Artery of Percheron Infarct — a diagnostic and prognostic conundrum!

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Abstract

A 39 year old male was found unconscious at home. On arrival to the hospital the patient was profoundly somnolent but easily rousable, with no focal neurological deficits. Imaging confirmed bilateral thalamic infarcts from the occlusion of Artery of Percheron, a rare anatomic variant which is a single arterial trunk supplying the thalamus and midbrain bilaterally. Anti-platelet therapy was initiated as soon as the diagnosis was established and the patient showed a rapid remarkable recovery over the next 48 hours. He continued to improve subsequently and was at baseline functional status at 6 months. Extensive investigations for etiologies were mostly unrevealing. In such patients presenting with drowsiness/somnolence, a posterior circulation stroke should be considered if no evidence of other more common causes are found. A CT head must be followed by an MRI to confirm the diagnosis and subsequent focus should be on eliciting risk factors and careful evaluation for etiologies.

Keywords: Artery of Percheron, Thalamic stroke, Cerebral Circulation, Posterior Circulation, Coma, dowsiness.

Background

The Artery of Percheron is a very rare variant of the posterior cerebral circulation where a single arterial trunk supplies blood to the paramedian thalami and the rostral midbrain bilaterally.[1] Occlusion of this artery results in a distinctive pattern of bilateral thalamic infarcts with or without midbrain infarction.[1,2] It has a variable presentation ranging from somnolence or transient loss of consciousness to as severe as coma in affected patients; accompanied by a wide variety of focal neurological findings.[3] Recognizing an acute PCA stroke can be both arduous and clinically challenging in terms of availability of radiological modalities and expertise for establishing diagnosis in a timely manner, for appropriate management and reverse any potentially permanent deficits.[4] Here we will discuss the case of a 39 year old man who presented to us in the ER after being found unconscious at home and was managed conservatively with subsequent excellent recovery.

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Case Report

A 39 year old male smoker presented in January 2015

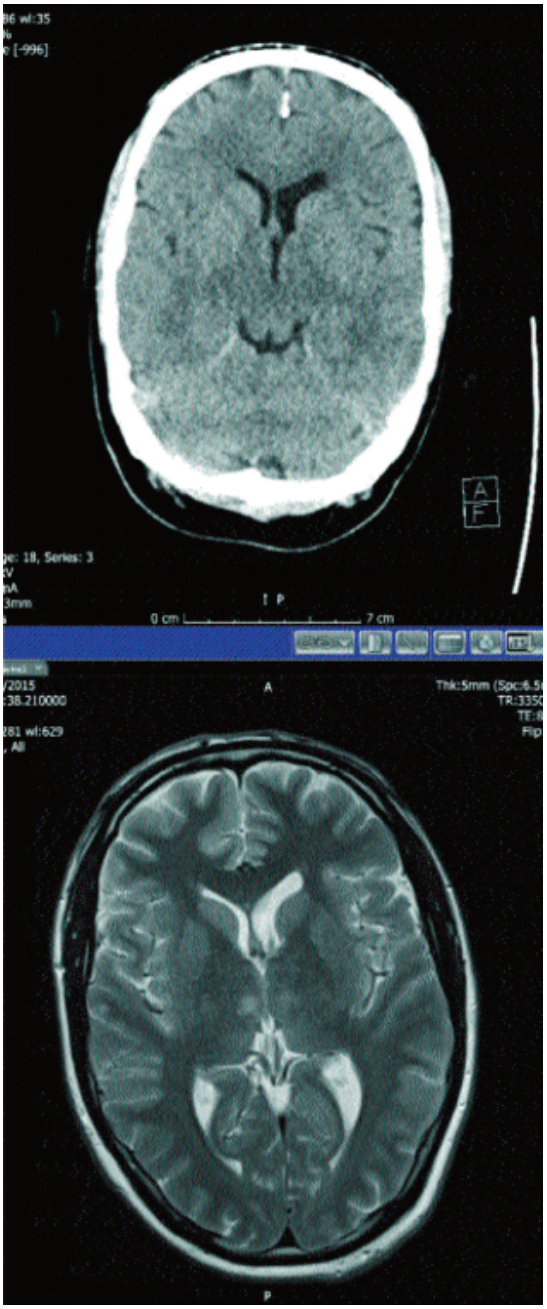

Figure-1: CT Head.

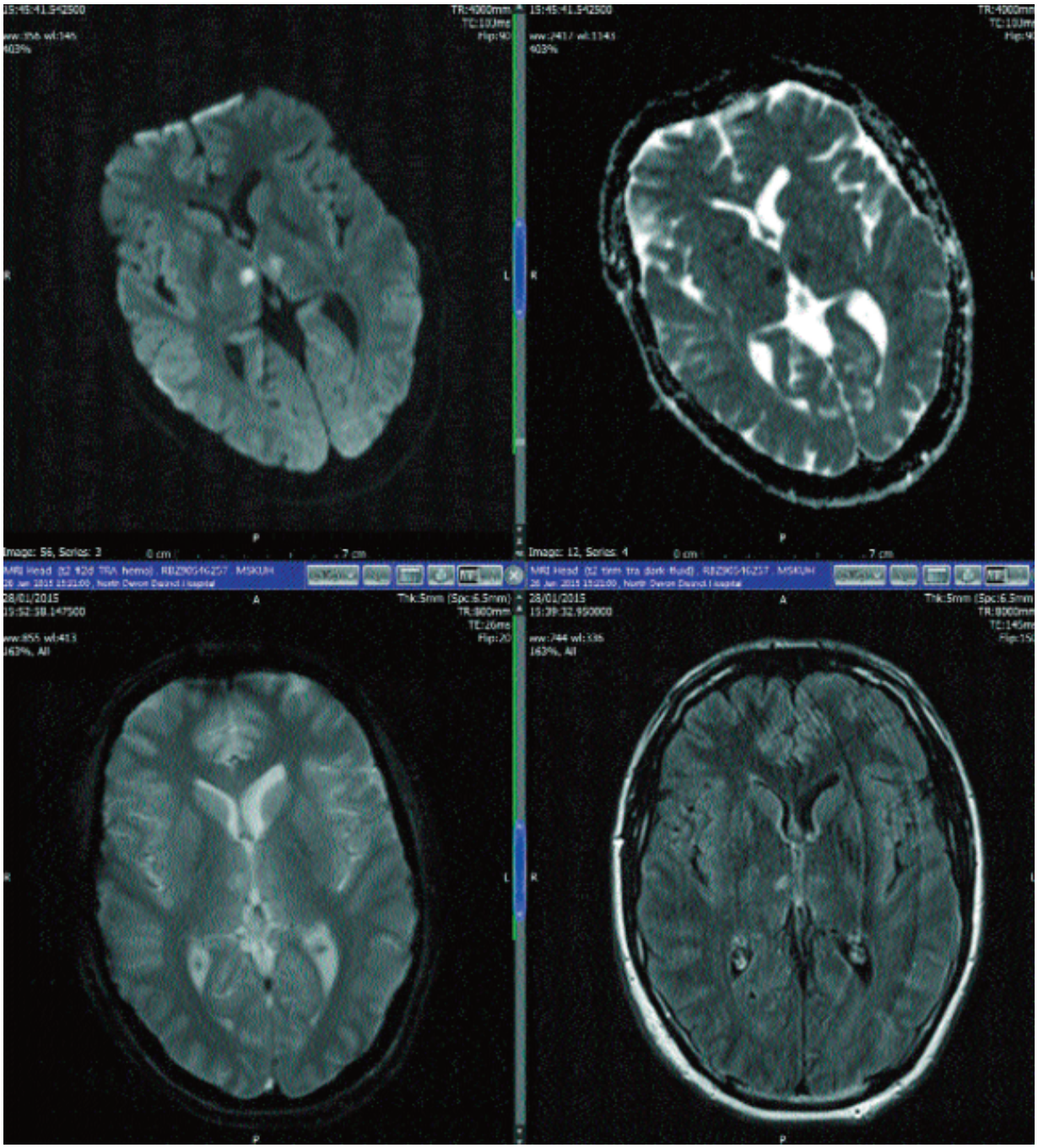

Figure-2: MRI Brain.

to North Devon District Hospital, Barnstaple, UK; after he was found unconscious by his wife at home. He was lying in the bath tub at the time of discovery. There was a vague history of a mild head injury sustained whilst he was wearing a helmet a couple of weeks ago and he had been suffering from some stress and low mood lately. The patient was otherwise previously completely fit and well; leading a healthy and active lifestyle, even frequently volunteering as a coast guard. There was no recent history of fever, headache, seizure, and no known toxic substance exposure. On arrival, the patient had a blood pressure of 109/67 mm Hg, HR 50 beats/min and respiratory rate 22 breaths/minute. On neurological examination, he was remarkably

somnolent but easily rousable and well-oriented in time, place and person. GCS was 13/15 at presentation (E3V4M6). There was no neck rigidity and pupils were bilaterally equal and reactive. No focal neurological sign could be elicited and systemic examinations were unremarkable.

Laboratory findings including blood glucose, full blood count, electrolytes; liver-, renal- and thyroid function tests, calcium, arterial blood gas and ammonia were unremarkable. Electrocardiogram (ECG) showed a normal sinus rhythm. Infective, metabolic and toxicology screens returned negative and therefore a preliminary diagnosis of Encephalitis was considered for which a lumbar puncture was carried out but also did not manifest any abnormality.

After an unremarkable head CT, an MRI scan was carried out the same day which revealed a high signal on T2 and FLAIR imaging in the medial portions of bilateral thalami demonstrating restricted diffusion consistent with acute ischaemic insult to these regions. There was no evidence of acute or previous intraparenchymal haemorrhage and ventricular system was also normal. Whilst this normal but rare variant of PCA was recognised to be artery of percheron infarct, an expert opinion from Neuroradiologist was awaited who later confirmed the findings.

CT Arteriogram from arch of aorta to Circle of Willis showed normally enhancing carotid and vertebral arteries without any evidence of dissection as well as normal enhancement of the basilar artery. Additional investigations included a 24 hour ECG which was normal and a Trans-thoracic Echocardiogram that showed overall good left ventricular systolic function and non-dilated cardiac chambers with no evidence of mural thrombus. All valves appeared structurally normal. However a Bubble Echo done at a later date was indicative of patent foramen ovale. In light of this finding, lower extremity doppler study was performed to look for Deep Venous Thrombosis but no signs were appreciated radiologically or clinically.

No evidence of coagulopathy or any vasculitides was obtained on extensive blood workup.

Soon after establishing diagnosis, patient was immediately started on Aspirin and was later shifted to Clopidrogrel for secondary prevention. He briskly continued to improve on anti-platelet therapy alone and had near complete recovery at 6 months follow-up.

Discussion

Artery of Percheron infarct, albeit a very rare occurrence; has been previously reported keenly for the rarity of the Artery of Percheron variant in the population and the radiological challenges it poses.[3,4] However it still remains a diagnostic conundrum in patients presenting with not-so-clear manifestations of posterior circulation stroke, such as the patient described here.

The most commonly reported presentation is a degree of impairment of conscious with vertical gaze palsies and/or other focal neurological findings.[3,5,6] However our patient presented with no focal neurological findings and exhibited only signs of disruption of reticular activating system. As such in a young patient like ours with no previous co-morbids presenting with only generalized CNS signs and symptoms; cerebrovascular infarcts would be low on a list of differential diagnosis. Generalized CNS insults such as infections, toxic insults and metabolic derangements would be more likely.[7] Hence valuable time for appropriate interventions might be lost; for example by the time a diagnosis of stroke is made the patient may be well past the therapeutic window for thrombolysis. To further confound matters, contrast and non contrast CT scans which are the initial radiological modalities in workup for stroke in most centers; are almost always unrevealing for thalamic strokes and even on MRI it may be difficult to appreciate changes consistent with acute ischaemic insults to thalami without supportive clinical findings.[3,4]

The second issue with such a presentation is the prognostic uncertainty it puts forward. As with our patient who was quite young and healthy, with no strong risks for thromboembolic disease, no etiologies identified on extensive investigations, the individual prognostic outcome cannot be reliably predicted.[8] Various therapeutic modalities exist, including thrombolysis for acute stroke (guidelines and recommendations vary), long-term anti-platelet therapy for secondary prevention and some anecdotal recommendations for even long term anticoagulation based on the fact that most strokes in young (including Artery of Percheron infarcts) are secondary to embolic phenomenon. However the extent of recovery and the risk of recurrence remain quite variable despite these measures.[1,2] This can present challenges to physicians with regards to counseling patients and discussing their prognostic outcomes.

Lastly we discovered evidence of likely Patent Foramen Ovale in our patients, yet no proof of an embolic source was substantiated. Conversely a patent foramen ovale can be found in upto 25-30% of the population and can be associated with cryptogenic strokes such as the one described here.[9,10] No consensus exists on the best treatment in such cases.[11] Our patient opted to not have closure of the patent foramen ovale.

Conclusion

In a patient presenting with drowsiness/somnolence, a posterior circulation stroke should be considered if no evidence of other more common causes are found. A CT head must be followed by an MRI to confirm the diagnosis and subsequent focus should be on eliciting risk factors and careful evaluation for etiologies.

To the best of our knowledge, our patient is the only one reported to have just dysfunction of arousal/conscious and no other focal neurological findings.

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Conflict of Interest: None to declare.

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Consent: Valid informed consent obtained from patient.

References

1. Raphaeli G, Liberman A, Gomori J, Steiner I. Acute bilateral paramedian thalamic infarcts after occlusion of the artery of Percheron. Neurology. 2006; 66:E7.
2. Gentilini M, de Renzi E, Crisi G: Bilateral thalamic artery infarcts: report of eight cases. J NeurolNeurosurg Psychiatry. 1987; 50:900-9.
3. Lamot U, Ribaric I, Popovic KS. Artery of Percheron infarction: review of literature with a case report. RadiolOncol. 2015; 49:141-6.
4. Menon MK, Jacob SM, Jalal MJ. Artery of Percheron occlusion: role of diffusion-weighted imaging in the early diagnosis. NeuroimmunolNeurolnflam. 2016; 3:14-6.
5. Monet P, Monet P, Garcia PY, Saliou G, Spagnolo S, Desblache J, et al. Bithalamic infarct: is there an evocative aspect? Radioclinical study. Rev Neurol. 2009; 165:178-84.
6. Lazzaro NA, Wright B, Castillo M, Fischbein NJ, Glastonbury CM, Hildenbrand PG, et al. Artery of percheron infarction: imaging patterns and clinical spectrum. AJNR Am J Neuroradiol. 2010; 31:1283-9.
7. Edlow JA, Rabinstein A, Traub SJ, Wijdicks EF. Diagnosis of reversible causes of coma. Lancet. 2015; 385:1179-80.
8. Li L, Yiin GS, Geraghty OC, Schulz UG, Kuker W, Mehta Z, et al. Incidence, outcome, risk factors, and long-term prognosis of cryptogenic transient ischaemic attack and ischaemic stroke: a population-based study. Lancet Neurol. 2015; 14:903-13.
9. Meissner I, Whisnant JP, Khandheria BK, Spittell PC, O'Fallon WM, Pascoe RD, et al. Prevalence of potential risk factors for stroke assessed by transesophagealechocardiography and carotid ultrasonography: the SPARC study. Stroke Prevention: Assessment of Risk in a Community. Mayo Clin Proc. 1999; 74:862-9.
10. Hagen PT, Scholz DG, Edwards WD. Incidence and size of patent foramen ovale during the first 10 decades of life: an autopsy study of 965 normal hearts. Mayo Clin Proc. 1984; 59:17-20.
11. Dalen JE, Alpert JS. Cryptogenic strokes and patent foramen ovales: what's the right treatment? Am J Med. 2016; 129:1159-62.