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Case Report

Cardiac cephalgia: A headache of the heart

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ABSTRACT

Atypical presentations of common diseases are often difficult to identify in time, nonetheless necessary, especially in cases of diseases like ischemic heart disease (IHD) which otherwise may progress into irreversible stage and ultimately, fatality, in the absence of timely administered medical treatment. We report a case of a 51-year-old male, presenting initially with only headache as the sole symptom, who later was diagnosed with severe coronary artery disease, and the symptoms resolved completely after coronary angioplasty with stenting.

<Learning objective: Atypical presentations, such as headache, can be more prevalent in ischemic heart disease (IHD) than was thought of, so awareness of the possibility of IHD while assessing headache and further study to see actually how commonly headache is associated with IHD are necessary.>

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Introduction

Classically, cardiac angina is well recognized as a precordial pain sensation with known radiation sites such as left upper limb, back, neck, jaw, and epigastrium. But pain may occur in abnormal sites too. Atypical presentations of common diseases are often difficult to identify in time, nonetheless necessary, especially in cases of diseases like ischemic heart disease (IHD) which otherwise may progress into irreversible stage and ultimately, fatality, in the absence of timely administered medical treatment. We report a case of a 51-year-old male, presenting initially with headache as the sole symptom, who later was diagnosed with having severe coronary artery disease, and the symptoms resolved completely after coronary angioplasty with stenting.

Case report

A 51-year-old Muslim male, hypertensive, non-diabetic, nonsmoker, with positive family history of ischemic heart disease

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that affected his siblings, presented in May 2013 with the complaints of post-stress, post-exertion headache which was self-limiting and episodic in nature. It used to originate in the pre-auricular area, spreading to forehead, and sometimes to vertex and occipital region. The headache had started two years previously in 2011, and initially used to last two to three minutes. The headache was not associated with aura, photophobia, watering of the eyes, nausea, vomiting, convulsion, or neck pain and was not related to change of posture. Later, as the severity of headache increased, he also started to experience mild chest tightness and sweating after a few moments after the start of headache. Repeated neurological examination failed to reveal any abnormal findings, and the extensive investigations which were done thinking the headache to be neurological in origin, also could not provide any clue for the cause of the headache. Magnetic resonance imaging (MRI) of the brain, performed in May 2011, showed only bilateral small old cerebral infarcts and carotid duplex study was normal.

Primarily he was diagnosed as having migraine by a neurologist. Propranolol, flunarizine, pizotifen, valporic acid, and nonsteroidal anti-inflammatory drugs (NSAIDs) were all tried as treatment, but his symptoms only deteriorated. Later, when he developed chest tightness, he was advised to undergo a treadmill

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test. During the test he again developed intense headache and mild chest discomfort with associated mild ST-T changes. The test was terminated and he was given sublingual nitrate, which cured the headache dramatically. After that, it was noted that every episode of headache could be relieved with sublingual nitrate.

In June 2011, a coronary angiogram was taken which revealed double vessel coronary artery disease [proximal left anterior descending artery (LAD) 70% and proximal left circumflex artery (LCX) 95% stenosis] as shown in Fig. 1. Percutaneous coronary intervention (PCI) with stenting was undertaken in the LAD and LCX at the same sitting, during which it was seen that each time when the percutaneous transluminal coronary angioplasty balloon was inflated at the diseased vessels, the patient complained of intense headache. After completion of the procedure, the patient was rendered totally free of symptoms (angiogram shown in Fig. 2).

In early 2013, he started having similar episodes of headache following exertion and was admitted again. His physical examination was normal. Routine blood test revealed dyslipidemia: total cholesterol – 195 mg/dL, low-density lipoprotein – 93 mg/dL, triglycerides – 472 mg/dL, high-density lipoprotein – 32 mg/dL. Electrocardiography and chest X-ray were normal. Echocardiogram showed no regional wall motion abnormality with good ejection fraction (66%). He underwent coronary angiogram again which revealed critical (90%) in-stent restenosis in proximal part of previously deployed LCX artery stent. Another intra-coronary stenting was done without any post-PCI complications. Before discharge, the patient was hemodynamically stable and completely symptom free.

Discussion

Headache is found in 6% of all cardiac ischemic cases [1]. According to the International Classification of Headache Disorders (ICHD), 3rd edition, cardiac cephalgia is a "migraine-like headache, usually but not always aggravated by exercise, occurring during an episode of myocardial ischemia. It is relieved by nitroglycerine" [2]. A set of diagnostic criteria has also been proposed:

- A. Any headache fulfilling criterion C.
- B. Acute myocardial ischemia has been demonstrated.

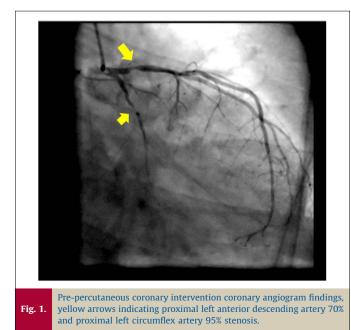


 Fig. 2.
 Post-percutaneous coronary intervention coronary angiogram findings.

- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to onset of acute myocardial ischemia
 - 2. either or both of the following:
 - (a) headache has significantly worsened in parallel with worsening of the myocardial ischemia
 - (b) headache has significantly improved or resolved in parallel with improvement in or resolution of the myocardial ischemia
 - 3. headache has at least two of the following four characteristics:
 - (a) moderate to severe intensity
 - (b) accompanied by nausea
 - (c) not accompanied by phototophobia 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.

Our case meets each of the mentioned criteria. This rare condition, cardiac cephalgia, is also called by several synonyms such as cardiac cephlalgia, angina capitis, angina cranialis, or simply as "headache angina" [3,4]. Not much literature is available, as the condition itself is rare and there is much deficit of awareness about this atypical presentation of ischemic heart disease. In 1971, Sampson reported in a group of 150 patients with angina pectoris, as much as 6% reported headache along with other symptoms [5], which is clearly not a negligible amount. In 2009, in a review including 30 case reports of cardiac cephalgia, Bini et al. found that in 27% of the cases, cardiac cephalgia was the only presentation of a cardiovascular ischemic event [3]. They also commented that cardiac cephalgia generally occurs after fifth decade of life, which matches our case. Prevalence of cardiac cephalgia in different sex groups is unknown. This form of headache is often confused with migraine for indistinguishable pattern of headache, but does not respond to NSAIDs [4,6], just like our case. Triptans are condraindicated [3], which is an important consideration during treatment, because triptans are often used for treatment of migraine. Vertebral artery dissection which also mimics migraine should be considered too, as a possible differential diagnosis [7]. The lack of other supportive clinical features such as insidiously onset headache, which is pulsating, persistent, or sometimes changing of location in nature exclude [8] vertebral artery dissection. Moreover, MRI of brain also did not show any sign of posterior ischemia. Cardiac cephalgia may occur again in event of coronary artery restenosis [9], also seen in our case.

There are four sets of theories [3] postulating the pathogenesis of headache angina:

- 1. Convergence theory: according to this theory, afferent somatic and visceral fibers converge on the same neurons, and stimulation of visceral afferents causes relaying of information corresponding to somatic region [4].
- 2. Cardiac ischemia causes reduction of cardiac output which results in elevated left ventricular and right atrial pressure, which again cause reduction of venous outflow of brain yielding to raise intracranial pressure and headache [10].
- 3. Cardiac ischemia causes release of neuromediators–serotonin, bradykinin, histamine, substance P, ANF (atrial natriuretic factor), which cause vasodilatation of the cerebral vessels and cause pain [11].
- 4. Cardiac cephalgia is caused by concurrent constriction of both cardiac and the cerebral vessels [12].

Although once considered rare, available literature suggests that headache may be present in significant number of IHD patients, and even may be the only presentation in some instances. Not considering this condition as a plausible differential diagnosis with enough importance may result in making misdiagnosis such as migraine, and prescribing the wrong treatment which will actually worsen the condition and cause severe suffering on part of the patient; and at the same time diagnosis of acute myocardial events will be delayed which can be fatal. After much discussion, it can be said that much awareness is needed among the concerned physicians, and further study is needed to see the prevalence of headache in IHD patients and to assess the true "atypicalness" of headache as a clinical feature of IHD.

Conflict of interest

None.

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