PROBABLE REVERSIBLE CEREBRAL VASOCONSTRICTION SYNDROME. A CASE REPORT

B CENSORI, T PATZIGUIAN, M GARDINETTI, E VENTURELLI, MR ROTTOLI
USC Neurologia, ***ASST Papa Giovanni XXIII - BERGAMO***

Introduction. The reversible cerebral vasoconstriction syndrome (RCVS) is included in section 6 of the 3rd International Classification of Headache Disorders "***Headache attributed to cranial or cervical vascular disorder***" (code 6.7.3) (1). The classification also allows a "probable" diagnosis when "Reversible cerebral vasoconstriction syndrome (RCVS) is suspected, but cerebral angiography is normal" (code 6.7.3.1).

Case Report. A 64-year-old woman had been using a corticosteroid-**sympathomimetic** (fluocinolone-**clonazoline**) **nasal spray** daily for one week, for a common cold. On the 7th day she awoke with a mild headache that suddenly increased to very high intensity in less than one minute, lasted about 45 minutes, and left behind a mild, diffuse headache. The same **"thunderclap" headache** occurred the following day, while she was coughing. Brain MRI the same day showed two small, recent, sulcal subarachnoid hemorrhages in the left and right convexities, without old cerebral microbleeds or superficial siderosis, confirmed by CT scan the following day **(Fig.1)**. Emergent CT-Angiography did not show intracranial vascular malformations or stenoses, but revealed narrowing and beading of both internal carotid arteries, suggestive of fibromuscular dysplasia (FMD) **(Fig. 2).** Neurological examination was normal. Oral nimodipine 15 mg 6 times per day was started. Follow-up MR-angiography (MRA) 7 days after the first headache did not show vasospasm. Transcranial Doppler up to 10 days after the first headache was normal. There were no thunderclap recurrences after admission, and the background headache ceased after 4 days. Brain MRI and MRA 2 months and 10 months after headache onset showed the hemosiderin deposits at the sites of bleeding.

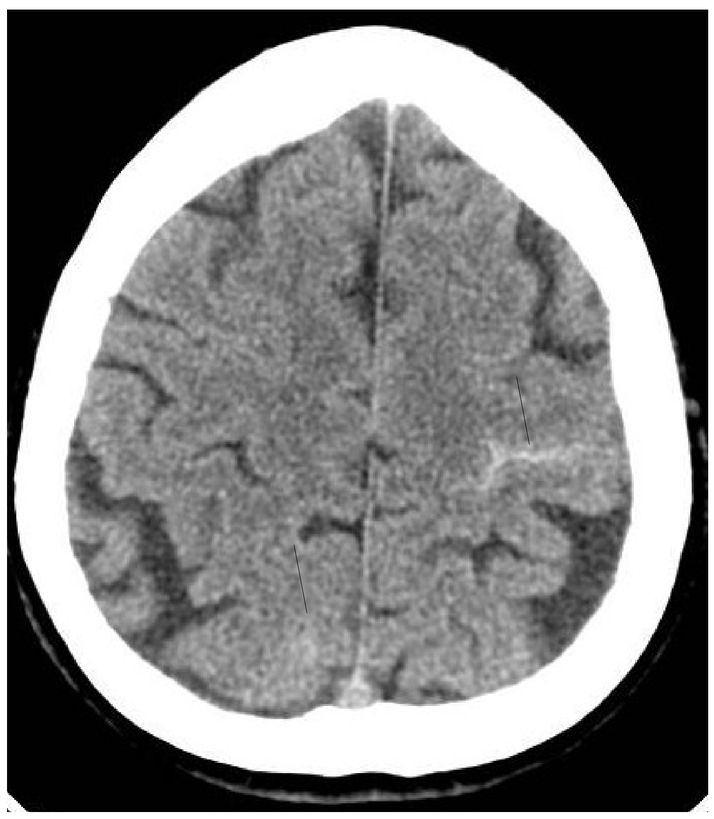

Fig. 1. Sulcal subarachnoid hemorrhage

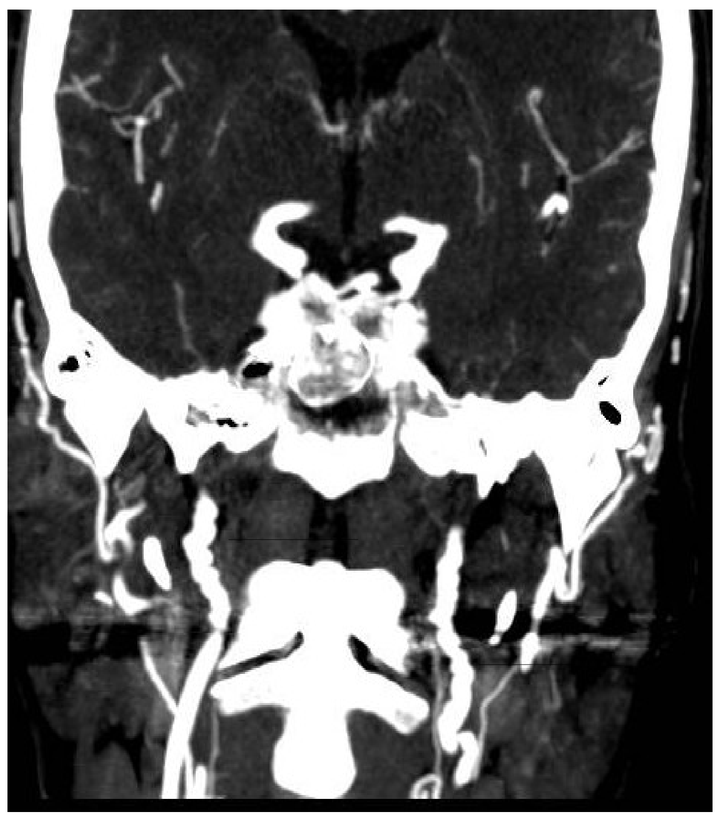

Fig. 2. Bilateral internal carotid beading

Discussion. This case fulfils the criteria for "***6.7.3.1 Headache probably attributed to reversible cerebral vasoconstriction syndrome***" (1). In these cases the explanation is that of a very distal vasoconstriction, not visible in conventional angiograms (2). Such a purely clinical diagnosis may be debated, because different conditions may produce this headache pattern. From this perspective, our case is interesting, because the clinical picture of probable RCVS was associated with one feature that is common in definite RCVS, such as atraumatic convexal subarachnoid hemorrhage, and was strongly time-linked with the use of a vasoconstricting agent (clonazoline), which is a "classical" cause of RCVS. Thus, the clinical construct of probable RCVS seems to be verified by the otherwise very typical neuroradiologic and etiologic features of our patients, although we did not carry out any intra-arterial angiography. The role of FMD is uncertain, but an association between RCVS and FMD has been described (3).

References

1. ICHD-3 beta. Cephalalgia 2013;33:629-808
2. Wolff V, Ducros A. Headache 2016;56:674-687

3.

Topcuoglu MA et al. Cephalagia 2016; Mar 7. pii: 0333102416637826. [Epub ahead of print]