CASE REPORT

Poor outcome associated with probable bilateral extracranial ICA vasospasm

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SUMMARY

We describe a woman with bilateral extracranial internal carotid artery (ICA) vasospasm. Initial MRI of the brain showed multiple areas of high-signal intensity in the white matter of both the frontoparietal lobes. She was alert, and the muscle strength of the four extremities was moderately decreased. Cranial CT on day 3 showed increased numbers of low-density areas in both the anterior cerebral arteries (ACA) and middle cerebral arteries (MCA), accompanied by neurological deterioration. Cranial and cervical CT angiography on day 9 showed that all areas of both the ACA and MCA had become low density. Both ICAs were markedly narrowed along their entire length and tapered. On day 16, the patient died. Three patients with bilateral extracranial ICA spasms have been described previously. To our knowledge, this is the first time to document bilateral ICA spasm causing elongated narrowing of the carotid arteries, leading to an unfortunate outcome.

BACKGROUND

Three patients with bilateral extracranial internal carotid artery (ICA) spasms have been described previously.^{1 2} One of these patients had a history of migraine, and the ICA spasm was segmental and reversible.^{1 2} We describe a woman presenting with bilateral extracranial ICA vasospasm, leading to a poor outcome.

CASE PRESENTATION

A 67-year-old woman with no risk factors for stroke, including migraine and no family history of stroke, cardiac or neurodegenerative diseases suddenly noticed difficulty in standing. She had no traumatic event and did not smoke or drink alcohol. On admission, the blood pressure was 168/84 mm Hg, the pulse rate was 115 bpm and regular and the body temperature was 36.1°C. She was alert, and the muscle strength of the four extremities was moderately decreased. The deep tendon reflexes were moderately increased in all four extremities, with a positive left Babinski's sign. Subjective numbness without decreased superficial sensation was evident in all four extremities. There was no cranial-nerve impairment.

On day 1, cranial MRI of the brain showed multiple areas of high-signal intensity in the white matter of both the frontoparietal lobes (figure 1A). Cranial MR angiography showed that both intracranial anterior cerebral arteries (ACA) and middle cerebral arteries (MCA) were poorly visualised, with right-sided dominance. Arrhythmias were absent on cardiac monitoring, repeated

electrocardiography and 24 h Holter monitoring; the echocardiogram was normal two times. The results of routine serum chemical analysis were normal, except for an elevated level of glucose (239 mg/dL). The serum levels of D-dimer, lactic acid and pyruvic acid were normal. Anti-DNA, antinuclear, antiphospholipid and anticardiolipin antibodies, and the results of lupus anticoagulant testing were negative. Cerebrospinal fluid showed no pleocytosis, and the results of bacterial culture were negative. The patient received intravenous argatroban (60 mg/day, 3 days) and edaravone (60 mg/day, 11 days) and was subsequently given intravenous heparin (10 000–15 000 units/day). Cranial CT on day 3 showed increased numbers of low-density areas in both the ACA and MCA, accompanied by neurological deterioration. On day 4, the consciousness level dropped to a coma with respiratory insufficiency, requiring oxygen inhalation. In addition, cranial CT showed a further increase in the number or area of low-density regions. Intravenous dexamethasone (12 mg/day, 5 days) was started for a suspected diagnosis of angitis. Blood pressure was measured three times a day or more frequently depending on the patient's condition. The systolic and diastolic blood pressure ranged from 107 to 190 mm Hg and 84 to 105 mm Hg, respectively. The blood pressure rose to 220/120 mm Hg on day 5, but there was no evidence of neurological deterioration. Sinus tachycardia was continuous, but arrhythmias were absent on cardiac monitoring and repeated electrocardiography. On the morning of day 6, decorticate posture occasionally developed for several minutes, and in the evening, paradoxical breathing characterised by swelling of the abdomen during inspiration and the reverse pattern during expiration was observed. On day 7, cardiopulmonary arrest occurred, and the heart rate and blood pressure were restored by immediate emergency life support. Mechanical ventilation was required, and the comatose state was unchanged. Cranial and cervical CT angiography on day 9 showed that all areas of both the ACA and MCA had become low density (figure 1B). Both ICAs were markedly narrowed along their entire length and tapered (figures 1A–D). Enhanced CT angiography of the chest showed no abnormality of the aorta or pulmonary artery, and there was no evidence of thrombosis within the left atrium. The patient's family requested that further treatment be withheld, and we therefore did not administer calcium antagonists or other medications.

To cite: Nobuhiro Sawa N, Kataoka H, Ueno S. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009767

Figure 1 Both internal carotid arteries (ICAs) were markedly narrowed along their entire length and tapered on cranial, CT angiography (A–D, blue arrow: right internal carotid artery, red arrow: left internal carotid artery). Multiple infarctions limited to the white matter consistent with the watershed between the territories of the anterior and middle cerebral arteries on diffusion-weighted MRI on day 1 (A). All areas of both anterior and middle cerebral artery territories became low density (B).

DIFFERENTIAL DIAGNOSIS

In our patient, infarction initially occurred in the watershed between the ACA and MCA and then extended to the ICA, causing severe bilateral stenosis of the extracranial ICA. Disorders potentially causing vascular stenosis include cerebral vasoconstriction syndrome characterised by narrowing of the cerebral arteries, and the stenotic lesion will nearly always be intracranial; an accompanying thunderclap headache is mandatory.³ Primary angiitis of the central nervous system associated with granulomatous or lymphocytic vasculitis affects the leptomeningeal and cortical arteries and responds to steroids.⁴ Arterial dissection often occurs after traumatic events. Our patient showed none of these characteristics, and there was no evidence of dissection on CT angiography, making these diseases unlikely. Diabetes mellitus does not seem to cause such long narrowing of the carotid arteries. Hypertension was not a likely cause of the bilateral stenosis of the extracranial ICA since the elevated blood pressure was apparently not associated with neurological deterioration, and the territory of the posterior cerebral arteries was spared on neuroimaging.⁵ We therefore gave our patient a diagnosis of vasospasm of the extracranial ICA, but we could not establish the cause.

OUTCOME AND FOLLOW-UP

On day 16, the patient died.

DISCUSSION

Three patients with bilateral extracranial ICA spasms have been documented, and the ICA spasm was reversible.^{1–2} Two patients

with bilateral extracranial ICA spasms showed no reduction in the frequency of ICA spasms or associated attacks despite anti-vasospastic treatments.^{1–2} Bilateral ICA spasms can have unfortunate outcomes, such as in our patient.

Learning points

- ▶ Four patients with bilateral extracranial internal carotid artery (ICA) spasms, including the present patient, have been described.
- ▶ Multiple areas of high-signal intensity in the white matter of the frontoparietal lobes on brain MRI suggested extracranial ICA spasm.
- ▶ Bilateral extracranial ICA spasms can rapidly progress to irreversible ICA spasms.
- ▶ Bilateral ICA spasms can have unfortunate outcomes.

Contributors NS and HK were responsible for the overall study design, and wrote the manuscript. HK and NS contributed to running the study and acquisition of data. HK and NS contributed to analysis and interpretation of the data. HK and SU contributed to drafting and critical revision of part of the submitted materials.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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