SCIENTIFIC LETTER

NATURAL EVOLUTION OF TYPE A AORTIC DISSECTION IN FOUR PATIENTS WITHOUT SURGICAL TREATMENT

ABSTRACT

Introduction and objectives: To describe the characteristics, form of presentation, clinics, treatment and evolution of four patients with type A aortic dissection on whom no surgical treatment was performed.

Methods: A descriptive study of 4 patients with type A aortic dissection is conducted.

Results:

Case 1 – 37-year-old male patient referred from another hospital with a clinical picture of 4-day aortic dissection. A tomography was performed and type A aortic dissection was diagnosed. He remained asymptomatic. Surgical treatment was proposed and the patient refused it. After 10 days, the patient was released with antihypertensive therapy. He has been on follow-up for a year, showing resorption of hematoma of ascending aorta and permeable dissection of descending aorta.

Case 2 – 49-year-old male patient referred from another hospital with a picture of 10-day type A aortic dissection. Upon admission, the patient was asymptomatic and refused surgical treatment, he had dissection and aneurysm of ascending aorta, arch and descending aorta. He was released after 7 days. He has been on follow-up for 22 months with no change of aortic diameters observed. He continues asymptomatic.

Case 3 – 75-year-old male patient with a history of two cardiac surgeries (MRS 12 years ago and AVR 7 years ago). He had had precordial pain radiated to the back 20 days ago. A CAT was performed showing type A aortic dissection. The patient was asymptomatic and refused surgical treatment given the risks. He has been on follow-up for 34 months.

Case 4 – 53-year-old male patient with a clinical picture of 36-hour type A aortic dissection. Upon admission, the patient was asymptomatic. Surgical treatment was proposed and the patient refused it. He was released after 10 days. He has been on follow-up, asymptomatic and with dilation of ascending and descending aorta.

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Conclusion: Type A aortic dissection is a high-mortality disease in its acute form. We presented 4 patients who refused surgery. Today, they remain asymptomatic after an average follow-up of 2 years, with aortic diameters similar to those presented in the acute phase.

Keywords: Aortic dissection. Treatment. Evolution.

INTRODUCTION

Etiology and classification of aortic dissections

The aortic dissection is characterized by the creation of a false lumen in the middle layer of the aortic wall. The types of dissections are classified according to the presence and location of early tears, as well as to the retrograde or antegrade extension of the dissection. The Stanford Group refers to types A and B, depending on whether the ascending aorta is affected by the dissection or not. DeBakey differentiates among type I, when the ascending and descending aortas are affected; type II, when only the ascending aorta is affected; and type III, when only the descending aorta is affected.

A typical aortic dissection (AD) begins with the formation of a tear in the aortic intima, exposing the sick underlying middle layer to pulsatile blood flow. This blood flow penetrates the middle layer, dissecting it and extending distally in variable length, creating a false lumen; occasionally, it extends proximally.

Shear forces may cause the tearing of the inner part of the dissected aortic wall (intimal flap), producing additional entry or exit sites. The distension of the false lumen may stenose and distort the true aortic lumen.

The most frequent location of the primary intimal tear is the ascending aorta between 1 and 5 cm above the right sinus of Valsalva in 65% of cases, the proximal descending aorta below the left subclavian vein in 20% of cases, the transverse aortic arch in 10% of cases, and the distal thoracoabdominal aorta in 5% of cases.

There are predisposing factors for aortic dissection such as age, systemic arterial hypertension (AHT), congenital abnormalities of the aortic valve, inherited disorders of the connective system, traumatic and other factors. AD affects patients between the fifth and seventh decades of life, with more frequency in males (3:1). For people under 40 years old, the frequency is similar in both sexes given the greater frequency of AD in women during the third trimester of pregnancy.

ATH is found in 80% of cases and is the second predisposing factor in terms of importance. Cystic medial degeneration is an intrinsic sign of various inherited disorders of the connective tissue, most notably the Marfan syndrome and the Ehlers-Danlos syndrome, in which there is collagen and medial elastin deterioration, which would be the main predisposing factor for most non-traumatic ADs. Patients with Marfan syndrome,

in addition to their propensity to the development of thoracic aortic aneurysms, have high risk of AD at a relatively young age. Annuloaortic ectasia with idiopathic dilatation of the ascending aorta and aortic insufficiency originates from cystic medial degeneration and also predisposes to AD.

Clinical presentation

The most common symptom is sudden-onset severe pain of a tearing, pulsating nature migrating in the direction of the dissection, located in the front of the chest, neck and jaw when the aortic dissection (AD) is in the proximal aorta or in the interscapular area and the abdomen if the AD is distal, with the incidence of shock and preserved or high arterial tension.

Less common symptoms upon presentation of the AD, with or without associated chest pain, would be:

-Cardiac arrest due to severe aortic insufficiency in proximal ADs;
-Syncope without focal neurological signs due to rupture of proximal AD in the pericardial cavity with tamponade or, less frequently, due to rupture of descending aortic dissection in the left pleural space;
-Stroke, peripheral neuropathies or paraplegia;
-Cardiac arrest or sudden death.

Physical examination

AHT appears in 80%-90% of distal ADs while is less common in proximal ADs. True arterial hypotension is more frequent in proximal ADs due to cardiac tamponade, although distal ADs also cause arterial hypotension due to intrapleural or intraperitoneal rupture. When the dissection occludes brachiocephalic vessels, it is possible to register arterial hypotension (pseudohypotension) inaccurately.

Typical physical signs associated with AD are more characteristic when the proximal aorta is affected. These would be:

-Pulse deficit (50% at proximal AD and 15% at distal AD) due to vascular lumen occlusion by the flap or due to the extension of the very dissection in the artery and compromise of the true lumen by the false channel. Sometimes, pulse deficits are transient due to distal reentry or movements of the intimal flap.
-Aortic insufficiency (AI) is an important sign of proximal AD (50%-66%) with a musical murmur at the right sternal border with intensity depending on blood pressure. Due to the associated cardiac arrest, the murmur and peripheral signs of the aortic insufficiency may disappear. Aortic root and ring dilatation, leaflet retraction, flap prolapse and ring torsion are involved in its origin.
-Neurological manifestations (6%-19%): stroke may occur in 3%-6% due to direct involvement of the common carotid or innominate artery; coma, paraplegia and paraparesis with less frequency.
-Inferior acute myocardial infarction (AMI) (1-2%) due to involvement of the coronary artery ostium because of the flap. The dissection may not be recognized, with catastrophic consequences if treated with thrombolytics.

-Renal infarction, renal failure and severe AHT due to compromised renal artery (5%-8%).
-Ischemia and mesenteric infarction (3%-5%).
-Femoral pulse deficit (12%) due to compromised iliac arteries, with minimum chest pain that may be confused with peripheral embolism.
-Other clinical manifestations may be hemothorax, hemoptysis and hematemesis due to ruptures in the pleural space, bronchi and esophagus. Ruptures in the left or right atrium, in the right ventricle with cardiac arrest have been described occasionally.
-Superior vena cava syndrome, sternoclavicular pulsation, pulsatile neck mass and Horner's syndrome.

The aortic dissection is characterized by the creation of a false lumen in the middle layer of the aortic wall. The types of dissections are classified according to the presence and location of early tears, as well as to the retrograde or antegrade extension of the dissection.

DIAGNOSTIC METHODS

As aortic dissection is an entity with very different forms of clinical presentation, the doctor needs to have a high level of suspicion to establish a quick and accurate diagnosis. The three clinical factors most frequently associated with aortic dissection are a history of arterial hypertension, sudden-onset intense chest pain and pain irradiation.1 These factors, together with a normal electrocardiogram, high blood pressure in anamnesis, the absence of any of the peripheral pulses, the murmur of aortic insufficiency and a widened mediastinum evidenced in the chest X-ray, force the doctor to rule out an aortic dissection.

There is no test of choice in the evaluation of aortic dissections because available techniques have advantages and disadvantages; for this reason, each technique will be more complete in the study of some of the different diagnostic aspects.

Electrocardiogram

The electrocardiogram is normal in most cases. In very hypertensive patients, it may show signs of left ventricular hypertrophy. Although it does not support the diagnosis of aortic dissection, it is essential to differentiate this entity from acute myocardial infarction, since the clinical picture may be very similar. It should be noted that, in cases where the flap dissects the coronary artery, the ECG can reveal signs of AMI.

Chest X-ray

Although it has been suggested that a chest X-ray properly performed and interpreted by experts has high diagnostic accuracy[2], in fact there are unmistakable signs of dissection in less than 30% of patients.[3] However, it is accepted that in the diagnostic process it is essential to have a chest X-ray because it can not only offer compatible signs and consequently support the need for another imaging test to confirm the diagnosis but also identify other causes of chest pain. On the other hand, high interobserver variability is recognized[2], so it should not be the only imaging test in case of a suspected dissection. Radiographic signs suggesting an aortic dissection are[2]: effacement or widening of the aortic knob, pleural effusion (usually left), tracheal deviation, distance over 6 mm between an intimal calcification and the

contour of the aortic wall, widened mediastinum, widened ascending or descending aorta, and widening of the paraspinal line. The separation between the intimal calcification and the aortic contour, a sign widely referred to in the literature, is much unspecified.

Laboratory data

Laboratory data are very non-specific. Leukocytosis is frequent and there may be anemia due to blood sequestration in the false lumen or to blood extravasation from the aorta. Creatinine may increase when renal perfusion decreases. Blood hemolysis in the false lumen causes a lactodehydrogenase rise in some patients[4]. Recently, the detection of specific anti-myosin antibodies has proven to have very high diagnostic accuracy for aortic dissection[5], although it is a method that is not yet available.

Echocardiography

The echocardiographic diagnosis is based on the detection of the intimomedial flap, which divides the aorta into the true and false lumens. The transthoracic echocardiography provides a sensitivity of between 50% and 80% and a specificity of between 70% and 90%.[6,7] The study must include the visualization of the aorta not only through the usual windows (left parasternal and apical) but also through the suprasternal, supraclavicular, subcostal, abdominal and, if there is pleural effusion, subscapular projections. Thus, the aorta is recognized in its entirety. Despite this, visualizing the descending aorta is particularly difficult.[8]

The transesophageal echocardiography (TEE) has changed the diagnostic attitude for aortic dissection radically. The esophagus is in close contact with the aorta, so its study is very comprehensive. The great limitation is the difficulty to study the highest part of the ascending aorta due to the interposition of the trachea and the left main bronchus, although it is very rare that dissections are exclusively located in that region. Adequate sedation of the patient is essential to prevent sudden blood pressure rises that may precipitate an aortic rupture. The sensitivity, specificity and predictive values are above 95%.[9] A transesophageal study must include the following aspects: identification of the flap and the true and false lumens; identification of the entry tear; thrombosis of the false lumen; involvement of the ascending aorta, arch and descending aorta; aortic diameter; aortic valve assessment; involvement of aortic branches including the proximal part of the coronary arteries and supra-aortic trunks; ventricular function study; and the presence of pericardial effusion. In all these aspects, the TEE has proven to have a high level of performance; therefore, if available, it could be the technique of choice. In addition, it is fast and can be performed with no need to transfer to the patient.

Computed tomography

The results offered by the computed tomography (CT) are comparable to those of the TEE. Its sensitivity and specificity are nearly 100%.[10] As compared to other techniques, it has some limitations such as the need for nephrotoxic contrast, the limited

possibility of detecting the entry tear[10] and the lack of hemodynamic information especially concerning the state of the aortic valve. Instead, the CT is widely available and is much less dependent on the operator than the other techniques. In those centers where TEE or magnetic resonance imaging (MRI) is not available, the combination of the transthoracic echocardiography and the CT provides rapid and accurate diagnostic information.[11] Undoubtedly, it is adequate to decide the urgent transfer of the patient to a reference center with cardiovascular surgery or to rule out the diagnosis of aortic dissection.

Magnetic resonance imaging

The MRI can be considered the most complete technique in the diagnosis of aortic dissection, since it allows for a thorough evaluation of the aortic morphology and its surrounding structures.[12] All the characteristics of the dissection studied by other techniques can also be properly evaluated with MRI. The cine MRI technique can also identify aortic regurgitation as it shows the areas of turbulent flow in the left ventricle during diastole.[13] Its great limitations are limited availability, the need for more time for its performance than other techniques and the difficulty of its performance on unstable and intubated patients. Also, it is contraindicated in patients with pacemakers and implantable cardioverter defibrillator.

Angiography

The use of angiography as a diagnostic tool in aortic dissections dates back to 1960. It has a sensitivity of 88%, a specificity of 94% and a diagnostic accuracy for aortic dissections of 98%.

The high-speed injection of iodinated contrast in the aorta allows to study its lumen and branches, including the coronary arteries, and to evaluate the aortic valve and its competence.

The angiographic diagnosis of aortic dissection is based on the demonstration of anatomical anomalies.

Direct signs are: *a)* Presence of an intimomedial flap, visualized as a linear and mobile intraluminal repletion defect; *b)* visualization of a false lumen with less density than the true one and slow washout of the contrast, and *c)* deformation of the border and normal curvature of the aorta due to the compression exerted by the false channel.

Indirect signs are: *a)* rigidity, lack of mobility in a segment of the aortic wall, which may express the presence of hematoma, and *b)* increase in the thickness of the aortic wall above 5 mm.

This technique may fail primarily for the following reasons: a) when the filling of the false channel is very good and the density of contrast is equal to that of the true channel, or when it is very bad and there is practically no passage of contrast; *b)* when the dissection is very small; and *c)* when with the angiography the aortic lumen "is seen" but the wall "is not seen", as in ultrasound images.

In brief, the angiography is a technique with good diagnostic performance for aortic dissections, safe, well tolerated even by patients in a critical situation, and with the advantage that it allows to visualize the state of aortic branches, including the coronary arteries.

MEDICAL TREATMENT FOR AORTIC DISSECTIONS

Once again, it is important to focus on the high index of suspicion, i.e. to think about this entity in order to establish the correct diagnosis of aortic dissection.

Once there is suspicion of this disease, and while waiting for the performance of the appropriate diagnostic studies, the patient must be admitted to the ICU, where blood pressure, heart rate, central venous pressure, urine volume and even pulmonary capillary pressure will be monitored. Pain and arterial hypertension will be carefully treated.

Pain is present in 90% of patients and is often unbearable. Morphine is a good drug given its blood pressure lowering effect, but other analgesics may also be used.

Arterial hypertension is also a very common finding, especially in abdominal aortic dissections. When hypotension is registered, the doctor will need to think about: *a)* cardiac tamponade; *b)* rupture of the aorta to the pleura or the peritoneum, and *c)* "pseudohypotension" due to the dissection of the brachiocephalic trunks.

The most recommended treatment for hypertension, in this entity, is fast-acting vasodilators, specifically nitroprusside. In order to try to avoid the progression of the dissection and the rupture of the aorta, in addition to the control of blood pressure by reducing the systolic level to 100-120 mmHg, the reduction of the speed and ejection force of the left ventricle with beta-blocker therapy was proposed. The most classic guideline is 1 mg of propranolol i.v. every 5 min to get a heart rate of 60-70 beats per minute. Naturally, other beta-blockers with fewer side effects may be used. Calcium antagonists, especially sublingual nifedipine, are also recommended for AHT control. ACE Inhibitors are useful in refractory AHT resulting from renal artery occlusion.

In brief, the medical treatment for aortic dissection is intended to control pain and arterial hypertension and to reduce the speed and ejection force of the left ventricle. All this is aimed at providing patient welfare and avoiding the progression of the dissection.

Once pain and hypertension are controlled and the contraction speed of the left ventricle decreases, the future therapeutic choice differs depending on the anatomical location of the dissection.

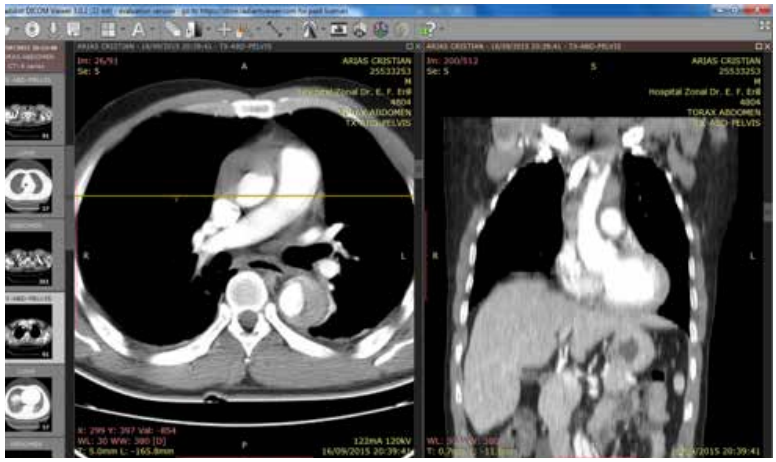
The treatment most widely used for a distal aortic dissection is also medical, with a hospital survival of 80%. This location occurs in older patients with a more advanced atherosclerotic disease and frequently also with a superimposed pulmonary disease. All these factors expose this kind of patients to high surgical risk. Naturally, this therapeutic recommendation is not universal and there are groups that advocate surgical treatment. However, there are studies showing the same results with the medical and surgical treatments of uncomplicated distal aortic dissections. A recent communication describes higher mortality in a group of patients with type B dissection treated with surgery as compared to those medically treated.

RESULTS

Case 1

37-year-old male patient referred from another hospital with 4-day precordial pain radiated to the back. A history of uncontrolled hypertension. A chest tomography was performed upon admission, diagnosing a type A aortic dissection from the arch to the aortic bifurcation. According to the echocardiogram, a dissection flap was observed in the ascending aorta 3 cm from the valve plane, which is thrombosed. Arch and descending aorta dissection flap with permeable false lumen.

He remained asymptomatic since the admission, with control of blood pressure. Surgical treatment was proposed and the patient refused it. After 10 days, the patient was released with antihypertensive therapy. He has been on follow-up for 12 months, presenting resorption of ascending aorta hematoma and permeable dissection in descending aorta without increase of aortic diameters. He continues asymptomatic.



Case 2

49-year-old male patient referred from another hospital with a diagnosis of 10-day type A aortic dissection. Upon admission, he was asymptomatic and presented dissection and aneurysm of ascending aorta, arch and descending aorta. An echocardiogram was performed and a dissection flap was observed in the ascending aorta 2 cm from the valve plane without compromising the valve. A computed tomography (CT) was performed and an aortic dissection was observed in the ascending aorta (with false lumen thrombosed), arch and descending aorta with permeable false lumen. The patient refused surgery. He was released after 7 days. He has been on follow-up for 24 months with no change observed in the aortic diameters. He remains asymptomatic.

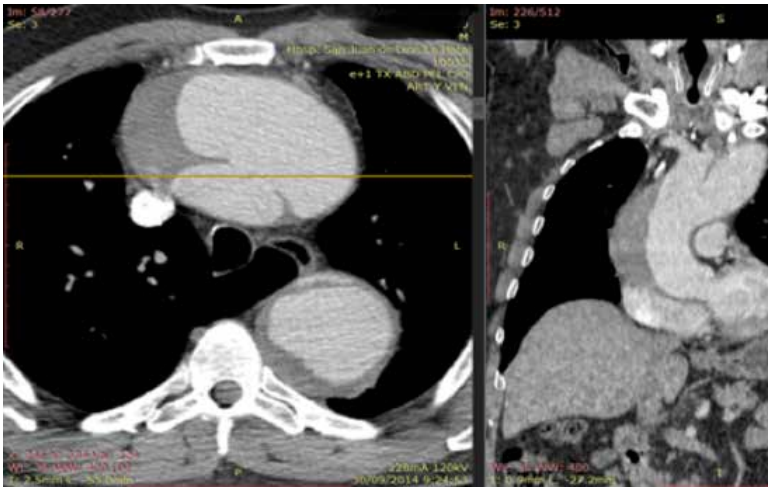
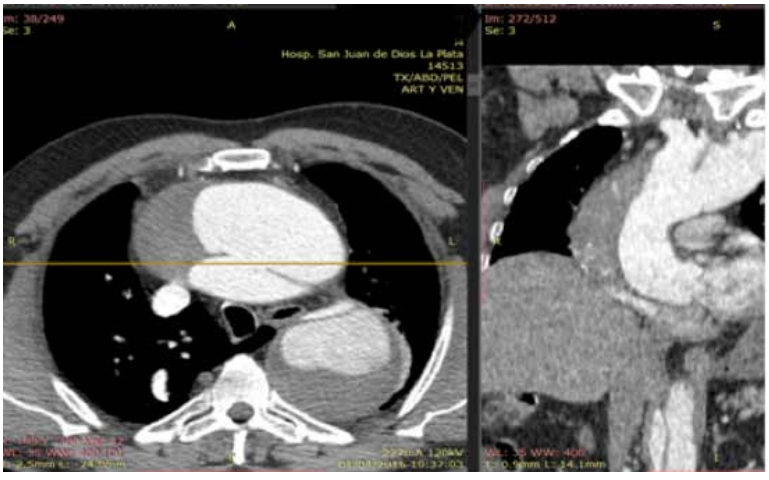


Image of admission in 2014.



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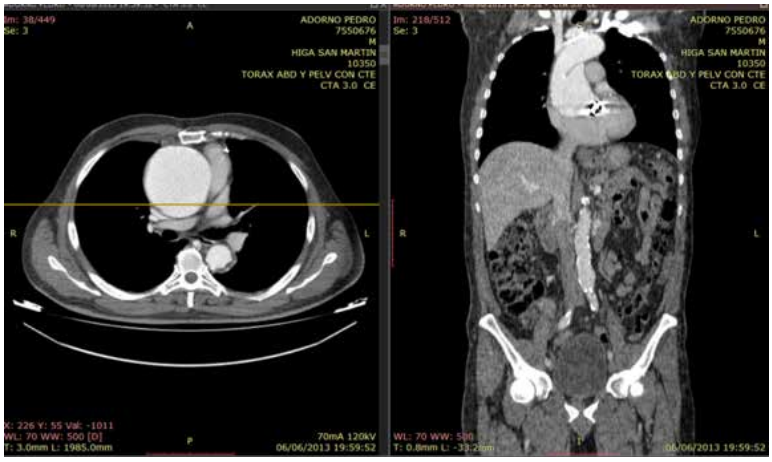
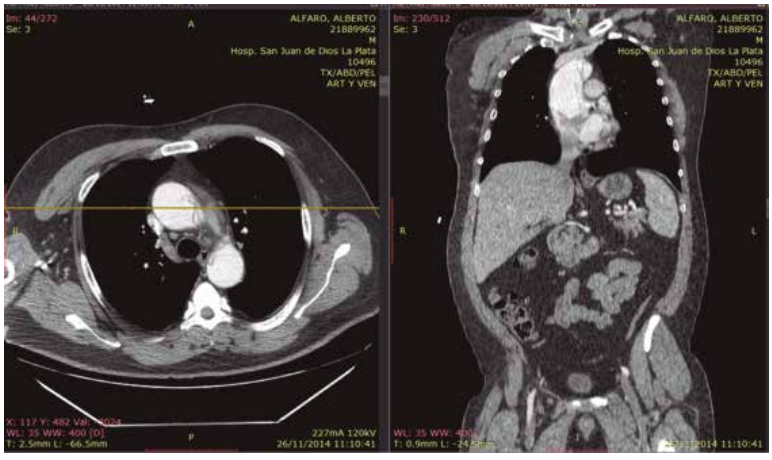
Case 3

75-year-old male patient with a history of two cardiac surgeries (myocardial revascularization with triple bypass 12 years ago and mechanical aortic valve replacement 7 years ago). Doctor's appointment due to 20-day precordial pain radiated to the back. Also a history of poorly controlled hypertension. A CT was performed and a type A aortic dissection was observed with no involvement of the descending aorta. The patient was asymptomatic and refused surgical treatment given the risks. He has been on follow-up for 34 months. He continues with similar aortic diameters.



Case 4

53-year-old male patient with a clinical picture of 36-hour type A aortic dissection. Upon admission, the patient was asymptomatic. A CT was performed and a type A aortic dissection was observed up to the aortic bifurcation. Echocardiogram: dissection flap in the ascending aorta, arch and descending aorta, permeable false lumen. Surgical treatment was proposed and the patient refused it. After 10 days, he was released. He continues on follow-up asymptomatic, with unaltered aortic diameters.



DISCUSSION

Today, no one questions the surgery indication for acute type A aortic dissection. Although our experience includes only 4 cases and there is no literature clarifying the chronic evolution of aortic dissections, these cases show that the aortic diameter remained unchanged, with an average follow-up of 24 months. Questions arise, like what happens with the aortic wall? or would the aortic diameter increase on long-term follow-up?

CONCLUSION

Type A aortic dissection is a high-mortality disease in its acute form. We presented 4 patients who refused surgery. Today, they remain asymptomatic after an average follow-up of 2 years, with aortic diameters similar to those presented in the acute phase.

REFERENCIES

1. Eagle KA, Quertermous T, Kritzer GA, Newell JB, Dinsmore R, Feldman L et al. Spectrum of conditions initially suggesting acute aortic dissection but with negative aortograms. Am J Cardiol 1986;57: 322-326.

2. Jagannath AS, Sos TA, Lockhart SH, Saddekni S, Sniderman KW. Aortic dissection: a statistical analysis of the usefulness of plain chest radiographic findings. Am J Radiol 1986;147: 1123-1126.

3. Luker GD, Glazer HS, Eagar G, Gutiérrez FR, Sagel SS. Aortic dissection: effect of prospective chest radiographic diagnosis on delay to definitive diagnosis. Radiology 1994;193: 813-819.

4. Slater AA, De Sanctis RW. The clinical recognition of dissecting aortic aneurysm. Am J Med 1976;60: 625-633.

5. Suzuki T, Katoh H, Watanabe M, Kurabayashi M, Hiramori K, Hori S et al. Novel biochemical diagnostic method for aortic dissection. Results of a prospective study using an immunoassay of smooth muscle myosin heavy chain. Circulation 1996;93: 1244-124.

6. Kasper W, Meinertz T, Kersting F, Lang K, Just H. Diagnosis of dissecting aortic aneurysm with suprasternal echocardiography. Am J Cardiol 1978;42: 291-294.

7. Victor MF, Mintz GS, Kotler MN, Wilson AR, Segal BL. Two-dimensional echocardiographic diagnosis of aortic dissection. Am J Cardiol 1981;48: 1155-1159.

8. Mathew T, Nanda NC. Two-dimensional and Doppler echocardiographic evaluation of aortic aneurysm and dissection. Am J Cardiol 1984;54: 379-385.

9. San Román JA, Vilacosta I, Fernández Avilés F. Disección de aorta: evaluación clínica, comparación de las técnicas diagnósticas y elección del tratamiento. Rev Esp Cardiol 1996;49 (Supl 4): 2-12.

10. Vasile N, Mathieu D, Keita K, Lellouche D, Bloch G, Cachera JP. Computed tomography of thoracic aortic dissection: accuracy and pitfalls. J Comput Assist Tomogr 1986;10: 211-215.

11. Tottle AJ, Wilde RP.H, Hartnell GG, Wisheart JD. Diagnosis of acute thoracic aortic dissection using combined echocardiography and computed tomography. Clin Radiol 1992;45: 104-108.

12. Panting JR, Norell MS, Baker C, Nicholson AA. Feasibility, accuracy and safety of magnetic resonance imaging in acute aortic dissection. Clin Radiol 1995;50: 455-458.

13. Sechtem U, Pflugfelder PW, Cassidy MM, White RD, Cheitlin MD, Schiller NB et al. Mitral or aortic regurgitation: quantification of regurgitant volumes with cine MR imaging. Radiology 1988;167: 425-430.

14. Doroghazi RM, Slater EE, DeSanctis RW, Buckley MJ, Austen WG, Rosenthal S. Long-term survival of patients with treated aortic dissection. J Am Coll Cardiol 1984;3: 1026-1034.

15. Masani ND, Banning AP, Jones RA, Ruttley MS, Frase AG. Follow-up of chronic thoracic aortic dissection: comparison of transesophageal echocardiography and magentic resonance imaging. Am Heart J 1996;131: 1156-1163.

16. Barbetseas J, Crawford S, Safi HJ, Coselli JS, Quinones MA, Zoghbi WA. Doppler echocardiographic evaluation of pseudoaneurysms complicating composite grafts of the ascending aorta. Circulation 1992;85: 212-222.