CASE REPORT

Cardiac cephalalgia: A deadly case report

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ABSTRACT

Cardiac cephalalgia is a nosologic entity that has only been acknowledged by the turn of the century, and is, consequently, often underdiagnosed, even by experienced neurologists. Unlike most headaches, however, failing to provide a proper and timely diagnosis can have deadly consequences. Report of a case of cardiac headache attended at the emergency department and literature review. This entity was first described in 1997; no studies have yet determined its prevalence, with the literature relying on case reports. The pathophysiology remains a mystery, with three main hypothesis: spinal convergence of cardiac visceral afferent nerves with somatic afferent nerves from the head, increase of intracranial pressure from decrease in cerebral venous return originated from the reduced cardiac output, and release of inflammatory markers during cardiac ischaemia, such as bradykinin, serotonin and histamin, causing vascular changes. Distinguishing this pathology from others, especially migraine, with which it shares many traits, is of paramount importance: vasoconstrictor drugs such as triptans are absolutely contraindicated, and the outcome can be dramatic. This case illustrates the need to promptly recognize this rare entity since failure to diagnose it can have devastating consequences.

Keywords: Cardiac Cephalalgia; Myocardial Ischemia; Cardiac Arrest.
INTRODUCTION

Cardiac cephalalgia is a nosologic entity that has only been acknowledged by the turn of the century, and is, consequently, often underdiagnosed, even by experienced neurologists. Unlike most headaches, however, failing to provide a proper and timely diagnosis can have deadly consequences.\(^1\)

CASE REPORT

We aim to report the case of ES, a 62 years old Caucasian male with no previous history of headache who went to the ER due to a aching, holocranial, and intense headache lasting over two weeks, with few moments of respite in the meantime, with nausea and emesis but no photo/phonophobia. He developed angina pectoris at the exact same time, and was subjected to a series of cardiac exams in the weeks before the pain, which appeared normal. His comorbidities included having been subjected to a kidney transplantation in 2004, being still in dialysis, cardiac pacemaker in 2012, as well as diabetes, heart failure and hypertension. His admission laboratory workup showed creatinine of 9.16, troponine of 1.95 and CK-MB of 24.24; six hours later, the exams showed an increase to 1.85 and 26.83, respectively. His EKG showed no ST-segment elevation.

The hypothesis of cardiac cephalalgia was raised and his care was transferred to the cardiology department. He was admitted to the Coronary Unit, received ASA and clopidogrel and the patient underwent a percutaneous intervention, which subsequently demonstrated critical lesions in anterior descending and right coronaries as well as thrombus in circumflex artery. After an emergency coronary artery bypass, he developed hyperkalemia and went into cardiac arrest, with unsuccessful reanimation attempts. He fulfilled criteria for cardiac cephalalgia, as the headache developed in close temporal relation to the ischaemia and had both moderate to severe intensity, nausea and absence of photophobia (Table 1).

DISCUSSION

This entity was first described in 1997\(^2\); no studies have yet determined its prevalence, with the literature relying on case reports.\(^3\) The pathophysiology remains a mystery, with three main hypothesis: spinal convergence of cardiac visceral afferent nerves with somatic afferent nerves from the head, increase of intracranial pressure from decrease in cerebral venous return originated from the reduced cardiac output, and release of inflammatory markers during cardiac ischaemia, such as bradykinin, serotonin and histamin, causing vascular changes.\(^4\) Distinguishing this pathology from others, especially migraine, with which it shares many traits, is of paramount importance: vasoconstrictor drugs such as triptans are absolutely contraindicated, and the outcome can be dramatic. In a review of seven cases, three had triple arterial lesion as well, but in all cases the patient survived.\(^4\)

CONCLUSION

This case illustrates the need to promptly recognize this rare entity since failure to diagnose it can have devastating consequences.

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**Table 1. Diagnostic criteria for Cardiac Cephalalgia.**

| International Headache Classification - 3rd edition | Part two - secondary headaches |
|---------------------------------------------------|-------------------------------|
| A. Any headache fulfilling criterion C.            |                               |
| B. Acute myocardial ischaemia has been demonstrated. |                               |
| C. Evidence of causation demonstrated by at least two of the following: |                               |
|   1. headache has developed in temporal relation to onset of acute myocardial ischaemia. |                               |
|   2. either or both of the following:              |                               |
|     a) headache has significantly worsened in parallel with worsening of the myocardial ischaemia. |                               |
|     b) headache has significantly improved or resolved in parallel with improvement in or resolution of the myocardial ischaemia. |                               |
|   3. headache has at least two of the following four characteristics: |                               |
|     a) moderate to severe intensity.               |                               |
|     b) accompanied by nausea.                     |                               |
|     c) not accompanied by photophobia or phonophobia. |                               |
|     d) aggravated by exertion.                     |                               |
|   4. headache is relieved by nitroglycerine or derivatives of it. |                               |
| D. Not better accounted for by another ICHD-3 diagnosis. |                               |
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