Mismatch in Brain Perfusion and Metabolism Detected with ^{99m}Tc-Hexamethyl Propylene Amine Oxime Single Photon Emission Computed Tomography and ^{18}F-Fluorodeoxyglucose Positron Emission Tomography in Moyamoya Disease

Justo Serrano Vicente, Luis Fernández Prudencio, José Rafael Infante Torre, Juan Ignacio Rayo Madrid

Department of Nuclear Medicine, Infanta Cristina Hospital, Badajoz, Spain

Abstract

We report a 47-year-old woman who developed an ischemic stroke with diplopia and dysarthria. Emergency computed tomography (CT) showed no pathological findings, and magnetic resonance (MR) showed mild ischemic-degenerative lesions. MR angiography and angiogram showed severe stenosis of both internal carotid and main intracranial arteries with plenty collateral vessels with "puff of smoke" suggesting a moyamoya disease (MMD). Brain perfusion single-photon emission CT showed global diminished perfusion in the brain lobes and a marked relative hyperperfusion in the cerebellum. However, brain ^{18}F-fluorodeoxyglucose-positron emission tomography showed physiological metabolism in the brain cortex with only slightly relative cerebellar hypermetabolism. MMD is a well-known arterial pathology that frequently develops with only mild symptoms until the middle age. Functional neuroimaging findings indicate a mismatch between brain glucose metabolism and brain perfusion, probably due to neuronal subclinical chronic ischemia in the brain cortex with preserved viability of neurons.

Keywords: *Brain ^{18}F-fluorodeoxyglucose-positron emission tomography, brain perfusion single-photon emission computed tomography, moyamoya disease, neuronal metabolism*

Introduction

Moyamoya disease (MMD) was first described in 1957 as a "bilateral hypoplasia of internal carotid arteries."[1] The name of the disease comes from Japanese and means "puff of smoke." Modern methods of brain vessel imaging, such as computed tomography (CT) angiography, magnetic resonance imaging (MRI) angiography, or rotational digital subtraction angiography, lead to a more frequent diagnosis of asymptomatic cases of this disease.[2] Functional neuroimaging such as positron emission tomography (PET) and single-photon emission CT (SPECT) can provide additional information that may be helpful in the management of this disease.[3]

Case Report

We present a 47-year-old female who was requested to the hospital due to decreased visual acuity and diplopia. After reading for a short time, she developed a headache, more acute on the left side. In the emergency room, the patient developed difficulty in speaking, a hypertensive crisis, and anxiety. Biochemical analysis showed no pathologic findings. A brain CT was performed and reported as normal.

One day later, a brain magnetic resonance (MR) was carried out showing a focal acute stroke located in the midbrain noted on diffusion-weighted imaging and in T2-fluid attenuated inversion recovery sequence [Figure 1].

Two months after hospitalization, a brain and aortic MR angiography was carried out showing bilateral occlusion of cervical segment internal carotid arteries. Based on these findings, MMD was suspected. Then, angiogram confirmed the extensive perforator collaterals from external carotid arteries giving the "puff of smoke" characteristic of this disease, involving both anterior and posterior circulation [Figure 2].

Four months later, a brain perfusion SPECT with ^{99m}Tc-hexamethyl propylene amine oxime was requested, showing a relative diminished perfusion of all brain lobes, more pronounced in occipital,

How to cite this article: Vicente JS, Prudencio LF, Torre JR, Madrid JI. Mismatch in brain perfusion and metabolism detected with ^{99m}Tc-hexamethyl propylene amine oxime single photon emission computed tomography and ^{18}F-fluorodeoxyglucose positron emission tomography in moyamoya disease. Indian J Nucl Med 2018;33:154-7.

Address for correspondence:
Dr. Justo Serrano Vicente, Hospital Infanta Cristina de Badajoz, Avda. Elvas SN. 06010, Badajoz, Spain.
E-mail: titoserrano@gmail.com

Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.IJNM_2_18

Quick Response Code:

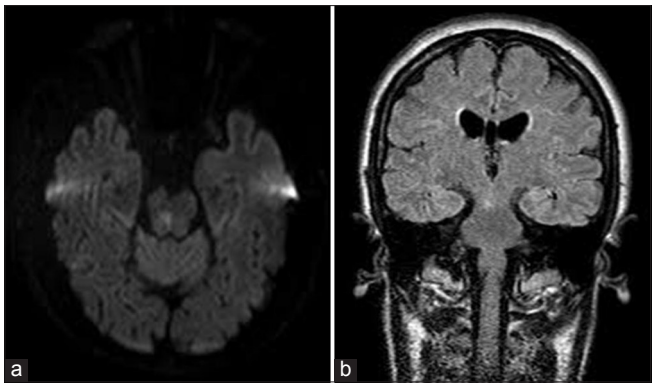

Figure 1: Magnetic resonance images performed 1 day after the stroke event. We can see a right medial mesencephalic lesion that exhibits restricted diffusion in diffusion-weighted imaging (a), hypointense signal in T1 sequence and hyperintense signal in coronal T2-fluid attenuated inversion recovery image (b). The appearance of the lesion described in the images is highly suggestive of an acute mesencephalic infarct

posterior temporal, and adjacent parietal cortex. The most relevant finding was a marked increase in perfusion in the cerebellum compared to the rest of the brain structures. Basal ganglia also showed preserved perfusion [Figures 3a and 4a].

Two weeks later, a revascularization was planned and the patient was referred to our department to perform a brain ^{18}F-fluorodeoxyglucose-PET (FDG-PET) to assess the viability of encephalic territories. Unexpectedly, despite the SPECT findings, the images showed a preserved metabolism of glucose in the brain lobes. The only finding was a slightly relative elevated metabolism in the cerebellum. Unfortunately, the revascularization procedure was ruled out due to technical impediments [Figures 3b and 4b].

Currently, the patient remains asymptomatic, with only mild dysarthria, attending periodic follow-up with the neurologist.

Discussion

Moyamoya is a well-known rare occlusive disorder of cerebral vasculature with poorly understood etiology. Genetic background is frequently mentioned because, in 15% of cases, the disease is found in other family members. These are most probably the cases of multifactorial autosomal inheritance.[1] The disease is characterized by progressive intracranial vascular stenosis of the circle of Willis, resulting in successive ischemic events. Hemorrhagic events can also occur. Diagnosis is established by the typical appearance on cerebral angiography, i.e., "puff of smoke" and refers to the appearance of multiple compensatory dilated striate vessels seen on angiography. CT and MRI play a major role in documenting the regions of infarction/hemorrhage. In these patients, chronic stenosis of the brain vessels causes a chronic hemodynamic stress and induces formation if new collateral vessels.[2] Functional neuroimaging studies with

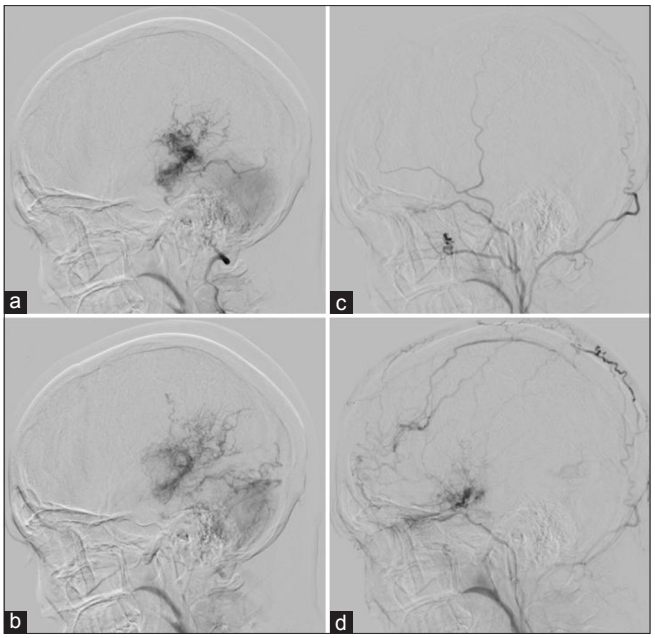

Figure 2: Angiogram performed from the left vertebral artery (a and b) and right common carotid artery (c and d) showing complete occlusion of the internal carotid artery. Enlarged perforator branches from the right external carotid artery giving rise to small corkscrew parenchymal vessels. This is the appearance of "puff of smoke" classical of moyamoya disease. The posterior circulation was also affected, involving both posterior cerebral arteries

PET are used in MMD to evaluate the cerebral perfusion employing radiotracers like $H_2(^{15}O)$ and the viability of the cerebral territories using FDG. Brain perfusion SPECT is also employed for the assessment of blood flow reserve of the brain (using acetazolamide challenge),[3] prediction of MMD progression,[4] patient follow-up,[5] prognostic before and after revascularization interventions, and detection of regional perfusion impairment of family members in familial cases.[6]

In this case, the unusual finding is the mismatch between perfusion and metabolism of the brain. Brain perfusion SPECT showed marked alterations in the brain cortical perfusion with relative elevated perfusion in the cerebellum while brain FDG-PET showed near normal distribution in cortical glucose metabolism and slightly relative elevated cerebellum metabolism [Figures 3 and 4]. Certain mismatch phenomena such as the "luxury perfusion" have been described in cerebrovascular diseases such as acute stroke,[7] but we have not found any similar case in literature. We can only describe relative decreases in perfusion since we do not have specific quantification software such as SPM. Our descriptions are qualitative compared to the normal distribution pattern of both tracers in the brain, but the SPECT findings suggested that the cerebellum showed a relative increased perfusion compared to the rest of the structures, whereas PET showed almost a normal distribution. Regarding the relative augmented perfusion in the cerebellum, it has been described that in advanced dementias, the cerebellar metabolism tends to

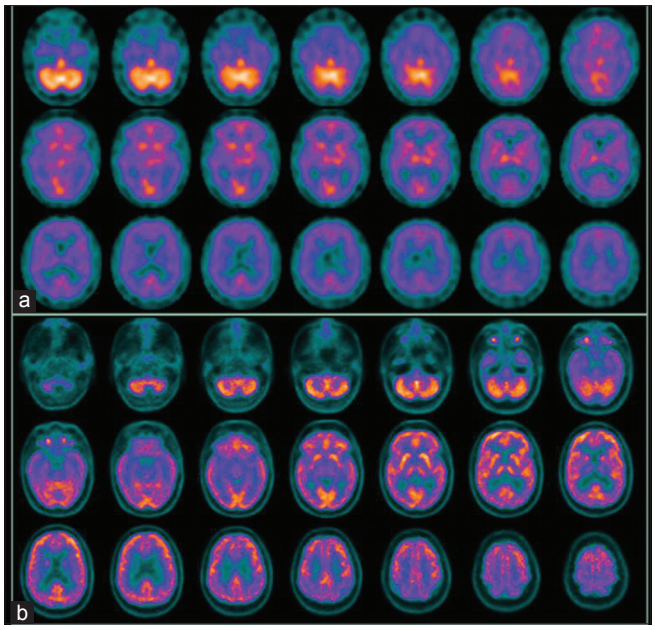

Figure 3: Brain perfusion single-photon emission computed tomography (a) and ^{18}F-fluorodeoxyglucose-positron emission tomography (b) performed 4 months after the stroke event and another 2 weeks, respectively. Transaxial images in three upper files (a) showing relative moderate hypoperfusion of the brain lobes, more marked in temporoparietal cortex. The cerebellum shows a relative elevated perfusion. In the three lower files (b), we see transaxial slices of ^{18}F-fluorodeoxyglucose-positron emission tomography showing conserved metabolism in the brain lobes with only a slightly increase in the cerebellum

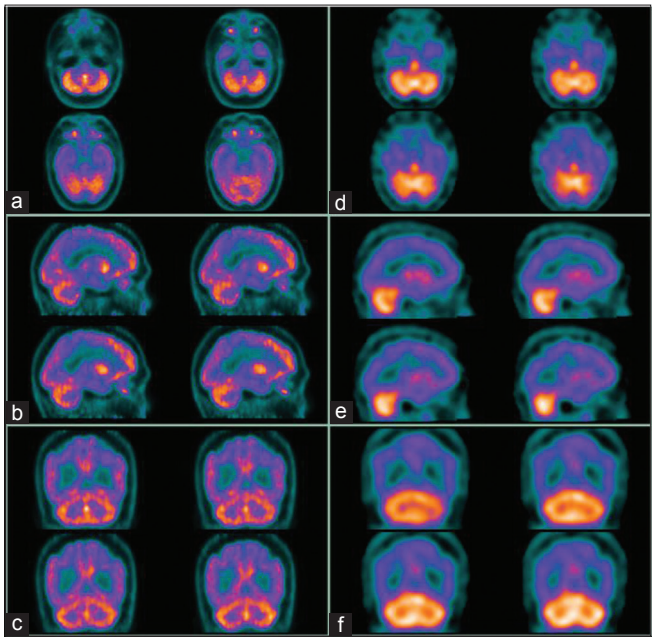

Figure 4: Two sets of three tomographic slices of positron emission tomography and single-photon emission computed tomography. On the left half, we see transaxial, sagittal, and coronal slices of brain ^{18}F-fluorodeoxyglucose-positron emission tomography (a-c, respectively). On the right half, correlative transaxial, sagittal, and coronal slices of cerebral perfusion single-photon emission computed tomography (d-f, respectively) are displayed. We note the difference between the cerebellum and cerebral cortex in the two different techniques with a marked relative hyperperfusion and only slightly relative hypermetabolism in the cerebellum when compared with other brain areas

increase progressively, likely as part of a compensatory mechanism that involves recruitment of alternate neuronal pathways.[8] In other neurodegenerative diseases such as Parkinson's disease, a progressive increase in cerebellar perfusion has been depicted, as a compensatory change that occurs as a consequence dysfunction in other neural structures.[9] This neural plasticity, developing and preserving some structures and circuits in a trend to compensate the dysfunction of other neural regions, has been described in the physiopathology of dementias.[10] MMD is a rare disease in which functional neuroimaging plays a minor role in its management, but in the case reported, SPECT and PET showed additional information that might help us understand the physiopathology of this disorder.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Kuroda S, Houkin K. Moyamoya disease: Current concepts and future perspectives. Lancet Neurol 2008;7:1056-66.
2. Yamada I, Suzuki S, Matsushima Y. Moyamoya disease: Comparison of assessment with MR angiography and MR imaging versus conventional angiography. Radiology 1995;196:211-8.
3. Kaku Y, Iihara K, Nakajima N, Kataoka H, Fukuda K, Masuoka J, *et al.* Cerebral blood flow and metabolism of hyperperfusion after cerebral revascularization in patients with moyamoya disease. J Cereb Blood Flow Metab 2012;32:2066-75.
4. So Y, Lee HY, Kim SK, Lee JS, Wang KC, Cho BK, *et al.* Prediction of the clinical outcome of pediatric moyamoya disease with postoperative basal/acetazolamide stress brain perfusion SPECT after revascularization surgery. Stroke 2005;36:1485-9.
5. Ibrahimi DM, Tamargo RJ, Ahn ES. Moyamoya disease in children. Childs Nerv Syst 2010;26:1297-308.
6. Volkan-Salanci B, Lay Ergün E, Genc Sel Ç, Yalnizoğlu D, Turanli G. The role of brain perfusion SPECT in moyamoya disease. Rev Esp Med Nucl Imagen Mol 2012;31:216-8.
7. Shishido F, Uemura K, Inugami A, Ogawa T, Fujita H, Shimosegawa E, *et al.* Discrepant 99mTc-ECD images of CBF in patients with subacute cerebral infarction: A comparison of CBF, CMRO2 and 99mTc-HMPAO imaging. Ann Nucl Med 1995;9:161-6.
8. Van Heertum RL, Greenstein EA, Tikofsky RS.

2-deoxy-fluorglucose-positron emission tomography imaging of the brain: Current clinical applications with emphasis on the dementias. Semin Nucl Med 2004;34:300-12.

9. Kapitán M, Ferrando R, Diéguez E, de Medina O, Aljanati R, Ventura R, *et al.* Regional cerebral blood flow changes in parkinson's disease: Correlation with disease duration. Rev Esp Med Nucl 2009;28:114-20.
10. Morbelli S, Perneczky R, Drzezga A, Frisoni GB, Caroli A, van Berckel BN, *et al.* Metabolic networks underlying cognitive reserve in prodromal alzheimer disease: A European alzheimer disease consortium project. J Nucl Med 2013;54:894-902.