ORAL PRESENTATION

Reversible cerebral vasoconstriction syndrome and primary angiitis of the central nervous system – the differential diagnosis challenge

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Abstract

Introduction: Primary angiitis of the central nervous system (PACNS) and reversible cerebral vasoconstriction syndrome (RCVS) are two possible etiologies in cases of cerebral arteriopathies. In spite of their contrasting prognosis and treatment, these two entities are not always easy to distinguish. We present a challenging clinical case, which illustrates the difficulty encountered in differentiating them.

Case Report: A 65-year-old woman was admitted with sudden onset of numbness of the left face and upper limb and dysarthria. Cerebral computed tomography-angiography showed discrete subarachnoid haemorrhage in central and postcentral sulcus, no aneurysms or vascular malformations were depicted. A cerebral magnetic resonance-angiography (MRA) (with venogram) performed later showed two small ischemic foci in the right fronto-insular territory, a subtle focal narrowing of the right adjacent M2 branch and hematic deposition in the right perisylvian sulci. Transcranial-doppler ultrasonography displayed increased left middle cerebral artery cerebral blood flow velocity, compatible with minor luminal stenosis (30%). She was discharged without focal neurologic symptoms, with the diagnosis of possible RCVS.

Three months later, the patient was readmitted due to aphasia and right central facial palsy. Brain MRA showed new left anterior insular and left frontal subcortical ischemic lesions and rarefaction of the left middle cerebral artery vascular tree, raising the hypothesis of PACNS. Transthoracic echocardiogram, 24h-Holter monitoring and cerebrospinal fluid study were unremarkable. She is still waiting the result of cerebral angiography.

Conclusion: This patient has several features that make the differential diagnosis between PACNS and RCVS difficult. In order to avoid other unnecessary diagnostic tests and to institute a correct therapy, we hope that the result of other ancillary exams, such as cerebral angiography, may help to distinguish them.

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