Diagnosis and Management of Acute Aortic Syndrome: A Literature Review

Dedy Irawan, Novi Kurnianingsih, Djanggan Sargowo, Anna Fuji Rahimah

Background: Acute aortic syndrome (AAS) is a potentially fatal disease involving the acute disruption of the aortic wall. Several conditions, such as intramural hematoma (IMH), aortic dissection (AD), penetrating atherosclerotic ulcer (PAU), as well as high mortality rates, are associated with this disease. Hypertension is an essential risk factor, and patients often present with pain. AAS may be challenging to be evaluated, as it has a pre-test probability and several diagnostic tests.

Objective: This literature review will discuss the proper diagnosis and management of Acute aortic Syndrome patients.

Discussion: Computed Tomography Angiography (CTA) is essential in managing AAS. The significance of prompt diagnosis and treatment of AAS is emphasized further by the current developments in imaging techniques and therapeutic interventions, ultimately leading to increased vigilance of this disease. Management of AAS included initial medical therapy and definitive therapy.

Conclusion: In A type AAS, surgical intervention is the definitive therapy, while type B is managed definitively by endovascular or medical treatment.

Keywords: Diagnosis; Management; Acute Aortic Syndrome.

One of the most perilous emergencies in cardiology is acute aortic syndrome (AAS), which is defined as acute disruption of the aortic wall. Several conditions, such as intramural hematoma (IMH), aortic dissection (AD), penetrating atherosclerotic ulcer (PAU), as well as high mortality rates, are associated with this disease. This condition affects 20 to 40 people/1000 000 population annually, with most of them in the form of aortic dissection (80%), followed by IMH and PAU (15% and 5%, respectively). According to the data, within the 20 years of 1990-2010, there was an increase in the overall global death rate due to aortic aneurysms and aortic dissection from 2.49 per 100 000 to 2.78 per 100 000 residents with a higher tendency for elderly males. However, despite improvement in surgical techniques, early mortality rates of type A Aortic dissection remains constant during the last 30 years at 20% to 35%. Corroboration of clinical suspicion, localization of tears, classification, and determination of urgency and the extent of dissection are the primary goals of diagnostic imaging studies upon clinical suspicion of AAS.

2. Discussion

2.1 Pathophysiology of Acute Aortic Syndrome

Histopathologic and genetic elements play essential roles in the diverse pathophysiology of acute aortic dissection. Acute aortic syndromes may develop during a buildup of blood within the tunica media of the aorta due to vasa vasorum rupture or whenever blood infiltrates the media through the lumen due to tear or ulcer (Figure 1). Dilation or rupture of the aorta may occur due to the inflammatory response in the media towards blood. This response will eventually end up in rupture or dilation of the aorta.

PAU was hypertension, hypercholesterolemia, and smoking. The significance of prompt diagnosis and treatment of AAS is emphasized further by the current developments in imaging techniques and therapeutic interventions, ultimately leading to increased vigilance of this disease.

Corroboration of clinical suspicion, localization of tears, classification, and determination of urgency and the extent of dissection are the primary goals of diagnostic imaging studies upon clinical suspicion of AAS.
In essence, every mechanism that compromised the media of the aorta may ultimately cause intramural hemorrhage and aortic dissection and rupture due to the formation of aneurysms and dilatation of the aorta caused by higher wall stress. In acute aortic dissection, medial wall degeneration or medial cystic necrosis is causing the tear in the intima layer of the aorta.1,9

The blood flow within the aortic wall causes the dissection to propagate in an antegrade and retrograde manner. Whenever there is an involvement of aortic side branches, complications in the form of aortic valve insufficiency, perfusion impairment, and tamponade arise [10]. Enhanced fluorodeoxyglucose (FDG) uptake on PET scan shows the degeneration of elastic tissue, necrosis, and apoptosis of smooth muscle cells.11 An increased risk of rupture in patients with concurrent inflammatory disorders such as Takayasu’s disease, Behçet’s syndrome, or polyarteritis nodosa, highlights the significance of this continued inflammation.12 There are multitudes of varying factors that promote AAS.

2.2 Classification of Acute Aortic Syndrome

2.2.1 Aortic Dissection (AD)

In essence, aortic dissection is the separation of aortic wall layers due to the disarrangement of media-induced by intramural hemorrhage, which leads to the formation of a true and a false lumen with or without communication. Accumulation of blood within the media in a dissection plane is instigated by the tear in the intimal layer. Blood then re-enters the aortic lumen via another tear in the intima, and aortic rupture may also occur due to the disruption of the adventitia layer. The risk of medial rupture is increased due to the degeneration of elastic tissue and necrosis and apoptosis of smooth muscle cells triggered further by inflammatory response to thrombus within the media.4 Type A and B AD may account for 62% and 38% of cases, respectively.13

2.2.2 Intramural Haematoma (IMH)

IMH is a condition where a hematoma develops in the media of the aortic wall in the absence of a false lumen and intimal tear. The finding of 5-mm circular or crescent-shaped thickening of the aortic wall in the absence of detectable blood flow is the basis for intramural hematoma diagnosis. Around 10%-30% of cases are attributable to Type A IMH (ascending aorta and aortic arch), while Type B (descending thoracic aorta) is responsible for 60%-70% of cases.4

2.1.3 Penetrating Atherosclerotic Ulcer (PAU)

Penetration of ulcerative atherosclerotic plaque in the aorta into the medial layer is termed as PAU. A pseudoaneurysm, IMH, acute AD, or aortic rupture may occur due to the extension of the ulcerative process. It is expedited in ascending aorta (Type A). The presence of multiple variable sized extensive atherosclerosis in the thoracic aorta with variable depth within the wall might increase the likelihood of encountering PAU. The Middle and lower descending thoracic aorta are the most prevalent location of PAU (Type B). The aortic arch or abdominal aorta might also be the sites for PAU, albeit less commonly, while ascending aorta is a much rarer sight to be involved.4

2.3 Diagnosis

2.3.1 Clinical Presentation

Irrespective of the underlying condition (e.g., contained aortic rupture, AD, PAU, or IMH), there is a similarity in the presentation in AAS patients. Acute aortic dissection patients most frequently present with a chief complaint of pain. Approximately 90% of over 1000 cases showed a severe pain pattern and occurred abruptly in 84% of cases. The pain is often described as stabbing or sharp, which fluctuates, contrasting with the classical description of acute dissection pain ripping or tearing. The location of initial disruption might be inferred based on the pain location and respective symptoms. The involvement of ascending aorta might be indicated by pain that radiates to the neck, throat, or jaw; especially those which occur concurrently with pulse differentials, signs of tamponade, or aortic regurgitation murmur; on the contrary, dissection of the descending aorta must be suspected when the pain is located at the back or abdomen. The pain of aortic origin must not be mistaken for acute coronary syndromes.10,13 Clinical findings alone might be insufficient to distinguish between classical dissection and IMH and PAU clearly. Patients often present with abrupt severe chest or back pain, especially in the elderly; however, this pain only seldomly manifests as signs of organ malperfusion and pulse deficit.7 Several major causes for delay in diagnosing AAS include heart failure, syncope, and abdominal pain.12 Pre-test probability might influence the outputs of diagnostic tests. A risk assessment tool was developed by the ACC/AHA. This tool scores patients’ pain features, clinical examination, and predisposing conditions on a scale of 0 (none) to 3.4

2.3.2 Laboratory testing

Upon clinical suspicion of CAD in patients presenting with chest pain, laboratory tests are needed to determine differential diagnosis and complications. These tests include complete blood count, procalcitonin, C-reactive protein, Troponin I or T, Creatine kinase, D-Dimer, Aspartate transaminase, Creatinine, lactate, blood gas, and glucose.4 One of the most promising diagnostic biomarkers to achieve the gold standard as a form point-of-care test is D-dimer. Clinical suspicion of AD is raised whenever there is an increase of D-dimer (cut off value: 500 ng/ml). There is also a piece of good evidence that states D-dimer’s potential ability to rule out AD.14,15

2.3.3 Electrocardiography

Electrocardiography (ECG) is mandatory testing for all patients. It may distinguish between different origins of pain, such as those due to acute myocardial infarction in which anticoagulant might be used in the management, from those due to aortic dissection in which anticoagulant is contraindicated. However, it is essential to note that both conditions may occur concurrently and can only be elucidated by thorough clinical evaluation and testing. Around 31% of the patients had normal ECG, followed by nonspecific ST and T-wave changes, ischemic changes, and evidence of an acute MI in 42%, 15%, and 5%, respectively.1

2.3.4 Chest X-Ray

Widening of the aorta is one of the possible findings in conventional chest radiography in cases of aortic dissection. Aortic kinking, widening of the aortic contour, opacification of the aorticopulmonary window, and displaced calcification are other examples of probable findings in this condition. Around 60%-90% of cases with suspected aortic dissection might display abnormalities in their chest radiography, while the remaining 10%-20% will have a normal chest x-ray. However, additional imaging studies need to be performed in almost all patients due to the limited sensitivity.1

2.4 Imaging Studies

2.4.1 Echocardiography

The use of transthoracic echocardiography (TTE) to facilitate prompt diagnosis in an emergency setting as a part of a multidisciplinary approach has been widely acknowledged.
Important information which might be helpful in scenarios where prompt decision-making is required might be obtained by performing focused TTE on the aortic valve and aorta segments along with a glimpse of left heart function. Potentially life-threatening complications of AD can be rapidly detected by TTE, and such complications include wall motion abnormalities, pericardial effusion, aortic regurgitation, and cardiac tamponade; however, TTE has relatively low accuracy in detecting AD (Type AAAD has a sensitivity of 78–100%, while Type B only has 31–55%). Thus, AAD cannot be ruled out based on a negative TTE.15

Transesophageal echocardiography (TEE) can visualize the aorta better than TTE, with higher image quality and spatial resolution to evaluate true lumen compression, secondary communications, and primary entry tear better. Combined with color flow patterns and flap movement, these parameters may have prognostic value. Intra-procedural monitoring can also be done by leaving the ultrasound probe in place.12

Thickening of the aortic wall can also be assessed using TEE. Even though it cannot differentiate between acute versus chronic hematoma, it is also valuable for establishing other differential diagnoses that involve aortic wall thickenings like aortitis or severe atherosclerosis. In any imaging modality, the thickness of greater than 5 mm in the presence of clinical symptoms suggesting AAS might point to a diagnosis of IMH. Echoluent or echo-free space inside the thickened aortic wall is a hallmark finding of IMH. There is a considerable amount of challenge in diagnosing IMH in its early state. Visualization of small communications and assessment of the intima is best performed using TEE with color Doppler. In TEE, PAU might appear a localized crater-like protrusion of the aortic lumen without intimal flap.7

2.4.2 Computed Tomography (CT)

CT is the most commonly used modality due to its speed and high sensitivity in detecting AD (95%). The overall accuracy is 96%. Findings of highly attenuated hemorrhagic collection within the mediastinum, pericardium, and pleura, as well as active contrast extravasation, are considered diagnostic.17 Usually, in an emergency department setting, a CT scanner is located within a reachable distance to enable unstable patients to be rapidly examined radiographically. Rapid interpretation upon completion of the examination is a must, as it will enable rapid triage decisions. Nonspecific acute chest pain may be evaluated by multi-detector helical CT.9

The presence of two lumens separated by the intimal flap is an essential finding on contrast-enhanced images. Multplanar reconstruction images are essential in helping to corroborate the diagnosis and determine the extent of involvement (mainly when aortic branch vessels are involved), even though AD can be diagnosed using transverse CT images. Multidetector CT plays a pivotal role in measuring the length, true lumen (TL) and false lumen (FL) of the aorta, the extent of dissection, the distance from the intimal tear to the vital vascular branches, and involvement of vital vascular structure in a precise and specific manner. Slower flow and a large diameter lumen that may contain thrombi are characteristics of FL. ‘Cobweb sign’ is a hallmark finding for FL. It is characterized by low attenuation of slender linear areas, which depicts the incompletely dissected media. FL in type A AD then extends distally, in a spiral fashion. In many instances, TL is the lumen that extends more caudally. It is essential to differentiate between FL and TL to decide when to consider endovascular therapy.4

The mainstay protocol for CT examination for chest pain is the triple rule-out protocol, the purpose of this protocol is to differentiate between acute aortic dissection, pulmonary embolism, and acute coronary syndrome. Future triple rule-out protocols may potentially diminish the number of diagnostic tests, time for triage, radiation exposure to the patient, and emergency department costs, even though limitations do exist. Motion artifacts that may be similar to findings of type A AD may pose a diagnostic challenge in the evaluation of AD, particularly in ascending aorta; however, ECG gating might solve this issue.8 The crescentic or circular highly-attenuated area on CT, which did not enhance after contrast medium injection, is diagnostic for IMH.7 Contrast-filled pouch-like protrusion in the thickened aortic wall is the typical finding for PAU in an atherosclerotic process. Besides ulceration, extensive atherosclerosis is also commonly found.7

2.4.3 Magnetic Resonance Imaging (MRI)

A comprehensive evaluation of aortic dissection might be obtained from an MRI examination, as it can merge anatomical and functional aspects in imaging. Pre- and post-treatment evaluation of the aorta is frequently done by contrast-enhanced MR angiography (CE-MRA). CE-MRA has a high tissue contrast and a short scan time. Labeled agents used with CTA are less preferred than gadolini-um-based contrast due to their higher nephrotoxicity. Rarely, nephro-genic systemic fibrosis as an adverse reaction may occur. Assessment of flow dynamics might be conducted using time-resolved MR angiography (MRA), which yields additional information and images. Sequential visualization of the true and false lumen is done by rapid acquisition of MRA sequences synchronized with the ECG. MRI is typically slower than CT, imaging, despite improved MR scan times due to image acceleration techniques. MRI is expected to be extensively used with better availability.7 CMR can easily detect crescentic aortic wall thickening without the intimal flap; methemoglobin formation within the hematoma causes the signal of the thickened aorta to intensify, which resulted in increased signal intensity on T1-weighted images in subacute IMH. However, it is important to remember that to correctly identify the extent of IMH and periaortic bleeding, the broad field of vision of CT and CMR must be taken into account. Several frequent findings of IMH include the presence of fluid extravasation, mediastinal hemorrhage, as well as pericardial and pleural effusion.7 Irregular contrast-illuminated ulcerative plaque outpouching beyond the aortic intima with a variable degree of associated IMH is a typical finding for penetrating ulcers.20

2.5 Other technique

2.5.1 Aortography

Historically, the gold standard for diagnosing AAS is retrograde aortography, with a sensitivity of up to 90% and specificity of > 95%. The angiographic diagnosis of AD is based upon ‘direct’ angiographic signs, such as the visualization of the intimal flap (a negative, frequently mobile, linear image) or the recognition of two separate lumens. The invasive, time-consuming, and high-cost natures of this modality are the reasons why this modality is no longer used to diagnose AAS.6,15

2.5.2 Intravascular ultrasound

Intravascular ultrasound is a reliable and safe tool to guide stent-graft positioning and provide real-time imaging of aortic pathology. However, the shortcoming of Doppler capabilities and the likelihood of off-center measurements continue to be an important limitation. Another possible approach to guide fenestration procedures is to identify communications between the true lumen and false lumen.15

2.5.3 Positron Emission Tomography

Currently, the role of 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography (PET) has grown in importance during
the evaluation of pathological processes of the aortic wall (i.e., inflammatory and/or infectious processes). The limited spatial resolution of PET has drastically improved due to PET/CT angiography or magnetic resonance angiography image acquisition, resulting in excellent anatomical visualization of mural or luminal abnormalities, enabling accurate localization of lesions. Increased levels of inflammatory serological markers (CRP and D-dimer), enhanced 18F-FDG uptake in the wall of an acute dissected aorta as shown by PET signal, and/or thrombus renewal/lysis (D-dimer, plasmin–alpha 2–antiplasmin complexes, thrombin–antithrombin III complexes, and P-selectin), may possess a potential added value of risk prediction for follow-up.12

2.6 Management of Acute Aortic Syndrome

2.6.1 Initial Medical Therapy

The goal of initial medical therapy in all AAS patients is to reduce wall stress (irrespective of the definitive treatment) to reduce the risk of developing end-organ damage and rupture and restrain the extension of the dissection. It is imperative to provide adequate pain relief (intravenous opiate analgesia) and control of systolic blood pressure ranges from 100 to 120 mmHg and heart rate less than 60 bpm. Intravenous beta-blockers (labetalol, propranolol, esmolol, or metoprolol) are the first-line drugs for these purposes. A feasible alternative for patients who cannot tolerate beta-blockers is non-dihydropyridine calcium channel antagonists (i.e., verapamil and diltiazem). Concurrent administration of vasodilators (intravenous sodium nitroprusside) and beta-blockers might be required to rapidly attain the optimal level of blood pressure.13 Other treatment includes restriction of strenuous physical activity, administration of long term sodium nitroprusside, pain control using opiate, achieving an LDL cholesterol target of less than 70 mg/dL by administration of statin (IIaA), and cessation of smoking (IB).12

2.6.2 Definitive Therapy

Asian, European, and American practice guidelines contain recommendations for the definitive treatment of AAS, which are slightly different among one another in terms of the level of evidence underlying these recommendations (Figure 8).14,17 AAS of the ascending aorta is considered surgical emergencies; one may consider a hybrid approach in selected cases, which is to combine endovascular and open approaches. On the contrary, medical treatment should be instituted in cases of acute aortic pathology limited to the descending aorta, except in the presence of complications (i.e., progressive dissection, organ or limb malperfusion, uncontrolled hypertension, intractable pain, or extra aortic blood collection (impending rupture)). However, most recommendations are based on Level C evidence because large RCTs regarding AAS are still lacking.10,18

The benefit of thoracic endovascular aortic repair (TEVAR) in managing patients with type B AD is still controversial. In order to be able to identify optimal indications for TEVAR, large-scale RCTs with long-term follow-up are still required. Investigation of Stent Grafts in Aortic Dissection trial found an improvement in 5-year survival, positive remodeling, and delayed disease progression upon the use of TEVAR along with optimal medical treatment; and yet, the results also revealed that there was no difference in total mortality.13,19,20

3. Conclusion

AAS is a potentially fatal disease besides ACS, which mandates timely management. Clinical features of AAS and its subsequent consequences are crucial to be comprehended. Imaging is an integral part of AAS management, with CTA being the most prominent modality. CTA has the advantage of being able to evaluate predictors of progression, ascertain complications and end-organ ischemia. Moreover, it can also visualize the anatomy and the extent of lesions well and can be performed swiftly. Recommendations for the management of AAS included initial medical therapy and definitive therapy. In type A AAS, surgical intervention is the definitive therapy, while type B is managed definitively by endovascular or medical treatment.

4. Declaration

4.1. Ethics Approval and Consent to participate

Not applicable.

4.2. Consent for publication

Not applicable.

4.3. Availability of data and materials

Data used in our study were presented in the main text.

4.4. Competing interests

Not applicable.

4.5. Funding source

Not applicable.

4.6. Authors contributions

Idea/concept: DI. Design: DI. Control/supervision: NK, DS, AF. Data collection/processing: DI. Extraction/Analysis/interpretation: DI. Literature review: DI. Writing the article: DI. Critical review: NK, DS, AF. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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References


