

Cardiac cephalalgia: First case from India

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Abstract

A 67-year-old male smoker had exertional headaches for 2 years. The headaches were holocephalic, very severe, excruciating, and occasionally accompanied by nausea. Physical examinations and neuroimaging were normal. Electrocardiogram (ECG) showed old infarct in inferior leads. Sublingual nitrate provided relief in headaches. Stress test was positive with recurrence of similar headaches with ECG changes suggestive of myocardial ischemia. Coronary angiogram revealed three-vessel disease. Coronary artery bypass surgery provided complete resolution of headaches.

Key Words

Angina, cardiac cephalalgia, coronary artery disease, exertional headache

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Ann Indian Acad Neurol 2016;19:252-254

Introduction

The clinical features of coronary artery disease (CAD) vary, and patients frequently present with symptoms other than chest pain.^[1] Patients with CAD who present without chest pain are frequently misdiagnosed and undertreated. These patients may have pain at other sites such as, arm, shoulder, back, jaw, or epigastrium. Pain at each of these sites may occur alone or in different combinations.^[1,2] Headache as the only symptom of myocardial ischemia is quite infrequent and there are only few case reports in the literature on cardiac cephalalgia (or cephalgia).^[3,4]

Case Report

A 67-year-old man presented with a 2-year history of episodic headaches. All attacks occurred only on exertion, initially after walking for about 10-15 min and later on after walking for just 1-2 min. The headaches were also provoked by some Valsalva-like activities such as lifting heavy objects and sexual activities. The headache was intense, excruciating in quality, and rated as 10 in severity on the visual analog scale. This intense and excruciating pain used to last for 10-60 min in each attack. His

routine and social activities were seriously hampered as he could not walk for more than 2 min.

The pain used to start posteriorly, becoming holocephalic within seconds. On a few occasions, the headache was associated with nausea. However, there was no association with vomiting, photophobia, phonophobia, or any aura. He never had any abdominal pain, chest discomfort (or atypical chest pain), jaw claudication, or breathlessness with the headaches.

He was a heavy chronic smoker and did not have diabetes, hypertension, or any vascular disease.

Physical and neurological examinations were normal including blood pressure and heart rate. Previous workup included routine hematological and biochemical investigations, all of which were normal. Erythrocyte sedimentation rate (ESR) was 26 mm/h. He had undergone magnetic resonance imaging (MRI) of the brain and cervical spine on many occasions, which did not reveal any abnormality. Prior treatments with various drugs such as, amitriptyline, sodium valproate, topiramate, flunirazine, paracetamol, indomethacin, and ibuprofen had no benefit.

A resting electrocardiogram (ECG) showed Q wave in D₂, D₃, and aVF leads, suggesting old inferior wall myocardial infarction. In view of exertional only headaches and ECG showing features of CAD, a possibility of cardiac cephalalgia was considered.

The patient got complete relief from headache after a single tablet of sublingual nitroglycerine in 2-3 min which he

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	DOI: 10.4103/0972-2327.165467

described as best response with any drug. Improvement with nitroglycerine raised a possibility of cardiac cephalgia. During a stress test, the patient had recurrence of similar headache, which was associated with 2 mm depression of ST segment in inferior leads. The patient did not feel any cardiac symptom during stress testing. Headache induced by stress test subsided by sublingual nitroglycerine. A coronary angiogram revealed three-vessel disease with 90% stenosis in right coronary artery [Figure 1], 75% stenosis in left anterior descending artery, and 70% stenosis in left circumflex artery [Figure 2]. Coronary artery bypass surgery was done at all sites of stenotic arteries. 2 weeks later following which the symptom of exertional headache completely disappeared.

Discussion

An exertional headache has broad differential diagnoses, which includes both primary and secondary headache disorders. Vast majority of exertional headaches are benign. The secondary causes associated with exertional headaches include space-occupying lesions (especially of posterior fossa), vascular abnormalities (aneurysm or arteriovenous malformation), Chiari malformation, and other obstructions of CSF flow.^[5] Cardiac cephalgia is an extremely rare type of exertional headache. There are about 32 cases of cardiac cephalgia in the literature. Knowledge of cardiac cephalgia and an early diagnosis is important to prevent any catastrophe.^[3,4]

A diagnosis of cardiac cephalgia is made according to International Classification of Headache Disorders (ICHD)-3 β criteria, which depend on the documentation of causation of headache by cardiac ischemia. Evidence of causation is suggested by the presence at least two of the following:

1. Headache developed in temporal relation to onset of acute myocardial ischemia;
2. Headache worsened with worsening of the myocardial ischemia or headache improvement with improvement in the myocardial ischemia;
3. At least two of the following four characteristics:
 - a. Moderate to severe intensity,
 - b. Accompanied by nausea,

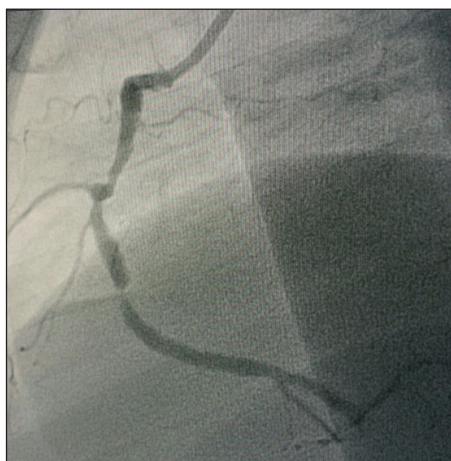


Figure 1: Coronary angiogram showing 90% stenosis in right coronary artery

- c. Not accompanied by photophobia or phonophobia, and
 - d. Aggravated by exertion; and
4. Headache is relieved by nitroglycerine or its derivatives. Our patient showed all four features.

Approximately 50% of cases of CAD are recognized because of the chest pain.^[6] However, it may arise in or radiate to the neck, jaws, tongue, teeth, throat, occiput, cheeks, tip of the nose, ears, shoulders, arms, hands, and to the epigastrium.^[1,2,6] The association of headache with myocardial infarction was noted in 1971 by Sampson and Cheitlin.^[7] They observed that about 6% patients with CAD had headache with chest pain. Headache as a presenting feature was first reported by Casky and Spierings in 1978.^[7,8] Thereafter, about 32 cases of cardiac cephalgia have been reported in the literature.^[3,4]

Our case presented with exertional headaches. Structural intracranial pathology was ruled out by neuroimaging. This case fulfilled the ICHD-3 criteria of cardiac cephalgia.^[9] Headache was always exertional and the patient had immediate symptomatic relief with sublingual nitrate. Moreover, he had complete relief by coronary bypass surgery suggesting that pain was cardiac in origin. To the best of our knowledge, this is the first case of cardiac cephalgia reported from India.

Most reported cases of cardiac cephalgia had headache as the main presenting feature.^[3,4]

However, only 27% patients reported headache as the only manifestation of cardiac ischemia.^[4] About 50% patients also had mild nonspecific pain in chest, epigastrium, arm, or mandible.^[4] Cardiac cephalgia may closely mimic migraine. Both cardiac cephalgia and migraine may produce severe exertional headache with autonomic features, particularly nausea. Up to 30% patients with cardiac cephalgia may have either nausea or vomiting.^[3,4] Our patient felt nausea on a few occasions, but he denied the presence of any other symptoms. It is extremely important to differentiate cardiac cephalgia with migraine as erroneous use of triptans may aggravate cardiac ischemia. Interestingly, nitroglycerine which induces migraine and other headaches provide relief in patients with cardiac cephalgia.^[4,10]



Figure 2: Coronary angiogram showing 75% stenosis in the left anterior descending artery and 70% stenosis in left circumflex artery

Majority of these patients have one or more risk factors for cardiovascular events, such as hypertension, diabetes, smoking, hyperlipidemia, and positive family history.^[3,4] Our patient was a heavy chronic smoker.

Cardiac pain is mediated by sympathetic and/or parasympathetic vagal fibers. These fibers converge to the somatic fibers of the various structures of the body. Anginal pain is mediated by sympathetic fibers from C8 to T5 in 50-60% of cases, by vagal fibers in 10-20%, and through both neural fibers in 30-40%.^[11] These variations in convergence of fibers are largely responsible for the variation of cardiac pain. If parasympathetic fibers are involved, the patients would get pain in the neck and head distribution. A recent observation on 326 patients with confirmed myocardial ischemia demonstrated increased prevalence of craniofacial pain with inferior wall ischemia.^[12] It is a well-known fact that symptoms of inferior wall ischemia are mediated by parasympathetic fibers of vagus nerve.

A few other hypotheses has also been suggested to explain the craniofacial pain in CAD. Elevations in intracranial pressure, release of neurochemical mediators, or spasm of cranial vasculatures are few other explanations for the headache in patients with CAD.^[3,4]

Unrecognized myocardial infarction is quite common in the elderly people. About 21-33% elderly men and 26-54% elderly female with myocardial infarction may escape clinical attention.^[13]

Therefore, it is possible that large number of patients with cardiac cephalgia remain undiagnosed. It can also be speculated that a subset of patient with myocardial ischemia who escape attention may have headache as a sole presentation.

Diagnose of cardiac cephalgia is very important. Review of the literature suggest mortality rate of about 12% for cardiac cephalgia.^[3] Therefore, it could be suggested that any elderly patient with the presence of cardiovascular risk factor having recent onset headache should be suspected for the presence of cardiac cephalgia. Review of the literature demonstrate that 57% patients with cardiac cephalgia had some pathological abnormalities in the baseline ECG trace.^[4] Therefore, a baseline ECG should be a must for elderly patients having headache and one of the risk factor for cardiovascular events.

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How to cite this article: Prakash S, Panchani N, Rathore C, Makwana P, Rathod M. Cardiac cephalgia: First case from India. *Ann Indian Acad Neurol* 2016;19:252-4.

Received: 26-02-15, **Revised:** 19-03-15, **Accepted:** 27-04-15

Source of Support: Nil, **Conflict of Interest:** None declared.