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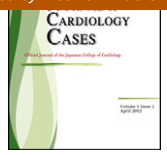

Case Report

A case of Prinzmetal angina diagnosed by Holter monitoring who survived a sudden cardiac death: Case report

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ABSTRACT

A 47-year-old female patient was admitted to our hospital after a syncope. She reported episodes of angina in previous weeks. On admission, there were no electrocardiographic changes but elevated troponin. Coronary angiogram showed minimal arteriosclerosis and normal left ventricle. Holter monitoring showed severe ST-segment changes during an anginal episode.

With calcium antagonists, the patient experienced no further episodes of angina or ST changes during telemetry.

Six weeks later, calcium antagonist was stopped for unknown reason. After that, the patient experienced a second prolonged syncope with cardiopulmonary resuscitation and defibrillation of ventricular fibrillation.

Discussion: "A variant form of angina pectoris" was first described by Myron Prinzmetal. He postulated coronary vasospasm as the underlying cause, however, after 50 years the exact pathophysiology is still not known.

Patients with "variant angina" usually present with "spontaneous" attacks of typical retrosternal anginal pain during rest or normal activities, but not with physical exercise.

Sudden cardiac deaths were reported in patients with Prinzmetal angina in only a few case reports.

Conclusion: In cases of variant angina accompanied by syncope, a provocation test and an electrophysiological study should be considered.

A 47-year-old female patient was admitted to our hospital after a first syncope while sitting in a bank. She had experienced chest pain and dyspnea before the syncope. She reported recurrent episodes of chest pain in the previous 2 weeks (about 1 episode/day). On admission, there were no electrocardiographic changes (Fig. 1) but the laboratory values showed elevated troponin level, therefore the decision to do an immediate invasive diagnosis was taken.

The coronary angiography showed only minimal, insignificant coronary atherosclerosis (Fig. 2), the left ventricular function was normal. Cardiac magnetic resonance imaging excluded myocarditis or a small (probably embolic) myocardial infarction.

Pulmonary embolism and aortic dissection were excluded by computed tomography scan.

While on the ward, the patient was still complaining of several daily episodes of anginal pain, however, exercise test showed no evidence of myocardial ischemia. Blood pressure and heart rate were also normal under exercise conditions.

The patient was put on a Holter monitor to exclude arrhythmias as a cause of syncope. An episode of anginal pain was accompanied by normal sinus rhythm, but surprisingly severe ST-segment changes were seen (Fig. 3).

Calcium antagonist therapy (amlodipine 5 mg bid) was started, and the patient was monitored by telemetry. Over the following 72 h, the patient experienced no further episodes of anginal pain or ST-segment changes.

The patient was discharged with the diagnosis of variant "Prinzmetal" angina. Syncope was assumed to be most probably due to a vasovagal reaction, atrioventricular block, or ventricular arrhythmia during an anginal episode.

Six weeks later, medication was changed by her physician for an unknown reason, calcium antagonist therapy was stopped and angiotensin-converting enzyme inhibitor therapy was initiated. Another two weeks later, the patient experienced a second, now prolonged syncope with cardiopulmonary resuscitation and defibrillation of ventricular fibrillation.

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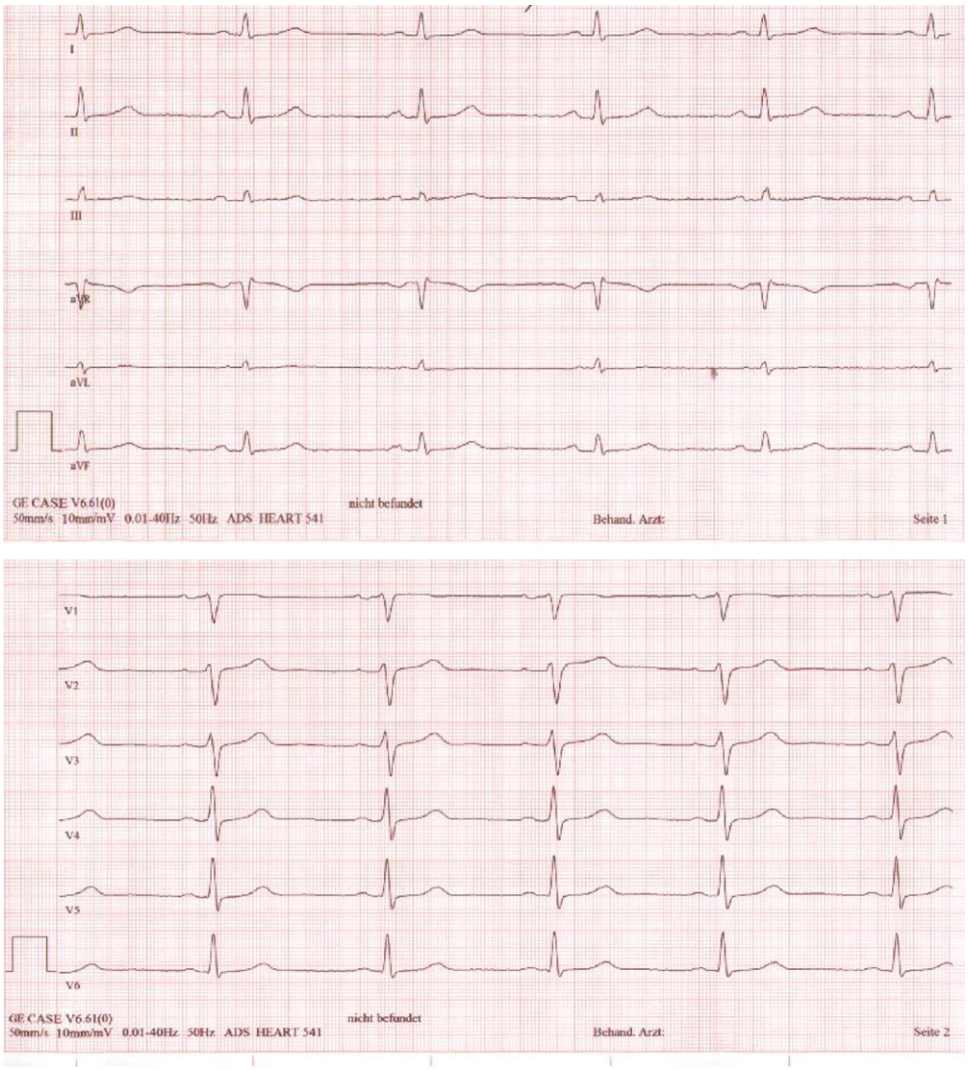

Fig. 1. Resting electrocardiogram showing normal ST-segment.

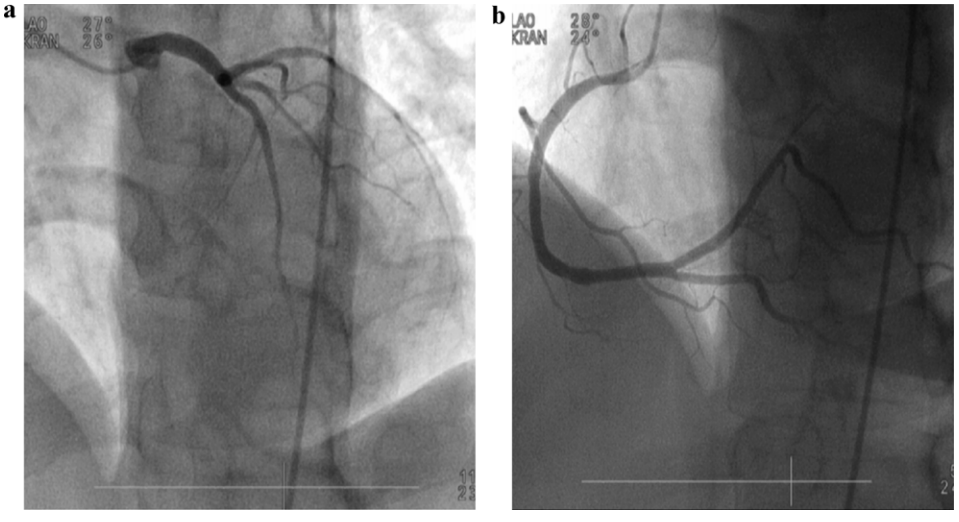

Fig. 2. (a) Left coronary artery in coronary angiogram. (b) Right coronary artery in coronary angiogram.

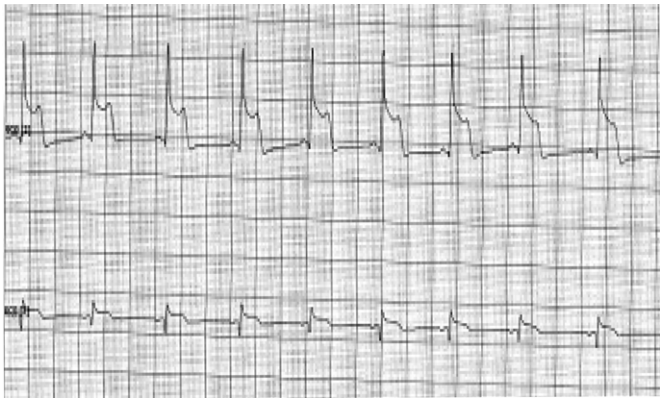

Fig. 3. Holter monitor showing ST-segment elevation.

She was admitted to our hospital, the calcium antagonist therapy was restarted, and an implantable cardioverter-defibrillator (ICD) was implanted. On discharge, the patient was free of angina symptoms and had no neurological deficits.

Discussion

"A variant form of angina pectoris" was first described by Myron Prinzmetal [1]. In his report, he illustrated 3 cases and summarized about 30 cases from the literature. He postulated coronary vasospasm as the underlying cause, however, even more than 50 years after the first report, the exact pathophysiology of this disease is still not fully known.

Risk factors for the development of "variant angina" include smoking, polymorphism of endothelial nitric oxide synthetase (eNOS), and low-grade inflammation [2,3]. In most patients with "variant angina", endothelial dysfunction can be found due to reduced bioavailability of nitric oxide [2]. In animal models, vasospasm usually affects coronary artery segments with no or only mild atherosclerosis [4].

Experimental studies indicate that the gene RhoA and its down stream effector ROCK/Rho-kinase (Rho-associated, coiled-coil containing protein kinase 1) are associated with regulation of eNOS activity and coronary artery smooth muscle hypercontraction [5].

Patients with "variant angina" usually present with "spontaneous" attacks of typical retrosternal anginal pain during rest or normal daily activities which cannot be induced by physical exercise. Sometimes angina symptoms are accompanied by palpitations.

Although intracoronary acetylcholine or hyperventilation are considered to have a major role in diagnosis of vasospastic angina [6], it was not performed in this patient, because we believed we already reached a diagnosis through the Holter monitoring, and appropriate therapy was started. Diltiazem is known to be effective in vasospastic angina, however, amlodipine is non-inferior [7].

Sudden cardiac death was reported in patients with Prinzmetal angina in only a few case reports [8,9], mainly due to ventricular arrhythmias or atrioventricular block during episodes of angina pectoris. However, current guidelines do not discuss or recommend implantation of ICDs or event recorders in patients with Prinzmetal angina and/or first syncope [11]. An electrophysiological study was not performed because the most likely trigger for the arrhythmia was myocardial ischemia, the left ventricular function was normal and there was no evidence of left ventricular scar. Therefore, the likelihood of inducible tachycardia was deemed low.

Takagi et al. discussed the importance of dual induction test in 10 patients who survived out-of-hospital cardiac arrest and found that all patients were positive for at least one test (acetylcholine provocation or electrophysiological study) [10].

Diagnosis in these patients is usually made by clinical symptoms, incident detection of electrocardiographic changes during an anginal episode, (invasive) provocation test showing coronary vasospasm, or a combination of the above with Holter monitoring reported in a few case reports [8,9], and not being used routinely in the diagnostic work-up of these patients [11]. However, the present case shows that (telemetric) Holter monitoring, especially with the possibility of additional ST-segment analysis, is useful in diagnosing these patients.

According to current guidelines [12], an ICD was implanted in our patient after she survived sudden cardiac death, even if it could be argued that effective calcium antagonist therapy will substantially reduce or even abolish the risk for future events. However, the risk of a second inappropriate change of medication was considered to be high.

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