CASE REPORT

Cardiac Cephalalgia Attacks Provoked by Prinzmetal Angina - A Case Report

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Abstract

Cardiac cephalalgia is a headache occurring as a manifestation of myocardial ischemia and is relieved by nitroglycerine. Cardiac cephalgia should be suspected in elderly patients with risk factors for atherosclerotic disease presenting with recent-onset headache. We present a 51-year-old man with no past medical history of headache or risk factors for vascular diseases who was diagnosed with supraventricular tachycardia (SVT) in 2005. In 2014 SVT attacks were longer and always followed by severe headache attacks. Accompanying symptoms were dizziness, non-rotational vertigo, confusion, episodes of double vision and tinnitus; these symptoms would last one day, followed by severe tiredness for 2 days. Neurologic and cardiologic evaluation as well as extensive diagnostic test between the attacks was normal. Catheter coronarography showed no stenoses but did show spasms in distal left anterior descending artery. The diagnosis of cardiac cephalalgia that manifested as reversible coronary vasospasm (Prinzmetal angina) has been established. A remarkable effect on SVT and headaches was shown after the introducement of diltiazem. Our case showed that cardiac cephalgia should also be suspected in middle aged patients with no past medical history of headache, and even no presence of risk factors for vascular disorders. CC should be distinguished from migraine, to avoid prescribing triptans and vasoconstrictors.

1. Introduction

Cardiac cephalalgia (CC) is a headache occurring as a manifestation of myocardial ischemia, and has been recognized as a distinct entity since 1997 [1] and included into the International Classification of Headache Disorders- 3 criteria (ICHD-3) version under Headache attributed to disorder of homeostasis as 10.6.[2]. Although 40 cases of CC have been reported since, the mechanism is still not understood.[3-6]

The clinical presentations of CC can be highly variable and the most consistent feature is a severe headache. The headache usually presents along with typical symptoms of angina pectoris; it may occasionally present as exertional headache without chest symptoms.[7,8] Within this headache subtype there is a high variability of clinical manifestations, so CC is probably not recognized in a number of cases and thus underdiagnosed.[6,9] Differential diagnosis with migraine is crucial to avoid the administration of vasoconstrictors.

We report a case of CC that manifested as reversible coronary vasospasm and review reported cases from the literature to highlight the importance of this condition in the differential diagnosis of a headache in a patient with a history of cardiovascular disease.

2. Case report

2.1. A 51-year-old man presented to the outpatient headache clinic in March 2017 with a three-year history of headache. In 2005 the patient began to experience episodes of a pressing feeling in the chest and was diagnosed with supraventricular tachycardia (SVT). The patient smoked from his teenage years to the age of 30, did not drink alcohol, had a normal weight, normal cholesterol values and was normotensive. In 2014 he experienced short-lasting heart palpitations followed by mild pressing headache (on visual analogue scale VAS 2-3) with no accompanying features, lasting a couple of hours, worsening under physical activity and with no need for medication. His headache attacks were always associated with the subjective sensation of palpitation and chest tightness, equivalent with SVT on electrocardiogram ECG.

In February 2017 the patient experienced episodes with SVT > 30 seconds, followed by severe headache attacks VAS 10, localized bilaterally but with greater intensity on the left side. Accompanying symptoms were dizziness, non-rotational vertigo, confusion, short episodes of double vision and tinnitus. These symptoms would last one day. In the subsequent 1-2 days, the patient was exhausted and was not able to perform daily activities. The frequency of these attacks was three per month. For the period leading up to November 2017 the headache attacks presented only during the daytime, thereafter the attacks also occurred at night, and on some occasions were accompanied by numbness in the arms. Since January 2019 the patient experienced almost daily attacks of angina, relieved by nitroglycerin.

2.2. Diagnostic work-up*:* Neurologic evaluation was normal between the attacks. Brain MRI, cerebrospinal fluid examination with pressure measurement, CT angiography of the neck and cerebral arteries, transthoracic echocardiography, daily blood pressure monitoring, tilt-test, CT scanning of the heart were all normal. Heart echocardiography showed EF 60%. Holter monitoring revealed sinus rhythm, with supraventricular extrasystole and SVT. An internal Loop Recorder (Medtronic

Linq) was implanted subcutaneously on the thorax to monitor the heart. Since January 2019 the patient was hospitalized several times due to chest pain; blood pressure ECG and blood tests including troponin were always normal. In January 2019 a catheter coronarography showed no stenoses but did show spasms in distal left anterior descending artery (LAD) and the patient was diagnosed with Prinz metal angina. Repeated catheter coronarography in August 2019 showed remarkable systolic "myocardic bridging" distally in LAD, but no stenosis was seen during systoles. Under the coronography the patient experienced spasms in LAD with subsequent ST elevation.

2.3. Medication: The patient´s daily medication included nitroglycerine spray, isosorbidmononitrat 60 mg, and atorvastatin. Medications such as indomethacin 150 mg, candesartan 32 mg, verapamil 480 mg had no effect on the headaches. When diltiazem 350 mg (calcium channel blocker) was introduced, it showed a remarkable (more than 50%) effect on SVT and headaches. The patient experienced severe headache attacks lasting 1-2 hours approximately 2 times per week, requiring no additional treatment for headache.

The patient has given informed consent for publishing the case report.

3. Discussion

According to the ICHD-3 criteria (2), CC is characterized as a migraine-like headache, usually, but not always aggravated by exercise, occurring during an episode of myocardial ischemia. Diagnostic criteria for CC are shown in Table 1.

Our patient fulfills the diagnostic criteria for CC: the headache is of severe intensity and has developed in temporal relation to the onset of acute myocardial ischemia, is aggravated by exercise, is not accompanied by phototophia or phonophobia, and the headache is relieved by nitroglycerine. The patient has been diagnosed with Prinz metal angina, coronarography has shown spasms in LAD with subsequent ST elevations. Upon introduction of diltiazem 350 mg in treatment, his cardiac manifestations, as well as headaches improved significantly.

Ischemic heart disease is a common cause of morbidity and mortality worldwide. Patients typically present with chest pain and breathlessness provoked by exertion or at rest. In several published cases, headache (extertional) could be the only manifestation of a myocardial infarction or angina pectoris, although this occurs very rarely.[7,8] Cardiac cephalgia manifests without a specific pattern of clinical features, it can present in a form of migraine, sometimes even accompanied by autonomic symptoms, or in a form of tension-type headache.[10] Pain location and accompanying symptoms are highly variable; however, the most consistent feature is severe headache intensity. The development of headache is usually subacute, but can present as a thunderclap headache.[11]

The recognition of myocardial ischemia as the cause of headache is important in clinical practice as not recognizing CC can have serious consequences. CC should be especially distinguished from migraine, in order to avoid prescribing medications such as triptans and other vasoconstrictors.[4] Both disorders can produce severe head pain accompanied by nausea, and both can be triggered by exertion. The difference is that migraine-like headache may be triggered by medications used for angina treatments such as nitroglycerine, and on the

contrary, headache due to CC is relieved after the application of nitroglycerine.

CC should be considered as one of the differential diagnoses in a new-onset, migraine-like headache, especially exertional headache when the patient is elderly, with a history of ischemic cardiopathy or has cardiovascular risk factors. CC should also be suspected even in patients when the chest pain is absent, and if the headache improves with nitrates.

However, our patient was middle aged, with earlier smoking history as the only risk factor, but had developed Prinz metal angina and CC.

In the cases where coronary angiography reveals occlusive disease, patients subsequently usually undergo coronary angioplasty, and their cardiac headaches resolve. [7,12,13]. However, our patient suffered from Prinzmetal angina with no indication for coronary angioplasty. Furthermore, his headaches and related symptoms could be confusing as he experienced confusion, tinnitus and vision difficulties as accompanying symptoms. After introduction of diltiazem, all his symptoms significantly improved. To the best of our knowledge, there is only one previously reported case (a 44 year old woman) where the coronary angiogram revealed reversible coronary vasospasm, without coronary occlusive disease.[14]

Knowledge of the pathophysiology of cardiac cephalgia is scarce, due to its rare clinical occurrence and variable clinical presentation. Headache developing in close relationship with symptoms of cardiac ischemia is highly suspectable of CC. In patients with known ischemic cardiopathy, its diagnosis depends on the presence of severe headache that is accompanied by nausea, worsened by physical exercise, and only ceases with nitrate administration. In some patients routine examinations such as cardiac enzymes, ECG and even exercise stress test can be negative. In such cases, only a coronary angiogram can provide evidence for diagnosis. Therefore, if the headache (especially exertional) occurs as the only manifestation of an acute coronary event, CC should be suspected in elderly patients with no past medical history of headache but with presence of risk factors for vascular disorders.

Our case showed that CC should also be suspected in even in middle aged patients with no past medical history of headache, and even no presence of risk factors for vascular disorders.

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Supplement 1: Diagnostic criteria for cardiac cephalalgia

A. Any criteria fulfilling criterion C
B. Acute myocardial ischemia has been demonstrated
C. Evidence of causation demonstrated by at least two of the following:
   1. Headache has developed in temporal relation to the onset of 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 phototophia or phonophobia
      d) Aggravated by exertion and
   4. Headache is relieved by nitroglycerine or derivatives of it
   5. Not better accounted for by another ICHD-3 diagnosis

