

CASE REPORT

Urgent decisions and a tight spot: embolic infarction of a herniated cerebellar tonsil

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SUMMARY

A previously well 30-year-old woman presented at 17:30 with a sudden onset of dizziness, ataxia and headache. She was initially investigated with a CT scan of the brain and lumbar puncture, which yielded no diagnosis.

Subsequent MR scan revealed multiple posterior circulation infarcts, along with a previously undiagnosed Arnold-Chiari 2 malformation with an associated syrinx of her cervical and thoracic spine. The infarct involved one of the herniated cerebellar tonsils. Oedema of an infarct in the herniated tonsils caused compression of the medulla at the foramen magnum, with associated neurological symptoms including Lhermitte's phenomenon and headache on Valsalva manoeuvre. Owing to these symptoms a surgical decompression was performed. The most likely aetiology of her stroke was determined to be a paradoxical embolus via patent foramen ovale.

BACKGROUND

Arnold-Chiari malformation is a common anatomical variant where the cerebellar tonsils herniate into the foramen magnum. It is frequently associated with the development of a cervical syrinx and occasionally with syringomyelia. In this report, we describe a case where acute embolic infarction of a tonsil in a young woman resulted in sudden presentation of symptoms or cervical cord compression. This hazardous complication of the interaction between two relatively common neurological problems has not been described.

CASE PRESENTATION

A 30-year-old Caucasian woman presented to the emergency department with sudden onset of dizziness and vomiting, followed by a headache. She had never experienced a similar episode previously. Her only medications were the combined contraceptive pill and occasional cyclizine for intermittent nausea. She was a lifelong non-smoker, drank alcohol occasionally and denied illicit drug use. There was no relevant family history.

On examination, she had brisk reflexes throughout and demonstrated dysmetria with her left upper limb. She had no tremor or nystagmus reported. Her saccadic movements were normal. Her gait was ataxic and her NIH Stroke Scale was reported as 3. Examination of other systems was normal.

INVESTIGATIONS

Full blood count, renal and liver profiles, and C reactive protein were normal. She had an urgent

cranial CT scan to look for evidence of stroke, which was reported normal initially; however, a later formal report suggested herniation of her cerebellar tonsils with a Chiari malformation (figure 1). A lumbar puncture was performed to rule out subarachnoid haemorrhage given the acute onset of headache which also precluded thrombolysis. The cerebrospinal fluid examination was normal with no evidence of infection or xanthochromia, with 47 red cells and normal protein. Opening pressure was not recorded. A prospective diagnosis of cerebellar stroke was made and she was transferred to the stroke unit. She underwent MRI of her brain and cervical spine, and cerebral angiography the following morning. On MR diffusion-weighted imaging she was found to have multiple small acute infarcts in the posterior thalamus and both cerebellar hemispheres (figures 2 and 3), all in a posterior circulation distribution. Medullary and cerebellar herniation through the foramen magnum was noted, representing an Arnold-Chiari type 2 malformation (figure 4). There was also a substantial syrinx in the spinal cord extending from C2 to T2 (figure 4), the extent of which was surprising given her relative lack of neurological signs. Radiology findings did not support dissection, vasculitis or fibromuscular dysplasia as potential aetiologies.

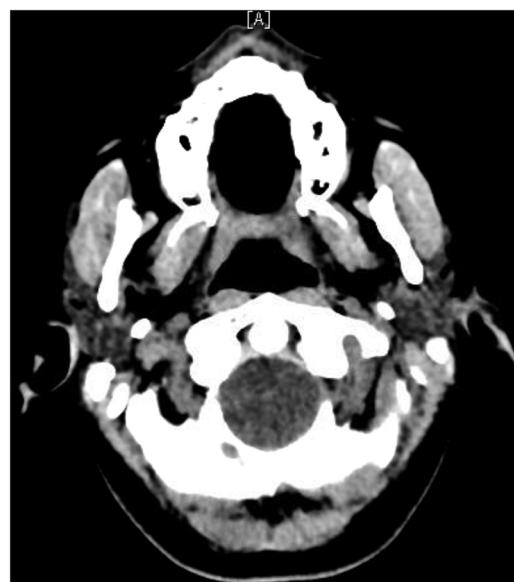


Figure 1 Neurological imaging of the patient. CT scan on admission showing crowding in foramen magnum due to herniation of cerebellar tonsils. Admission non-contrast CT scan of the brain. Foramen magnum appears crowded with no visible cerebrospinal fluid.



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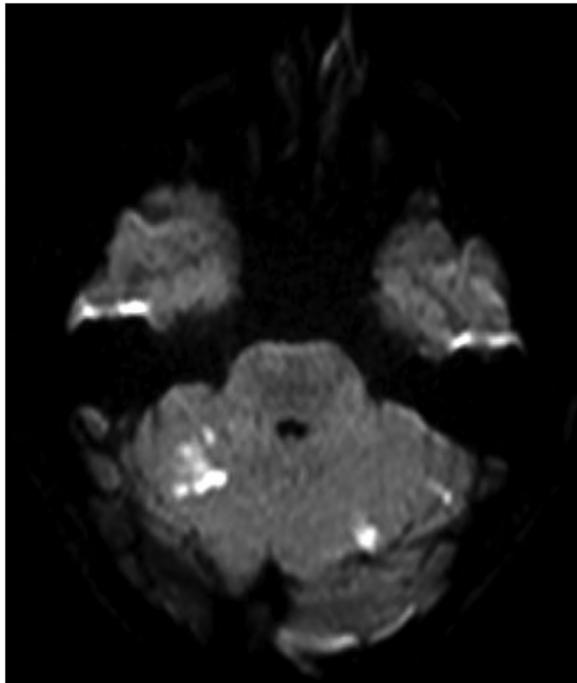


Figure 2 Axial diffusion-weighted imaging (DWI) MR showing areas of acute infarction in both hemispheres of the cerebellum. DWI sequence MR scan of the brain. Multiple small infarcts were visible in both cerebellar hemispheres.

Physiological monitoring demonstrated that she remained in sinus rhythm throughout her hospital stay.

The patient's headache initially improved following admission; however, 2 days after admission she reported worsening of her headache on sitting up and coughing. This was thought to be concerning, as she had a lumbar puncture performed in the context of an undiagnosed Arnold-Chiari malformation. Repeat MR brain scan and review of her initial MRI showed acute infarct within her herniated right cerebellar tonsil on both fluid-attenuated inversion recovery (FLAIR) and diffusion-

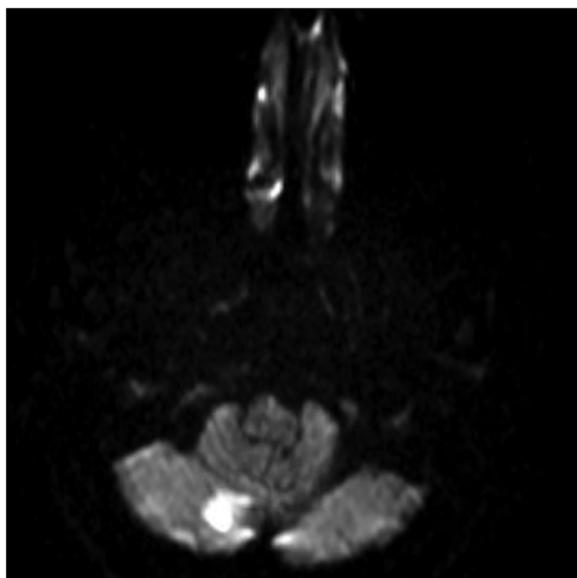


Figure 3 Axial diffusion-weighted imaging MR from just above foramen magnum showing acute infarct in the right inferior cerebellum.

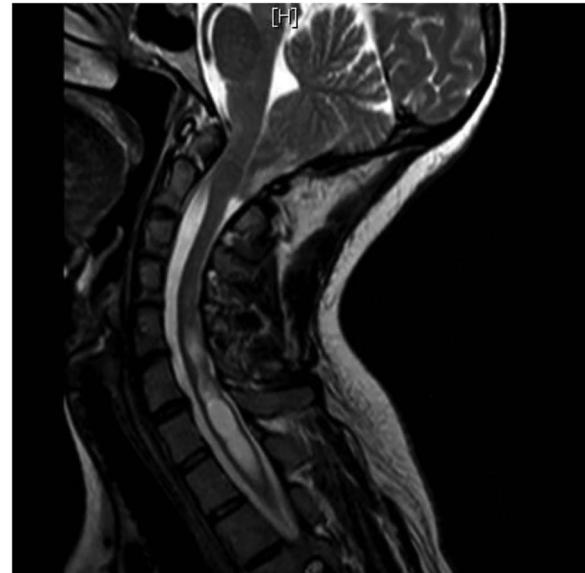


Figure 4 Sagittal T2 MR scans of the brainstem and cervical cord. Syrinx present in cervical and upper thoracic cord. Cerebellar tonsil shows evidence of infarction.

weighted imaging (figures 5 and 6) thus increasing her risk of spinal cord or medullary compression. She had no new focal neurology at that time, but some hours later she described bilateral paraesthesiae in her upper limbs on laughing or neck flexion, characteristic of Lhermitte's phenomenon.

TREATMENT

She was reviewed by a neurosurgical service and underwent craniocervical decompression of the Chiari malformation. This was uncomplicated and she was transferred back to our care to complete her workup on her symptoms having resolved.



Figure 5 Axial T2 fluid-attenuated inversion recovery MR scan showing infarction of the right cerebellar tonsil.

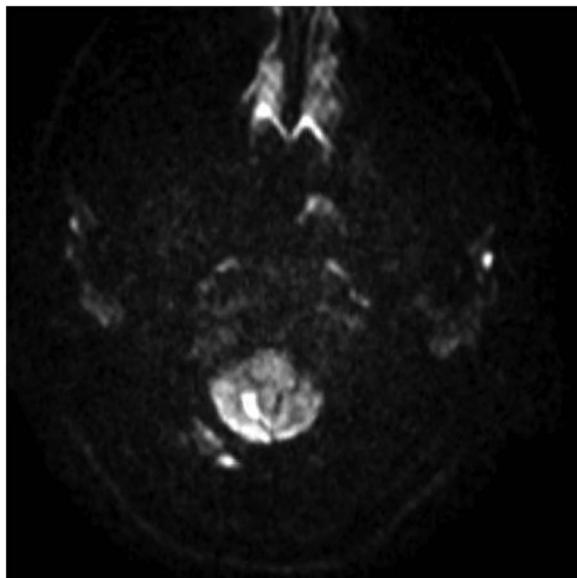


Figure 6 Axial diffusion-weighted imaging MR at level of foramen magnum showing hyperintensity of the right cerebellar tonsil consistent with the infarct.

OUTCOME AND FOLLOW-UP

Postoperatively, our patient was discharged on antiplatelet treatment (aspirin 75 mg daily).

Subsequent contrast echocardiography revealed significant flow across the interatrial septum without valsalva, consistent with a large patent foramen ovale or atrial septal defect which is presumed to be the most likely cause of her infarct.

DISCUSSION

This report is of an unusual interaction of two fairly common neurological conditions resulting in a potentially very serious consequence. The assessment of dizziness in the emergency department is frequently challenging¹ with particular difficulty for the emergency staff in distinguishing between central and peripheral causes. MR scanning is not easily available out of ordinary working hours and in the context of the availability of acute interventions for stroke decisions are often made quickly on the basis of CT scans where structural subtleties like the presence of herniated cerebellar tonsils may often be missed.

Even following our patient's MR scan it was only when she developed new symptoms that the infarction of the tonsil was recognised.

The failure to recognise the Arnold-Chiari malformation and herniated tonsils led to the medical team on admission performing a lumbar puncture to rule out subarachnoid haemorrhage. While there was no absolute contraindication to lumbar puncture, the identification of a Chiari malformation may have led to a decision to defer the procedure in the context of no blood having been seen on CT scan. Recent studies have suggested that sensitivity CT may be approaching 100% if performed in the first 6–12 hours after onset of symptoms of subarachnoid haemorrhage² and thus lumbar puncture may not be necessary if a

serious caution or contraindication exists.³ MR is clearly the investigation of choice in a case such as this, where a patient presents with acute neurological signs suggestive of a potential posterior circulation stroke.^{4,5} However, MR remains unavailable outside standard working hours in many hospitals.⁶ Eventually it was not the imaging findings that led us to her diagnosis but the symptoms and signs she developed after 48 hours, probably when oedema of the infarcted tonsil had progressed, resulting in compression of the posterior columns and causing Lhermitte's sign. This emphasises the continued importance of clinical neurological history taking and examination in the era of hyperacute stroke.

Learning points

- ▶ Arnold-Chiari malformation is a relative contraindication to lumbar puncture; however, it may be performed where it is necessary to rule out a serious pathology, such as meningitis or subarachnoid haemorrhage.³
- ▶ Urgent brain imaging and rapid neurological assessment of suspected stroke is now the norm in most centres to facilitate potential intervention. In circumstances of more complex neurological presentations caution should be exercised before engaging in more invasive investigations such as a lumbar puncture, with detailed neurological examination and formal evaluation of brain imaging carried out before the start.
- ▶ While historically stroke was characterised by signs that were 'maximal at onset', neurological deterioration occurs in 20–30% of cases. It is important that patients are kept under observation and fully re-evaluated if this occurs to identify remediable causes. This may be one reason for the effect of stroke unit care in reducing death and disability.⁷

Contributors RMD, DB and JAH were all involved in diagnosing and managing the patient. All contributed to the writing of the paper.

Competing interests None declared.

Patient consent Obtained.

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