Reversible Cerebral Vasoconstriction Syndrome

Makarand Kulkarni 1, Vinay Chauhan 2, Sudheer Shetty 3

Abstract

Reversible cerebral vasoconstriction syndrome (RCVS) is a disease characterized by thunderclap headache with severe vasospasm of middle sized vessels of circle of Willis or the extracranial circulation which spontaneously revert back. We report a middle aged female with severe headache and vasospasm of the vertebral arteries and vessels of circle of Willis causing multiple cerebral infarcts. The vasospasm resolved within 3 months.

Introduction

Reversible cerebral vasoconstriction syndrome (RCVS) is a rare entity characterized clinically by thunderclap headache with or without neurological deficit or convulsions. Radiologically the disease is characterized by multifocal vasoconstriction of cerebral arteries which can lead to multiple infarcts. The disease involves medium size vessels. The vasoconstriction usually spontaneously resolves within weeks to months which is an important diagnostic feature. We report a case of a middle aged female with thunderclap headache with multiple acute infarcts. MR angiography showed diffuse vasospasm of A1 segment of bilateral anterior cerebral artery territory and branches of bilateral middle cerebral arteries. The vasospasm resolved after 3 months.

Case Report

35 years old female presented with sudden onset of thunderclap headache followed by visual disturbance and weakness in all the four limbs. On examination patient was conscious but disoriented. There was no significant past history apart from a spontaneous abortion 6 months ago or no significant history of drug intake.

Magnetic resonance imaging (MRI) showed multiple acute infarcts in bilateral water shed territory between anterior and middle cerebral artery (Figure 1). There was no subarachnoid hemorrhage. Magnetic resonance angiography (MRA) showed diffuse spasm of the vessels of circle of Willis with beaded appearance of branches of middle cerebral artery (MCA) (Figure 2A). Similar beaded appearance was noted in bilateral vertebral arteries (Figure 2B). The patient was subjected to digital substruction angiography (DSA) which confirmed that there was diffuse vasospasm. Intra-arterial Nimodipine was given into the vertebral arteries which failed to cause vasodilatation (Figures 3A and B).
Routine investigations including CBC, ESR, electrolytes, CRP were negative. CSF examination and vasculitic workup were negative.

On the basis of clinical presentation of thunderclap headache and radiological findings of diffuse vasoconstriction with beaded appearance of vertebral arteries and vessels of circle of Willis a possibility of reversible cerebral vasoconstriction syndrome versus primary CNS angiitis (PCNSA) were made.

Patient was treated with antiplatelets, verapamil, nimodipine, low molecular weight heparin with vitamin b12 and folic acid. She also received injectible 1 gm daily for 5 days in the initial stages of admission.

Methyl prednisolone patient gradually improved over a period of 4 -5 weeks, regained consciousness, started recognizing the relatives with improvement of speech and vision, spasticity decreased in all 4 limbs and regained sphincter control.

After 3 months a repeat MRI and MRA showed complete resolution of the vasospasm (Figure 4A, B) with chronic infarcts in bilateral high fronto parietal region (Figure 4C). The vessels were found to be of normal size and calibre.

Discussion

Reversible cerebral vasoconstriction syndrome first described by Call and colleague is (1) and is characterized by multifocal reversible vasoconstriction. It is an under-recognized cause of thunderclap headache. Its pathophysiology is unknown however abnormality in regulation of vascular tone is thought to be responsible which can be primary or secondary to exogenous factors including drugs, trauma or endocrine abnormality.¹ There are many precipitating factors which include postpartum state, use of nasal decongestant, cocaine, antidepressants, ergot, catecholamine secreting tumors such as phaeochromocytoma, glomus or carotid tumors, immunosuppressant or blood transfusions.¹² Relationship with postpartum state may lead to suggestion of association with sex hormones.¹ The disease usually occurs in young patients (20-40 yrs) with female predominance.¹³

The disease usually presents with thunderclap headache (TCH). Thunderclap headache is characterized by sudden onset of severe headache which reaches its peak within one minute.¹⁴ It is a presenting feature in subarachnoid hemorrhage, pituitary apoplexy and cerebral venous sinus thrombosis. RCVS is a recently recognized entity with such presentation.¹³ The headache in RCVS is recurrent and initial imaging investigations may be negative. Recurrent TCH and negative initial CT and MRI has a high predictive value for possibility of underlying RCVS.³ Other neurological symptoms include transient visual or sensory symptoms, aphasia, hemiparesis, ataxia, seizures and stroke. Our patient presented with TCH, visual disturbance and altered sensorium due to development of large stroke in bilateral water shed territories between middle - anterior cerebral artery and territories supplied by anterior cerebral arteries. CSF examination in most of the cases is negative except for mild elevation of CSF protein and mild increase in WBC count.¹ In our case CSF examination was normal.
Neuroimaging findings in RCVS show diffuse vasoconstriction which involves large and medium size vessels. There may be involvement of intra and extracranial vessels as was seen in our case. Vasospasm gives a characteristic beaded appearance to the vessels. The beaded appearance may also be seen in Primary CNS angiitis. However, RCVS can easily distinguished from PCNSA by dramatic onset, recurrent thunderclap headache and normal CSF examination. The most specific finding is resolution of angiographic findings within days to weeks (usually 12 weeks) as seen in our case. There are some reports about use of intra-arterial Nimodipine which causes reversal of vasospasm and is a helpful differential diagnostic tool from other vasculitis. In our case there was no reversal of vasospasm.

The other neuroimaging findings in RCVS are multiple infarcts, subarachnoid hemorrhage, reversible cerebral oedema as seen in posterior reversible encephalopathy (PRES) and intracerebral hemorrhage. In our case there were multiple acute infarcts in bilateral watershed territory.

RCVS is treated with short course of steroids, calcium channel blockers and magnesium sulfate. The disease is reversible with a good outcome. There are reports of use of intra arterial Nimodipine in multiple sessions with good outcome. Our patient was treated with single intraarterial Nimodipine in the acute stage at the time of DSA which failed to dilate the vessels. Subsequently the patient was treated with antiplatelet and low molecular weight heparin as the patient developed multiple infarcts in bilateral cerebral hemispheres. She was also treated with verapamil. methylprednisone was injected, though the role of steroid in RCVS is controversial. Vitamine B12 and Folic acid were given for stroke.

In conclusion, RCVS is an important cause of recurrent thunderclap headache, characterized by multifocal vasospasm giving beaded appearance on imaging which should be differentiated with primary CNS angiitis. It has a benign course with reversal of vasospasm.

References