Unusual presentation of more common disease/injury

Occlusion of the artery of Percheron: an unusual cause of bilateral stroke

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Summary

The artery of Percheron is a rare anatomical variant whereby a single vessel arising from the proximal segment of one posterior cerebral artery supplies both medial thalami. This is a rare example of a single arterial supply to brain structures on both sides of the midline. Occlusion of the artery of Percheron results in bilateral medial thalamic infarction, which is manifest clinically as gaze paresis, cognitive disturbance and altered consciousness. The presentation can mimic subarachnoid haemorrhage, drug intoxication, encephalitis and other inflammatory or infective conditions. The presentation is similar to the 'top of the basilar syndrome' and early recognition should prompt further investigation for underlying stroke aetiologies and consideration can be given to thrombolysis if vascular occlusion can be confirmed.

BACKGROUND

Ischaemic stroke is very common, but despite this posterior circulation ischaemic stroke can sometimes be difficult to recognise clinically as its presentation can be very varied. It often mimics other pathological processes including intracranial haemorrhage, infection and inflammation. Clinicians need to be vigilant to uncommon presentations of common disease so that appropriate investigations and management can be initiated. This patient's clinical presentation not only highlights the varied and staggered onset which is often described as part of the 'top of the basilar syndrome' but also identified an unusual anatomical variant which contributed to his presentation. In this case, occlusion of the artery of Percheron was responsible for bilateral infarction of the medial thalami.

CASE PRESENTATION

A 46-year-old Caucasian man presented to the emergency department having been found collapsed by his colleagues. Initially, he complained of a headache, and was confused and disorientated with slurred speech. In the ambulance he became increasingly drowsy and dysarthric. His Glasgow Coma Scale was 8 and he was noted to have pinpoint pupils bilaterally. His condition did not improve following intravenous naloxone administration by the paramedics. He was hypertensive with blood pressure (BP) 159/104 mm Hg. Blood glucose was normal at 6.9 mmol/l.

No significant medical history or family history was elicited and he was taking no regular medication. He had received the seasonal flu vaccination 3 weeks prior to his presentation. His colleagues and family denied the possibility of illicit drug use. He was a non-smoker and consumed alcohol in moderation.

A provisional diagnosis of subarachnoid haemorrhage was made and he was intubated prior to transfer for urgent CT head imaging.

INVESTIGATIONS

Cranial CT did not identify any acute intracranial abnormality. ECG confirmed a sinus rhythm. Initial biochemistry and haematology were unremarkable, including normal erythrocyte sedimentation rate and C reactive protein. Treatment for encephalitis was started pending cerebrospinal fluid (CSF) analysis. CSF protein was mildly elevated at 0.64 g/l, but CSF white blood cells were normal at $<1 \times 10^6/l$ and there was no evidence of subarachnoid haemorrhage. CSF glucose was normal at 3.8 mmol/l. EEG did not demonstrate any evidence of seizure activity. An MRI scan of his brain was organised. This demonstrated the presence of bilateral areas of high signal in both thalami on T2-weighted imaging with restricted diffusion, consistent with acute bilateral paramedian thalamic infarction, without midbrain involvement (figure 1). Urine toxicology was unremarkable, and there was no evidence of vasculitis on serum immunology. Transthoracic and transoesophageal echocardiography were normal. CSF PCR testing was negative for herpes simplex, enterovirus and varicella zoster viruses. CT and MR vascular imaging did not show any evidence of intracranial venous thrombosis or significant occlusive disease of any of extracranial or intracranial arteries, although the left vertebral artery was dominant.

DIFFERENTIAL DIAGNOSIS

The history available at the time of assessment was limited, but having presented with sudden onset headache and progressive neurological deficit, subarachnoid haemorrhage was the initial working diagnosis. Drug intoxication and viral encephalitis were also considered within the early differential diagnosis. Progressive onset of ophthalmoplegia, motor dysfunction and behavioural abnormalities raised the possibility of 'top of the basilar' syndrome with a vascular aetiology being responsible for this

Figure 1 Axial MRI of brain showing high signal on T2-weighted (A) and diffusion-weighted images (DWI) (B) with corresponding low signal on adjusted diffusion co-efficient (ADC) sequences (C) due to acute bilateral thalamic infarction (arrows). T2 (D), DWI (E) and ADC (F) sequences at the level of the midbrain showing no evidence of acute infarction.

gentleman's symptoms. The diagnosis of bilateral paramedian thalamic infarction was confirmed by MRI.

TREATMENT

Following initial cranial CT, the patient was transferred to the intensive care unit. Although modern cranial CT, when performed early, is highly sensitive for identifying subarachnoid haemorrhage the diagnosis was confidently excluded following CSF examination.¹ The patient received treatment for viral encephalitis, although this was stopped following receipt of negative CSF PCR results. The key to diagnosis for this patient was the MRI brain scan, following which standard secondary prevention with antiplatelet agents, lipid lowering therapy and BP control was started in addition to further investigation for the underlying stroke aetiology.

OUTCOME AND FOLLOW-UP

Following successful extubation, neurological assessment demonstrated upward gaze deviation, dysarthria and slow-reacting pupils. Initially, no obvious motor deficit was identified in his limbs but both plantar responses were extensor. Over the following days he was noted to have a variable pyramidal weakness in his left upper and lower limbs with associated hyperreflexia and he was hypersomnolent. He also experienced visual hallucinations, paranoid ideation and impaired cognition (Mini-Mental State Examination 18/30). BP remained elevated although no underlying cause of this was identified, and BP lowering therapy was started. Over the following week the hallucinations and paranoia resolved and the limb weakness significantly improved. Vertical gaze paresis persisted but gradually improved over the course of several months, as did his cognitive impairment. Twelve months following presentation, he has made a near-complete recovery. With

the exception of hypertension, further investigation did not identify any proximal source of embolism or other underlying pathology in our patient.

DISCUSSION

The clinical presentation of this patient gives rise to a number of possible aetiologies including subarachnoid haemorrhage, drug intoxication and viral encephalitis, all of which were excluded by subsequent investigation. The key to diagnosis in this patient was the MRI scan. The progressive onset was typical of the 'top of the basilar' syndrome, suggesting occlusion at the top of the basilar artery but this was excluded by CT and MR angiography. Cerebral venous thrombosis can give rise to a similar clinical and radiological picture although intracerebral haemorrhage is often a feature of intracranial venous thrombosis and the abnormalities are not confined to specific vascular territories.² Venous occlusive disease was excluded on CT venography in our patient. Occlusion of a single arterial supply to both thalami was therefore responsible for this patient's presentation. This is known as the artery of Percheron and is one of only a few examples of a single arterial supply to structures of both cerebral hemispheres.

The arterial supply to the thalamus is complex and typically comprises four vascular territories: the anterior territory supplied by the polar artery which arises from the posterior communicating artery; and the paramedian, inferolateral and posterior territories arising from the posterior cerebral arteries.³

Percheron^{4 5} described a number of anatomical variations of the paramedian arterial supply to the thalamus, the most common of which is a pair of arteries arising from each P1 segment of the posterior cerebral artery (PCA), type I. Type II variants occur when both pairs of

perforating arteries arise from the P1 segment of a single PCA, with type IIb variants occurring when a single vessel (the artery of Percheron) arises from the proximal PCA which then supplies both thalami. A vessel bridging both PCAs with an arcade of perforating vessels forms a type III variant.

The true prevalence of the artery of Percheron is unknown.^{3 6} Although the artery of Percheron has previously been directly visualised on angiography⁷ it is unusual to directly visualise it on CT or MR angiography, particularly in the presence of occlusive disease.^{3 6} Successful recanalisation of the artery of Percheron with intra-arterial thrombolysis has however been described.⁷

Although this patient's angiography demonstrated a patent basilar artery, the onset and progression of symptoms and signs suggested posterior circulation ischaemia to be a possible differential diagnosis. Clinicians need to be vigilant for the constellation of clinical features which contribute to the 'top of the basilar syndrome'. These include ophthalmoplegia, dysarthria, visual disturbance, altered consciousness and motor paresis indicating ischaemia of the structures of the upper brainstem and posterior cerebral artery territories.⁸ The onset of symptoms can often be staggered. Early identification of basilar artery occlusion may allow recanalisation and reperfusion via intravenous and/or intra-arterial thrombolysis although data from large randomised controlled trials to support this are lacking.⁹ Basilar artery occlusion without recanalisation has a high associated mortality of approximately 90%.⁹

Our patient's clinical features were consistent with bilateral paramedian thalamic infarction, which typically include altered consciousness, memory impairment and various vertical gaze palsies. In particular, vertical gaze palsies have previously been described in the absence of midbrain involvement.¹⁰ The clinical deficits are in keeping with pathology affecting the five major functional classes of nuclei which are found within the thalamus: reticular/intralaminar, sensory, effector, associative and limbic.³ As in our patient's case, the prognosis following thalamic infarction is favourable and appears to be better than after thalamic haemorrhage although this is largely based on small case series.^{3 11} Complete recovery following bilateral medial thalamic infarction has previously been reported.¹²

Learning points

- ▶ Sudden and staggered onset of ophthalmoplegia, dysarthria, altered consciousness and motor paresis should alert the clinician to the possibility of ischaemia within the upper brainstem and posterior circulation; the 'top of the basilar syndrome'.
- ▶ The artery of Percheron is an uncommon anatomical variant where both medial thalami are supplied by a single vessel arising from one posterior cerebral artery.
- ▶ Occlusion of the artery of Percheron results in infarction of both medial thalami, a rare example of a single artery supplying structures in both cerebral hemispheres.
- ▶ Early recognition and treatment of posterior circulation strokes should prompt further investigation for underlying stroke aetiologies.

Competing interests None.

Patient consent Obtained.

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