CASE REPORTS

A case of recurrent acute aortic syndrome: Beyond emergency diagnosis and treatment

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Received: March 11, 2018 **Accepted:** May 31, 2018 **Online Published:** June 27, 2018
DOI: 10.5430/crim.v5n3p18 **URL:** https://doi.org/10.5430/crim.v5n3p18

ABSTRACT

Introduction: Acute aortic syndromes (AAS) are emergency conditions with a common pathway but various clinical manifestations. In order to reduce the extremely poor prognosis, these syndromes require a rapid diagnosis and decision making.
Case report: We report the case of a young black woman with recurrent aortic dissection (RAD), presenting to the Emergency Department (ED) with an atypical symptom: ankle soreness. After the surgical treatment, several exams were performed in order to investigate underlying conditions for recurrence: after a first suspicion of tuberculous aortitis, the final diagnosis was Takayasu's arteritis.
Discussion: The aim of this article is to underline the extremely heterogeneous presentation of AAS that worsens the already complicated process in diagnosing the syndromes. Despite uncommon signs and symptoms, identifying patients with a high pre-test likelihood for the disease is crucial to promptly get a correct diagnosis. Once the diagnosis has been confirmed, since AAS may be a spy for important systemic diseases, conditions such as congenital disease, autoimmune and infectious aortitis need to be excluded and treated to prevent any recurrence or systemic implications.

Key Words: Recurrent aortic dissection, Aortitis, Tuberculous aortitis, Takayasu's arteritis

1. INTRODUCTION

Acute aortic syndromes (AAS) are emergency conditions with a common pathway but various clinical manifestations. They affect more frequently adult males with atherosclerotic risk factors.[1] We report the uncommon case of a young black woman with recurrent aortic dissection (RAD).

AAS is still a clinical diagnostic challenge upon initial presentation: only 15%-43% of cases are diagnosed on initial evaluation and up to 30% remain undiagnosed antemortem.[2] With a mortality of 1% per hour after symptom onset in untreated patients, early diagnosis is mandatory, followed by appropriate surgical or medical interventions.[3] Hence a multidisciplinary approach is requested to investigate all the possible aetiologies, beyond the well-known risk factors, especially among young patients with recurrency.

2. CASE REPORT

JR, a 25-year-old African lady was admitted to the ED with a green code triage. She was complaining for acute leg pain, in particular at the right ankle, for a week. Her blood pressure in the last week had an average level of 80/50 mmHg.

Six months before the patient had undergone an intervention

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of ascending aortic arch and descending aorta substitution with tubular prosthesis, after acute dissection of a saccular aneurysm of aortic arch and descending aorta. In the past medical history, a mild anaemia had been recorded. She was on pharmacologic treatment with bisoprolol 2.5 mg, acetylsalicylic acid 100 mg, and pantoprazole 20 mg.

On the last ED admission she was alert, and afebrile. She had good bilateral air entry, and oxygen saturation of 99% in room air. Blood pressure was 80/40 mmHg, and pulse rate at 60 beats/min.

On physical examination she presented rhythmic heart sounds, with a murmur located in aortic focus (Grade 3/6 Levine). No abnormalities were recorded in lung and abdominal examination. Her limbs were warm to the touch. Pulses were present, symmetrical, and synchronous. She didn't have peripheral oedema. There was no sign of deep vein thrombosis. Also, there were no focal neurological signs. However, she was walking with difficulty.

No abnormalities were found in the EKG, or in the bed-side ultrasonography performed in the ED. Blood gas analysis and first line laboratory tests were normal.

Although the patient reported hypotension for more than a week, in the past medical records her blood pressure values were higher, while being on the same therapy, and the systolic murmur wasn't reported. Given the known history of previous AD, the hypotensive state and the new systolic murmur of the heart sounds, we referred the patient to abdomen and thoracic CT-scan to exclude AAS. The radiologist described a left subclavian artery from the right common trunk with opacification defect of about 15 mm and downstream reentry. The right subclavian artery was dilated with tortuous course. In the thoracic-abdominal aortic passage multiple images of parietal ulcers and dissections with ectasy of aortic lumen and a maximum transversal diameter of 36 mm were recorded. There were no signs of active bleeding. Multiple big necrotic lymphnodes in periaortic-caval area, in the mesenteric root, medially to the small gastric curvature, to hepatic and splenic pedicle, with a maximum diameter of 3 cm were also noted (see Figures 1 & 2).

The patient underwent a surgical vascular intervention with the implant of endovascular prosthesis between descending thoracic and abdominal aorta, on top of the celiac trunk (see Figure 3).

During her permanence in ICU several exams were performed in order to explore underlying conditions for recurrence.

In the suspicion of great vessels vasculitis, she underwent a rheumatological consultancy which suggested measurement of several autoantibodies (ANCA, ANA, anti-ENA, anti-DNA and total IgG and IgG4), which were negative, and a CT-PET that showed abnormal concentration of the radiopharmaceutical at the apex of the aortic arch, which, for its focal characteristic, was suspect for inflammatory pathology in the active phase.

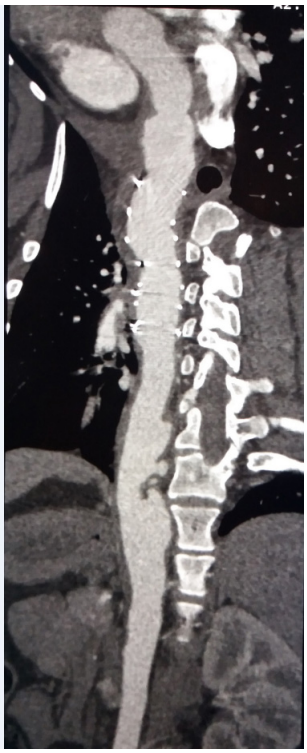

Figure 1. Coronal TC scan showing, in the thoracic-abdominal aortic passage, multiple images of parietal ulcers and dissections with ectasy of aortic lumen and a maximum transversal diameter of 36 mm. The previously implanted prosthesis is evident.

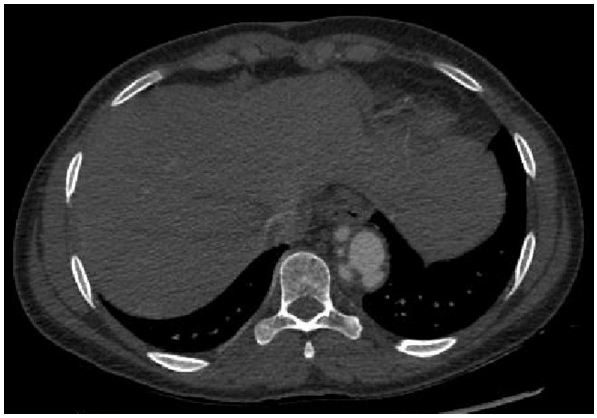

Figure 2. Axial TC scan of the abdomen confirms the presence of AAS

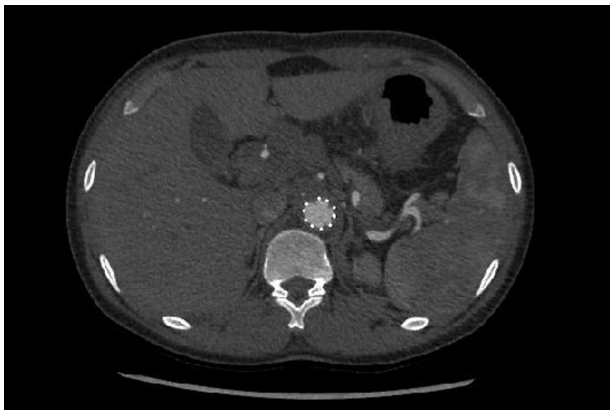

Figure 3. Axial TC scan performed after treatment shows the endovascular prosthesis between descending thoracic and abdominal aorta, on top of the celiac trunk

Lab tests for viruses – such as HCV, HBV, and HIV – were negative, as well as Treponema, and urinary acid-alcohol resistant bacilli (BAAR). TB-quantiferon and IgG Ab anti-Chlamydia turned out to be positive.

Because of the CT evidence of abdominal necrotic lymph nodes, she had one surgically removed. Microbiologic analysis did not find BAAR, and the histologic examination described non-specific granulomatous lymphadenitis necrotizing with areas of confluent necrosis. PCR for M. Tuberculosis complex done on lymph nodes tissue resulted positive; therefore an empiric antibiotic therapy for abdominal tuberculosis started. Finally, cultural analysis confirmed the presence of Mycobacterium tuberculosis.

After six months follow-up, a PET-TC was repeated showing the same abnormal concentration of the radiopharmaceutical at the apex of the aortic arch compared to the one performed at the beginning. An Eco-colour-power Doppler ultrasonography and an angio-RMN of neck and supra-aortic trunks were performed, showing thickening with enhancement of the wall of the thoracic aorta as an active process, and the stenosis of the right brachiocephalic trunk, left common carotid, left subclavian was recorded, while the right subclavian was aneurysmal and irregular.

The distribution of the disease, together with the patient's age and her symptoms allowed to formulate the diagnosis of Takayasu's arteritis. The patient was then subjected to specific immunosuppressive therapy. Currently, she is followed closely, is in good general conditions and no further episodes of acute aortic syndrome have occurred.

3. Discussion

AAS are considered rare diseases, with an incidence of 3-5 cases on 100,000 inhabitants.[4] Acute aortic dissection (AD) comprises 85% to 95% of all AAS,[5] the incidence increases with age and is higher in males. When compared to men, women affected by AD present more frequently atypical symptoms leading to delayed diagnosis and higher mortality (30.1% for women versus 21.0% for men).[5] Black patients with AD are younger than white patients.[5]

In the IRAD registry (International Registry of Acute Aortic Dissection) people younger than 40 years are only 7%.[6] RAD[6] is reported in younger patients[7] who generally experience proximal followed by distal AD.[8] They are more likely to have Marfan syndrome (MFS) and, between non-MFS patients, infectious and non-infectious aortitis need to be considered.[1]

The atypical clinical presentation of our young patient did not help us in the diagnostic process since she came to the ED for a symptom apparently not related with the final diagnosis (ankle's soreness). The in depth and meticulous examination of the patient in the ED led to consider the whole signs (the systolic murmur not recorded previously and the hypotension) as branches of the same tree, together with a past significant medical history.

The ischemic genesis of leg pain, reported in less than 10% of patients with aortic AD,[1] was excluded as both anterior tibial pulses were present without sign of ischemia. Other possible causes are the occlusion of spinal arteries, which can be responsible of acute paraplegia, painless or similar to Leriche syndrome;[1] in 10% of patient, a lower limb ischemic neuropathy is present, caused by a deranged perfusion of the femoral territories.[1] Furthermore, aortitis is connected with limb claudication, usually of rapid onset, progressive and bilateral.[1] Therefore, lower limb claudication, associated with the multifocal aortic lesions, the gender and the younger age, strengthened the suspicion of an inflammatory aortic disease.

Even if aortitis, defined as an inflammatory thickening of the aortic wall from varying etiologies associated with specific histopathological lesions,[9] is a rarer risk factor for aortic dissection, 1% to 5% of patients develop AD(3). Among autoimmune aortitis, the most frequent are Takayasu's arteritis and giant cell arteritis.[10] Also medium-vessel vasculitis (polyarteritis nodosa and Kawasaki disease), ANCA-associated vasculitis and a new category called variable vessel vasculitis (Behcet's disease and Cogan's syndrome) can affect aorta, although seldom.[11] Furthermore, aortitis may be part of the spectrum of IgG4-related disease, an heterogeneous group of disorders, recently described, that can be responsible of 50% of isolated aortitis, periaortitis or retroperitoneal fibrosis.[11] In our patient IgG4 antibodies were normal.

Laboratory inflammatory parameters and autoantibodies are useful in differential diagnosis but CT or MRT are mandatory to reveal aortitis. PET may be even more sensitive. The definitive diagnosis is obtained with biopsy.[11]

In our case, elevated levels of hsCRP and procalcitonin strengthened the hypothesis of infectious aetiology. Our patient had positive IgG Ab anti-Chlamydia Pneumoniae, which seems to be responsible of aortic wall degeneration through the production of metalloproteinases that destroy the matrix proteins of the aortic wall resulting in aneurysmal dilatation.[12,13] Between infectious aortitis, the most frequent are Staphylococcal and Streptococcus pneumonia associated, usually affecting the thoracic aorta. Salmonella species usually affect the abdominal aorta.[11] Also Treponema Pallidum can be responsible of aortic localizations almost entirely in the ascending aorta or in the aortic arch.[11] Negative VDRL-test made the diagnosis of syphilitic aortitis unlikely in our patient. Aortitis by Mycobacterium tuberculosis is a rare disease. It involves generally descending aorta or aortic arch and is responsible of aneurysmal dilation with secondary dissections or rupture, or stenotic lesions with coartation. Mycobacterium tuberculosis may reach the aortic wall by hematogenous spread with direct implant on the intima, or to the adventitia and media by the vasa vasorum; indirectly via the lymphatics or by direct extension from a contiguous focus such as a lymph node or paraspinal abscess or pulmonary lesions.[14] A review of cases of mycobacterial aneurysm of the aorta found only 41 cases between 1945 and 1999, and in the 63% of cases a countiguous tubercular lymph node was described.[15]

The suspicion of tuberculous aortitis must arise in any patient presenting with aortitis or aortic aneurysm who has evidence of pulmonary or extra-pulmonary tuberculosis at present or in the past.[16] The definitive diagnosis is most often established by histology in surgical specimens, with evidence of granulomatous aortitis with caseous necrosis.[16]

The histologic observation of granulomatous necrotizing abdominal lymphadenitis and positivity of PCR for M.Tuberculosis led to start empiric therapy for TBC. Finally, positive cultural exams for M. Tuberculosis confirmed the diagnosis of abdominal tuberculosis.

The presence of an active abdominal tuberculosis suggested at the beginning that tubercular aetiology was the most likely diagnosis for RAD in our patient, even if the definitive histologic confirmation of granulomatous aortitis lacks in our case and is objectively difficult to be obtained.[14]

After 6 months of treatment, a resolution of aortic inflammation was expected. The persistence of uptake in the aortic arch imposed the need to look for other possible diagnoses. A more detailed study of the large arterial vessels was mandatory, and the anomalies highlighted in the course of the epiaortic vessels, together with the age and the history of our patients made Takayasu's arteritis the most probable etiology.[17] In fact, this autoimmune disease defined "the woman's pulseless disease" has usually two clinical stages: the first, paucisymptomatic due to chronic inflammation, and the occlusive phase, when organ damage is already evident. In this second stage, signs of peripheral hypoperfusion are evident, such as limb ischemia manifested as claudication in lower limbs but, more frequently, in upper limbs. American College of Rheumatology published the classification criteria of Takayasu arteritis as follows: age 40 years or younger at disease onset; claudication of extremities; decreased brachial artery pulse; systolic blood pressure difference of greater than 10 mmHg between arms; bruit over subclavian arteries or abdominal aorta; and patognomonic angiographic findings.[17] In our patient, we found almost three of this criteria, so that diagnosis is highly probable (sensitivity 90.5%, specifity 97.8%[17]). Also Ishikawa criteria are satisfied.[9]

Differential diagnosis between tuberculous aortitis and Takayasu's aortitis is very difficult without the anatomopathological data. Tuberculous aortitis usually causes true or false aneurysms of the descending thoracic and abdominal aorta, through a mechanism of erosion of the vessel wall. While dissection and rupture are important complications of tuberculous aortitis, stenoses are typical of Takayasu's arteritis.[18] Although it is commonly thought that aortic involvement is a late complication of systemic vasculitis, this case, together with other rare cases series, shows that the aortic damage can be the first sign leading to the diagnosis of vasculitis.[19–21]

4. Conclusion

The first key message that this case can teach is that AAS can have an extremely heterogeneous presentation, worsening the already complicated process in diagnosing the syndromes: it is mandatory to suspect it to reach the diagnosis. The second one is that a recurrent AAS may be a spy for important systemic diseases. Therefore a multidisciplinary approach to this pathology is essential, aimed at a quick diagnosis and treatment first, and at researching and treating the triggering causes later, to avoid complications and recidivism.

Conflicts of Interest Disclosure

The authors declare no conflicts of interest.

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