Cough headache as a presenting feature of posterior reversible encephalopathy syndrome (PRES)

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A 23-year-old man with chronic renal failure on maintenance haemodialysis was referred to the neurology outpatient clinic with new onset cough headache. On evaluation, his blood pressure was 220/120 mmHg and outpatient fundus photography showed grade IV hypertensive retinopathy. Urgent magnetic resonance imaging showed features of posterior reversible encephalopathy syndrome (PRES). Emergent treatment of hypertension led to a rapid resolution of his cough headache. New onset cough headache may be a marker of PRES.

KEYWORDS: cough headache, PRES, posterior reversible encephalopathy syndrome

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Case presentation

A 23-year-old man on haemodialysis presented to us with a new onset headache of 10 days duration. He had undergone a live related-donor renal transplant 10 years previously for bilateral multi-cystic dysplastic native kidney disease. With triple immunosuppression (tacrolimus, mycophenolate mofetil and prednisolone), he maintained stable graft functions for 7 years. After this, he developed a progressive worsening of renal functions. In April 2019, a graft biopsy showed features of chronic antibody mediated rejection, and haemodialysis had been initiated 4 months earlier. His current headache appeared during bouts of coughing or sneezing. It was a generalised, severe, brief (<1 minute) holo-cranial headache that subsided spontaneously, and it occurred 2–3 times a day. He had a history of hypertension for the past 1 year and was taking metoprolol, hydralazine and prazosin. His medications included subcutaneous epoetin beta (RhEPO; a synthetic, recombinant of erythropoietin) 4,000 IU three times per week. His blood pressure was 220/120 mmHg. A neurology outpatient fundus photograph revealed grade IV hypertensive retinopathy without any other neurological findings. Serum creatinine levels were 663 μmol/L and electrolytes were normal.

Diagnosis

A diagnosis of secondary cough headache was considered. The possibilities were a hypertensive urgency, posterior reversible encephalopathy syndrome (PRES) or a structural posterior fossa lesion.

Initial management and prognosis

He was admitted to the intensive care unit. Intravenous labetalol and nitroglycerin infusions were used to lower his blood pressure. He also required escalation of his oral antihypertensives with addition of clonidine 0.6 μg/day and nifedipine 20 mg three times a day. Magnetic resonance imaging (MRI) of the brain showed extensive fluid-attenuated inversion recovery (FLAIR) hyperintensities involving the tempo-occipital cortex and cerebellar white matter without diffusion restriction, MR angiography was normal and the MRI findings were compatible with PRES syndrome.
**Posterior fossa lesion**

Treatment of accelerated hypertension may be sufficient to cure meningioma.

**Subdural hematoma.**

**Posterior reversible encephalopathy syndrome**

**Spontaneous intracranial hypotension.**

**Chiari type I malformation.**

**Acute sphenoid sinusitis.**

**Cerebral aneurysms.**

Cough headaches could indicate an evolving PRES.

**Meningitis.**

Severe PRES can lead to cerebral infarction or haemorrhage.

Prompt recognition and treatment of the underlying condition can lead to good outcomes and reversibility of the MRI changes on follow-up. Most patients experience a good outcome within weeks, although up to 15% of patients can develop morbidity or mortality with severe PRES.

**Case progression and outcome**

A diagnosis of cough headache secondary to PRES was considered. RhEPO was identified as a potential culprit and discontinued. Over the next 3 days, his blood pressure reduced to 160/100 mmHg and his cough headache disappeared. Levetiracetam was started pre-emptively for seizures. He was discharged uneventfully after 5 days.

**Discussion**

Cough headache is a well described, but rare headache. Headaches are characteristically triggered by a rapid increase in intra-abdominal pressure which occurs with coughing, sneezing or straining. Two types of cough headache are described: primary and secondary (symptomatic) cough headache. Primary cough headache is defined by the International Headache Society (IHS) as a headache, precipitated by coughing or straining in the absence of any intracranial disorder and lasting up to 30 minutes. Almost any other type of underlying headache (such as migraine or tension type headache) can be exacerbated by coughing, but are not included in the rubric of cough headache. Secondary (symptomatic) cough headache (SCH) comprises around 40% of cough headaches and is associated with a wide variety of aetiologies. Prominent among them are posterior fossa lesions and Chiari malformations, which account for nearly 80% of SCH (Box 1).

PRES is a characterised by headache, seizures, altered mental status or cortical visual disorders (such as cortical blindness and hemianopsia). Headaches are reported in up to 50% of patients with PRES. Most often, headaches are diffuse and gradual in onset, or of the thunderclap variant (in which MR angiography changes of reversible cerebral vasocstriction accompany PRES). Cough headache has not been described in PRES. MRI shows white matter vasogenic oedema predominantly affecting the subcortical and cortical regions of the occipitoparietal lobes, however, oedema can involve the cerebellum, brainstem or even frontal regions in more severe cases. These changes are well appreciated on T2 or T2-FLAIR MRI sequences.

**Key points**

- Cough headache should prompt neuro-imaging to identify symptomatic causes.
- Cough headaches could indicate an evolving PRES.
- Treatment of accelerated hypertension may be sufficient to cure cough headaches in certain cases.

**References**


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