Aortic aneurysm dissection – diagnosis may be hard to make

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Abstract

The authors report the case of a 70-year-old woman admitted with intense asthenia, anemia and increased erythrocyte sedimentation rate following an acute and sudden episode of anterior thoracic pain six weeks before. Clinical investigation was conclusive for the diagnosis of subacute aortic dissection type A. Surgical repair and placement of endovascular prosthesis was performed, allowing the disappearance of general manifestations.

Aortic dissection is usually associated with an acute and highly symptomatic clinical presentation. This case report illustrates the diagnostic difficulties when aortic dissection presents with prominent features characterized by systemic symptoms and elevated acute phase response laboratory parameters. Knowledge of this atypical presentation, a high degree of suspicion, and investigation using an adequate imaging method will help improve our capability for a correct diagnosis of this entity.

Key words: aortic aneurysm; aortic dissection; erythrocyte sedimentation rate.

Introduction

Acute aortic dissection is a rare pathology, with an incidence of 0.5 to 1/100 000/year in the United States, affecting mainly male individuals from the 5th life decade onwards.[1]

This is a medical emergency with high mortality, which can only be reduced by the quick recognition and subsequent starting up of the appropriate therapy. If it is not timely diagnosed, mortality rate will grow of 1 to 2% per hour, up 90% in the first few weeks.[2]

Dissection is defined as acute in the first fortnight of progress, subacute up to two months and chronic after the second month. After the acute stage, both mortality as the progression risk reduce over time.[3]

Rare are the cases where dissection is presented with a chronic and atypical form featured by a delayed febrile condition associated to constitutional symptoms and an increase on the Erythrocyte Sedimentation Rate (ESR).[4,5]

The clinical case we present aims to show how a less common presentation and evolution, the scarcity of available diagnostic means and, probably not including this hypothesis in the differential diagnosis, can make difficult both the diagnosis as the appropriate therapy onset.

Clinical Case

Female patient, 70 years of age, living in a rural area in Grandola district, with a personal history of hypertension and dyslipidemia, treated with medicine. She was apparently well up to six months before being admitted, time in which she referred an episode of anterior torachalgy anterior, intense, sudden onset, irradiating to the dorsal, followed by a general malaise. The patient went first to the Permanent Assistance Service of her residential area, where she was seen, underwent an ECG and received analgesic therapy, being discharged with a (NSAID) non-steroidal anti-inflammatory drugs. On the following days, due to a persistent thoracic pain, already less intense, the patient went several times to Grandola SAP, to Sao Bernardo Hospital Emergency Unit and to her Assistant Physician. She always got an indication for NSAID and analgesics, having also started, physiotherapy. On the following days she has noticed a pain relief, but also had complaints of easy fatigue, intense asthenia and anorexia. The supplementary exams already performed have revealed an anemia of 108 g/L and ESR of 106 mm/hr being at this moment referred to

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the Hospital with an urgent request for a Medicine consultation. She denied dyspnea, wheeze, orthopnea or other breathing symptoms, epigastralgia, symptomatology suggesting gastrointestinal hemorrhage, osteoarticular complaints, as well as fever, diaphoresis or significant weight loss. She denied tuberculosis personal history.

The objective exam, on admission showed a good general condition and nutrition, apyrexia, eupnea, normal BP, dysrhythmic cardiac sounds, without changes on the pulmonary auscultation; the abdominal exam, the palpation of peripheral pulses, the exam both of joints and neurology were normal.

Some lab tests were made *(Table 1)*. The abbreviated urine test was normal. The thorax X ray has shown an enlargement of the heart and mediastine shadow, small left pericardial effusion, without changes in the pulmonary fields. The electrocardiogram (ECG) made on admission has shown an auricular fibrillation with a quick ventricular response, QS from V1-V4 with upper ST unlevel in the same derivations and a bad progression of R waves of V4-V6. The abdominal ultrasound has shown a liver with normal dimensions and without echostructural changes, with the remaining abdominal structures also without relevant structural changes.

A transthoracic echocardiogram (TTE) showing a dilated aorta root and ascending aorta, slightly thickened aortic cuspid with a conservative systolic opening, non hypertrophic left ventricle, depressed global systolic function, anterior wall hypokinetic and interventricular septum, anterior moderate pericardial effusion, posterior and lateral, without hemodynamic commitment, light mitral and moderate aortic failure.

Diagnosis of aorta aneurysm, myocardial infarction in pericarditis complicated subacute stage – Dressler syndrome – or, eventually, another etiology pericarditis – namely infectious, neoplasm or associated to a collagenosis. She started therapy with acetylsalicylic acid concomitant with angiotensin-converting enzyme ACE, with benefit.

The analytical evaluation was supplemented, obtained with serum to echovirus, coxsackievirus and human immunodeficiency virus 1 and 2, which were negative, as well as the auto-immunity markers, namely the antinuclear antibody (ANA), Anti-DNA and Anti-neutrophil cytoplasmic antibodies ANCA, VDRL and tuberculin test (5 PPD Un).

TABLE I

Analytic results on admission

Hematologic Values	
Hemoglobin (g/L)	108
Hematocrit (%)	31,1
Mean corpuscular volume (fL)	96,4
Leukocytes (109/L)	7,8
Differential count (%)	
Neutrophiles	69%
Lymphocytes	19%
Monocytes	9%
Eosinophiles	1%
Basophils	2%
Platelets (109/L)	362,000
Erythrocytary sedimentary rate (mm/hr)	114
Serum iron (µmol/L)	32
Ferritin (mcg/L)	2227
Transferrin (mcg/L)	132
Other laboratorial value	
Fastening glucose (mg/dL)	127
Urea (mg/dL)	47
Creatinine (mg/dL)	0,9
Ionogram	Normal
Aspartate aminotransferase (U/L)	99
Alanine aminotransferase (U/L)	113
Alkaline phosphatase (U/L)	525
Gamma glutamyl transferase (U/L)	391
Lactate dehydrogenase (U/L)	668
Total bilirubin (mg/dL)	0,79
Direct bilirubin (mg/dL)	0,38
Creatinine kinase (U/L)	34
Troponin	0,02
Total Cholesterol (mg/dL)	198
HDL Cholesterol (mg/dL)	52
LDL Cholesterol (mg/dL)	144
Tryglicerides (mg/dL)	176
C reactive Protein (mg/dL)	3,4

In order to clarify the mediastine enlargement, a thorax CAT scan revealing an aneurysmatic dilation of the thoracic aorta, left pleural effusion and pericardial effusion, absence of pleural-parenchymal lesions,

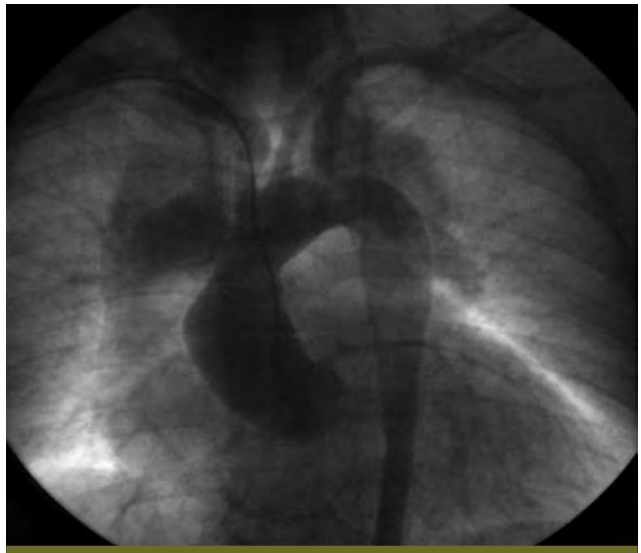
being impossible a better characterization because a endovenous contrast was not used.

It was then requested a Magnetic Resonance Angiography (Angio-RM), showing a type A aortic dissection, associated to a hematoma of the tunica media type A, on the initial portion of the aortic ascending segment and valve insufficiency, hemopericardium along the anterior wall of the right auricle and the bilateral pleural effusion with a minimum volume.

Afterwards it was made a transophageal echocardiogram (ETE), confirming the dissection 6 cm of the valvular plan (two flap zones limiting two fake lumen around the true lumen), spreading to the aorta arch and descending aorta, being the fake lumen, at the descending aorta level, thrombotic, keeping a perimeter higher than the true lumen; aortic regurgitation of a light degree.

In the pré-surgical evaluation, the patient underwent cardiac catheterization. Left ventriculography revealed an apical dyskinesia, postero-basal akinesia and antero lateral and important diaphragmatic hypokinesia, being the fraction of global ejection of the left ventricle estimated in 38%. Aortography has displayed a Grade III/IV aortic failure and moderate dilation of the ascending aorta, confirming a Type A aorta dissection, visualizing a fake lumen before the origin of the left subclavia *(Fig. 1)*.

The patient was directed to the referring hospital, where she has undergone a surgical intervention with an endovascular prothesis placement. At present, it is asymptomatic being registered a regression of the laboratorial changes clear on admission, showing a light aortic valvular insufficiency, and subsequently a follow-up of the cardiothoracic surgery and cardiology.



Aortography showing a contrast passage from the lumen interior to the vessel wall where some contrast is retained after its passage.

FIG. 1

Discussion

In spite of the extensive literature on the aorta dissection, the natural history of this pathology is yet to be clarified.[6] Traditionally, the clinical diagnosis of the aortic dissection is inadequate, being correctly suspected in just 15 to 43% of the cases, in the patient initial evaluation.[2,7] When the diagnosis is established, it is not uncommon that this is an accident, decurrent of the investigation of other diagnosis hypothesis. Post-mortem studies show the correct diagnosis is not even reached in around 10% of cases.[8]

In Stanford classification considering a type A dissections, those involving the ascending aorta (proximal dissection), regardless of the origin local, and type B all those not involving the ascending aorta (distal dissection). In the DeBackey classification, three types of aortic dissection are considered: (a) type I, with an origin in the ascending aorta, spreading at least until the aortic arch and often to the descending aorta, (b) type II limited to ascending aorta and the Type III, with origin in the descending aorta, usually with distal dissection spreading, and can, seldom have a retrograde progression.[9]

Most patients with a thoracic aortic dissection referring lancinating pain (90% sensitivity) of sudden onset, reduces the probability of dissection [predictive value (pv) negative 0.3 confidence interval (CI) 95% 0.2.-0.5].[10] More often the pain is located on the thorax (32%), dorsal (32%) or abdomen (23%).[10]

Other presentation forms include: syncope (13%), cerebral ischemia (6%), pericardium rupture with cardiac tamponade, involving one or more coronary artery (usually the right coronary), causing an acute coronary syndrome, hoarseness, dysphagia, superior vena cava syndrome, Horner's syndrome.[10] Rare manifestations of dissection, in particular chronic dissection, include a condition of persistent fever, nocturnal hyperhidrosis, weight loss and a general malaise associated to an increase of parameters in acute stage, namely ESR and anemia of chronic disease,

emulating other clinical entities, namely an infectious disease, neoplasm or auto-immune.[4,5]

In the objective exam, 49% patients are hypertensive; however, an equal proportion is hypo- or normotensive. In 31% of patients is evident the reduction of one or more pulses (carotid, radial, femoral) or an asymmetry of blood pressure (BP) higher than 20 mmHg. A diastolic blow or aortic regurgitation may be found in 28% of cases. Around 17% of patients have neurologic symptoms. The absence of any of these signs does not change significantly the diagnosis probability.[10]

The thorax X ray usually is abnormal (sensitivity 90%), and can show a mediastine enlargement, dilation of the aortic arch, change in the diameter between the ascending and descending aorta, indefinition of the aortic margin, unilateral or bilateral pleural effusion, calcium sign (consisting in the separation of the tunica intima with calcification of the external margin at the arch level ≥ 1cm present in a minority of cases). The finding of an aorta or a mediastine without morphologic changes reduces the probability of dissection (negative pv 0.3 CI 95% 0.2-0.4).[10]

Regarding electrocardiographic changes, in the literature is referred that the appearance of Q waves, *again* or ST segment elevation is evident in 7% of patients, at the time of admission. However, several changes can arise, namely auricular fibrillation, unspecific repolarization changes, evidence of ventricular hyperthrophy. On the other hand, normal ECG occurs in 8 to 31% of patients, i.e., either the presence or absence of electrocardiographic changes has a low predictive value.[7,10]

It seems to be the conjugation of several factors that contributes to increase the diagnosis probability. The presence of a sudden thoracic pain, peripheral pulse asymmetry and mediastine enlargement, a triad present in 27% of patients increases significantly the probability of dissection (positive pv 66.0 CI 95% 4.1-1062.0).[10]

Predisposing factors are: arterial hypertension present in 75% of cases, aorta coarctation, bicuspid aortic valve, vasculitis of the giant cell arteritis type, congenital diseases of the connective tissue as the Marfan syndrome and, less often Ehlers-Danlos syndrome, Turner syndrome, pregnancy, cocaine, intra-aortic catheter, cardiac surgery history.[10]

The main causes of death are aortic rupture with hemopericardium and/or hemothorax, cerebral infarction, visceral ischemia and acute cardiac failure.[10]

Once raised the dissection hypothesis, the supplementary means of diagnosis must be selected appropriate to confirm the diagnosis, according to the available resources and the patient clinical stability. Thorax X Ray, ECG and TTE are compulsory.[11,12] The choice between ETE, thoracic CAT scan, Angio-RM and aortography with a coronariography is not consensual[11,12]. In the IRAD (International Registry of Acute Aortic Dissection) study, the initial test most often was a thoracic CAT scan (61%), followed of a TTE associated to the ETE in 33%, aortography in 4% and the RM in less than 2%.[11] The second diagnosis method was the ETT/ETE in 56% of cases, a thoracic CAT Scan in 18%, the angiography in 17% and the MR in 9%. Most patients have been undergoing several imagiology exams, with an average of 1.8% per patient.[10]

Once established the diagnosed and characterized the dissection, it has a surgical indication in the following situations: proximal aorta dissection, complex distal aortic dissection, distal dissection of the Marfan syndrome.[9]

In the clinical case we present, the final diagnosis was dissection of Stanford Type A and DeBakey Type I associated to an hematoma of the tunica media in the initial portion of the aorta ascending segment and aortic valvular insufficiency. It was further complicated with a myocardial infarction already in subacute stage, result of the compressive effect of the fake lumen associated to the intramural hematoma on the coronaries, which were revealed normal.

In this context, the finding of a pericardial effusion and an increase on the inflammatory endpoints sustain the hypothesis, initially considered of the Dressler's syndrome. This is a possible complication in which the pericardial involvement emerges between 2 to 12 weeks after an acute myocardial infarction, through an auto-immunity mechanism, often associating pericardial effusion to a pleural effusion. It can be associated the general malaise condition to unspecific systemic symptoms, as it was the case of our patient.

An age above 50 years old, the association of systemic symptoms and a marked increase of ESR it still leads to considering the diagnosis of giant cell arteritis, also as a risk factor to the occurrence of aneurysm and aortic dissection. The absence of headache or any abnormality of the temporal artery, the negativity

of auto-immunity markers and the unfolding of the clinical and laboratorial condition, with a regression on the laboratorial changes evident on admission, enable to excluding this diagnosis.

The final hypothesis it is to be a case of atypical aortic dissection, in which all the clinical and laboratorial condition result of the chronic progression of an aortic dissection whose initial condition, not allowing a diagnosis of acute stage, is subsequently masquerade by a constellation of unspecific and unsuspected clinical and laboratorial changes on that diagnosis.

It was due to its rarity, in the form of presentation, that we decided to present and share our experience in this case, highlighting the difficulty that we face when uncommon situations, but serious, as the aortic dissection, manifest themselves in an unsuspected and subacute form. ■

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