REVERSIBLE CERVICAL VASOCONSTRICTION SYNDROME PRESENTING AS RECURRENT THUNDERCLAP HEADACHE

M.R. Mazza[1], L. Rapisarda[1], A. Quattrone[1,2], F. Bono[1,2].

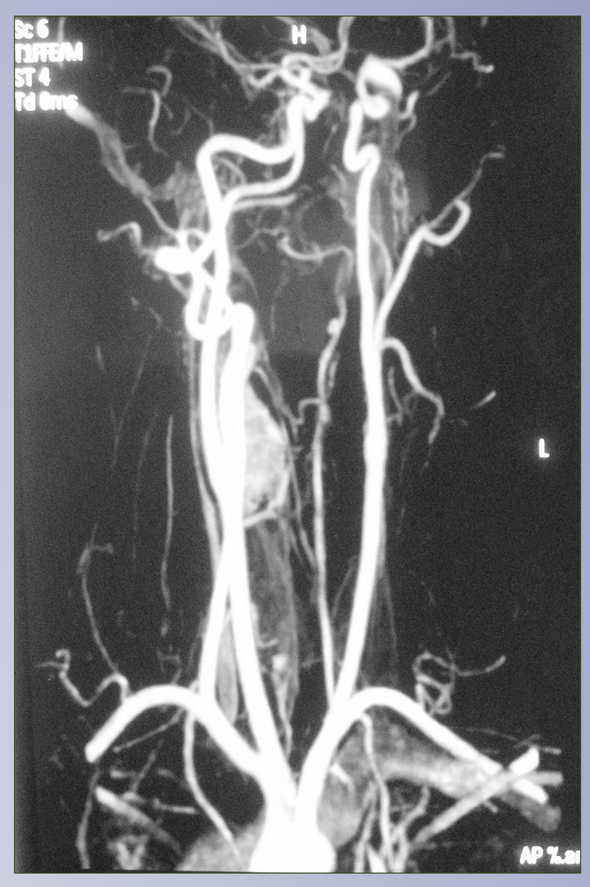
[1]Headache Center, Institute of Neurology, Department of Medical and Surgical Science, "Magna Graecia" University, Catanzaro, Italy .
[2] Neuroimaging Research Unit, Institute of Molecular Bioimaging and Physiology- National Research Council- Catanzaro, Italy

Introduction

Thunderclap headache (TCH) associated with reversible cerebral vasoconstriction syndrome (RCVS) is well recognized, while TCH associated with cervical reversible vasocostriction syndrome (RCeVS) has been rarely reported. We here describe two patients with recurrent TCH due to RCeVS.

Case description

Case 1. A 25-year-old woman had a history of episodes of sudden and severe head pain, unilateral, peaking in less than 1 minute, lasted few hours. During headache she experienced elevated blood pressure. An important physical and emotional stress caused an acute myocardical infarction without thrombosis. Two years later the patient complained of recurrent episode of TCH associated with decrease in visual acuity in right eye and numbness in the right arm, these symptoms disappeared spontaneously after few hours. Brain MRI was normal; cervical MR angiography (MRA) with gadolinium revealed a right laterocervical mass with intense vascolarization, compatible with paraganglioma, and the stenosis of ipsilateral carotid artery. These data allowed us to make a diagnosis of recurrent TCH associated with RCeVS due to paraganglioma. The administration of nimodipine and then the surgical removal of paraganglioma led to resolution of headache. Repeated cervical MRA showed the resolution of radiological RCeVS.



MRA with gadolinium shows a right laterocervical mass with intense vascolarization compatible with paraganglioma and the stenosis of ipsilateral carotid artery

Case 2. A 36-year-old woman developed an acute, severe thunderclap headache, located primarily in the right frontal-parietal area, associated with numbness and weakness in the right arm and than in the right leg. Headache and the other neurological symptoms resolved spontaneously in a few days. After a month she developed two further episodes of severe thunderclap headache, associated with unilateral or bilateral blurring vision lasted about one hour. Neurological examination revealed only the hypoesthesia in the right side of body. Brain MRI was normal; cervical MRA revealed a stenosis of both right vertebral artery and left carotid artery. The administration of nimodipine improved her neurological symptoms. During the follow up cervical MRA was normal.

Discussion and Conclusion

The first case describes TCH due to RCeVS associated with paraganglioma; the second one shows TCH associated with RCeVS. These cases provide the evidence that recurrent TCH may also occur in patients with RCeVS. We realize that RCeVS is not uncommon and should be considered in patients presenting recurrent TCH and focal neurological signs in the absence of other etiologies.

References:
1-Verillaud B. et al. Reversible cerebral vasoconstriction syndrome in two patients with a carotid glomus tumour. Cephalalgia 30(10) 1271-1275
2-Ducros et al. The clinical and radiological spectrum of reversible cerebral vasoconstriction syndrome. A prospective series of 67 patients. Brain (2007), 130, 3091-3101.

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