

An incidental finding of localized aortic arch dissection in a polytraumatized patient. A case report and state of the art of *non-A non-B* aortic dissection

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Abstract

Non-A non-B aortic dissection is considered a rare nosological entity, included in the Stanford classification, representing a small percentage of the total aortic dissections that occur annually. Regarding this form, the literature reports a more complicated disease course compared to other types of dissection. We describe the case of a 76-year-old patient who accessed the triage section of an emergency department for a polytrauma picture and, after surgical treatment of a leg fracture, received a diagnosis of *non-A non-B* aortic dissection, "localized" to the arch and incidentally detected. The angio-computed tomography (CT) showed that the only intimal tear was located in the central portion of the aortic arch, with no exit tear. No signs of malperfusion or clinical symptoms related to the aortic finding were evident. The Aortic Team decided on a conservative approach, whereby the patient was started on medical therapy to control blood pressure in a monitored bed of a semi-intensive care unit. The persistent asymptomatic state, a condition of hemodynamic stability, and an unchanged angio-CT picture enabled discharge on day 7 and the assignment to a close follow-up.

Introduction

Non-A Non-B type aortic dissection is considered a rare nosological entity, accounting for a small percentage of the total number of dissections that occur annually. Although it is included in the Stanford classification (Figure 1) [1], it does not fall within other standardized classifications for aortic dissection, despite medical literature reporting a more complicated disease course for this form compared to other types of dissection. In the case reported here, we describe a "localized" *non-A non-B* type aortic dissection, found incidentally in a polytraumatized patient and treated conservatively with medical therapy.

Case Report

A 76-year-old man was taken to the Emergency Department of the "Madonna del Soccorso" Hospital in San Benedetto del Tronto (Italy) following a car accident. His medical history included arterial hypertension, type 2 diabetes mellitus, dyslipidemia, and

smoking. Clinical evaluation showed a normal state of consciousness but with signs of blunt right hemithorax trauma and right knee pain. Thoraco-abdominal and neurological imaging tests were normal. Physical examination demonstrated a blood pressure of 135/65 mmHg, a heart rate of 75 bpm in sinus rhythm, and an oxygen saturation of 96% in room air. Abnormal laboratory values included: white blood count 18,200, hemoglobin 11.4 g/dL, platelet count 610,000, uric acid 10.6 mg/dL, creatinine 1.9 mg/dL, and blood urea nitrogen 161 mg/dL. Radiological skeletal tests documented a proximal, multifragmentary, intra-articular fracture of the tibia and fibula with an external tibial plateau depression and a simple fracture of the right IX rib's lateral arch. The patient underwent open reduction and internal fixation of the tibia and fibula fractures. Furthermore, due to a nodular opacification in the right upper hilum (approximately 1.7 cm in diameter) on chest X-ray, a high-resolution chest computed tomography (CT) scan was scheduled for further in-depth analysis.

The chest CT confirmed the nodular formation with irregular and “spiculated” edges (maximum diameter of approximately 2.5 cm) in the right upper lung lobe and revealed the presence of an intimal dissection flap in the aortic arch, approximately 4 cm in longitudinal diameter, starting just distally to the emergence of the left subclavian artery, without evident involvement of the supra-aortic vessels. The subsequent thoracic aorta angio-CT diagnosed a *non-A non-B* dissection, associated with focal dilation of the aortic arch (maximum diameter of 40 mm) with regularity of the supra-aortic vessels at their origin and of the descending aortic tract. The intimal tear was very unusual and located in the central part of the aortic arch; no exit tear was found (Figures 2 and 3).

Considering the division of the aortic arch into three parts as reported by von Segesser LK (Figure 4) [1], the entry tear was located in segment 2B (including the origin of the left common carotid vessel). No evident signs of malperfusion and/or clinical symptoms related to the aortic finding were present. Thus, this was an E2/M0 type *non-A non-B* aortic dissection, according to the Type-Entry-Malperfusion classification (E2: Entry-Tear in the arch; M0: absence of malperfusion) [1,2].

During hospitalization, transthoracic echocardiography revealed a left ventricle of normal dimensions and volumes, with mild concentric wall hypertrophy (indexed mass of 125 g/m²), normal systolic function indices (left ventricular ejection fraction by Simpson's biplane method of 55%), and no abnormalities of regional wall motion. The aortic root had a normal size (maximum diameter of 35 mm at the Valsalva sinuses), while a dilation of the ascending aorta was evident (maximum diameter of 42 mm in the first segment). At right parasternal and suprasternal views, the distal portion of the ascending aorta and the aortic arch were poorly

visualized, and no significant valvular dysfunctions were noted. We proposed a transesophageal echocardiogram, but the patient refused esophageal probe introduction. Considering the clinical picture and hemodynamic stability, the aortic team decided on a conservative approach consisting of complete monitoring in a semi-intensive care unit, medical therapy to control blood pressure (systolic pressure was always maintained below 120 mmHg) and heart rate (always 60-80 bpm in sinus rhythm). β -blockers, calcium antagonists, and angiotensin II antagonists were used. Following an unchanged picture at a check-up thoracic aorta re-angio-CT 7 days after admission, given the persistent asymptomatic state and after consultation with the aortic team, the patient was discharged in stable hemodynamic conditions, with antihypertensive therapy to be continued at home and a follow-up re-angio-CT scheduled in 3 months.

Discussion

As clearly reported in a recent article by Di Marco *et al.* [2], *non-A non-B* type aortic dissections, within the Stanford classification framework, are defined as an intimal tear occurring beyond the ascending aorta, initially involving the aortic arch and either remaining isolated to it or extending distally (Figure 1) [1]. Forms that remain isolated to the arch are also referred to as “localized”. There is considerable debate on the definition of *non-A non-B* aortic dissection. Koechlin *et al.* state that for the diagnosis of *non-A non-B* dissection, it is mandatory to locate the entry tear within the aortic arch, and extensions to the arch from entry tears originating in the descending aorta are not to be considered as *non-A non-B* forms [3]. For these reasons, the *non-A non-B* dissection is also identified as aortic arch dissection, meaning the part of the thoracic aorta that goes from the pericardial sac to the isthmus, identified as the attachment point of the arterial ligament, distally to the left subclavian artery [4]. However, *non-A non-B* type aortic dissections are considered rare forms and have a mortality rate that is between that of type A and type B Stanford forms, with a lower incidence compared to type A but higher than type B (from 3% to 11% of all acute aortic dissections). The population most at risk for such lesions tends to be younger than that affected by type A dissections. Medical literature reports that about 88% of patients with *non-A non-B* aortic dissection progress to disease complications, and 29% of these patients may present signs of malperfusion [4]. The “gold standard” for diagnosis remains thoraco-abdominal angio-CT for its reliability in confirming the presence of the dissection and its extension, the exact localization of the entry tear, the

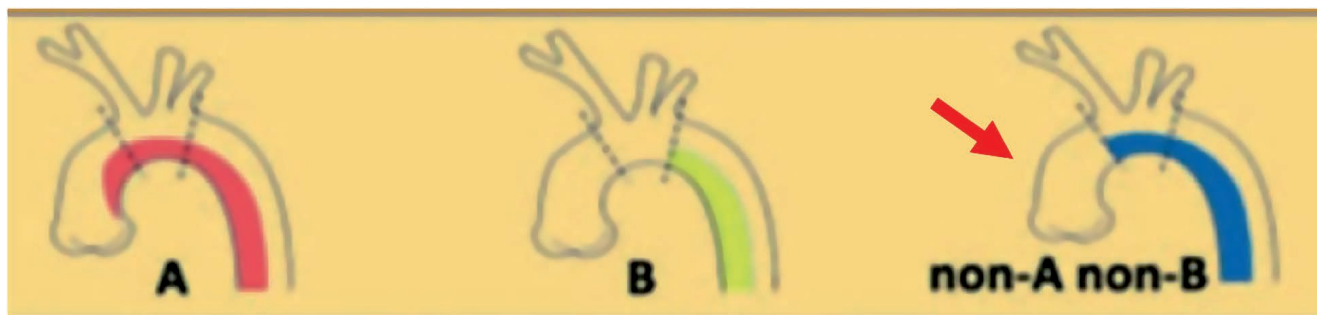


Figure 1. *Non-A non-B* dissection according to the Stanford classification. Modified from: Sievers *et al.* (2020).

anatomy of the flap, the morphology and relationships of the true and false lumen, the origin of the visceral vessels from the true or false lumen, and thus the presence of malperfusion. In the same way, the number and location of possible re-entry tears, the diameters of all aortic segments, and any multiple tears are correctly evaluated by the same radiological technique. All this information is essential for surgical planning [2]. Currently, there is no established gold standard for the treatment of acute *non-A non-B* type aortic dissection, as the medical literature available to date is still significantly lacking on the topic. Compared with type A dissection, in which surgical treatment is mandatory and with time-dependent effectiveness, in *non-A non-B* forms, we may have multiple choices of management. Among the possible approaches, surgical treatment is more commonly

utilized, while medical therapy alone is usually reserved for more stable cases. Thoracic endovascular aortic repair constitutes a significant portion of endovascular treatments for *non-A non-B* dissections, while medical therapy consists of the control of blood pressure levels through careful monitoring and management of antihypertensive therapy. The medical literature reports a significant difference in outcomes between the two types of management, with a documented 30-day mortality rate of 14% and 3.6% for patients treated with conservative medical therapy and endovascular or open surgery, respectively [5].

In a case series of 31 patients, Koechlin *et al.* sought to determine whether there was a significant difference in outcomes between conservative and surgical treatment in isolated aortic arch dissection [3]. The authors concluded that, in the

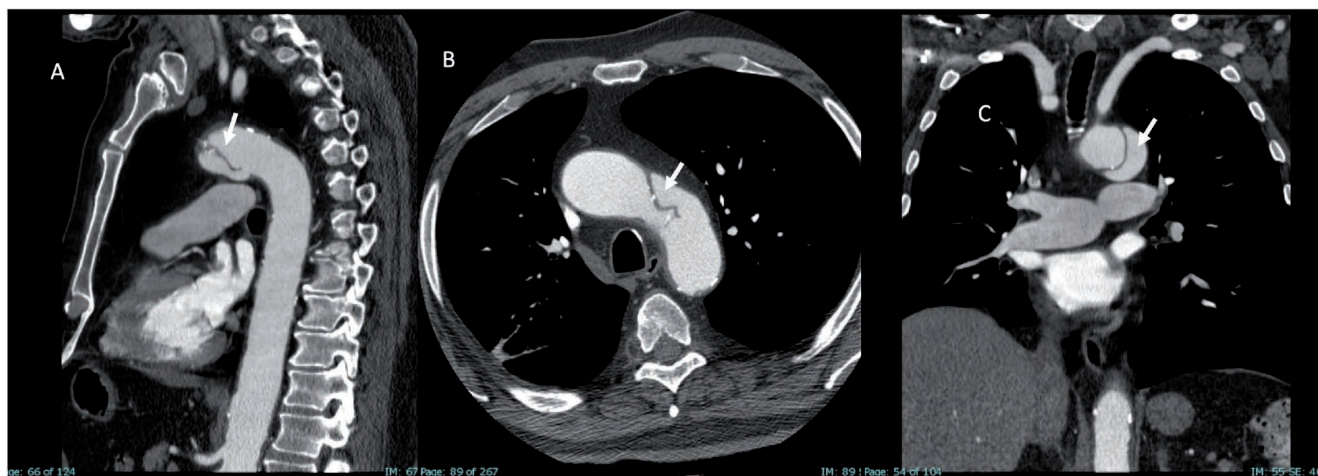


Figure 2. Angio-computed tomography representation of the thoracic aorta showing (arrows) the dissection flap in the aortic arch in longitudinal (A-B) and transverse (C) projections.

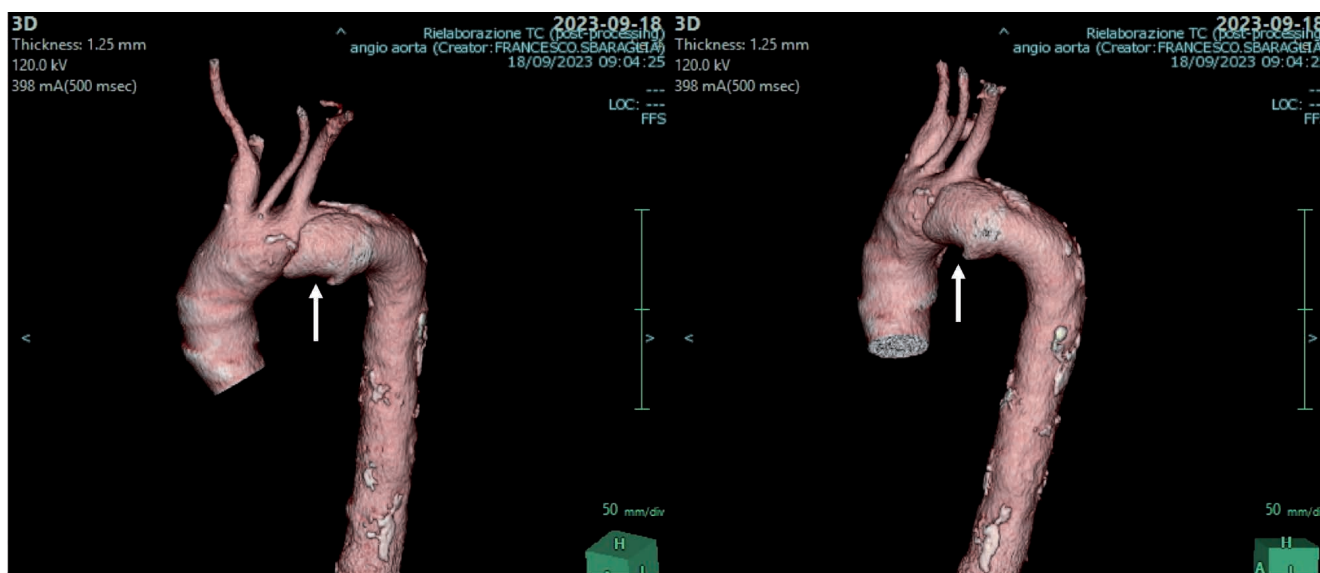


Figure 3. 3D volume-rendering processing of the thoracic aorta angio-computed tomography, showing the “localized” dissection of the aortic arch (arrows).

mid-term follow-up (up to 2 years), there was no significant difference between the two types of treatment in terms of survival or acute neurological events. However, in the long-term follow-up (beyond 2 years), there was a high percentage of referrals to surgery in the group initially assigned to conservative treatment.

A study conducted by Urbanskia *et al.* [6], which compared two groups of subjects undergoing medical and surgical therapy

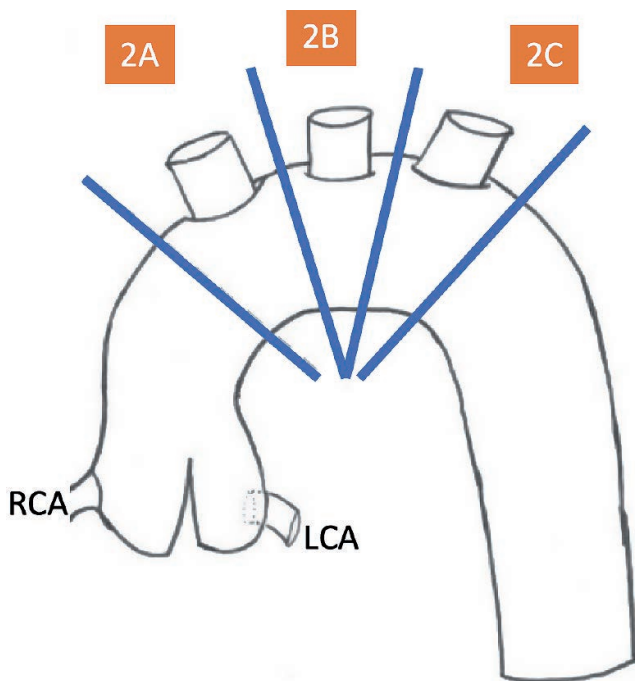


Figure 4. Aortic arch anatomy with subdivision into three parts. Modified from: Sievers *et al.* (2020).

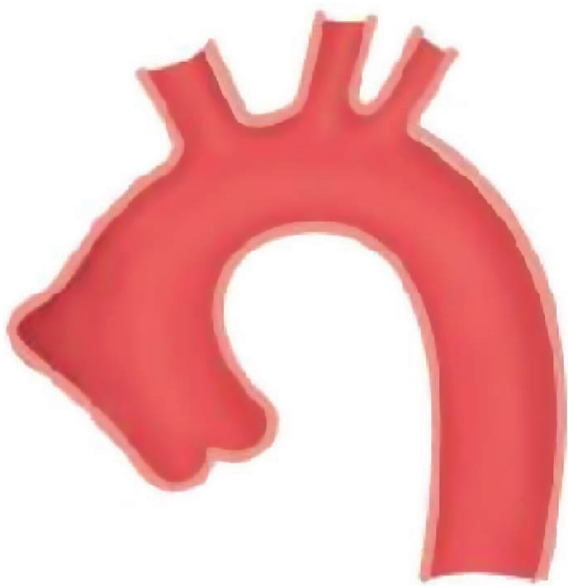


Figure 5. Class III of acute aortic syndromes. Modified from: Erbel *et al.* (2014).

respectively, concluded that open surgery or endovascular treatment should be the preferred option in acute *non-A non-B* type aortic dissection, where the dissection extends through most of the aortic arch, regardless of whether the tear is in the arch or in the descending aorta with subsequent retrograde propagation.

In our case, the entry tear is identifiable in the tract just distal to the emergence of the left subclavian artery, not involving either the ascending aorta or the origin of the supra-aortic vessels. In addition to the entry tear, no exit tears are visible, and, importantly, there is no involvement of the descending portion. Therefore, this case can be defined as a “localized” form of *non-A non-B* aortic dissection, uncomplicated, in a patient asymptomatic for chest pain and hemodynamically stable, in which the diagnostic finding is thus incidental. The evidence thus far reported supported the decision to treat the patient conservatively with medical therapy, in a hospital setting, with an excellent short-term outcome. The possibility of consulting the aortic team, consisting of a cardiologist, cardio-surgeon, vascular surgeon, angiologist, anesthesiologist, and radiologist, allowed for a reasoned, evidence-based, and thus far effective decision. The patient was nevertheless assigned to a careful and close follow-up with radiological re-evaluation in 3 months, in addition to the prescription of home antihypertensive therapy with strict monitoring of blood pressure values and ensuring open access to the reference Cardiology Unit in case of symptom occurrence. The 2014 European Society of Cardiology guidelines reported *non-A non-B* aortic dissection as class III of acute aortic syndromes - subtle or discrete aortic dissection with bulging of the aortic wall (Figure 5) [7].

In 2019, the European Association for Cardio-Thoracic Surgery and the European Society for Vascular Surgery published an expert consensus document for the treatment of thoracic arch pathologies, in which they added a third category called “non-A-non-B dissection”, to be used for patients whose proximal dissection flap begins in the aortic arch [8]. The 2022 American College of Cardiology/American Heart Association aortic disease guideline cited this document [9].

The same guideline cited a paper published by Rylshi *et al.* in which *non-A non-B* dissection was classified as descending-entry type with entry distal to the left subclavian artery and dissection extending into the aortic arch, and arch-entry type with entry between the innominate and left subclavian arteries [10]. The authors concluded that the acute *non-A non-B* aortic dissection frequently requires emergency aortic repair due to organ malperfusion or aortic rupture. Most descending-entry and arch-entry *non-A non-B* dissection patients undergo aortic repair within 2 weeks after dissection onset.

Conclusions

In conclusion, *non-A non-B* aortic dissection is a rare clinical entity and is rarely reported in the literature, requiring further prospective studies both to improve our understanding of the mechanism underlying its occurrence and to obtain more substantial evidence on the correct management and referral to the different surgical techniques currently available and/or medical therapy. The case reported here can be considered rare and unusual because the dissection appeared to be of limited extent, isolated to the aortic arch, with a single-entry tear and no exit tear. The activation of an aortic team, indispensable in such cases, allowed for reasoned management based on the scientific evidence available today.

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