Reversible cerebral vasoconstriction syndrome: a case series of 7 patients

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BACKGROUND AND AIMS

Reversible cerebral vasoconstriction syndrome (RCVS) is characterized by recurrent ‘thunderclap’ headaches with reversible multifocal narrowing of the cerebral arteries, with or without focal neurological deficits. “Reversible” refers to the dynamic arterial changes seen on angiography, but clinical deficits resulting from infarction or haemorrhage may be persistent. We analysed clinical, neuroimaging and outcome data of patients with angiographically confirmed RCVS.

METHOD

Retrospective study of patients admitted with the diagnosis of RCVS between August/2015 and September/2017 in our institution.

RESULTS

7 RCVS patients

- 4 male
- mean age: 41 years (23-67)
- negative history of hypertension or migraine

Admission blood pressure > 140/90 mmHg: 3 patients

- mean systolic blood pressure: 187,7 mmHg
- mean diastolic blood pressure: 96,3 mmHg

ASSOCIATED CONDITIONS	n
No identifiable precipitating factor / spontaneous	3
Exposure to vasoactive substances	2
Cannabinoids	1
Selective serotonin reuptake inhibitors	1
Puerperium	1
Autonomic dysreflexia (Guillain-Barré syndrome)	1

CLINICAL MANIFESTATIONS	n
Recurrent ‘thunderclap’ headache	6
No identifiable trigger	3
Triggered by sexual activity	3
Triggered by Valsalva maneuvers	1
Triggered by exercise	1
Seizures	1
Focal neurological deficits	3
Persistent (>24h)	3
Transient (<24h)	0

4 patients with isolated headache

DIAGNOSTIC INVESTIGATION	n
Angiographic study	7
CT angiography + MRI angiography	4
CT angiography + conventional angiography	1
CT angiography only	1
MRI angiography only	1
Brain imaging	7
CT scan + MRI	5
CT scan only	1
MRI only	1
Lumbar puncture	4

➤ Normal first angiographic study in 2 patients

- Mean time [symptoms onset - diagnosis]: 8 days (2-16)
- Mean time [diagnosis - arterial normalization]: 107 days (12-254))

➤ Three patients with abnormal brain imaging:

- 2 patients with subarachnoid hemorrhage + intra-parenchymal hemorrhage
- 1 patient with cerebral infarction

TREATMENT	n
Nimodipine	5
Acetylsalicylic acid	3
Corticosteroid / Immunosuppressant	2
Antiepileptic	1
Anti-edematous	1

- No symptomatic recurrence in follow-up
- No occurrence of death

CONCLUSIONS

- ✓ RCVS diagnosis is not always obvious, particularly when headaches are the sole clinical presentation or when the first angiographic study is normal.
- ✓ Some cases assumed as primary headaches may be unidentified RCVS, which is why this syndrome should be included in the differential diagnosis of ‘thunderclap’ headaches, even if the initial investigation is negative.