

国際頭痛分類第3版 可逆性脳血管攣縮症候群による頭痛

- A. 新規の頭痛で、Cを満たす。
- B. 可逆性脳血管攣縮症候群と診断されている。
- C. 原因となる証拠として、以下のうち少なくとも1項目が示されている。
1. 頭痛は局在神経学的欠損または痙攣発作（あるいはその両方）を伴うことも伴わないこともあり、血管造影で「数珠（string of beads）状外観を呈し、可逆性脳血管攣縮症候群の診断の契機となった。
 2. 頭痛は以下の項目のいずれかまたは両方の特徴を持つ。
 - a. 雷鳴頭痛として発現し、1ヶ月以内は繰り返し起こる。
 - b. 性行為、労作、ヴァルサルバ手技、感情、入浴、シャワーなどが引き金となる。
 - c. 発現から1ヶ月を超えると、著明な頭痛は起こらない。
- D. 次のいずれかを満たす。
1. 頭痛は初発から3ヶ月以内に解消した。
 2. 頭痛は解消していないが、初発からはまだ3ヶ月経っていない。
- E. 他に最適な国際頭痛分類第3版の診断がなく、動脈瘤性クモ膜下出血が適切な検査で除外されている。

【ICD-3】

6.7.3.1 Acute headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)

Description:

Headache caused by reversible cerebral vasoconstriction syndrome (RCVS), typically thunderclap headache recurring over 1-2 weeks, often triggered by sexual activity, exertion, Valsalva manoeuvres and/or emotion. Headache can remain the sole symptom of RCVS or be a warning symptom preceding haemorrhagic or ischaemic stroke.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Reversible cerebral vasoconstriction syndrome (RCVS) has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:
1. headache, with or without focal deficits and/or seizures, has led to angiography (with “string of beads” appearance) and diagnosis of RCVS
 2. headache has one or more of the following characteristics:
 - a) thunderclap onset
 - b) triggered by sexual activity, exertion, Valsalva manoeuvres, emotion, bathing and/or showering
 - c) present or recurrent during ≤ 1 month after onset, with no new significant headache after > 1 month
- D. Either of the following:
1. headache has resolved within 3 months of onset
 2. headache has not yet resolved but 3 months from onset have not yet passed
- E. Not better accounted for by another ICHD-3 diagnosis¹.

Note:

In particular, aneurysmal subarachnoid haemorrhage has been excluded by appropriate investigations.

Comments:

Reversible cerebral vasoconstriction syndrome (RCVS) is a poorly understood condition, characterized clinically by severe diffuse headaches that typically are of the thunderclap type, mimicking aneurysmal subarachnoid haemorrhage.

RCVS is the most frequent cause of thunderclap headache recurring over a few days or weeks. 6.7.3.1 Acute headache attributed to reversible cerebral vasoconstriction syndrome may rarely have other modes of onset: progressing rapidly over hours or more slowly over days.

Large series of patients with confirmed RCVS have shown that up to 75% present with headache as the only symptom, but the condition can be associated with fluctuating focal neurological deficits and sometimes seizures. 6.7.3.1 Acute headache attributed to reversible cerebral vasoconstriction syndrome may be a warning symptom preceding haemorrhagic or ischaemic stroke. Headache is absent in a minority of cases of RCVS.

Angiography in RCVS is, by definition, abnormal, with alternating segments of arterial constriction and dilatation (“string of beads” or “sausage on a string” appearance). However, MR-, CT- and even catheter-angiography can be normal during the first week after clinical onset. Patients with recurring thunderclap headache and a normal angiogram, but fulfilling all other criteria for RCVS, should be considered as having 6.7.3.2 Acute headache probably attributed to reversible cerebral vasoconstriction syndrome. Brain MRI is abnormal in 30% to 80% of cases, showing various patterns of lesions including intracranial haemorrhages (convexity subarachnoid, intracerebral and/or subdural), cerebral infarctions and/or cerebral oedema corresponding to “posterior reversible encephalopathy syndrome”.

At least half of cases of RCVS are secondary, mainly postpartum and/or attributable to exposure to vasoactive substances including illicit drugs, alpha-sympathomimetics and serotonergic drugs. The disease is self-limiting in 1-3 months, with disappearance of the arterial abnormalities (hence “reversible”) and, almost always, resolution of the headache. However, strokes due to RCVS can produce permanent impairment.