Case 1/2014 - Syncope Due to Cardiogenic Shock in a 25-year-old Male Patient

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The patient was a twenty-five-year-old male individual, who suddenly started feeling faint and malaise at fainted on February 18th, 2009. He was taken to the medical emergency service of the local hospital by family members approximately one hour after symptom onset. The patient was previously healthy and asymptomatic. There was no history of hypertension, diabetes, dyslipidemia, heart disease or use of illegal drugs.

At physical examination the patient had lowered level of consciousness, 60/50 mmHg blood pressure, heart rate (HR) of 150 beats per minute (bpm), no palpable pulses in the upper limbs and symmetrical pulses in the lower limbs.

Several tests were performed on the day of hospital admission. The electrocardiogram (ECG) showed sinus tachycardia, with a HR of 150 bpm, PR interval of 120 ms, QRS duration of 80 ms (Figure 1). Chest radiography showed cardiomegaly, mediastinal enlargement and clear pulmonary fields (Figure 2). The transthoracic echocardiography showed dissection of the ascending aorta with cardiac tamponade (compression of the right atrium).

Laboratory assessment showed hemoglobin of 14.2 g/dL, hematocrit 42.9%, leukocytes 12,400/mm^3 (78% neutrophils, 2% eosinophils, 14% lymphocytes and 6% monocytes), platelets 202.000/mm^3, urea 30.5 mg/dL, creatinine 1.2 mg/dL, potassium 3.8 mEq / L, sodium 146 mEq/L, glucose 132 mg/dL, alkaline phosphatase 46 IU/L, gamma-glutamyl transpeptidase 37 IU/L, aspartate aminotransferase 21 IU/L and alanine aminotransferase 34 IU/L.

Due to the patient's clinical instability, tracheal intubation for ventilatory support was required, as well as volemic expansion with 0.9% saline solution and a vasoactive drug (norepinephrine) to elevate blood pressure.

After volume expansion and noradrenaline administration, blood pressure increased to 126/40 mmHg, with a heart rate of 135 bpm, and due to the severity of the clinical picture, his transfer to the Emergency Department of Instituto do Coração (InCor) was requested in the same evening.

Keywords

Syncope; Shock, Cardiogenic; Aortic Aneurysm; Aneurysm, Dissecting.

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Upon admission to InCor on February 19, 2009, the patient was sedated, assisted by mechanical ventilation and with pallor (++/6). The cardiorespiratory examination, showed a heart rate of 110 bpm, blood pressure 110/70 mmHg, normal pulmonary auscultation; however, cardiac auscultation showed muffled heart sounds (no accessory heart sounds, murmurs or pericardial friction rubs were identified). Abdominal examination was normal and there were no arterial pulses in the upper limbs, with bilateral cyanosis in the fingers. The arterial pulses were palpable and symmetrical in the lower limbs.

The laboratory assessment performed on February 19, 2009 showed: hemoglobin of 17.1 g/dL, hematocrit 54%, 26,900/mm^3 leukocytes (1% rods, 84% segmented neutrophils, 7% lymphocytes and 8% monocytes), 107.000/mm^3 platelets, urea 39 mg/dL, creatinine 2.6 mg/dL (glomerular filtration, 32 mL/min/1.73m^2), sodium 134 mEq/L, potassium 4.9 mEq/L, lactate 33 mg/dL, prothrombin time (International Normalized Index, INR) of 3.8 and activated partial thromboplastin time ratio of 1.53. Arterial blood gas analysis showed a pH of 7.22, partial pressure of Carbon Dioxide (pCO_2) of 32.9 mmHg, partial pressure of oxygen (pO2) of 206 mm Hg, O_2 saturation of 99.9%, bicarbonate 13 mEq/L and excess of base (-) of 13.7 mEq/L.

The ECG was suggestive of electrical alternans (Figure3).

Transesophageal echocardiography showed left ventricular hypertrophy with normal systolic function and a 62-mm aneurysmal dilation in the ascending aorta with the dissection lamina starting 1.7 cm from the valve. The dissection extended up after the aortic arch and affected the innominate artery, the left common carotid artery and the left subclavian artery. Severe aortic valve regurgitation and large pericardial effusion were also observed, with signs of right ventricular restriction to diastolic inflow.

Surgical treatment of the aortic dissection was indicated; however, the patient had bradycardia in the perioperative period, followed by asystole unresponsive to resuscitation maneuvers and died.

Clinical aspects

This is a case of a male patient, 25 years of age with no previously described clinical history that sought medical attention complaining of malaise and feeling faint. The clinical picture had started 1 hour before he sought the local hospital emergency room. The patient clinical evolution was dramatic, presenting at the examination as shock, initially of undetermined etiology, going to aortic dissection and cardiac tamponade, and culminating in death within hours of symptom onset, before any surgical intervention. Acute dissection of the ascending aorta was then identified, with rapid and catastrophic clinical deterioration, a determining factor for the diagnosis to be attained as early as possible, as well as an early and accurate intervention.

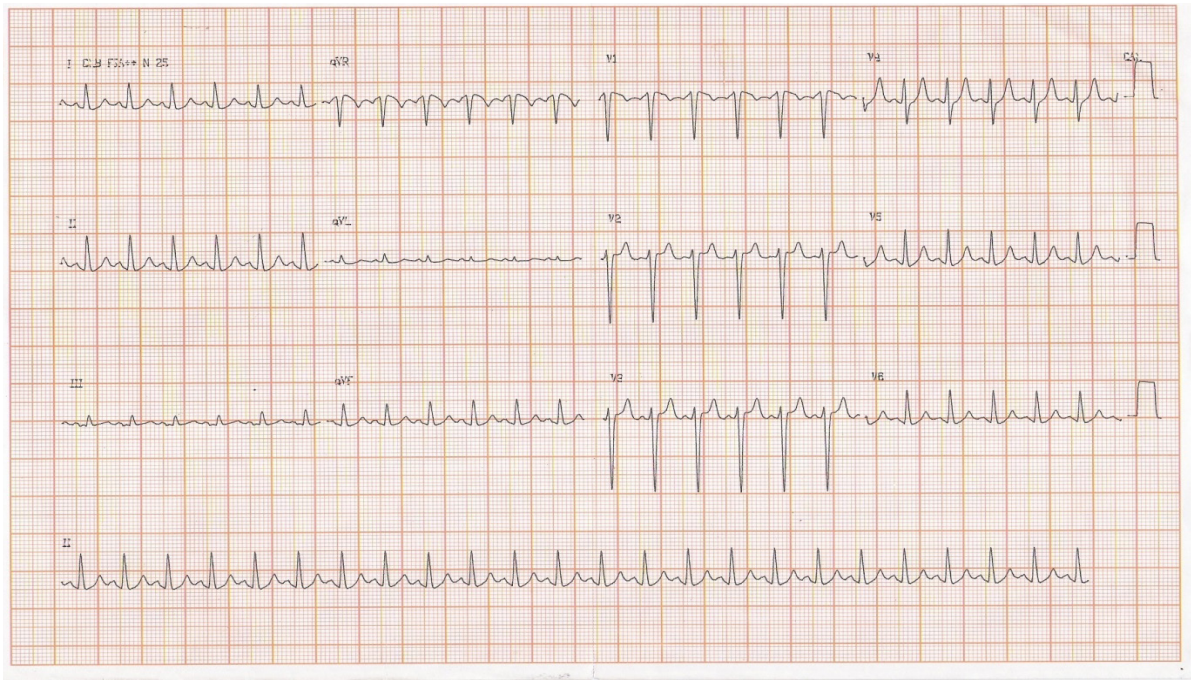

Figure 1 – Resting electrocardiogram showing sinus tachycardia.

Acute aortic dissection is a cardiovascular emergency with high mortality, ranging from 50 to 68% in 48 hours, 70% at 1 week and 85% in 1 month[1]. The intimal tear is transversal, located in 70% of cases in the aorta ascending (just above the aortic valve), 20% in the descending aorta after the aortic arch, and 10% in the transverse aorta. Once started, the dissection progression can be distal, retrograde or occur in both directions and be of varying extension[1,2]. In the present case, the aortic dissection started in the ascending portion, retrograde, with bending of the anterior plan between the true and false lumen, generating a mechanical obstruction to the left ventricular outflow tract (Figure 4).

Aortic dissection is more common in male individuals and the male/female ratio ranges from 2:1 to 5:1, with the highest incidence between 45 and 70 years of age, with a peak age between 50 and 55 years for cases of proximal dissection and between 60 and 70 years for the distal cases[3,4]. The preponderant factor at the dissection is the involvement of the medial layer, as in the genesis of aneurysms in general, and the medial disease that most often predisposes to dissection is medial cystic necrosis. The dissection can be triggered by "traumatic" factors of the arterial wall, such as hypertension, aortic coarctation, pregnancy and bicuspid valve[5].

There are also other clinical entities that compromise aortic endothelial integrity and function, such as aortitis, hereditary connective tissue disorders (Marfan syndrome and Ehlers-Danlos syndrome), congenital malformations of the aortic valve, trauma, aortic valve replacement and situations that facilitate endothelial injury, such as a simple catheterization[6]. The clinical picture and therapeutic approach are sufficiently different to justify the separation of this entity into two groups: type-A Stanford dissection, involving the ascending aorta extending or not to descending aorta and type-B Stanford dissection, which involves only the descending aorta, with this classification being the most widely accepted[1,4]. From a purely surgical standpoint, De Bakey et al[3] classified dissecting aneurysms into three groups: type I corresponds to the dissection starting in the ascending aorta extending for a varying distance, distally; type II is the dissection limited to the ascending aorta; and type III is the dissection started after the emergence of the left subclavian artery and that progresses to the abdominal aorta with a varying extent. In this case, we found a type-A Stanford aortic dissection, with probable hypertensive etiology, as demonstrated by echocardiography, in addition to the lack of evidence of any risk factors described in the literature, whether in the past medical history, at the clinical evaluation or in the tests performed, to justify another etiology.

The main clinical manifestation of aortic dissection is high-intensity chest pain, accompanied by symptoms of sympathetic activity. The onset of pain is almost always sudden, being characterized as a sharp, stabbing or throbbing pain, associated with sweating and may be limited to the thorax or be retrosternal, radiating to the back, abdomen, upper or lower limbs[4]. It can also be associated with dyspnea

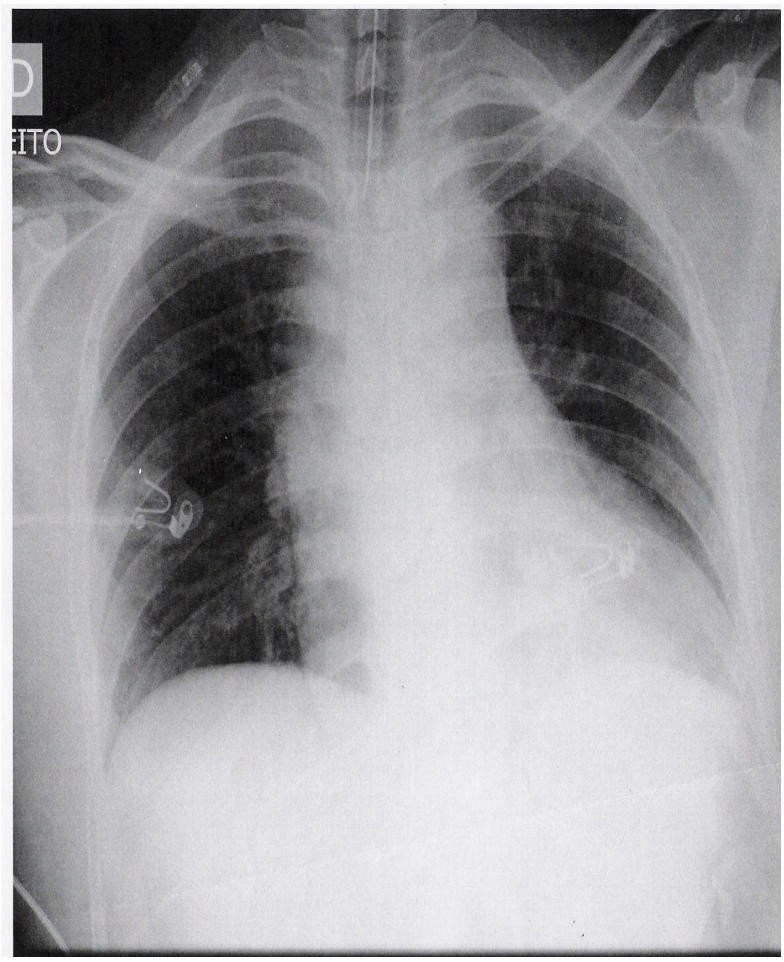

Figure 2 – Chest radiography showing cardiomegaly, mediastinal enlargement and clear pulmonary fields.

and pulmonary edema[4-7]. However, the clinical manifestation may be subtle or atypical and aortic dissection without prior chest pain has been previously described[8,9] and occurs in approximately 5 to 15% of all aortic dissections[10,11]. In the present case, the patient reported no chest pain and two possible explanations are offered for the painless aortic dissection: first, the dissection can cause a protrusion (bulging) of the intima into the inner layer and reinsert the lumen of true aorta, without moving the adventitia to the outer layer; and second, the dissection can occur very slowly, without sudden or violent distension of the adventitia, and thus it can be painless[12]. Fever may accompany a third of the patients with dissecting aneurysms, and is generally not associated with endocarditis[11]. Physical examination may disclose asymmetry of peripheral pulses, and in 50% of cases with type-A dissection, alterations in one or more pulses, and in only 16% in type-B dissections[1,7].

Imaging diagnostic methods include echocardiography (transthoracic or transesophageal), computed tomography or angiography. Many services use echocardiography as the sole preoperative assessment for surgical indication in acute type A dissections. In general, the transesophageal echocardiography is preferred, which can reach 97-100% of sensitivity and 77-97% of specificity[13,14]. It is also of importance the fact that the echocardiography provides information on the presence and magnitude of aortic regurgitation, aortic annulus dimensions, evidence of aortic regurgitation prior to the dissection; these are key data for surgical management, regarding the aortic valve preservation[14].

In the present case, due to the patient's clinical instability, transesophageal echocardiography was performed. CT and angiography are useful for special situations or in indeterminate cases, as these tests are more time-consuming and mainly the angiography may precipitate aortic rupture. Patients with a history of coronary artery disease or patients undergoing cardiac surgery or special procedures for myocardial revascularization should undergo preoperative coronary angiography.

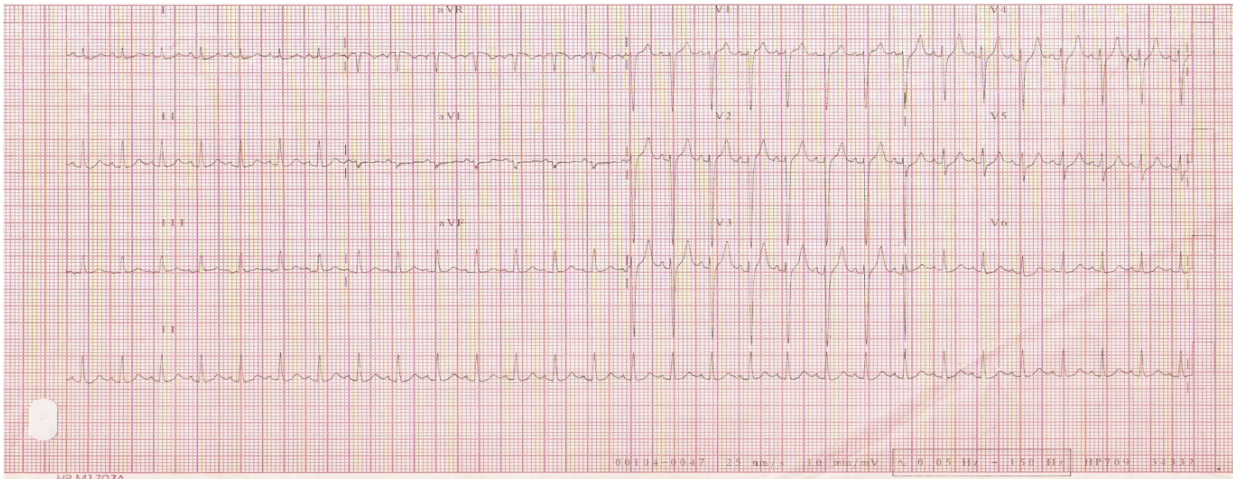

Figure 3 – Resting electrocardiogram showing electrical alternans.

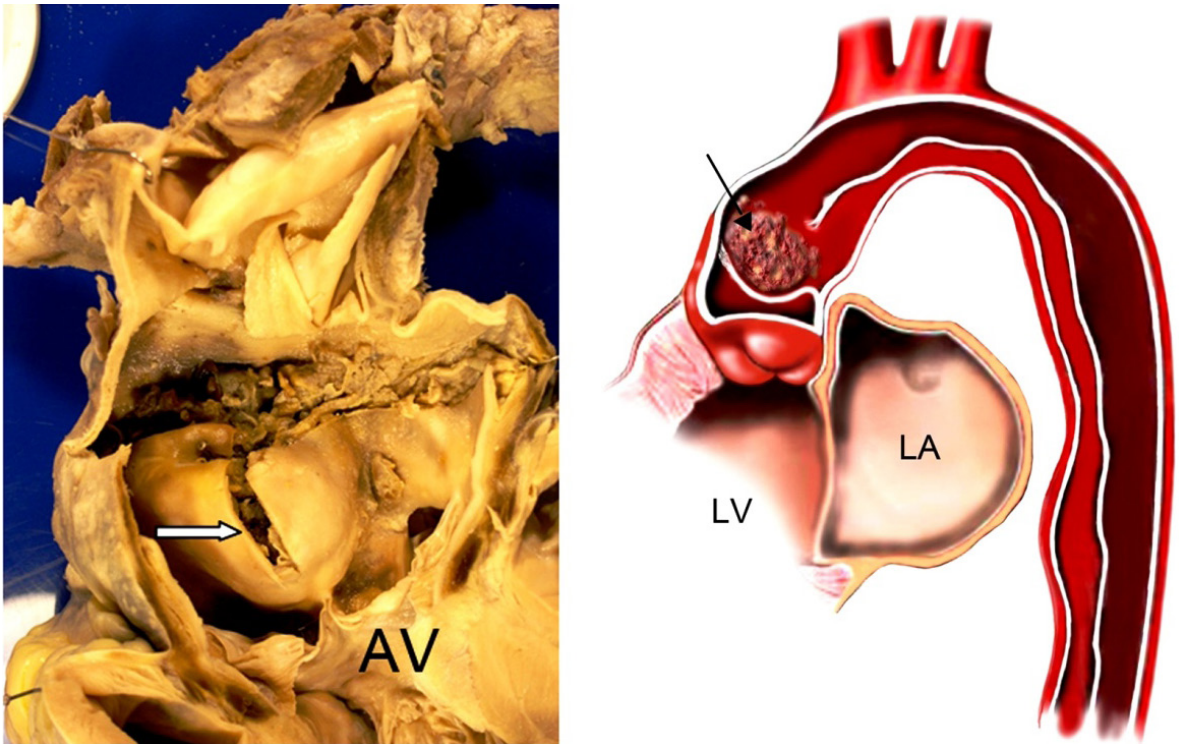

Figure 4 – *(A) Open ascending aorta showing the dissection with the inner layer of the arterial wall protruding into the valve plane. The white arrow shows this region, with the section taken for histological analysis. The light blue arrow indicates the tear that defines the dissection orifice, in this case affecting the entire circumference of the artery - artistic illustration of the pathological process. The arrow indicates a clot in false lumen, pushing down the inner part of the aortic wall. LA: left atrium; AV: aortic valve; LV: left ventricle.*

Anatomopathological Session

Thus, as this is a condition in which mortality is very high, surgical treatment should be immediate, and late survival is not free of complications (mainly due to the high persistence of patent false lumen in the segments of the aortic arch, descending and abdominal aorta). The need for surgical treatment immediately after diagnosis aims at preventing its life-threatening complications, such as cardiac tamponade secondary to rupture of the aorta, as reported in this case, acute myocardial infarction, symptomatic aortic insufficiency, neurological complications, among others, with a dramatic impact on mortality[15,16]. The gold standard treatment is the surgical intervention and the traditionally proposed surgery is the interposition of the supracoronary tube, of which in-hospital mortality rate varies, ranging between 10 and 30%[17,18]. Unfortunately, this patient did not survive long enough to undergo such treatment.

(Dr. Tiago Rodrigues Politi)

Diagnostic hypothesis: ascending aorta dissection. (Dr. Tiago Rodrigues Politi)

Other diagnoses: Systemic arterial hypertension, ascending aorta aneurysm, cardiac tamponade. (Dr. Tiago Rodrigues Politi).

Necropsy

Necropsy disclosed dissection of the ascending aorta, starting approximately 4 cm above the valve, affecting the entire ascending aorta in retrograde fashion. As it affected only the ascending aorta, the dissection was characterized as type II, according to the DeBakey classification, and Stanford type A. The dissection orifice affected the entire arterial circumference, being perpendicular to its axis. There was protrusion of the inner layer of the dissected aorta, leading to artery obstruction (Figure 4). This obstruction caused multiple organ ischemia, including almost total infarction of both kidneys and cerebral infarction in the right parietal region (the carotid arteries had no dissection). There was rupture of the aorta into the pericardial sac, leading to cardiac tamponade, which was the final event which led to the patient's death.

The patient had signs of hypertension such as alterations in renal microcirculation, as well as in the afferent arterioles, and left ventricular concentric hypertrophy.

The patient's age suggested a genetic syndrome underlying the dissection, but he had no clear phenotype of Marfan syndrome and, moreover, showed indicators of hypertension described above.

It is noteworthy, in this case, the prolapse of internal layer of the dissection, leading to blood flow obstruction. It is an unusual complication of acute aortic dissection, which does not appear even in some fairly comprehensive anatomopathological descriptions of this disease[19,20]. (Dr. Paulo Gutierrez)

Anatomopathological diagnosis

Acute aortic dissection and aortic obstruction by the internal lamina of the sliced arterial wall; probable systemic arterial hypertension.

(Dr. Paulo Gutierrez)

Commentary

Cause of death: cardiac tamponade. (Dr. Paulo Gutierrez)

Acknowledgement

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