

CASE REPORT

A conversion disorder or a stroke? A proximal basilar artery thrombosis induced 'locked-in' syndrome in a young Caucasian woman

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SUMMARY

The incidence of the basilar artery occlusion is relatively low among all strokes. Clinical presentation varies depending on the location of the occlusion. The symptoms include mild dysarthria to coma or sudden death. The initial subtle clinical presentation could lead to misdiagnosis. Psychogenic diagnosis in the differential could make the timely diagnosis more difficult. This case involves a 34-year-old woman presenting with a gradual onset of slurred speech. The initial CT scan of head did not indicate any intracranial pathology, and she was initially treated for an anxiety/conversion disorder. With progression of the pathology, the patient quickly developed a 'locked-in' syndrome, with preserved high cognitive function and vertical eye movement, but otherwise total loss of motor function. The diagnosis was confirmed with MRI/MR angiography studies, which indicated thrombosis of the proximal basilar artery. Serological studies did not disclose any relevant risk factors.

BACKGROUND

Basilar artery occlusion is found in 2 of 1000 autopsy studies. The percentage of basilar artery occlusion among all posterior circulation strokes ranges from 11% to 39% as cited in the Boston Posterior Circulation Registry¹ and Lausanne Stroke Registry, respectively.² The clinical presentation of basilar artery occlusion varies significantly dependent on the location and time elapsed.³ The risk factors that would lead to an infarct in the basilar artery region are not different from those that lead to cerebrovascular accidents (CVA) elsewhere. These risk factors include hypertension, diabetes mellitus, hyperlipidaemia, smoking, hypercoagulable states and cardiac disease. CVA are more common in elderly of late 60s–70s and the incidence is higher in men (2:1) as compared with women. Because of the low incidence and varied clinical presentations, many healthcare professionals may misdiagnose this pathology in young patients as either a psychogenic disorder or substance abuse.

The clinical presentation of a basilar artery infarct can vary from a gradual presentation to a sudden onset of bulbar or motor symptoms. These symptoms include: dysarthria, bilateral facial and extremity weakness as well as emotional lability. As one of the clinical presentations of basilar artery infarct, locked-in syndrome is characterised by

preserved consciousness, eye movements along with loss of speech and quadriplegia.^{4 5}

CASE PRESENTATION

The patient is a 34-year-old woman who was in her usual state of health and had been to swimming with her children the day before presentation. After the pool activities the patient complained of feeling fatigued, weak and had three episodes of emesis which did not involve any blood. As time progressed, the patient went to bed. Around 4:30, she awoke and went to the bathroom, complaining at that time of feeling unsteady. Around 7:30, the patient was found on the floor in the bathroom, with slurred speech and left-sided weakness. Once at the emergency department, the patient's family offered a medical history of attention-deficit disorder treated with Adderall. Her husband also mentioned that the patient was under more stress than usual. The initial neurological evaluation took place at the primary care hospital, the patient scored 15 of 15 on the Glasgow Coma Scale (GCS); however, a more detailed neurological assessment was not available. The initial CT scan of head did not indicate any intracranial abnormality. While the patient was being evaluated, it was noted that she was progressively deteriorating, becoming less able to communicate and increasingly frustrated. At this point, the patient was administered sedatives to treat her anxiety. Hours later, the patient's condition worsened and she was transferred to our facility for higher level of care.

On arrival to our facility, her GCS was 11. On physical exam it was noted that the patient followed commands inconsistently and therefore it was difficult to complete a thorough neurological exam. It was noted that the patient's pupils were 2 mm bilaterally and reactive to light and accommodation and gag reflex was present. Deep-tendon reflexes were hyper-reflexic in all extremities, with an upgoing Babinski sign, and decerebrate posturing with painful stimuli. At this point, it was evident that the patient was in a locked-in syndrome. She had preserved higher cognitive function, blinking her eyes to express yes or no, but unable to move any part of her body.

The diagnosis was confirmed with an MRI of brain without contrast, which indicated she had sustained a large acute pontine infarct involving the width of the pons (figure 1). An MR angiography of the brain without contrast with maximum intensity projections and using three-dimensional

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Figure 1 MRI of the brain showing the large acute pontine infarct involving most of the width of the pons, predominant on the left.

time-of-flight imaging technique identified the occlusion to be at the proximal area of the basilar artery with some flow being maintained at the uppermost portion of the basilar artery (figure 2). After evaluation by a neurologist, neither thrombolysis nor thrombectomy was recommended because the patient's symptoms had been present for more than 12 h. There is no definitive recommendation on the time frame for reperfusion therapy in a basilar artery stroke; however, aggressive treatment of patients presenting with symptoms for more than 12 h may not be justified.⁴ Patient was treated with antiplatelets agents: aspirin and plavix (clopidogrel). During her inpatient stay, the patient underwent an extensive workup to find the cause of her stroke; meanwhile receiving supportive care along with physical, occupational and speech therapies. The patient's hypercoagulable workup was essentially negative except for a mild elevation of rheumatoid factor (16, normal range 0–14) and a decrease in complement component 3 (45, normal range 88–201). In addition to this workup, an ECG and a transthoracic ECG were within normal limits. Three weeks of her hospital stay, the patient was stabilised and transferred to a neurological rehabilitation centre for further physical and occupational therapies.

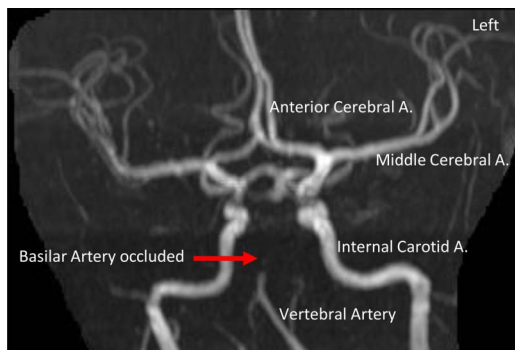


Figure 2 MR angiography of the brain showing the occlusion of the proximal basilar artery. There is some flow in the uppermost basilar artery, which is probably retrograde, via the posterior communicating arteries, which also appeared to provide flow to the posterior cerebral arteries.

OUTCOME AND FOLLOW-UP

This case report was written 3 months after the patient went through the extensive neurological rehabilitation programme. On the follow-up of the patient's progress it is known that she recovered part of her motor function and was able to mobilise in a wheelchair with assistance. Patient is able to use a 'speak and spell' device to help with communication. She is now receiving speech therapy and occupational therapy to help improve specific activities of daily living. Actually, the patient signed the consent by herself although the handwriting looks like scribble.

DISCUSSION

This is an unusual case of an acute basilar artery thrombosis occurring in a young Caucasian woman. The clinical presentation and her lack of common risk factors for stroke led to an untimely diagnosis. Though the incidence of basilar artery thrombosis is higher in elderly population, cases of basilar artery occlusion have been reported in young population,⁶ even in paediatric patients.⁷ The clinical investigation of hypercoagulability and cardiovascular abnormalities did not disclose any contributing factors. Therefore, we must consider other unknown risk factors. For example, the previous swimming pool activity could contribute to the dissection of the basilar artery. The recent stress in patient's life could be a significant risk factor for stroke in young population. A case control study by Egido *et al*⁸ disclosed that stressful habits are significantly associated with high risk of stroke. Other case reports conclude that oral contraceptives may be implicated as a cause of basilar artery thrombosis.^{7–9} In this case, the patient did have an intra-uterine device but in previous months, she had been on oral contraceptive pills. There are articles that implicate methamphetamine abuse as a cause of cerebrovascular pathology, including those that lead to strokes.¹⁰ This patient was taking Adderall 20 mg daily to treat her attention deficit disorder. Adderall is a brand name medication of a combination drug that is composed of amphetamine salts. However, there is no direct evidence that Adderall can cause CVA.

Infarctions are usually treated with medical therapy alone and early reperfusion should be initiated in the emergency department if indicated. However, instead of the reperfusion, this patient was initially treated with sedative drugs because of the mimics of the early neurological focal sign and symptoms with conversion disorder. It has been well known that drowsiness and fluctuation in level of consciousness are common findings in brainstem strokes or lesions. Sedative treatment not only masks those critical sign and symptoms, it also worsens the already depressed respiratory and circulatory activity. In addition, sedatives reduce blood pressure, which decreases perfusion of the brain stem.⁴

Though the risk factors of stroke in this patient are obscure, we should not let that mislead our clinical differential and diagnosis. A complete neurological examination by the first treating physician might have raised suspicion of basilar artery occlusion. A retrospective study by von Campe *et al* showed that 15 of 24 (63%) patients with basilar artery occlusion presented with articulatory speech difficulties. The other initial presentations include: motor deficits, vertigo, nausea or vomiting and headaches. The most frequent objective initial findings include: motor deficits, facial palsies, eye movement abnormalities, lower cranial nerve deficits, altered level of consciousness and bilateral extensor plantar responses. Although the onset of the disease was gradual, some warning signs could present for couple of months in advance. Only 8% had an acute course without any warning signs. In another words, about 92% of patients with

basilar artery occlusion present with unequivocal brainstem signs.¹¹ In this patient, the initial assessment occurred at another facility and detailed documentation of the neurological evaluation is not available to us.

Learning points

- ▶ It is indispensable to have a complete neurological physical examination in every patient with focal symptoms.
- ▶ Awareness of atypical stroke signs or symptoms in low-risk populations.
- ▶ Neurological rehabilitation can significantly improve poststroke outcomes.

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Competing interests None.

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