tc-ECD was injected when the boy was free of symptoms [SPECT-A], and during the EEG changes while the boy was studying arms up on the table [SPECT-B] (Fig. 2). The patient’s parents were informed about imaging procedures after an academic patient based meeting in which the patients’ course was discussed and detailed informed consent was received from the family, afterwards.

The SPECT images were interpreted visually and semiquantitatively. On visual interpretation SPECT-A indicated the perfusion defect (infarcted region) on the right frontal cortex, which was in concordance with the previous SPECT obtained 2 years ago. Additionally, SPECT-A showed hypoperfusion on the right parieto-occipital region and slightly increased perfusion on the left parietal cortex. In addition to SPECT-A findings, SPECT-B displayed significant hyperperfusion on the left parietal cortex; diffuse hypoperfusion on the right parietal, right temporal and left frontal cortex.

We calculated $^{99m}$Tc-ECD uptake in selected cortical regions by a semiquantitative study. This analysis has been obtained from both SPECT-A and SPECT-B. Regions of interests (ROIs) were drawn over the selected cortical areas and bilateral cerebellum on the sum of the three consecutive transverse SPECT slices according to Talairach coordinates. Brain perfusion SPECT of the child’s 4-year-old younger brother was also obtained since the child had similar symptoms. Brain perfusion SPECT showed decreased perfusion on left frontal and left temporal lobes. Afterwards, a multiple burr-hole surgery was planned for him as well.

### Discussion

In the presented case, a child with the diagnosis of MMD developed numbness of right arm after a successful burr-hole operation. Although the symptoms were first thought to be due to ischemia, the EEG changes were suspicious for seizure. Therefore, two subsequent brain perfusion SPECT studies were performed to evaluate whether the patients’ symptoms and EEG changes were

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<th>Table 1</th>
<th>Semi-quantitative analysis of brain perfusion studies.</th>
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<td>Right</td>
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<tr>
<td>SPECT-A</td>
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<td>0.62*</td>
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<tr>
<td>Right parietal/left parietal</td>
<td>0.65*</td>
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* ROI results which are concordant with visual interpretation.
due to ischemic attacks or an epileptic discharge. The SPECT studies showed that the symptoms were due to seizures rather than ischemia.

In patients with MMD, chronic stenosis of brain vessels causes a chronic hemodynamic stress and induces formation of new collateral vessels. Any increase in brain oxygen demand or a forced vasodilatation such as acetazolamide challenge paradoxically reduces blood flow of the areas under threat, because increased global brain perfusion due to the vasodilatation steals blood from the affected brain cortex. Brain perfusion SPECT is used to evaluate cerebral perfusion in patients with MMD. Acetazolamide challenge is of great value in these patients because it reveals the cortical areas that are under ischemic risk. Brain perfusion SPECT can be used to follow-up of these patients especially when they develop new symptoms.

In the presented patient, the initial brain perfusion SPECT (Fig. 1) was carried out to identify whether the patient had any chronic ischemic areas and to have baseline brain perfusion information for patient follow-up. Afterwards, the patient had bilateral multiple burr-hole surgery involving frontal, parietal and occipital areas. Following surgery he was presented with right arm numbness and stated that sometimes weakness of the left leg also accompanied. In children with MMD, ischemic attacks occur as a result of hypocapnia which is induced by vasoconstriction in related brain regions. Although the patient was represented with ischemic symptoms, his EEG monitoring was also suspicious for seizure. In our opinion, hypoperfusion of the right parieto-occipital region presented in SPECT-A was a new ischemic area and slightly increased perfusion of the left parietal cortex was because of the burr-hole effect. However, SPECT-B revealed markedly increased perfusion of left parietal lobe which was compatible with seizure focus as supported by the semiquantitative analysis (Table 1). The results of these concurrent brain perfusion scintigraphies showed epileptic seizure focus which was probably induced by reduced blood flow of carotid arteries due to a specific arm and head position which precipitated ischemia and started an epileptic discharge of neurons in our patient. Additionally, in the ictal-SPECT, ictus resulted in a kind of steal phenomenon from the right temporoparietal and left frontal cortex as supported by the semiquantitative analysis (Table 1), which was similar to acetazolamide challenge. Ictal focus, probably the left parietal lobe, stole blood from areas of low blood flow reserve in the right temporal, right parietal and left frontal cortex; and marked all these brain regions where vasoreactivity was lost under ischemic risk.

So et al. reported that brain perfusion SPECT results, performed 6–12 months after indirect by-pass operation, could predict the clinical outcome in pediatric MMD patients. It helps to monitor the progressive stages of the disease. They also stated that brain perfusion SPECT findings were correlated with the results of intelligence test in children. Although they performed a relatively delayed control angiography, they did not find any correlation between angiography results and brain perfusion SPECT. Brain SPECT can detect regional perfusion instability before treatment and to determine the extent of improvement in functional perfusion after therapy.

As discussed above, seizures can be present in the course of MMD or can be seen as a complication of surgery. In MMD patients underlying mechanism of epileptic discharges are usually hypoperfusion due to chronic ischemia. Identification of such epileptic focus would change the surgery plan and reperfusion surgery might improve patient symptoms. In the presented patient, brain perfusion SPECT studies not only indicated the infarct area and the recent cortical areas under ischemic risk, but also showed that the child’s symptoms were due to epileptic attacks. Furthermore, brain perfusion scintigraphy can be used for the assessment of brain perfusion in the symptomatic relatives of patients with MMD. The literature knowledge and indications of brain perfusion SPECT can be listed as given in Table 2.

### References