Reversible Cerebral Vasoconstriction Syndrome

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<table>
<thead>
<tr>
<th>Reversible Cerebral Vasoconstriction Syndrome</th>
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<tbody>
<tr>
<td>• Isolated benign cerebral vasculitis or angiopathy</td>
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<tr>
<td>• Call-Fleming syndrome</td>
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<td>• CNS pseudovasculitis</td>
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<td>• Benign angiopathy of the central nervous system</td>
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<td>• Postpartum angiopathy</td>
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<td>• Migrainous vasospasm</td>
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<td>• Migraine angiitis</td>
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<tr>
<td>• Idiopathic thunderclap headache with reversible vasospasm</td>
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<tr>
<td>• Drug induced cerebral vasculopathy</td>
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<td>• Fatal vasospasm in migrainous infarction</td>
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Reversible Cerebral Vasoconstriction Syndrome

Characterized by the association of severe headaches with or without additional neurologic symptoms, and constriction of cerebral arteries which resolves spontaneously in 1–3 months

The most common clinical feature is a severe acute headache, often thunderclap in nature—a sudden excruciating headache that peaks in less than 1 min, like a “clap of thunder”—mimicking that of a ruptured aneurysm.

Reversible Cerebral Vasoconstriction Syndrome

Cortical subarachnoid hemorrhage (20–25%)

Cerebral infarction and parenchymal hematoma (5–10%)

Reversible Cerebral Vasoconstriction Syndrome

Mild form of angiitis?
\[\downarrow\]
Focal symptoms

Migraine continuum?
\[\downarrow\]
Purely cephalalgic

Reversible Cerebral Vasoconstriction Syndrome

Increasingly recognized as a distinct syndrome due to a transient and reversible disturbance of arterial tone regulation, without inflammation of the arteries, mainly characterized by severe headaches, which are secondary and symptomatic of the underlying vascular abnormality.

Reversible cerebral vasoconstriction syndrome has been reported in patients aged 13–70 years. Mean age of onset is around 45 years with a female to male preponderance from between about 2 and 10:1.

Reversible Cerebral Vasoconstriction Syndrome

The exact incidence is unknown

Probably still underdiagnosed, particularly the pure cephalalgic form

Reversible Cerebral Vasoconstriction Syndrome

<table>
<thead>
<tr>
<th>Delay from headache onset to</th>
<th>No of days (mean (SD) [range])</th>
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<tbody>
<tr>
<td>Diagnosis of cerebral haematoma</td>
<td>1.7 (2) [0–4]</td>
</tr>
<tr>
<td>Diagnosis of subarachnoid haemorrhage</td>
<td>5 (5) [0–20]</td>
</tr>
<tr>
<td>First seizure</td>
<td>3 (1.4) [2–4]</td>
</tr>
<tr>
<td>Posterior reversible encephalopathy syndrome</td>
<td>4 (1.9) [1–6]</td>
</tr>
<tr>
<td>Last recurrent thunderclap headache</td>
<td>7.4 (5.6) [0–28]</td>
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<tr>
<td>Transient neurological deficit</td>
<td>11.6 (4.9) [0–23]</td>
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<tr>
<td>Symptoms of cerebral infarction</td>
<td>13.5 (2.1) [12–15]</td>
</tr>
<tr>
<td>Diagnosis of cerebral infarction*</td>
<td>12 (3) [9–15]</td>
</tr>
</tbody>
</table>

*Diagnosis of infarction may precede symptoms of infarction because an asymptomatic infarct was found in one patient who had a repeat MRI at day 9.

Reversible Cerebral Vasoconstriction Syndrome

Retrospective American series of 16 patients hospitalized for suspected CNS angiitis, of whom 10 had repeat angiography to assess reversibility of vasoconstriction.

Reversible Cerebral Vasoconstriction Syndrome

Prospective Taiwanese series of 56 patients with recurrent thunderclap headaches of whom 22 had proven initial vasoconstriction, then a series of 32 patients (including 12 from the first series) with the proven syndrome

Reversible Cerebral Vasoconstriction Syndrome

Prospective French series of 67 cases, seen in a single institution between 2004 and 2007, who all had vasoconstriction and repeat angiography showing its resolution

The pathophysiology remains unknown

Transient disturbance in the control of vascular tone leading to segmental and multifocal arterial constriction and dilatation

25–60% secondary to exposure to vasoactive, sympathomimetic or serotonergic substances, and/or to the postpartum state

Reversible Cerebral Vasoconstriction Syndrome

Selective serotonin reuptake inhibitors
Sympathomimetics
Over the counter nasal decongestants
Weight loss supplements
Herbal medications
Illicit drugs

Alcoholic intoxication may be an additional precipitating factor but has only been incriminated in association with exposure to other drugs, such as cannabis, ecstasy or cocaine.

Postpartum RCVS starts in two-thirds of cases during the first week after delivery.

Usually after a normal pregnancy.

In 50–70% of cases, it is associated with the intake of vasoconstrictors, mostly ergots used to treat postpartum hemorrhage or to inhibit lactation.

Reversible Cerebral Vasoconstriction Syndrome

- Postpartum
  - With or without exposure to vasoactive substances, eclampsia/pre-eclampsia

- Exposure to vasoactive substances
  - Cannabis, cocaine, ecstasy, amphetamines, LSD, binge drinking
  - Selective serotonin reuptake inhibitors
  - Nasal decongestants—phenylpropanolamine, pseudoephedrine, ephedrine
  - Ergotamine tartrate
  - Methergine
  - Bromocriptine, lisuride
  - Triptans
  - Isometheptine
  - Nicotine patches
  - Ginseng

- Catecholamine secreting tumours
  - Phaeochromocytoma, bronchial carcinoid tumour, glomus tumours

- Exposure to immunosuppressants or blood products
  - Tacrolimus (FK-506), cyclophosphamide, erythropoietin, intravenous immunoglobulin, red blood cell transfusion, interferon-α

- Miscellaneous
  - Hypercalcaemia, porphyria, head trauma, subdural spinal haematoma, carotid endarterectomy, neurosurgical procedures, CSF hypotension

- Extra or intracranial large artery disorders
  - Cervical dissection, unruptured intracranial aneurysm, dysplasia

Reversible Cerebral Vasoconstriction Syndrome

- Catecholamine secreting tumors
- Head trauma
- Neurosurgical procedures
- Carotid endarterectomy
- Carotid stenting
- Intracranial hypotension.

Reversible Cerebral Vasoconstriction Syndrome

History of migraine is found in only 16–20% of RCVS cases, no different from the prevalence of migraine in the general population.

Overlap with the Posterior Reversible Encephalopathy Syndrome

RCVS is a frequent if not a constant feature of PRES

20–30% of cases are normotensive

Normotensive cases have vasogenic edema that is more extensive

Symptomatic treatment
Analgesics?
Antiepileptics?
Blood pressure?
Intensive care unit?

Reversible Cerebral Vasoconstriction Syndrome

Steroids? -------→ probably not

Nimodipine?

Intra-arterial verapamil?

Intraarterial milrinone?

Reversible Cerebral Vasoconstriction Syndrome

Diagnostic criteria for reversible cerebral vasoconstriction syndrome (adapted from the International Headache Society diagnostic criteria for "acute reversible cerebral angiopathy" and the criteria proposed in 2007 by Calabrese et al.):

- Acute and severe headache (often thunderclap headache) with or without focal neurological deficits or seizures
- Monophasic course without new symptoms more than 1 month after clinical onset
- Segmental vasoconstriction of cerebral arteries demonstrated by angiography (MRA, CTA or catheter)
- Exclusion of subarachnoid haemorrhage due to a ruptured aneurysm
- Normal or near normal CSF (protein <1 g/l, white cells <15/mm³, normal glucose)
- Complete or marked normalisation of arteries demonstrated by a repeat angiogram (MRA, CTA or catheter) after 12 weeks, although they may be normal earlier

Now what?

WE’VE SHOWN THAT THE RECENT UNEXPLAINED BEHAVIOR IS DIFFERENT THAN THE PREVIOUS UNEXPLAINED BEHAVIOR.
Thank You