Abstract

The patient, MSM, a 69-year-old man, sought medical care due to left dorsal and right lower limb pain.

The chest x-ray showed mediastinal enlargement.

He was undergoing examination when he lost consciousness and went into shock. Subcutaneous emphysema was observed in the left hemithorax, as well as abolition of breath sounds at auscultation. Tracheal intubation was performed with draining of blood-tinted fluid from the left hemithorax.

Echocardiography showed left ventricle with 44/29 mm; septum, 12 mm; posterior wall, 13 mm; mild aortic root dilation, dissection of the lamina and periaortic hematoma. The valves and pericardium were normal.

The patient was transferred to Instituto do Coração - InCor.

Physical examination (21 Oct 2004: 10:45) showed that the patient was sedated with tracheal intubation, pale, heart rate at 90 bpm, blood pressure 130 x 80 mmHg, bloody drainage in the chest tube.

Electrocardiogram - frequency 90 bpm, sinus rhythm, low voltage in the frontal plane and decreased voltage in left leads (Fig. 1).

Computed tomography showed bilateral subcutaneous emphysema, thoracic aorta with inaccurate borders in its descending portion (from the subclavian artery to the middle portion), collapsed left lung and extensive collection of hematic effusion with underlying parenchymal alterations. The analysis of the heart was impaired by the presence of hemothorax.

While undergoing computed tomography, the patient showed no pulse, mydriasis, with asystole unresponsive to resuscitation and died (21 Oct 2011; 15:00 h).

Keywords

Acute Coronary Syndrome; Pulmonary Embolism; Pneumothorax; Aorta, Thoracic / abnormalities.

Section Editor: Alfredo José Mansur (ajmansur@incor.usp.br)
Associated Editors: Desidério Favarato (dclfavarato@incor.usp.br)
Vera Demarchi Aiello (anpvera@incor.usp.br)

Clinical aspects

The case described is a previously healthy, 69-year-old male patient, treated in a hospital with pain in the left back region and right lower limb. Based on this complaint, chest pain protocol was initiated and a chest radiography was performed, which showed mediastinal enlargement, first element suggestive of a specific etiology for the case (present in 63% of cases of ascending aortic dissection and in 56% of cases of descending aorta involvement). It is worth noting that, in relation to the initial symptoms, pain in the right lower limb may arise in the context of acute aortic dissection with progression to distal arterial segments, although not sufficient, however, to confirm the diagnosis for the case, requiring specific additional investigation.

During the observation period there was a sudden worsening of the picture, characterized by decreased level of consciousness, circulatory shock and subcutaneous emphysema, with pulmonary propedeutics of left pleural effusion, requiring invasive measures of support for stabilization. At this point, it is appropriate to review the initial hypotheses when chest pain is accompanied by high mortality characteristics, such as those presented in this case (pain associated with circulatory shock):

- Acute Coronary Syndrome: Acute Myocardial Infarction (AMI), either through the involvement of the right ventricle, large areas of the left ventricle (extensive anterior AMI, for instance), or the onset of mechanical complications, could justify this picture. However, this evolution is not associated with findings such as subcutaneous emphysema, and its diagnosis is accompanied by electrocardiographic findings (ST-segment elevation or pathological Q waves, for instance).
- Massive Pulmonary Thromboembolism: It is associated with pleural effusion, as described in this case, in up to 40% of cases, but is very rarely accompanied by pneumothorax and subcutaneous emphysema. The literature has publications in the form of case reports; however, they stress the subacute characteristic of these complications (two to four days after the event). Noteworthy is also the primary presentation of electrocardiographic signs of right chamber overload and electrical axis deviation, not shown in this case.
- Spontaneous Hypertensive Pneumothorax: The association of this entity with chest pain, shock and subcutaneous emphysema is well described; however, important clinical findings not described in this case, such as tracheal deviation, hypertympanism and overall decreased pulmonary auscultation are striking. In addition, it often affects slim individuals with lung diseases and it possibly would have been identified at the initial radiograph in the emergency room.

Mailing Address: Vera Demarchi Aiello

Av. Dr. Enéas C. Aguiar, 44, Sub-solo, bloco I, Cerqueira César.
Postal Code 05403-000, São Paulo, SP – Brazil
E-mail: demarchi@cardiol.br, vera.aiello@incor.usp.br

DOI: 10.5935/abc.20130037
Complicated Aortic Dissection (rupture and ischemia of lower limb): It may be present in the context of this case, presenting with similar characteristics, including decreased level of consciousness, either by circulatory shock (complication of 13% of cases), or by the extension of dissection to the brain circulation (8% - 15% cases). Moreover, its rupture can course not only with pleural effusion (hemorrhagic or serohematic, with this being attributed to the underlying presence of pleural inflammatory process in 16% of cases), but also, albeit rarely, with sudden pneumothorax and subcutaneous emphysema, related to its expansion into the lung parenchyma and subsequently, to the pleural cavity. Kurosaki et al. described the case of a 79-year-old patient with a history of atrial fibrillation and previous transient ischemic attack, who was a victim of sudden death, in whom a hypertensive pneumothorax occurred as a result of ruptured thoracic aortic aneurysm.

Based on the clinical case presentation and literature data, the last two clinical entities would be the main diagnostic hypotheses until this phase of evolution.

The echocardiogram (probably transesophageal), well indicated considering the clinical instability of the case, showed dissection of the lamina and periaortic hematoma and signs of ventricular hypertrophy, establishing not only the case diagnosis, but also being sufficient to define the appropriate therapeutic measures. The sensitivity and specificity of transesophageal echocardiography in this context are, respectively, 98% and 95%. Furthermore, arterial hypertension, present in about 72% of cases of aortic dissection, emerges as a possible underlying disease and a risk factor for this complication based on the echocardiographic findings.

Thoracic aortic dissections are more common in males (2:1) and at the age range of 60 to 70 years, being classified, particularly by duration of symptoms, as acute or chronic, with a cutoff of 15 days to differentiate them; and by location as Stanford type A, when they involve the ascending portion of the artery, or Stanford type B when they spare this segment.

At admission in InCor, the heart rate and blood pressure were noteworthy, which were outside the established targets for effective clinical management of acute aortic dissection. However, these findings could be related to the possible use of vasoactive agents. Furthermore, the patient was sedated, which in addition to hindering the assessment of focal neurological deficits that would contribute to the diagnosis of complications related to the underlying disease, would also prevent the definition of the surgical therapeutic approach, as the comatose state is a formal contraindication to the invasive approach.

The initial electrocardiogram showed sinus rhythm and low voltage in the frontal plane and left precordial leads. This finding may suggest evolution of the case to one of the possible complications of aortic dissection: pericardial effusion, which occurs in about 2% of cases. Also through the analysis of the ECG, other diagnostic possibilities become less likely, as described above.

The chest angiotomography, a key element in the diagnostic investigation and anatomic evaluation of thoracic aortic dissection in patients with hemodynamic stability, has a respective sensitivity and specificity of 98% and approximately 100% in this entity. In this case, in addition to demonstrating the inaccurate borders of the aorta (indirect...
sign of rupture), it showed left lung collapse, with massive ipsilateral hemothorax, in addition to involvement of the middle and posterior mediastinum, which may be due to aortic rupture into the pleural and mediastinal space and corroborate the diagnosis of complicated aortic dissection. It should be noted, at this moment, the rare but well-described association of these cases with pneumothorax and subcutaneous emphysema7.

Aortic dissection carries multiple associations. In addition to the advanced age and hypertension (the only risk factors suggested for this patient), we could cite, for example, bicuspid aortic valve (5%-7% of cases), previous surgical manipulation, aortic coarctation and cocaine use (1% of the cases), not easily identifiable in this case. Moreover, it is worth mentioning Marfan syndrome (5% of cases), which can contemplate during its natural history, the association of aortic dissection and pulmonary involvement in the form of pneumothorax, but which are very rarely clinically concomitant19,10.

(Dr. Carlos Vinícius Abreu do Espirito Santo, Dr. Rafael Alves Franco)

Diagnostic hypothesis

The patient developed Cardiopulmonary Arrest (CPA) with asystole during chest CT. This evolution leads us to possible etiologies for the outcome11:

• Hypovolemia: due to aortic rupture and massive bleeding.
• Cardiac tamponade: due to retrograde dissection into the pericardial sac (not shown in the initial echocardiographic evaluation).
• Acute Myocardial Infarction: due to retrograde dissection of the coronary ostia.
• Hypoxia: due to left lung collapse and pleural effusion to the right demonstrated at the tomography, associated with risks of transporting patients.

Finally, for the clinical picture described here, there are two diagnostic hypotheses to be considered: acute dissection of the thoracic aorta (Stanford type A), progressing to shock due to aortic rupture with hypovolemia and / or hypertensive pneumothorax.

(Anatomopatologico Session)

(Anatomopathological diagnoses)

Coarctation of the aortic arch at the isthmus region; ruptured acute aortic dissection; benign and mild nephrosclerosis; gallbladder calculosis; left hemothorax (cause of death).

(Anatomopatologico Session)

(Comments)

This is the case of a 69-year-old man who presented with acute aortic dissection, confirmed by echocardiography, associated with subcutaneous emphysema and left hemothorax, evolving with loss of consciousness and hemodynamic shock. The presence of systemic arterial hypertension is not mentioned in the history, but the patient had no known medical follow-up.

The necropsy confirmed the diagnosis of acute aortic dissection, with rupture into the left hemithorax. The dissection originated in the descending thoracic aorta and did not affect the proximal segments of the vessel, and was therefore classified as DeBakey type IIIb or Stanford type B. Coarctation of the aortic arch was also detected, located in the isthmus region, with moderate luminal stenosis. The aortic valve was normal. We found anatomopathological evidence of systemic arterial hypertension, with renal hyaline arteriolsclerosis, but there was no significant left ventricular hypertrophy, heart weight was within normal limits, and probably the blood pressure alteration was a minor one.

The most common risk factor for acute aortic dissection is hypertension, but one should also remember the association with connective tissue diseases, particularly Marfan syndrome and bicuspid aortic valve. However, when considering only the type III dissections, i.e., those that originate in the descending thoracic aorta, the frequency of systemic arterial hypertension is even higher and there is no statistical association with bicuspid aortic valve12.
Figure 2 – Cross-section of the heart showing mild left ventricular concentric hypertrophy.

Figure 3 – Aorta opened longitudinally showing the extremity of the ascending segment of the aorta, aortic arch and beginning of the descending thoracic segment. There is an abrupt narrowing of the vascular lumen at the isthmus of the aortic arch (asterisk), with mild poststenotic dilatation of the beginning of the descending thoracic aorta, in which one can observe the site of acute dissection entry (arrow). Observe the wall dissection and adventitial hemorrhage present at the beginning of the descending thoracic aorta (double asterisk).
On the other hand, there are even rarer conditions associated with acute aortic dissection, such as pregnancy, heart surgery, chest trauma, giant cell aortitis, coarctation of the aorta and others. In fact, there have been several previous publications that reported an association between coarctation of the aorta with acute aortic dissection, as in this case.\textsuperscript{13-15}

Obviously the correction of the condition (aortic coarctation and acute dissection) is surgical and may occur at different times or even simultaneously.\textsuperscript{16} Finally, it is interesting to mention that in this case the aortic valve was not bicuspid, an alteration often associated with coarctation of the aorta.

(\textit{Dr. Luiz Alberto Benvenuti})

References


Figure 4 – Renal hyaline arteriolosclerosis. Renal cortical arterioles showing thickened, eosinophilic walls, with hyaline aspect (arrows). Hematoxylin-eosin staining.