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AORTIC DISSECTION - TWO-STAGE RUPTURE

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Abstract: Ankylosing spondylitis (AS) is a chronic inflammation with a wide clinical spectrum of articular and extra-articular changes that occurs in genetically predisposed individuals and is triggered by environmental factors. Cardiovascular involvement in HLA-27 positive spondyloarthropathies has been demonstrated by multiple working groups. All studies share findings that include conduction abnormalities and aortic root pathologies (aortitis-type) as well as aortic valve disease (1-4). Several authors (5-10) have highlighted pathological similarities between AS and Marfan syndrome (MS), suggesting that there may be a common tissue susceptibility factor.

We present the case of a patient with AS who suffered a spinal cord injury and a two-stage bleeding aortic dissection.

Keywords: aortic dissection, ankylopoietic spondylitis, aortitis, collagenosis.

INTRODUCTION

The aorta is the ultimate vascular conduit, carrying nearly 200 million liters of blood through the body during a lifetime. In addition to its conduit function, the aorta plays an important role in the control of systemic vascular resistance and heart rate through pressure receptors located in the ascending aorta and aortic arch. The aorta plays the role of a "second pump" (Windkessel function) during diastole, with a lot of importance for coronary perfusion, but not only. In healthy adults, the diameter of the aorta usually does not exceed 40 mm, and it progressively narrows downstream. Diameters are influenced by several factors, including age, gender, body surface area, and blood pressure. The upper diameter limit is 40 mm for men and 34 mm for women. The rate of aortic diameter expansion is approximately 0.9 mm in men and 0.7 mm in women for each decade of life.

Diseases of the aorta consist of aneurysms, acute aortic syndromes including aortic dissection, intramural hematoma, penetrating aortic ulcer and traumatic aortic injury, pseudoaneurysms, aortic rupture, atherosclerotic and inflammatory conditions, as well as genetic (e.g., Marfan syndrome) and congenital (e.g., coarctation) conditions. The evaluation of the aorta is mainly based on imaging methods: ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI). Endovascular therapies play an increasingly important role in aortic disease, while surgery remains necessary in many situations.

Aortic stiffness is one of the earliest manifestations of adverse structural and functional changes occurring in the vascular wall and is highly recognized as marker in cardiovascular disease. Currently, several non-invasive methods are used to assess aortic stiffness, such as pulse wave velocity and augmentation index. Increased aortic stiffness leads to increased pulse wave velocity in the artery. Recent ESC/ESH guidelines recommend measuring arterial stiffness as part of a comprehensive evaluation of patients with hypertension. The diagnosis of aortic pathology requires increased attention, as the literature presents limited data on the histological patterns of aortic pathology

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in terms of medial degeneration (DM), atherosclerosis (ATS) and aortitis in relation to potential risk factors, as well as their distribution in different aortic segments. While DM is reported to be the main histological finding in cases of aneurysm, the roles of ATS and inflammatory processes seem to be underestimated. A clinical manifestation of all these aortic lesions is aortic aneurysm. As a permanent dilation of the aortic wall, an aortic aneurysm can affect all aortic segments, including the thoracic aorta and abdominal aorta. These aneurysms are classified according to the aortic segments involved, such as aortic root, ascending aorta, arch, or descending aorta for thoracic aneurysms, and adrenal, juxtarenal, and infrarenal aorta for abdominal aneurysms. The etiology, natural history, histology, and treatment differ for each of these segments of aneurysms because the aorta is subject to a variety of diseases, degenerative, inflammatory, infectious, and idiopathic.

Ankylosing spondylitis (AS) is a prototypical disease of the so-called spondyloarthropathies, which also includes other diseases such as undifferentiated spondyloarthropathy, psoriatic arthritis, and inflammatory bowel disease-ankylosing spondylitis related. AS as a definite entity is easy to diagnose, although at the beginning of the disease, symptoms and complementary investigations can be inconclusive. It is known that many undifferentiated

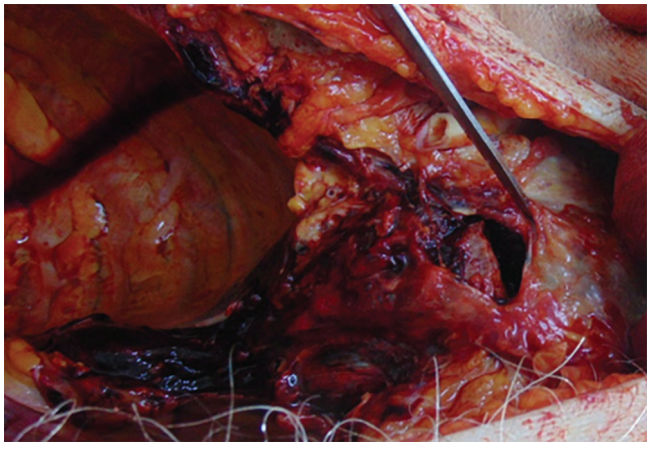

Figure 1. Cervical fracture C5-C6 level.

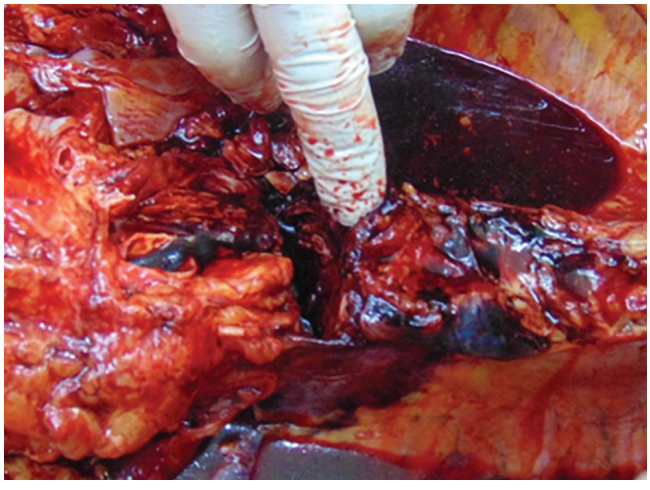

Figure 2. Thoraco-lumbar fracture T12-L1 level.

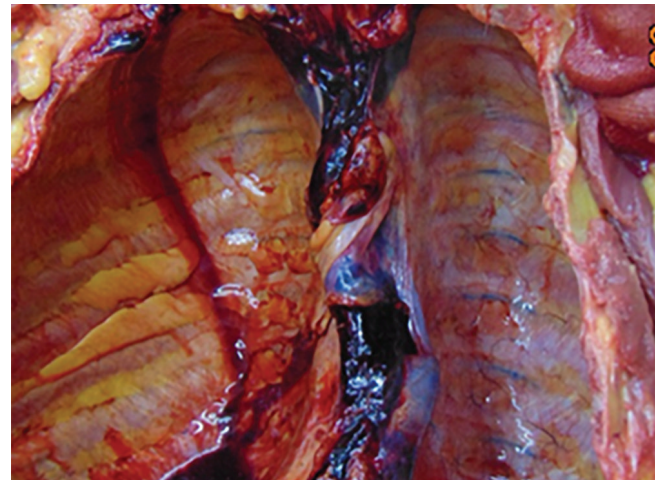

Figure 3. "S" shaped spine.

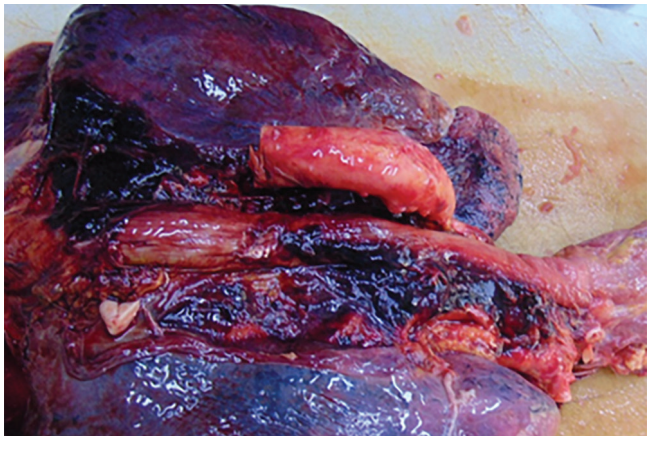

Figure 4. Rupture of the aorta.

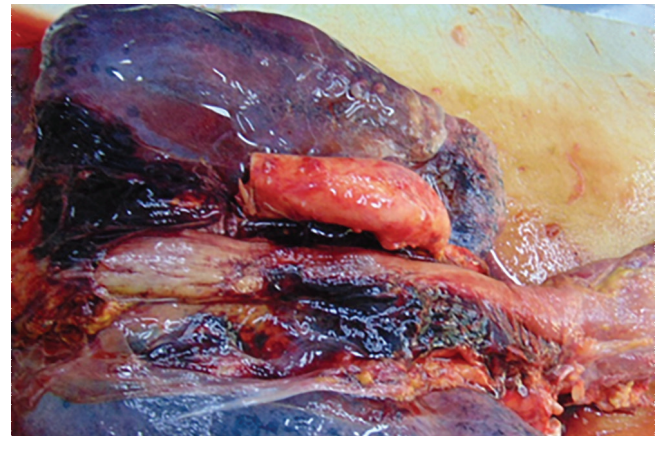

Figure 5. Periaortic hemorrhagic infiltration.

spondyloarthropathies can progress to ankylosing spondylitis (4, 5).

Multiple connective tissue disorders share mutations in the fibrillin-1 (FBN1) gene, so they have acquired the collective name of "fibrillinopathies". Fibrillinopathies comprise a group of tissue disorders in which fibrillin or transforming growth factor-ß (TGF-ß) mutations cause clinical changes predominantly located in the skeleton, aorta, and eye (11). Fibrillin-1 is found in the extracellular matrix having multiple locations (e.g., tendons, ligaments, periosteum, skin, heart valves, aorta, eyes) and has a role in maintaining the structural integrity of tissues and organs. The associated cardiological pathology in patients with AS reveals a specific behavior towards fibrosis of the connective tissue in areas subjected to an increased degree of stress, such as the ascending segment of the aorta artery. Histological studies revealed adventitial scarring, intimal proliferation and aortic wall thickening (12-13).

CASE PRESENTATION

We present the case of a 64-year-old man, known to have ankylosing spondylitis, who suffered a vertebro-medullary trauma through aggression (fall from the same level). After the trauma, the patient loses consciousness for a short period of time (minutes), remains on the ground, he is later helped to get up and goes home. He remains at home for approximately 12 hours with a worsening condition, after which he calls for a medical team and he is taken to a hospital unit. In the emergency room, the patient had a Glasgow score of 15 points, a blood pressure of 143/74 mmHg, peripheral oxygen saturation of 98%, temperature 36oC, he was conscious, cooperative, complained of neck pain and presented with quadriplegia. The CT scan performed reveals a 10 mm diastasis fracture at the cervical C5-C6 level, as well as ankylosing spondylitis changes. The patient is transferred to a specialized hospital unit.

At admission, the patient had a Glasgow score of 15 points, blood pressure of 85/50 mmHg, peripheral oxygen saturation of 99%, ventricular rate of 60 beats per minute. He is admitted to the Intensive Care Unit with the diagnosis of polycontusion due to a fall from the same level, cervical trauma, C5-C6 level cervical spine fracture with diastasis, ankylopoietic spondylitis and quadriplegia. At the clinical examination, the patient was quadriplegic, conscious, cooperative, hemodynamically and respiratory stable. The patient's medical history included stage IV ankylopoietic spondylitis, dementia, bilateral coxarthrosis, strong myopia, amblyopia, chronic duodenal ulcer. The events that followed were hepatocytolysis syndrome and rhabdomyolysis syndrome and on the 4th day of hospitalization, he developed bradycardia, became hemodynamically unstable, and he went into carrdiac arrest that did not respond to CPR.

In this case, a forensic postmortem examination was performed. During the external examination, injuries of violence (ecchymoses and excoriations) were found on the cephalic extremity and in the posterior cervical region. The internal examination reveals significant cerebral edema, C5-C6 cervical fracture with diastasis with subdural blood blade (Fig. 1), right hemothorax in the amount of 2500-3000 ml predominantly liquid blood, rupture of the thoracic aorta in the lower 1/3 with hemorrhagic infiltrated edges and fringes (Figs 4, 6, 7) as well as a massive periadventitial aortic hemorrhagic infiltrate (Fig. 5), the thoracic spine had the appearance of the letter "S" (dextroconvex scoliosis) (Fig. 3), with hemorrhagic

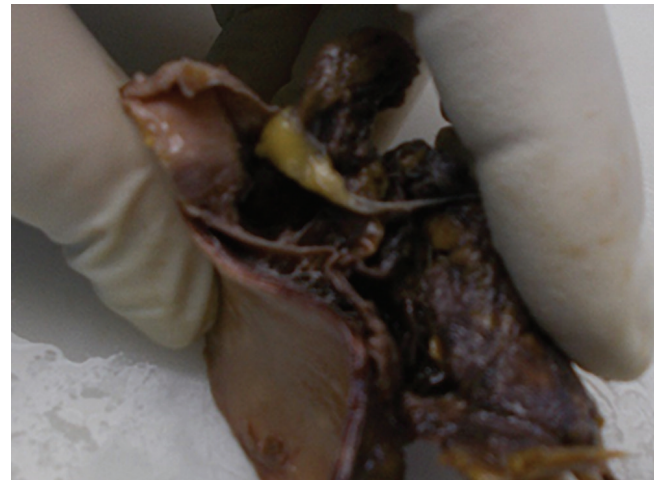

Figure 6, 7. The dissection fold of the aorta after formalization.

infiltrate at the level of perivertebral and periaortic soft tissues, the aortic artery was aligned after the curvature of the thoracic spine, as well as a complete fracture of the T12-L1 vertebral body (Fig. 2). The forensic postmortem examination report concluded the following: the death was violent, due to acute cardio-circulatory insufficiency, the consequence of the rupture of the thoracic aorta in two stages with massive hemothorax in the context of cervical and thoraco-lumbar spine trauma on a background of ankylopoietic spondylitis and aortic atherosclerosis. The established causal link was directly conditional.

DISCUSSIONS AND CONCLUSIONS

In the previously presented case, a traumatic event was added on the preexisting aortic pathology but the moment in time did not coincide with the complete rupture of the aortic wall. This rupture can occur even after a minor event (low intensity trauma, increased intravascular pressure for any reason - including emotional background, especially in association with an increased aortic wall stiffness) or spontaneously, through the progressive weakening of the aortic wall. Aortic rupture is usually fatal because the blood in the aorta is under very high pressure and can quickly exit the vessel through a rupture, leading to profuse hemorrhage and death in about 80% of cases. In the previously presented case, we can take into account a hemorrhage in two stages, if the first stage is considered the traumatic event that contributed to the formation/ aggravation of the aortic dissection on a modified vessel by the pre-existing pathology. The second stage can be both traumatic and non-traumatic, being represented by a hypertensive episode or by an increase in intrathoracic pressure for any reason. The installation of second stage is sudden, well evident by the acute degradation of the patient's condition, the accumulation of a large amount of blood in a short time leading to unsuccessful CPR attempt. We can also subject to the discussion a time 0 represented by the changes of the aortic wall, of inflammatory, atherosclerotic cause or a combination of the two.

Among the extra-articular changes, SA associated with aortitis, this being the most life-threatening manifestation. Aortitis associated with SA involves extensive intimal and periaortic fibrosis phenomena, with the final result being stiffening of the vascular wall, which becomes much more susceptible to ruptures and dissections. In the previously presented case, the patient also associates aortic atheromatosis as an additional risk factor. The two pathologies, aortic changes generated by AS and atherosclerotic changes, constitute in this case time 0 of aortic dissection.

Conflict of interest

The authors declare that they have no conflict of interest.

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