Vasoconstriction Syndrome

The Worst Headache Ever: A Case of Postpartum Reversible Cerebral Vasogenic Syndrome

Case Report
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Introduction/Abstract

Reversible cerebral vasoconstriction syndrome (RCVS) is an important cause of recurrent thunderclap headaches characterised by reversible segmental vasoconstriction of cerebral arteries.

RCVS is diagnosed by the demonstration of segmental constriction of cerebral arteries on CTA, MRA or cerebral angiogram (‘string of beads’ appearance) in the context of a thunderclap headache, with complete resolution of symptoms and signs on imaging within 3 months.

10% occur in the postpartum period. Other common triggers include hypertensive crises and vasoconstricting agents.

Investigations and Differential Diagnosis

Non-contrast CT brain 2 days after first headache normal.

MRI brain 1 day after third headache normal.

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MRA (3D, time of flight) showed abnormal narrowing at A1 and P1 segments of the circle of Willis, and P3 and P4 segments of the left posterior cerebral artery.

This confirmed the diagnosis of RCVS.

Differential Diagnoses: migraine, subarachnoid haemorrhage, eclampsia, Venous sinus thrombus.

Treatment and Follow-up

She received paracetamol and codeine for her headaches until they resolved spontaneously.

Follow-up MRI and MRA 8 months later revealed complete resolution of the previously noted vasoconstriction, i.e. the focal stenoses in the bilateral A1 and P1 segments, and left P3 and P4 segments had opened up and were now of normal calibre.

Case Presentation

A 35 year old nurse (gravida 0, para 2) was admitted with recurrent thunderclap headaches following the birth of her first child.

She experienced a prolonged labour, received an epidural (bupivacaine/fentanyl) and lost 1.5L blood, which required treatment with a IVI phenylephrine and a bolus of ephedrine for symptomatic hypovolaemic hypotension. She was also given ergometrine, misoprostol, carboprost, oxytocin, 2U packed red cells, (Hb dropped to 6) and 2U fresh frozen plasma shortly after delivery.

Her 3 headaches were 30 minutes after birth, 5 days and 11 days postpartum. There was associated nausea but no vomiting, photophobia, phonophobia or focal neurological deficit. Each episode reached maximal intensity within minutes and settled over 48-72 hours.

She had a history of migraine and 3 episodes of vagovagal syncope during pregnancy.

Discussion

When associated with the puerperium, RCVS typically occurs within 7 days postpartum, usually following uneventful pregnancy and delivery.

Classical presentation: recurrent severe occipital thunderclap headaches over 1-3 weeks +/- accompanying confusion, seizures and focal neurological deficits. Usually benign and reversible, but can rarely cause ischaemic and haemorrhagic complications.

Pathophysiology is poorly understood. The characteristic segmental and multifocal narrowing and dilatation of 1+ cerebral artery is presumed to result from a transient non-inflammatory vasospastic disturbance in the control of cerebrovascular tone.

RF: pregnancy, hypertensive crises, vasoactive (adrenergic or serotoninergic) drug use, insertion of an epidural, and genetics. No common structural abnormalities in have been observed in the cerebral vasculature of sufferers.

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Pregnancy is associated with increased sympathetic tone which causes vasoconstriction known to persist beyond the duration of the headache.

Genetic polymorphisms (Val55Met) in the gene for brain-derived neurotrophic factor (BDNF), have been associated with more severe vasospastic in patients with RCVS. BDNF is important for neuronal survival and synaptic plasticity, and is also associated with disorders of abnormal vascular tone such as unstable angina.

RCVS has clinical and radiological similarities to eclampsia and pre-eclampsia. All three conditions may thus be part of the same disease spectrum, with common pathophysiological mechanisms.

Patient Perspective

I was half alive and half dead… I was totally terrified. I am too scared to read my notes because I don’t like remembering. Nowadays I am very cautious. If I have a headache at all I am very paranoid…. I wanted to have another child so that my daughter could have a brother, but I am too terrified about ever experiencing this again to even consider it.

References