Unusual association of diseases/symptoms

An unusual cause of blackout with transient loss of consciousness: Prinzmetal angina

Mohammed Majid Akhtar,¹ Rashid Akhtar,² Abid Akhtar,³ Javed Akhtar⁴¹Department of Cardiology, St Thomas' Hospital, London, UK²Department of Intensive Care, Broomfield Hospital, Chelmsford, UK³Department of Medicine, Queen Mary and Westfield Medical School, London, UK⁴Department of Cardiology, Queens Hospital, Romford, UK

Correspondence to Dr Mohammed Majid Akhtar, gogetmajid@gmail.com

Summary

The authors present the case of a 61-year-old woman who was troubled by regular episodes of throat discomfort, headache, dyspnoea and tingling sensation in the upper limbs. These were associated with occasional episodes of transient loss of consciousness accompanied by urinary incontinence over a period of 5 years. As these episodes became increasingly frequent, she was referred to a neurologist. Initial neurological assessment and investigations had a negative diagnostic yield and she was therefore referred for cardiac review. A repeat 24 h Holter revealed intermittent episodes of significant ST-segment elevation associated with a Mobitz type II atrio-ventricular block correlating with her symptoms. Her echocardiography and coronary angiography were normal; hence a diagnosis of Prinzmetal angina was made. She was treated appropriately with nitrates and a calcium channel blocker and followed up in cardiology clinic with no further recurrence of symptoms.

BACKGROUND

Prinzmetal angina is an important variant of angina that can cause catastrophic symptoms in affected patients but is often missed in view of its infrequency, the low diagnostic yield of standard investigations for coronary disease and because it is ignored in many acute coronary syndrome protocols. As such, patients may be falsely reassured that there is no cardiac aetiology to their complaints especially if their echocardiogram, perfusion stress test and coronary angiogram are normal.

In a case like this, where blackouts are associated with multiple other symptoms including urinary incontinence, patients may be diagnosed as being epileptic and given anti-epileptic drugs that will have no efficacious benefit to their symptomatology.

This case is important as a reminder to keep Prinzmetal angina as a differential diagnosis for transient loss of consciousness or for throat/chest pain when usual investigations for coronary artery disease are normal. Treatment is relatively straightforward and can have 'life-changing' results for the patient who might be suffering from the problem for a long time.

CASE PRESENTATION

We present the case of a woman in her early 60s with a history of sero-positive rheumatoid arthritis diagnosed at the age of 26, gastro-oesophageal reflux, hypertension, smoking and hypercholesterolaemia and a several-year history of episodes of tingling sensation in the upper limbs, throat pain, sweating and blackouts lasting in total for 5–10 min.

These episodes would occur randomly at rest, especially late at night, thereby waking up the patient. Sometimes these episodes led to loss of consciousness and urinary incontinence. There was no history of fits during one

witnessed event. Her brother died of myocardial infarction at the age of 42 and her father died of ruptured aortic aneurysm at the age of 75.

She was referred to the neurologist by her general practitioner; however, initial neurological assessment and investigations including ECG, EEG, brain CT scan and 24 h ECG monitoring were all unremarkable. She was then referred for cardiac review.

Despite multiple risk factors, her symptoms did not fit with a diagnosis of coronary artery disease. Echocardiography, a cardiac MRI (CMR) scan and repeat 24 h ECG monitoring were subsequently arranged. Her echocardiogram was normal; however, a repeat 24 h ECG revealed episodes of significant ST elevation leading to a Mobitz type II atrio-ventricular (AV) block with the onset of symptoms.

INVESTIGATIONS

A baseline Holter ECG recording showed normal sinus rhythm, as shown in figure 1. A 24 h ECG tape revealed sinus rhythm with two episodes of significant ST-segment elevation associated with a Mobitz type II AV node block, as shown in figures 2 and 3. These ECG changes correlated with the timing of the patient's symptoms of burning pain in the throat, tingling sensation in the arms, sweating, nausea and dizziness.

The CMR scan revealed focal fibrosis in the basal infero-lateral wall with late gadolinium enhancement. This suggested an area of previous infarct (likely related to coronary vasospasm) that was not seen on the conventional transthoracic echocardiogram (TTE). CMR is not a necessary investigation in patients with Prinzmetal angina but has the benefit of being able to identify structural heart diseases that may be missed on the TTE, particularly if

Figure 1 Rhythm strip from 24 h ECG monitoring, showing normal baseline ECG recording.

suboptimal views are obtained on the echocardiogram. CMR can better assess for arrhythmogenic aetiology and exclude other conditions such as apical hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy that may not be seen well on the TTE owing to suboptimal images and may account for presentations with blackouts.¹

Coronary angiography revealed unobstructed coronaries with dominant right coronary artery and normal left ventricular systolic function. Hence, a diagnosis of Prinzmetal angina was made and the patient was treated

with a calcium channel blocker (amlodipine 10 mg) and nitrates (isosorbide mononitrate—ISMN 60 mg). After commencing treatment, the patient did not have further blackouts and her throat pain and sweating episodes disappeared.

DIFFERENTIAL DIAGNOSIS

Given the patient's history, the 24 h ECG revealing significant symptomatic ST elevation and a Mobitz type II AV block with normal coronary angiogram, a diagnosis

Figure 2 Rhythm strip showing sinus rhythm with dynamic ST elevation on 24 h recording, associated with burning throat pain, tingling sensation in the arms and dyspnoea.

Figure 3 Rhythm strip showing ST elevation and Mobitz type II atrio-ventricular block on 24 h ECG monitoring, associated with the patient feeling dizzy and pre-syncope symptoms.

of Prinzmetal angina was made. It is most likely that she used to develop intermittent coronary artery vasospasm causing dyspnoea, throat pain associated with sweating and, on several occasions, transient loss of consciousness. Apart from significant ST elevation she developed episodes of high-grade AV block, which is most likely the explanation for her blackouts as all other relevant tests including neurological investigations were unremarkable.

TREATMENT

The patient was given aspirin, amlodipine and modified-release ISMN in view of her episodes of Prinzmetal angina. Since the patient started taking amlodipine and nitrates, she made a full recovery with no further episodes of throat pain or loss of consciousness. This led to significant improvement in her life.

OUTCOME AND FOLLOW-UP

On follow-up in the clinic, the patient was found to be doing well and was asymptomatic from a cardiac point of view. A repeat 72 h ECG monitor while on ISMN and amlodipine showed sinus rhythm with no abnormal ECG changes. She remained in sinus rhythm with occasional atrial and ventricular ectopic beats.

DISCUSSION

Prinzmetal angina has been documented in about 2–10% of patients. It is an important but infrequent cause of cardiac chest pain. In Prinzmetal angina, the symptoms typically occur at rest and in waves or cycles. Prinzmetal angina is caused by coronary artery vasospasm from contraction of the smooth muscle causing luminal obstruction. This is entirely different from luminal narrowing and occlusion caused by atherosclerotic coronary artery disease.

This form of coronary vasospasm was coined Prinzmetal angina after its characterisation by Prinzmetal *et al* in the late 1950s.²

Chest discomfort unrelated to effort with transient ST elevation on ECG in the absence of significant

coronary artery disease is the key to the diagnosis of coronary artery vasospasm.³ Occasionally, pharmacological testing with intravenous Ergonovine may be required if Prinzmetal angina is suspected on clinical grounds without documentation of typical ECG changes.⁴

Ergonovine is an ergot alkaloid and has a powerful stimulant effect on smooth muscles and can induce coronary artery vasospasm in susceptible individuals. One study found that more than 50% of patients with normal coronaries and a history of angina can be diagnosed with coronary vasospasm if intravenous Ergonovine is administered.⁵ Ergonovine provocation testing can also be used to detect spontaneous remission of variant angina in patients on long-term calcium channel antagonists for Prinzmetal angina.⁶ However, other research into regular Ergonovine testing shows that its sensitivity in diagnosing vasospastic angina is lower than previously emphasised.⁷ In some cases, intracoronary acetylcholine provocation testing is used as Ergonovine tends to produce more diffuse vasospasm which is undesirable.⁸ Intracoronary acetylcholine has been shown to provoke epicardial coronary artery spasm in patients with Prinzmetal angina, although its use is limited to those with a high clinical suspicion of Prinzmetal angina.⁹

Arrhythmias have been documented to be associated with coronary artery spasm including VT, VF and high-grade AV block/complete AV dissociation, which was noted in our case (Mobitz type II AV block).^{2 10–13}

Calcium channel blockers and long-acting nitrates are effective in the treatment of Prinzmetal angina with their smooth muscle relaxant and vasodilatory effects. There are reports of two cases of medically intractable Prinzmetal angina that have been successfully treated by coronary artery bypass grafting using the internal mammary artery despite patent native coronaries.¹⁴

The incidence of Prinzmetal angina may be reduced now because of the widespread availability and use of calcium channel blockers for various indications including hypertension. In patients where there is life-threatening ventricular arrhythmias associated with the coronary vasospasm, defibrillator insertion may be indicated.¹³

Learning points

- ▶ Consider Prinzmetal angina as a differential diagnosis in a young, usually female, patient with symptoms classically occurring at rest (especially at night) rather than on exertion.
- ▶ In patients presenting with blackouts it is important not to 'jump' to conclusions and label patients as having neurogenic syncope or epilepsy just in view of loss of consciousness and urinary incontinence as cardiogenic syncope can also cause similar symptoms.
- ▶ Nitrates and calcium channel blockers are useful in resolving the symptoms of Prinzmetal angina.
- ▶ A 24 h ECG tape can be a very useful means of assessing for arrhythmogenic aetiology if the symptoms occur frequently; otherwise prolonged Holter recordings or Internal Loop Recorder (Reveal) implants may be indicated, particularly if the clinician is suspicious of a malignant arrhythmia causing the presentation.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Akhtar MM, Akhtar R, Akhtar A, Akhtar J. An unusual cause of blackout with transient loss of consciousness: Prinzmetal angina. *BMJ Case Reports* 2012;10.1136/bcr.01.2012.5539, Published XXX

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