Brain single photon emission tomography and hypercapnia test in testing cerebrovascular reserve capacity, in Moya moya disease

Abstract

Moya moya is a progressive cerebral occlusive vasculopathy, rare in European countries. We describe a case of a young woman with right-hand side hemiparesis, mixed expressive aphasia, organic psychosyndrome and cognitive malfunction. Detailed imaging methods displayed bilateral stenosis of the internal carotid artery, bilateral ischemic cerebral changes and bilateral perfusion deficit, which guided us to the final diagnosis. Before the bypass surgery, cerebrovascular reserve capacity (vasoreactivity), by the brain single photon emission tomography and hypercapnia, were assessed and the lower cerebrovascular reserve was demonstrated. Bilateral bypass surgery with extracranial-intracranial anastomosis, improved the neurological deficit. Diagnosis was confirmed by histological examination of the vessel wall specimen.

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Introduction

It is necessarry to think of Moya moya disease in younger patients with cerebrovascular events. Our patient was originally treated with psychiatric drugs as her physician did not think of an ischemic cerebral etiology. Correct diagnosis and effective treatment moderated symptoms and improved the quality of life. We present among other findings, the imaging methods which supported differential diagnosis.

Case presentation

We have examined a 30 years old, single, childless woman, who complained for more than a year for upper limbs acral paresthesia, right-hand weakness and double vision. Moreover, the patient complained for difficulty in words expression. From her medical history, she mentioned only insignificant head injuries without commotion. For a decade she was treated in a ambulatory psychiatric unit for "prefrontal psychopathy". Her mother, living in a common household with her, noted psychical difficulties that resulted in an anamnestic suicidal attempt. The neurologist, diagnosed: ischemic stroke with right-hand hemiparesis, mixed and mostly expressive aphasia, psychosyndrome with organic inhibition and cognitive function disorder.

Ultrasound detected bilateral intracranial, internal carotid and anterior cerebral arteries stenoses, normal findings of both vertebral arteries and increased flow velocity in the area of the posterior cerebral arteries. Brain computed tomography (CT) scan and magnetic resonance imaging (MRI) examination showed ischemic changes of the left frontal and right parietal lobes. Perfusion CT scan showed reduced blood flow, blood volume and prolonged time-to-peak values on the left hemisphere. Brain angiography verified both internal carotid arteries stenoses and stenoses of the anterior cerebral arteries. The posterior cerebral circulation was normal. Before stenotic parts, the vessels were elongated and coiled. After having been filled with contrast media, they created a picture of the "smoke puffs carried away by a breeze", "Moya moya like" as said in the Japanese (Fig. 1. a,b).

Suspicion of Moya moya disease which is rare in Europe with progressive cerebral vasculopathy was expressed [1, 2]. Psychiatric disease was not considered as a possibility. Serum laboratory markers did not provide evidence of vasculitis. Bypass cerebral neurosurgery was considered and examination of cerebrovascular reserve capacity (vasoreactivity) using cerebral single photon emission tomography (SPET) and the hypercapnic test, were performed. Under native conditions, these tests showed obvious defects in the frontal left and parietal right (Fig. 2a) hemispheres, relevant to the ischemic changes also shown by CT and MRI. After ap-

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plying CO_2 load, cerebral perfusion deteriorated in the frontal left and a very heavy hypoperfusion occurred bilaterally in the parietal-temporal region (Fig. 2b). Thus cerebrovascular reserve capacity was significantly reduced.

The patient underwent a bypass neurosurgery procedure. Anastomosis was performed on the left side between the superficial temporal artery and the middle cerebral artery branches supplemented with encephalo-duro-myo-synangiosis. Four months later a similar bypass procedure took place on the right side, i.e. a extra-intracranial (EC-IC) anastomosis between the superficial temporal artery and the middle cerebral artery.

Histological examination of the resected stenotic vessel segment, showed lipid deposits without any signs of simultaneous phlegmasia (Fig. 3). Based on the above tests and histology, the diagnosis of Moya moya disease was confirmed.

Three and four months after surgery, the function of the bypasses was bilaterally checked by ultrasound and also by angiography, during which, neoangiogenesis was shown on the left side of the brain confirming the diagnosis (Fig. 4).

Postoperation we noticed regression of the right hand hemiparesis, the expressive aphasia and the psycho-organicity regression. A cerebral SPET examination showed improvement of the perfusion on the right parietal lobe, while the perfusion on the left frontal lobe remained unchanged (Fig. 5).

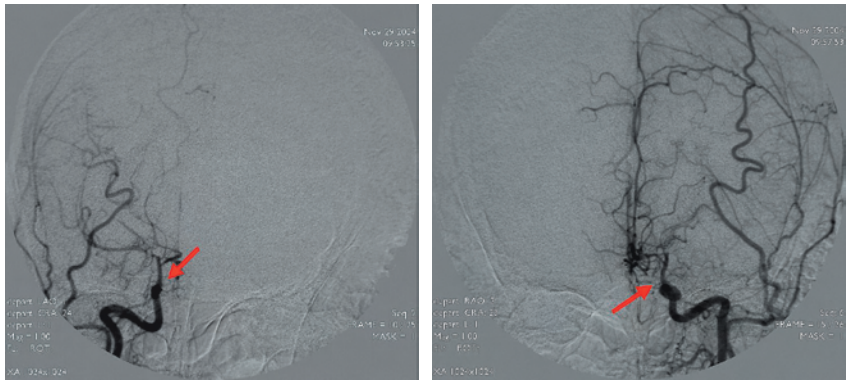

Figure 1. Carotid angiography of the right (1a) and left (1b) common carotid artery shows severe stenosis of both internal carotid arteries and bifurcation of the anterior cerebral and the middle cerebral arteries (arrows). Elongated and coiled vessels like ("smoke puffs carried away by breeze") are seen bilaterally.

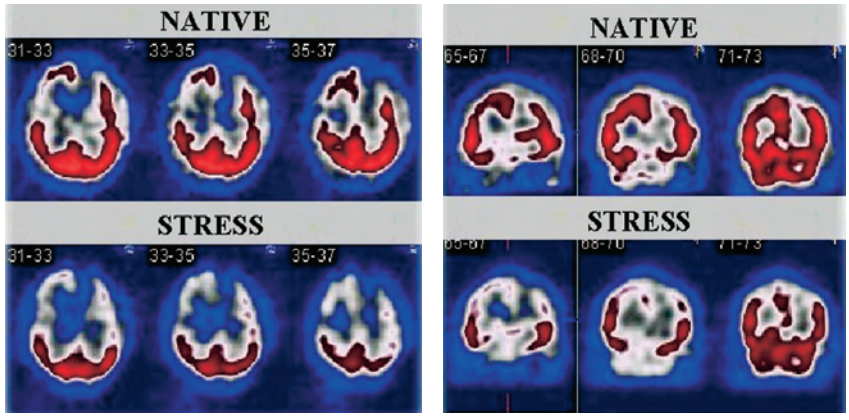

Figure 2. Cerebral SPET in selected sections at the transversal (2a) and at the coronary planes (2b). The first line shows sections made under native conditions with defect in accumulation of radiopharmaceuticals on the frontal left and parietal right arteries. On the second line, there are sections made after CO_2 load. They show perfusion deterioration of the left frontal artery and bilaterally of the parietal-temporal area. Cerebrovascular reserve capacity was significantly reduced.

Discussion

Moya moya obtained its name from the characteristic findings on cerebral angiography. Contrast media shows elongated and coiled central collateral vessels due to stenoses or obstructions of ICA bifurcation: a characteristic image of "smoke carried by breeze" or "Moya moya" in the Japanese language [3]. The disease is manifested by cerebral vascular bouts, mostly ischemic ones [1, 11]. Imaging methods like primarily brain angiography, sonography and histology, are essential for the diagnosis [4, 7].

Conservative treatment using acetylsalicylic acid, steroids, vasodilatation drugs, mannitol or antibiotics seems to be ineffective [5, 6, 8-10]. In several studies, symptomatic improvement was documented in patients with ischemic Moya moya disease following the i.v. influsion of calcium channel blockers (varapamil, nimodipine), but this effect is rather partial and not permanent [11, 12]. Surgical revascularisation treatment

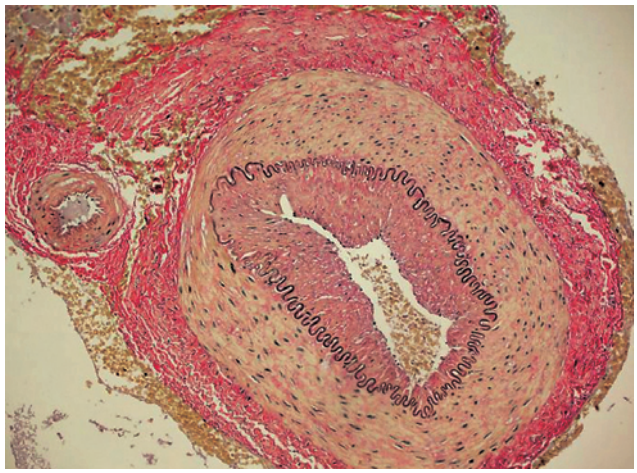

Figure 3. Cerebral vessel in a cross section, with lipid deposits placed in the vascular wall, without phlegmasia. (Verhoeff, 200x).

has the most signifiant effect [6, 13 -16]. Before contemplated bypass procedures, it is appropriate to examine the cerebrovascular reserve capacity using cerebral SPET with the hypercapnic or the acetazolamide test and with ultrasound or

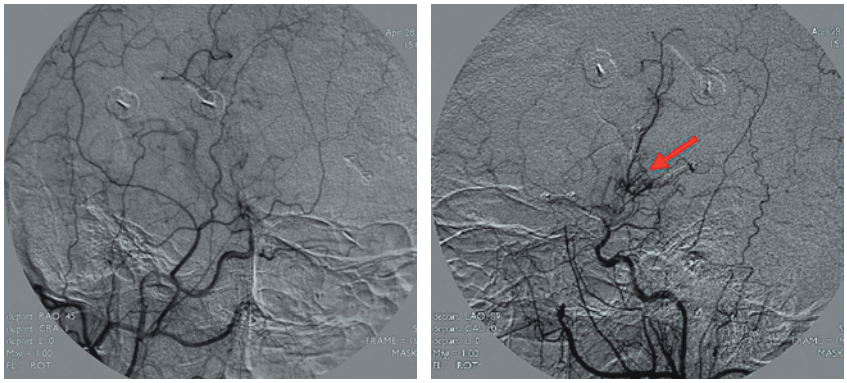

Figure 4. Post-operative carotid angiography of the left side (4a: 3 months after surgery) and of the left common carotid artery (4b: 4 months after surgery) shows functionality of both EC-IC anastomosis and moreover neoangiogenesis on the left side (arrow).

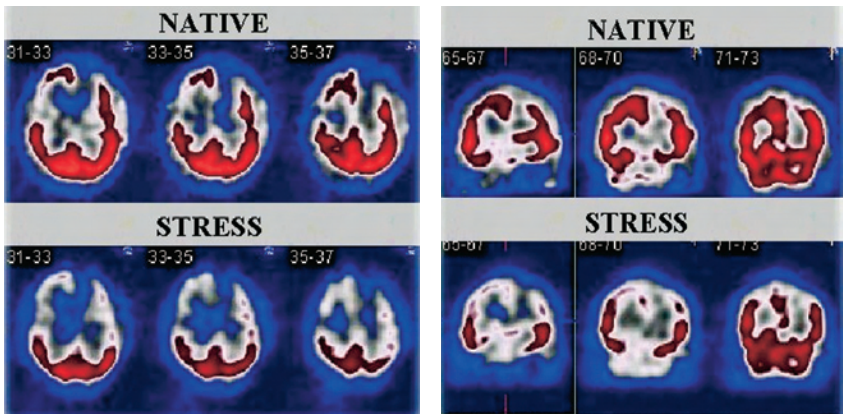

Figure 5. Cerebral SPET examination under native conditions-selected sections in the transverse (5a) and the coronary planes (5b). The first line shows sections made under native conditions before surgery, the second line shows the post-operative sections. Radionuclide accumulation on the right parietal lobe was improved while on the left frontal lobe remained unchanged.

perfusion CT. If reduction of cerebrovascular reserve capacity is found, the improvement of neurological findings, following a successful operation, is highly probable. Surgery under these conditions also reduces the risk of new significant cerebral vascular events or their recurrences [5, 8]

In concordance with the literature, we chose in our patient to test cerebrovascular reserve capacity using SPET brain scintigraphy [7, 17]. Surgical treatment is widely recommended. The right choice of treatment was confirmed by improvement of the patient's symptoms.

In conclusion, we have described a young woman with decreased vasoreactivity. She underwent bilateral bypass surgery (EC-IC) anastomosis which improved her neurological deficit. Angiographic and SPET findings as well as examination of the histological specimen from a blood vessel, confirmed the diagnosis of Moya moya disease.

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