Editorial: The first step of diagnosis is to know of the disease and question its presence, lessons from cardiac cephalalgia

Cardiac cephalalgia is one of the symptoms of ischemic heart disease expressed as a headache at the time of the onset of myocardial ischemia. The term, cardiac cephalalgia, was first reported as a separate clinical entity in 1997 [1]. One of the typical symptoms of ischemic heart disease is chest pain, and 6% of patients with angina complain of headache as a symptom [2], which is often accompanied by chest pain. On the other hand, in cardiac cephalalgia, headache is the only primary symptom with no accompanying chest pain. This condition is so rare that many cardiologists are assuming to have never encountered it. Moreover, nitrates and derivatives used to treat angina may cause headache as a side effect, which makes the diagnosis difficult in some cases [3].

A typical case of cardiac cephalalgia was reported by Chowdhury et al. in Journal of Cardiology Cases [4]. The case report was of a 51-year-old male patient who experienced headache as his only initial symptom, and thus a definitive diagnosis was difficult to make. As the symptom progressed, he started to experience chest discomfort, eventually leading to a suspicion of ischemic heart disease. Coronary angiography revealed coronary stenosis, and the diagnosis of coronary artery disease was confirmed. Sublingual administration of nitroglycerin was reported to relieve the headache. Symptoms including headache disappeared after successful revascularization with stent placement. The patient had a subsequent recurrence of headache, and coronary angiography identified in-stent restenosis. In this case, the onset of myocardial ischemia and the development of headache as well as the resolution of ischemia and the disappearance of headache are demonstrated in the same patient with repeatability. Although there are many case reports of cardiac cephalalgia, this is the most impressive case to show the relationship between myocardial ischemia and headache in such a compelling way.

Bini et al. conducted a systematic review of cardiac cephalalgia in 2009 [5]. This is the most comprehensive and detailed description of the concept of the disease to date. The review closely investigated 30 cases presented in 26 case reports that were searched using PubMed. Interestingly, of these 26 case reports, 17 (65%) were published in journals in the fields of neurology and neurosurgery, and only 5 (19%) were published in journals in the cardiovascular field. It is understandable that patients who report only headache do not visit cardiologists, but instead visit their primary care physician first most of the time. Even if the primary care physician refers the patients to a specialist for headache with unknown cause, they are usually referred to a neurologist or a neurosurgeon. In the published cases, while the neurologist or neurosurgeon was having a hard time making a diagnosis, the case was found to be ischemic heart disease with headache being its only clinical symptom. This was probably a surprise to the specialists, which led them to describe the cases in the reports. Meanwhile, in “the second edition of the International Classification of Headache Disorders (ICHD-II, 2004),” which is known as a classification of causes of headache, cardiac cephalalgia is classified as one of the secondary headaches in the group of “Headache attributed to disorder of homoeostasis” [6]. ICHD-II, 2004 is a classification standard made by physicians in the neurology field with no involvement of cardiologists, which indicates a problem, that awareness of cardiac cephalalgia remains low among cardiologists. In addition, a case was reported in which a delayed diagnosis of cardiac cephalalgia led to a fatal course [7]. It is necessary that cardiologists increase awareness of cardiac cephalalgia and try to disseminate its concept to family doctors and physicians in the neurology field around them, because the first step of diagnosis is to know the existence of the disease and question its presence.

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