

A fatal headache with normal brain imaging

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Abstract

Headache at rest or on exertion could rarely be the only presenting symptom of an acute coronary syndrome. This is referred to as “cardiac cephalgia”. A healthy 37-year-old man presented with a headache of one week. He was initially treated with an analgesic and, a Magnetic Resonance Imaging of the brain was performed on day five of the illness as the headache was persistent. The next day he developed a generalised seizure, followed by unresponsiveness. He was rushed to hospital within 15 minutes and was pronounced dead on admission. Cause of death was established as acute myocardial infarction at post-mortem examination. This patient fulfilled the ICHD-3 diagnostic criteria for cardiac cephalgia. Postulated mechanisms of cardiac cephalgia include referred pain to cervical nerve roots, increased intracranial pressure from sudden reduction of cardiac output, cerebral vascular changes caused by chemical mediators released in myocardial ischaemia and hypoperfusion. Cardiac cephalgia should be suspected in any unexplained headache, especially if the headache occurs in relation to exertion. Prompt diagnosis and treatment will reduce the morbidity and mortality in cardiac cephalgia.

Key words: cardiac cephalgia, angina cranialis, myocardial infarction, headache

Introduction

Headache at rest or on exertion could rarely be the only presenting symptom of an acute coronary syndrome. This is referred to as “cardiac cephalgia” or “angina cranialis”. Cardiac cephalgia is a serious cause of secondary headache and is under-diagnosed

and under-reported. Failure to diagnose cardiac cephalgia may lead to severe morbidity and even death.


Case presentation

A 37-year-old man had persistent occipital headache for one week (based on the history obtained from the wife). There was no worsening of the headache with exertion. There was no past history of hypertension, ischaemic heart disease, diabetes or dyslipidaemia. He was treated with an analgesic, pantoprazole and domperidone by a neurologist. Clinical examination findings from initial consultation records revealed normal blood pressure and normal central nervous system examination. However, as the headache did not improve, Magnetic Resonance Imaging (MRI) of the brain was performed on day five of the illness and was advised to review with the results. The headache persisted and he went on to develop blurred vision and unsteady gait over the next 24 hours. He was observed to have sudden jerking of the arms and eyes rolling up followed by unresponsiveness. He was rushed to an emergency treatment unit within 15 minutes and was pronounced dead on admission. MRI scan of the brain was normal on subsequent review.

Post mortem examination revealed a grossly enlarged heart. All major coronary arteries were occluded (80-90%) with atheromatous plaques. On macroscopic examination, there was evidence of acute and previous myocardial infarctions with fibrosis in the left ventricle and the papillary muscles (Figure 1). In addition, there were extensive atheromatous plaques in the aorta and the circle of Willis. However, there was no thrombosis of the carotid, vertebral, basilar arteries and arteries of the circle of Willis. The brain parenchyma did not show any evidence of infarctions or haemorrhage.

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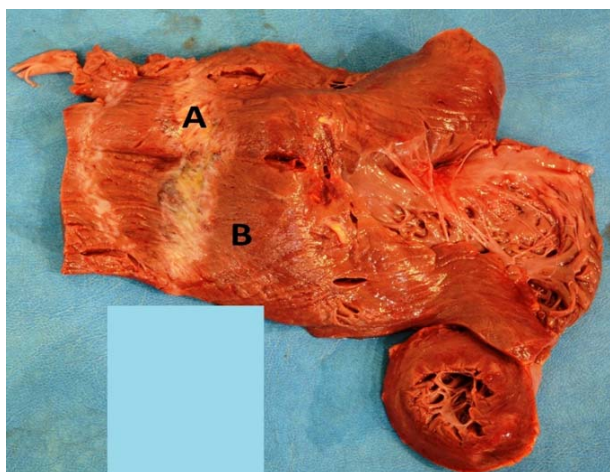


Figure 1. Macroscopic appearance of the heart showing, (A) areas of old infarction and fibrosis and (B) acute infarction of the left ventricle.

Discussion

The most common differential diagnosis encountered here by the clinician at first point of contact was possibly a primary headache, most likely a new onset migraine or tension headache. International Classifi-

cation of Headache Disorders (ICHD-3) describes cardiac cephalgia as “a migraine-like headache, usually but not always aggravated by exercise, occurring during an episode of myocardial ischaemia”.¹ Headache as the only presenting symptom of myocardial ischaemia has been described in less than 6% of patients.² This patient fulfilled the ICHD-3 diagnostic criteria for cardiac cephalgia (Table 1).

Postulated mechanisms of cardiac cephalgia include referred pain to upper cervical nerve roots, increased intracranial pressure caused by reduced venous return as a result of sudden reduction of cardiac output and cerebral vascular changes caused by chemical mediators such as serotonin, bradykinin, substance P and atrial natriuretic factor released during myocardial ischaemia.³ Cerebral hypoperfusion has been implicated in cardiac cephalgia and hypothesized to be due to cerebral vasoconstriction and cortical spreading depression following myocardial ischaemia.⁴

Red flag signs described in cardiac cephalgia include headache brought on with exertion, new or different headache, age over 50 years and history of vascular risk factors.⁵ Apart from a new onset headache,

Table 1. ICHD-3 Diagnostic criteria for cardiac cephalgia¹

A. Demonstrated acute myocardial ischaemia	
B. Evidence of causation demonstrated by at least two of 1 – 4.	<ol style="list-style-type: none"> 1. Onset of headache is in temporal relation to the onset of myocardial ischaemia 2. Improvement, resolution or worsening of headache occurs in parallel with improvement, resolution or worsening of myocardial ischaemia 3. Presence of at least two of the following characteristics: <ul style="list-style-type: none"> • Moderate to severe intensity • Accompanied by nausea • Not accompanied by photophobia or phonophobia • Aggravated by exertion 4. Relieved by nitroglycerine or its derivatives
C. Not better accounted for by another IHCD-3 diagnosis.	

this patient was not known to have any of these features. Cardiac cephalgia presenting with headache not related to exertion has been reported.⁶ Most of the patients reported in literature are elderly and has been diagnosed and treated.⁶⁻⁸ Although cardiac cephalgia is still considered a hypothetical condition, this fatal case illustrates the need to consider cardiac cephalgia in any unexplained headache not responding to headache-specific treatment, even in the absence of red flag signs.

Conclusion

Cardiac cephalgia is a serious cause of secondary headache which can masquerade as a primary headache. Clinicians should consider cardiac cephalgia in any unexplained headache, even if the headache is unrelated to exertion, in younger age groups and even in the absence of a history of vascular risk factors. High index of suspicion, prompt diagnosis and treatment will help to reduce cardiac morbidity and death.

Conflicts of Interest

The author has no conflicts of interest to declare.

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