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Acute Aortic Intramural Hematoma Presenting with Painless Recurrent Syncope

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ABSTRACT

A patient presented with recurrent syncope due to transient severe hypotension. The patient's history, physical examination, and initial baseline investigation did not suggest a cardiovascular cause. After fluid resuscitation, a raised jugular venous pulse was noted. Bedside transthoracic echocardiogram showed a pericardial effusion and a proximally dilated aorta. Computed tomography of the thorax confirmed these findings and also demonstrated an intramural hematoma of the proximal aortic wall.

The patient was transferred to a cardiothoracic center, where he was at first treated medically. He then developed sudden cardiogenic shock due to pericardial tamponade and was successfully operated on.

It is important to recognize an acute intramural hematoma of the proximal aortic wall as a cardiothoracic emergency. This condition can present atypically, but nevertheless warrants urgent surgical intervention, equal to type A aortic dissection. Echocardiography can help in making the diagnosis.

INTRODUCTION

The acute aortic syndrome is defined as the collective term for life-threatening acute aortic conditions and includes aortic dissection, intramural hematoma, and penetrating atherosclerotic ulcer of the aortic wall [Vilacosta 2009]. An acute aortic dissection, the commonest entity of the acute aortic syndromes, can present without pain in many forms, eg, as a stroke or with aortic valve regurgitation and rarely as painless syncope [Hagan 2000; Nallamothu 2002; Shen 2010]. Recently the first case report of a painless aortic dissection presenting with recurrent syncope has been published [Shen 2010].

An intramural hematoma of the aortic wall (IMH) can be present in 5% to 20% of patients with an acute aortic syndrome [Hagan 2000; Maraj 2000; Evangelista 2006]. This is the first case report to describe an acute intramural hematoma of the proximal aorta presenting with painless recurrent syncope.

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CASE PRESENTATION

A 71-year-old Caucasian man was brought to the emergency department (ED) after suddenly losing consciousness for 2 minutes while sitting in a chair. He denied any pain in his chest before or after the event, and did not complain of any respiratory, abdominal, genitourinary, or neurological symptoms. His past medical history included hypertension and esophageal reflux disease. He took amlodipine and lansoprazole as his only medications.

The paramedic crew measured his blood pressure at 60/39 mmHg and his pulse at 70 beats per minute. He received an intravenous crystalloid fluid bolus on transfer to hospital.

In the ED, his blood pressure normalized and the remainder of his physical examination was unremarkable.

During his assessment in the ED, he had 2 further events of sudden loss of consciousness. On each occasion, his systolic blood pressure was recorded below 60 mmHg, and his heart rate remained normal.

Repeated measurements of hemoglobin were normal as well as test for renal and liver function, serum glucose, and inflammatory markers. An arterial blood gas showed no hypoxemia. Serial electrocardiograms were normal, and his chest radiograph was reported as normal. After fluid resuscitation, a raised jugular venous pressure was noted, and it was decided to investigate for a pericardial effusion. A bedside transthoracic echocardiogram in the ED showed a small- to moderate-size pericardial effusion with spontaneous echogenic contrast. The echocardiogram also showed a significant dilatation of the proximal aorta, but only mild aortic regurgitation (Figure 1).

A subsequent computed tomography of the thorax, including contrast aortogram, confirmed a dilated proximal aorta (6 cm) and a pericardial effusion, suspicious of a hemopericardium. Additionally, an intramural hematoma of the proximal aortic wall of 1.5 cm diameter was demonstrated, but no aortic dissection flap (Figure 2).

The patient was transferred to a cardiothoracic center. He was treated conservatively for the first 3 days. On the morning of the fourth day, he developed cardiogenic shock

due to a rupture of the proximal aorta, causing pericardial tamponade. He underwent emergency graft replacement of the proximal aorta and aortic valve replacement (Bentall procedure). A large intramural hematoma was found intraoperatively, which had ruptured into the pericardial sac. No evidence of an aortic dissection was detected. Thereafter the patient made a full recovery.

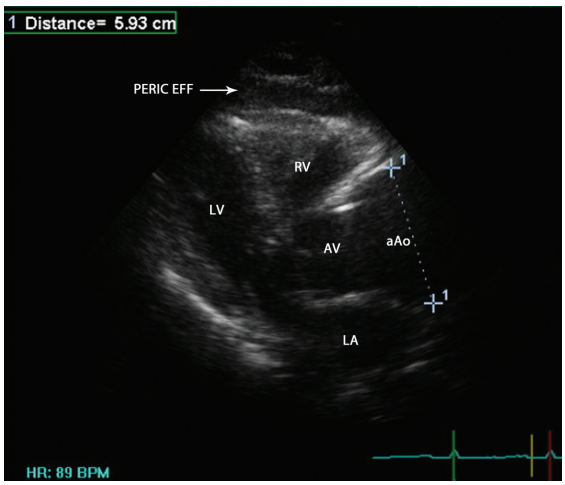

Figure 1. Echocardiography image of dilated aorta. Shown is a still image of a long axis view of the left ventricle (LV), the right ventricle (RV), the left atrium (LA), the aortic valve (AV). The ascending aorta (aAo) is dilated, and there is also a pericardial effusion (PERIC EFF) visible above the RV (arrow).

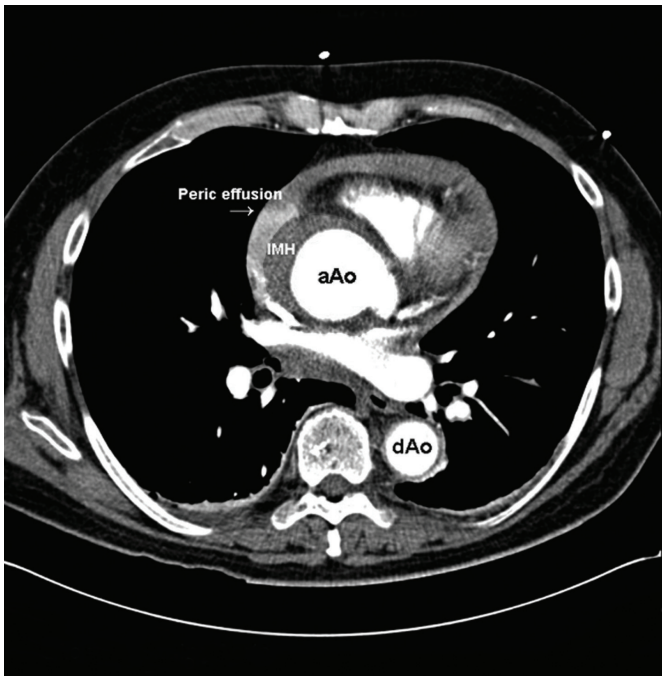

Figure 2. Computed tomography of the thorax. Note the proximally dilated ascending aorta (aAo), compared to the descending aorta (dAo) size. The aortic wall is increased in diameter from 7:00 to 12:00 due to the hematoma. IMH indicates intramural hematoma; peric effusion, pericardial effusion.

DISCUSSION

The typical presentations of aortic dissection and IMH are undistinguishable clinically, usually with severe pain in the chest, back, neck, or upper abdomen [Hagan 2000; Maraj 2000; Wvangelista 2005; Nienhaber 2010]. In previous studies of up to 977 patients with aortic dissection, 5% to 10% had presented painless and 3% painless, but with syncope [Hagan 2000; Nallamothu 2002].

Recently, a case of a painless aortic dissection presenting with intermittent syncope was reported, and because there seemed to be a great overlap in the clinical presentation of the 2 conditions, it is to be expected that an intramural hematoma of the aortic wall can also present atypically, without or with only little pain and with recurrent syncopes, as in our patient [Shen 2010].

Most patients with IMH are men and above 65 years old [Hagan 2000; Evangelista 2005]. Hypertension is the main risk factor [Evangelista 2005]. IMH is thought to be a precursor of dissection and caused by rupture of the vasa vasorum, leading to accumulation of blood inside the media of the aortic wall [Hagan 2000; Evangelista 2005; Song 2011]. Classification into proximal type A and distal type B IMH is similar to the Stanford classification of aortic dissections [Hagan 2000; Evangelista 2005; Nienhaber 2012].

Several pathophysiological mechanisms could be responsible for syncope in an acute aortic syndrome, including hypotension due to blood loss into the pleural or pericardial space, the latter potentially resulting in cardiac tamponade [Hagan 2000; Nallamothu 2002; Evangelista 2005], or a vaso-vagal reaction due to intense pain or stretching of the baro-receptors of the aortic wall [Nallamothu 2002].

The 2 main radiological differences between IMH and classic aortic dissection are the presence of an aortic wall hematoma and the absence of a dissection flap in IMH [Hagan 2000; Maraj 2000; Nallamothu 2002; Evangelista 2005; Song 2011; Nienhaber 2012]. It is conceivable that in IMH a very small tear could occur in the intima that cannot be seen with current imaging techniques and is not recognized as a dissection. This would make the IMH part of the spectrum of aortic dissections and not a separate entity. Further research may clarify this issue [Song 2011]. The role of biomarkers, eg, D-dimer, in the diagnosis of aortic dissection is still controversial, and their role in diagnosing IMH is even less clear [Evangelista 2005; Brown 2011; Citro 2011].

Nevertheless, it is imperative to make the diagnosis of type A IMH in a timely manner, since untreated it has a similar poor prognosis as an acute type A aortic dissection [Hagan 2000; Maraj 2000; Evangelista 2005; Nienhaber 2012].

Because it is a rare condition, clinicians do not always treat an acute type A IMH with the same urgency as type A aortic dissection [Citro 2011]. This can potentially lead to fatal complications [Hagan 2000; Maraj 2000; Evangelista 2005; Citro 2011; Nienhaber 2012]. Hence, for acute type A aortic intramural hematoma, most published papers advocate emergency or urgent open surgical repair, especially in patients with complications, eg, cardiac tamponade [Zhang 2011; Nienhaber 2012]. For type B IMH medical management or

an endovascular interventional approach is recommended [Evangelista 2005; Zhang 2011; Nienhaber 2012].

In the acute setting, especially in a hemodynamically unstable patient, transthoracic echocardiography is safe and can be of diagnostic use [Citro 2011; Cecconi 2012; Nienhaber 2012].

If, however, a patient presents with typical features of an acute aortic syndrome, immediate computed tomography of the aorta or transesophageal echocardiography are needed to confirm the diagnosis [Hagan 2000; Evangelista 2005; Cecconi 2012; Nienhaber 2012].

This case report illustrates that patients with an acute aortic syndrome, including IMH, can present with atypical symptoms and signs, such as syncope, and are at a high risk of potentially fatal complications if the diagnosis and emergency management are delayed.

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