

Images of the month 2: No laughing matter: Symptomatic Chiari malformation type I

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Case presentation

A 29-year-old woman was referred to the general medicine clinic for a 3-month history of worsening headache. It was generalised and squeezing in nature, and occurred daily with coughing, laughing or straining, leading to sudden inability to move and embarrassment in work and social life. This was associated with giddiness and nausea but there were no other neurological symptoms or deficits. As her headaches happened with Valsalva-like activities, a magnetic resonance imaging of the brain was performed to exclude a secondary cause of headache

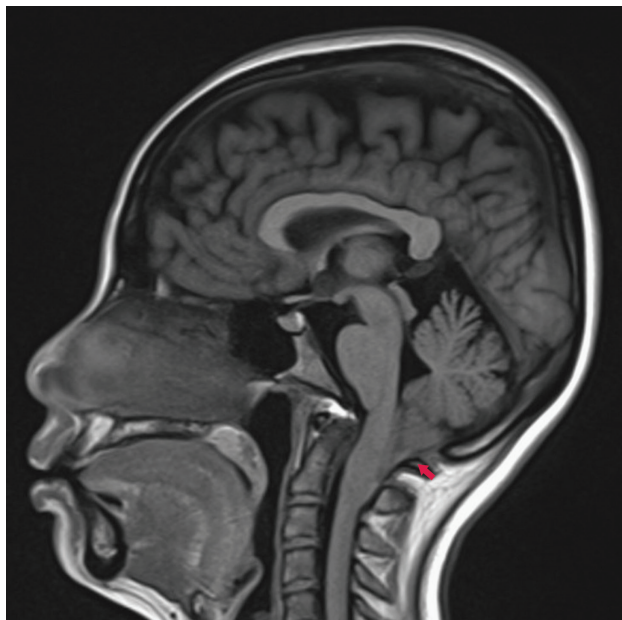


Fig 1. Sagittal image from the magnetic resonance imaging of the brain showing tonsillar descent below the foramen magnum (arrow).

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and it showed marked tonsillar descent below the foramen magnum without syrinx and hydrocephalus (Fig 1). A diagnosis of Chiari malformation type I (CM-I) was made. She underwent decompression of the foramen magnum and C1 laminectomy for symptomatic CM-I and the tonsils were confirmed to be well below the C1 arch intra-operatively. Her symptoms' frequency and severity improved significantly post-surgery with enhanced quality of life.

Discussion

CM-I is the most common type of Chiari malformation. It is often asymptomatic but can present with headache, syringomyelia, hydrocephalus, cerebellar dysfunction, cranial neuropathies or brainstem compression symptoms in adolescence or adulthood.¹ Decompressive surgery is indicated for symptomatic patients with CM-I.¹

Headache is one of the commonest medical complaints and should be approached in a systematic manner to differentiate between primary and secondary headaches; the latter can account up to 10% of all headaches.^{2–4} The SNOOP4 mnemonic (Table 1) is a useful acronym in history taking to elicit red flags

Table 1. The SNOOP4 mnemonic for excluding red flags (secondary causes of headache)

Clinical features	Need to exclude
S Systemic symptoms: fever, chills, myalgia or weight loss	Metastasis and infection
N Neurological symptoms or deficits	Stroke, mass lesion and encephalitis
O Older age at onset (>50 years)	Temporal arteritis, glaucoma and mass lesion
O Onset: thunderclap headache	Bleed
P Papilloedema	Raised intracranial pressure
P Positional	Intracranial hypotension
P Precipitated by Valsalva manoeuvre or exertion	Raised intracranial pressure
P Progressive headache or substantial pattern change	Any secondary cause

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suggestive of secondary causes of headache that will warrant further investigations, such as neuroimaging, lumbar puncture, and specialist review.⁴ Although this patient's demographics fit the typical profile of a patient with migraine, prompt recognition of red flags and early diagnosis of the underlying problem with timely intervention can lead to high-value care and better clinical outcomes, as seen in this case.² ■

References

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