Cardiac Cephalalgia Presented with a Thunderclap Headache and An isolated Exertional Headache: Report of 2 Cases

Pei-Yu Cheng[1], Hiu-Ngar Sy[2], Wei-Liang Chen[3], Yen-Yu Chen[2,4,5]

Abstract- Headache could be the only manifestation of a myocardial infarction or angina pectoris. The recognition of myocardial ischemia as the cause of headache is important in clinical practice. We report two cases of cardiac cephalalgia, defined as headache attributed to myocardial ischemia. The first patient presented with a thunderclap headache probably secondary to a myocardial ischemia and the second patient presented with isolated headaches secondary to angina pectoris triggered by exertions. The clinical presentations of cardiac cephalalgia are highly variable and the most consistent feature is severe in intensity. Cardiac cephalalgia should be considered one of the differential diagnoses of exertional headache and thunderclap headache when the patient is older or has cardiovascular risk factors.

Key Words: Cardiac cephalalgia, Cardiac cephalgia, Exertional headache, Thunderclap headache, Angina headache, Myocardial ischemia

Acta Neurol Taiwan 2010;19:57-61

INTRODUCTION

Headache is an uncommon presentation of myocardial ischemia. In 1971, Sampson et al.[1] reported that only 6% of 150 patients with angina pectoris complained of headaches and these were not the sole symptoms. In 1997, Lipton et al.[2] described 2 patients with myocardial ischemia presented with exertional headache and proposed the term "cardiac cephalgia". The second edition of the International Classification of Headache Disorders (ICHD-II, 2004[2]) includes the diagnostic criteria of "cardiac cephalalgia", coded 10.6 and classified under the group of headache attributed to the disorder of homeostasis (Table 1).

It is usually not difficult to diagnose myocardial ischemia if typical chest symptoms are apparent. However, when the chest symptoms such as oppression, tightness, or pain of chest, or diaphoresis are lacking, the diagnosis of headache secondary to a cardiac origin is usually difficult and problematic. With the increasing number of the relevant literatures, it is considered that cardiac cephalalgia has a great diversity of clinical presentations[3].

In this article, we report two additional cases of car-

From the [1]Division of Endocrinology and Metabolism, Department of Internal Medicine, [2]Department of Neurology, and [3]Department of Radiology, Changhua Christian Hospital, Changhua, Taiwan; [4]Department of Neurology, Taipei Veterans General Hospital, Taipei, Taiwan; [5]Institute of Clinical Medicine, National Yang-Ming University, Taipei, Taiwan.
Received July 3, 2009. Revised July 24, 2009.
Accepted September 7, 2009.

Reprint requests and correspondence to: Yen-Yu Chen, MD. Department of Neurology, Changhua Christian Hospital, Changhua, Taiwan, No. 135, Nansiao St., Changhua City, Changhua County 500, Taiwan.
E-mail: yychen@mail2000.com.tw

Table 1. *The ICHD-II diagnostic criteria for cardiac cephalalgia, coded 10.6*

A. Headache, which may be severe, aggravated by exertion and accompanied by nausea and fulfilling criteria C and D
B. Acute myocardial ischaemia has occurred
C. Headache develops concomitantly with acute myocardial ischaemia
D. Headache resolves and does not recur after effective medical therapy for myocardial ischaemia or coronary revascularization

ICHD-II: The second edition of the International Classification of Headache Disorders[2]

diac cephalalgia who presented with a thunderclap headache and an isolated exertional headache, respectively.

CASE REPORT

Patient 1

A 52-year-old woman was referred to this hospital because of a sudden onset of chest pain with radiation to neck accompanied by a severe throbbing headache immediately after local anesthesia when she was receiving a plastic eyelid surgery at another hospital. The 12-lead electrocardiography (ECG) showed an quivocal ischemic pattern. However, the levels of troponin-I (1.70 ng/mL, reference: <0.04 ng/mL) and creatine kinase-MB (7.5 ng/mL, reference: 0.6-6.3 ng/mL) were elevated. An acute coronary syndrome was suspected. Coronary angiography did not showed a stenotic lesion and an ergonovine provocation test also showed a negative result. The echocardiogram showed a regional wall motion abnormality with a preserved left ventricular systolic function. Myocardial ischemia secondary to coronary spasm was considered because of having chest pain and elevation of cardiac enzymes. She was treated with nitroglycerin. Her chest pains and headaches gradually decreased from the second hospital day but had not completely resolved on the day 3.

A neurologist was consulted. The headaches had an abrupt onset, with maximum intensity at the onset, and simultaneously associated with her chest pain. The headaches were severe, bilateral, and throbbing in nature, and accompanied by nausea but no vomiting. Physical activity or exertion aggravated her headaches as well. Thunderclap headache secondary to myocardial ischemia was considered. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) of the brain were performed to exclude possible serious intracranial lesions such as an aneurysm or cerebral vasoconstriction. A suspicious small acute infarct in the left superior frontal gyrus was noted (Fig A). Neither aneurysm nor definite lumen narrowing of major cerebral vessels was found.

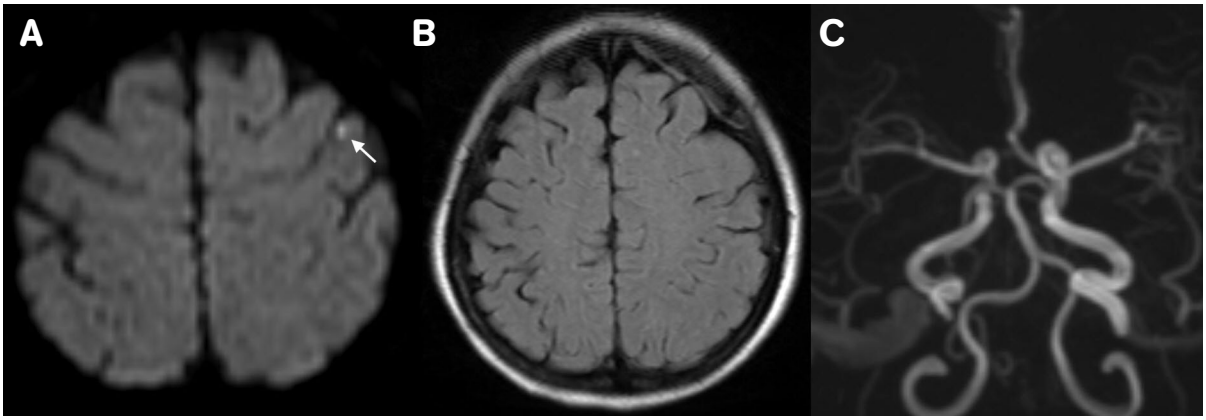

Figure. (A) The diffusion weighted imaging (DWI) showed a bright spot in the left superior frontal gyrus (arrow). (B) The Fluid Attenuated Inversion Recovery (FLAIR) did not show a corresponding signal change. (C) The magnetic resonance angiography (MRA) did not show segmental vasoconstriction.

Because of simultaneous occurrence of severe headache and myocardial ischemia, cardiac cephalalgia was diagnosed. Nimodipine (30mg every 4 hours) was administered via oral route since the third hospital day3 for her thunderclap headache. She reported no more headaches since the fourth hospital day4 and she remained headache free inat the follow-up after discharge.

Patient 2

A 67-year-old woman came to the neurology clinic of this hospital because of numerous headaches for 2 months. She described the headaches were severe in intensity, throbbing in characteristics, and consistently starting from her jaw, then extending up to bilateral mandibles and temporoparietal regions. They were always immediately triggered by exertion such as a walk or physical activity and relieved within 5 minutes after a rest. No nausea or vomiting was noted. She had no baseline headache between attacks.

She had a historiesy of hypertension, type 2 diabetes and hyperlipidemia and was treated at another hospital. Her medicines included bisoprolol 5mg a day, atorvastatin 10mg a day and glibenclamide 5mg a day. Her body mass index was 32 (Kg/m^2).

Presence of multiple vascular risk factors raised a the suspicion of cardiac cephalalgia. Asking about chest symptoms in detail, she reported exertional dyspnea, but there was no chest tightness, chest pain or cold sweating. Because the index of suspicion for myocardial ischemia was high, she was referred to a cardiologist immediately. A brain CT was performed at the same time because posterior fossa lesion should be excluded for a patient with exertional headaches. Brain CT scan showed no significant structural lesion. ECG did not show an ischemic pattern. Aspirin, isosorbide-5-mononitrate and sublingual nitroglycerin were prescribed.

Aspirin and isosorbide-5-mononitrate provided modest benefits for her symptoms (a decrease in headache frequency) while sublingual nitroglycerin relieved each exertional headache sooner (within 2 minutes) than just taking a rest. Therefore, at another hospital she received a coronary angiography at another hospital. About 3 weeks after the procedure, she returned to the neurology clinic and told that she had 2-vessel coronary occlusive disease and single stent was implanted successfully. Her exertional headache completely resolved after the successful coronary angioplasty.

DISCUSSION

The first case we reported in this article presented with a thunderclap headache with a concomitant severe chest pain. A myocardial ischemia is suspected straightforward. However, making a diagnosis of headache secondary to a myocardial ischemia could be problematic. Previously, reported cases of thunderclap cardiac cephalalgia had no chest symptoms at the onset of headache). In these situations, the diagnosis would be more difficult[4-5].

The thunderclap headache is an acute and severe headache that is maximum in intensity at the onset[6]. There are many potential causes of such headaches including subarachnoid hemorrhage, reversible cerebral vasoconstriction syndrome, arterial dissection, pituitary apoplexy and intracerebral hemorrhage[2,6]. Therefore, it is mandatory to perform appropriate and full investigations for every patient with thunderclap headache[2]. Myocardial infarction has not been considered as a differential diagnosis of secondary causes of thunderclap headaches[2,6]. When the thunderclap headache is the sole presentation of a myocardial infarction, the diagnosis could be a great challenge because a routine neuroimaging survey could be negative result and a primary thunderclap headache could be erroneously diagnosed.

Diffusion weighted imaging (DWI) of the brain in the first patient we reported showed a bright up spot suggestive of a focal cerebral ischemia. Small emboli, cerebral vasoconstriction, or vasculitis are possible causes. Although the MRA (Fig. C) did not show segmental vasoconstriction in major cerebral vessels, small-sized vessel spasm could not be completely excluded. The administration of nimodipine is based on the report of successful treating patient with primary thunderclap headache[7]. The patient's headache and chest pain improved with initial nitroglycerin treatment. This sup-

ported the diagnosis of cardiac cephalalgia. Furthermore, her headache improved more after additional nimodipine treatment. This suggested that nimodipine might be beneficial in relieving the thunderclap headache of this patient.

In contrast to the first case, the second patient had exertional headaches as the sole presentations of her angina pectoris. The headaches differed from primary exertional headaches in several aspects. First, the headaches of this patient started from her jaw and extended to bilateral mandibles, not typically seen in primary exertional headaches[2,8]. Second, the age of this patient is older than the average age of patients with primary exertional headaches[8]. Third, the duration of headaches in this patient is short, almost immediately relieved with rest whereas the duration of primary exertional headaches is usually longer, about 5 minutes to 48 hours[2].

On the first occurrence of exertional headache, subarachnoid hemorrhage and arterial dissection should be excluded[2]. Similar to the thunderclap headache, exertional headache secondary to a cardiac origin is hard to be recognized. Because myocardial ischemia is a serious and potentially fatal illness, early recognition and appropriate treatment are important. Therefore, myocardial ischemia should be added to the differential diagnosies of the secondary causes of exertional headaches.

The clinical presentations of cardiac cephalalgia are highly variable among patients of previous reports. Headache may be the only manifestation in 27% of cases[3]. It usually occurs after exertion, but 33% of the patients appeared at rest[3]. Nausea, the associated symptoms of cardiac cephalalgia defined in ICHD-II, was found the least frequent presentation (27%)[9]. The most consistent feature is severe in intensity[3,9]. The inhomogeneous features and the rarity of this headache syndrome frequently lead to a diagnostic difficulty and delay treatments.

Although headache as the sole presentations of angina pectoris or myocardial infarctions has been increasingly recognized and reported[4,10-15], it is still difficult to be recognized in clinical practice. The clinical clues for suspicion include (1) headache pattern as exertional or thunderclap headache; (2) older age; and (3) having cardiovascular risk factors. It is essential for physicians, neurologists, cardiologists, headache specialists, and doctors of the emergency department to be alert when facing these patients.

The pathogenesis of cardiac cephalalgia remains unclear and has been thoroughly reviewed by Bini. et al.[3]. There were four proposed theories. In brief, the first theory is based on the convergence of autonomic sensory fibers and trigeminal or spinal somatic fibers[3,14]. The second theory is based on a reduced cardiac output associated with myocardial ischemia that results in a reduction of the cerebral venous return and subsequently an increase in intracranial pressure[3,14]. The third theory is based on the release of neurochemical mediators after myocardial ischemia resulting in vasodilatation and headache[3,14]. The fourth theory assumes those vasospasms occur in both coronary and cerebral vessels. Further studies focusing on the pathogenesis are needed for the better understanding of this rare headache syndrome[3].

In conclusion, cardiac cephalalgia could be a secondary cause of exertional and thunderclap headache and should be considered one of the differential diagnoses when the patient is older or having cardiovascular risk factors, even without chest symptoms.

REFERENCES

1. Sampson JJ, Cheitlin MD. Pathophysiology and differential diagnosis of cardiac pain. Prog Cardiovasc Dis 1971; 13:507-31.
2. Headache classification subcommittee of the International Headache Society. The International Classification of Headache Disorders 2nd Edition Cephalalgia 2004;24 Suppl 1:1-160.
3. Bini A, Evangelista A, Castellini P, et al. Cardiac cephalgia. J Headache Pain 2009;10:3-9.
4. Broner S, Lay C, Newman L, et al. Thunderclap headache as the presenting symptom of myocardial infarction. Headache 2007;47:724-5.
5. Dalzell JR, Jackson CE, Robertson KE, et al. A case of the heart ruling the head: acute myocardial infarction present-

ing with thunderclap headache. Resuscitation 2009;80:608-9.
6. Schwedt TJ, Matharu MS, Dodick DW. Thunderclap headache. Lancet Neurol 2006;5:621-31.
7. Lu SR, Liao YC, Fuh JL, et al. Nimodipine for treatment of primary thunderclap headache. Neurology 2004;62:1414-6.
8. Pascual J, Iglesias F, Oterino A, et al. Cough, exertional, and sexual headaches: an analysis of 72 benign and symptomatic cases. Neurology 1996;46:1520-4.
9. Chen SP, Fuh JL, Yu WC, et al. Cardiac cephalalgia. Case report and review of the literature with new ICHD-II criteria revisited. Eur Neurol 2004;51:221-6.
10. Auer J, Berent R, Lassnig E, et al. Headache as a manifestation of fatal myocardial infarction. Neurol Sci 2001;22: 395-7.
11. Seow VK, Chong CF, Wang TL, et al. Severe explosive headache: a sole presentation of acute myocardial infarction in a young man. Am J Emerg Med 2007;25:250-1.
12. Gutierrez Morlote J, Fernandez Garcia JM, Timiraos Fernandez JJ, et al. Cardiac cephalgia: an underdiagnosed condition? Rev Esp Cardiol 2005;58:1476-8.
13. Martinez HR, Rangel-Guerra RA, Cantu-Martinez L, et al. Cardiac headache: hemicranial cephalalgia as the sole manifestation of coronary ischemia. Headache 2002;42:1029-32.
14. Lipton RB, Lowenkopf T, Bajwa ZH, et al. Cardiac cephalgia: a treatable form of exertional headache. Neurology 1997;49:813-6.
15. Blacky RA, Rittelmeyer JT, Wallace MR. Headache angina. Am J Cardiol 1987;60:730.