CASE SERIES

Acute aortic dissection type A: case series and insights on incidence, management and outcomes

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Abstract

Background: Acute aortic dissection (AAD) is a life-threatening condition with high mortality rates, despite significant advances in surgical approaches. The understanding of the clinical presentation and outcomes is crucial in order to upgrade management strategies. However, epidemiological data regarding AAD occurrence are scarce in Europe, highlighting the gap of evidence in the existing guidelines.

Case Series: We investigated 197 consecutive patients admitted to our institution from January 2018 to December 2019 with suspicion of type A AAD, conducting a retrospective case series. All demographic characteristics, as well as the outcomes of these patients, were recorded and further analyzed to deliver data on the epidemiology of AAD. A total of 197 patients were admitted to our hospital with a suspected AAD. Forty-one (25.9 %) patients presented with a dilated aortic lumen or with a previously repaired aortic dissection, while 28 patients (14.2 %) were diagnosed with AAD (14 patients with type A AAD, 13 with type B AAD and 1 with intramural hematoma). Among 14 patients with type A AAD, nine patients (64.0 %) were treated surgically, while the rest were managed conservatively due to futile clinical status or inability for immediate transportation to a surgical facility. The most frequent initial symptom was chest pain in 86.0 % of patients, followed by dyspnea in 42.9 %. Post-surgical mortality was 33.0 %, while all patients that were managed conservatively did not survive. D-dimers on arrival were significantly lower among patients who survived compared to those who did not.

Conclusion: The incidence of type A AAD in our case series was consistent with the one demonstrated in other international cohorts; however, the mortality in our patient group was higher. Our results encourage surgical treatment due to a lower in-hospital mortality rate when compared to conservative treatment. HIPPOKRATIA 2021, 25 (1):42-46.

Keywords: Acute aortic syndromes, acute aortic dissection, aortic aneurysm, Stanford type A

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Introduction

Acute aortic dissection (AAD) is a devastating condition of the thoracic aorta with critical clinical presentation and extremely low survival rate even after surgical or endovascular treatment[1-3]. AAD is the most common form of acute aortic syndromes (AAS) constituting an emergency condition; AAS also include intramural hematomas or hemorrhages, subtle dissections without hematoma, penetrating atherosclerotic ulcers, and iatrogenic or traumatic AAD[4]. The rapid symptom onset and the similar to myocardial infarction clinical manifestation of type A AAD may lead to incorrect diagnosis and mistreatment as an acute coronary syndrome[5]. The delayed or missed diagnosis composes the primary factor leading to a low survival rate, accompanied by the non-specific clinical signs and symptoms and the limitations in the existing diagnostic and therapeutic strategies. Cardiac tamponade, heart failure, and myocardial ischemia are the primary causes of death among patients with AAD[6].

High clinical suspicion after an initial physical examination and prompt initiation of surgical treatment after rapid diagnosis are of paramount importance with an eye towards the decrease of in-hospital mortality rates[7]. Transthoracic and transesophageal echocardiography, widely used in the emergency department (ED), can contribute to the rapid diagnostic approach of AAD. However, chest computed tomography angiography (CCTA) is an indispensable imaging method for the conclusive diagnosis of AAD, with 100 % sensitivity and 98 % specificity[8]. Magnetic resonance imaging (MRI) and invasive aortography are rarely used as diagnostic imaging techniques, as they are more time-consuming than the aforementioned examination methods[3].

Understanding the clinical presentation, epidemiolog-

ical characteristics, and outcomes of AAD patients is crucial in developing targeted therapeutic strategies. However, epidemiological data concerning the occurrence of AAD are lacking both in Europe and worldwide, thus the gap of evidence in the existing guidelines is broad[4]. Herein, we present a case series of patients presenting to the ED of our hospital with AAD, reflecting the incidence, clinical presentation, management, and outcomes of this particular emergent cardiovascular condition in a local cohort. For all we know, no epidemiological data exist so far regarding AADs in Greece. Thus, this is the first attempt to enhance current evidence regarding this critical medical condition.

Methods

We retrospectively analyzed consecutive patients presenting to the ED of our tertiary hospital, from January 2018 to December 2019, with a suspicion of AAD leading to diagnostic imaging. The clinical suspicion of AAD was high in patients with abrupt onset of chest or back pain, focal neurological deficit, hypotension, shock or tamponade, and signs of end-organ hypoperfusion. The CCTAs of patients were analyzed, and all available clinical data from patients were recorded. The widely accepted Stanford system[9] was used for the anatomical classification of the affected aorta: type A if the ascending aorta was affected and type B if the ascending aorta was not affected[10]. Only patients with the first incidence of type A AAD were selected for our case series, while patients with extension of the dissection or patients under reevaluation of previously repaired aortic dissection were excluded. The dissection was considered acute when symptoms occurred within 14 days prior to the ED admission, and the surgical repair was noted as urgent when conducted within three days after symptom onset[11]. Our center was a tertiary hospital without an in-site cardiothoracic surgery department; therefore, patients selected for surgical treatment were transferred to affiliated hospitals. The postoperative outcome of these patients was followed up using hospital records and telephone-based interviews.

Statistical Analysis

Numeric variables are presented as numbers for categorical characteristics, as mean ± standard deviation (SD) for data showing normal distribution, and as median [interquartile range (IQR)] for data with skewed distribution. Differences between groups were analyzed using Fisher's exact test for categorical variables, and continuous variables were compared using Student's t-test or the Mann-Whitney U test, as appropriate. All reported p-values were two-sided, and those <0.05 are considered to be statistically significant. We performed data analyses with the IBM SPSS Statistics for Windows, Version 25.0 (IBM Corp., Armonk, NY, USA).

Case Series

Patient characteristics

Between January 2018 and December 2019, a total of 197 patients were admitted to our hospital with a suspected AAD. Among them, 118 patients (60 %) had normal aortic lumen, and 41 cases (25.9 %) presented with a dilated aortic lumen or a previously repaired aortic dissection. Individuals with normal aortic lumen or with previously surgically repaired aortic dissection were excluded from further analysis. In total, 28 patients (14.2 %) were diagnosed with AAD. Among these, type A AAD was the most common disease entity (n =14/28; 50 %), followed by type B AAD (n =13/28; 46.4 %) and intramural hematoma (n =1/28; 3.6 %) (Figure 1). We, therefore, located a total of 14 patients with type A AAD; demographics and other baseline characteristics, as well as clinical features of patients included in our case series, are presented in Table 1.

Incidence of type A AAD

Our case series involves patients presenting with a type A AAD to a tertiary university hospital, part of a healthcare system covering a geographic area with an estimated population of 1,105,176 during the period 2018-2019. The estimated incidence of AAD was five cases per 100,000 [95 % confidence interval (CI): 3.74 to 6.38], while the incidence of type A AAD was 2.53 per 100,000 (95 % CI: 1.75 to 3.67). At the same period, 221,094 patients were admitted to the ED of the hospital, while only 11,612 required cardiological consultation. Cases holding high clinical suspicion of AAD, patients with AAD diagnosis, and patients with type A AAD diagnosis constituted the 1.7 %, 0.25 %, and 0.12 % respectively of cases who needed cardiological consultation.

Aortic dissection type A patients

The mean age of individuals with type A AAD was 58.9 ± 15.8 and 69.4 ± 11.4 for males and females, respectively, while the male-to-female ratio was 9:5. Nine patients (64.3 %) were treated surgically, while 5 (35.7 %) were managed conservatively due to futile clinical status or inability for immediate transportation to a surgical facility. Eleven cases (78.6 %) were observed from November to April, which implies a seasonal distribution (p =0.013). The most commonly observed extension of aortic dissection involved the whole aorta until the foramen of the diaphragm in five cases (35.7 %) or both the ascending aorta and aortic arch in five cases (35.7 %), followed by extension only to the ascending aorta in four patients (28.6 %).

Symptoms on arrival

The most frequent initial symptom was chest pain in 12 cases (85.7 %), followed by dyspnea in six patients (42.9 %). Four (28.6 %) individuals presented to the ED in circulatory shock, with the following clinical signs: pallor, perspiration, and clammy extremities. The median elapsed time from symptom onset to admission in the ED was 2 hours (IQR: 1-8.9 h). Hemodynamically unstable patients at hospital admission did not survive until the transportation to the cardiothoracic surgery unit, and the median time from symptom onset to death was 8.7 hours

Table 1: Demographic Characteristics of patients, with acute aortic dissection type A (n=14)

	#Total pts (n=14)	#Surg pts (n=9)	#Cons pts (n=5)
Demographic Characteristics			
Age, y	62.6 ± 14.9	59.7 ± 10.6	68.0 ± 21.0
M:F	9:5	6:3	3:2
Time from symptom onset to hospital arrival, h	2 (1.0-8.9)	4 (1.0-13.5)	3.02 (0.9-3.7)
Clinical Manifestations			
Chest pain, n/N (%)	12/14 (85.7 %)	9/9 (100 %)	3/5 (60.0 %)
Dyspnea, n/N (%)	6/14 (42.9 %)	5/9 (55.6 %)	1/5 (20.0 %)
Shock, n/N (%)	4/14 (28.6 %)	0/9 (0 %)	4/5 (80.0 %)
Neurologic deficit, n/N (%)	4/14 (28.6 %)	2/9 (22.2 %)	2/5 (40.0 %)
Heart murmur, n/N (%)	4/14 (28.6 %)	3/9 (33.3 %)	1/5 (20.0 %)
Vomiting, n/N (%)	3/14 (21.4 %)	2/9 (22.2 %)	1/5 (20.0 %)
Clinical Findings and Blood Laboratory Assessment			
SBP on arrival, mmHg	134 ± 41.5	145 ± 45.2	116.0 ± 30.5
HR/min	92.8 ± 24.1	91.9 ± 19.1	94.4 ± 33.0
SpO_2, %	94.0 ± 4.76	95.6 ± 2.3	91.4 ± 6.7
D-dimers, ng/ml	5.248 (2.430-11.410)	2.455 (1.457-4.523)	11.410 (7.740-34.690)
D-dimers >500 ng/ml	12/14 (85.7 %)	7/9 (77.8 %)	5/5 (100 %)
ECG normal, n/N (%)	9/14 (64.3 %)	6/9 (66.7 %)	3/5 (60.0 %)
Comorbidities			
Hypertension, n/N (%)	6/14 (42.9 %)	4/9 (44.4 %)	2/5 (40.0 %)
Diabetes, n/N (%)	1/14 (7.1 %)	0/9 (0 %)	1/5 (20.0 %)
Smoking, n/N (%)	3/14 (21.4 %)	2/9 (22.2 %)	1/5 (20.0 %)
Prior CV surgery, n/N (%)	2/14 (14.3 %)	0/9 (0 %)	2/5 (40.0 %)
Renal Failure, n/N (%)	3/14 (21.4 %)	2/9 (22.2 %)	1/5 (20.0 %)
Outcomes			
Death, n/N (%; 95%CI)	8/14 (57.1 %; 32.6- 0.78.7)	3/9 (33.3 %; 11.7- 64.9)	5/5 (100 %)

Values that follow normal distribution are expressed as mean ± standard deviation, while values that follow skewed distribution are expressed as median and in brackets interquartile range. AAD: acute aortic dissection, n: number of patients with the parameter, N: total number of patients, CI: confidence interval, y: years, M: male, F: female, SBP: systolic blood pressure, mmHg: millimeter of mercury, HR: heart rhythm, h: hours, pts: patients, ng: nanogram, ml: milliliter, SpO_2: saturation of hemoglobin with oxygen, Surg: surgical management, Cons: conservatively management, pts: patients; CV: cardiovascular.

(IQR: 6.4-13.4 h). The median time from the admission to our hospital to the arrival to the cardiothoracic surgical center was 6.6 hours (IQR: 4.0-20.1 h) (Table 1). Six patients (42.9 %) presented with systolic blood pressure (SBP) higher than 130 mmHg, four patients (28.6 %) had SBP lower than 100 mmHg, and three patients (21.4 %) had normal SBP. The standard 12-lead electrocardiogram (ECG) confirmed normal findings without ischemic changes in nine patients (64.3 %).

Past medical history - laboratory examinations on ED arrival

Among the patients' cardiovascular risk factors, the most frequent were hypertension in six (42.9 %) and chronic renal failure in three cases (21.4 %), while smoking was prevalent in three patients (21.4 %). Interestingly, six patients (42.9 %) were free of medical treatment. Data on the past medical history of included patients are presented in Table 1. The most frequent laboratory finding was the elevated D-dimers (>500 ng/ml), observed in 12 patients (85.7 %). Patients who underwent surgery had significantly lower D-dimer levels (2,455 ng/ml; IQR: 1,457-4,523) compared to those treated conservatively (11,410 ng/ml; IQR: 7,740-34,690; p =0.011). Patients in circulatory collapse had significant higher D-dimer levels (23,050 ng/ml; IQR: 10,281-35,163) compared to hemodynamically stable patients (2,480 ng/ml, IQR: 1,728-5,248; p =0.02). D-dimers on arrival were significantly lower among patients who survived (2,079 ng/ml; IQR: 916-2,467) compared to those who did not survive (11,410 ng/ml; IQR: 7,317-25,635; p =0.002).

In-hospital mortality

Post-surgical mortality was 33.0 %, while the in-hospital mortality rate of the conservatively managed patients was 100 %. Only medical treatment with arterial blood pressure (ABP) control agents was administered in patients who did not survive until surgery and those with high risk for surgical repair. Among seven patients that were admitted within the critical first 2 hours, only two finally survived (28.6 %). Patients who were admitted with disturbance of consciousness, hemodynamic instability, or SpO_2 less than 92 % had a higher mortality rate when compared with the rest of the patients [odds ratio (OR):

35; 95%CI: 1.74 to 703; p=0.026]. The overall survival rate was 42.9 % (n =6/14), and patients who survived were younger than 70 years (OR: 13; 95 % CI: 0.55 to 306; p =0.085). Data on the outcomes of the enrolled patients are presented in Table 1.

Discussion

Recent epidemiological data on the prevalence of AADs are inadequate, and, as highlighted by current guidelines, more evidence is needed on the caseload-outcome relationship in this field[4]. Our case series aims to highlight the incidence, the clinical presentation, the management strategies, and the outcomes of this particular emergent cardiovascular disease for the first time in a representative Greek cohort.

Interesting findings emerged from comparing our research results with the existing bibliographic evidence; we conducted a systematic review of the literature, reporting data on a total of 31,160 patients, extensively described in the Supplementary Appendix. Data analysis of our cases demonstrated a significantly lower proportion of surgically treated (64.3 % vs 80.3 %) and a higher proportion of medically treated patients (35.7 % vs 16.9 %) compared to the literature. Additionally, our data concerning the in-hospital mortality were not consistent with those of the published literature. The in-hospital mortality of both surgically (33 %) and medically treated patients (100 %) was higher in our patient group than the existing data, which demonstrate 11 % and 37.8 % in-hospital mortality, respectively. A possible explanation

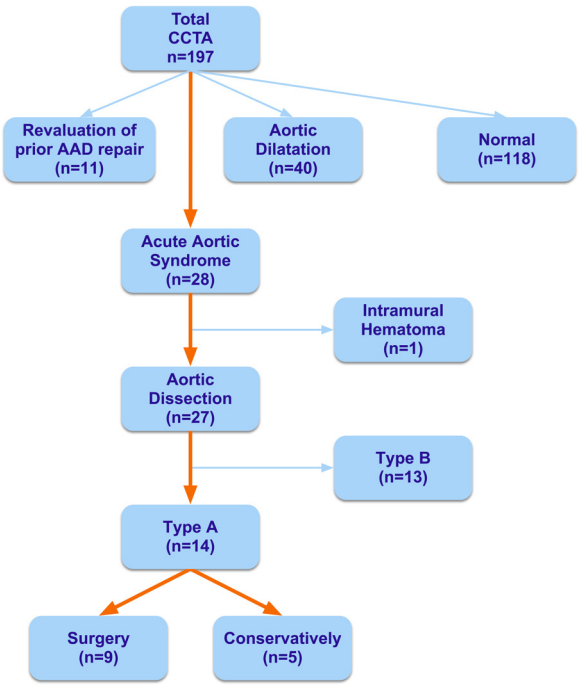

Figure 1: Screening of chest computed tomography angiography of patients with clinical suspicion of acute aortic dissection in the emergency department.
CCTA: chest computed tomography angiography, AAD: acute aortic dissection; ED: emergency department.

for this phenomenon could be the lack of an in-site surgical department and a selection bias, i.e., surgical deference of patients with a futile prognosis. The remarkably high mortality between patients treated conservatively underlines the need for surgical approach in the majority of cases.

The incidence of AAD in our patient group was 5 per 100,000, and the incidence of type A AAD was 2.53 per 100,000 population. These rates were in accordance with previously published international studies reporting that the incidence of AAD ranges from 2.5 to 15 per 100,000 and the incidence of type A AAD from 1.7 to 8 per 100,000[6,12,13]. Likewise, the baseline characteristics of our patient group did not differ from those presented in international registries. Hypertension and male gender were the most prevalent cardiovascular risk factors among type A AAD patients, a fact that was in line with the IRAD study[1]. Connective tissue disorders like Marfan syndrome, pregnancy, cocaine use, peripheral artery disease, and valvular heart disease are proven risk factors for AAD[14] that were not reported in the past medical history of our patients. Additionally, our data were in harmony with a recent metanalysis, reporting an increased frequency of AAD occurrence during the cold winter months[15].

Despite recent improvements in AAD diagnosis leading to the diagnosis of more than 80 % of cases within the first 24 hours after symptom onset, total mortality remains alarmingly high[16]. The mortality of surgically treated patients included in our case series was comparable with the Spanish registry[16] (RESA-II) (33 % vs 34 %), thus being higher than other international registries[17,18]. The surgical approach of our AAD patients showed a clear benefit regarding mortality. In this way, our results encourage rapid surgical treatment due to a lower in-hospital mortality rate than initial conservative treatment.

D-dimer levels have been underlined as a useful laboratory test to rule out AAD in the ED since values below the reference limit lead to the exclusion of AAD presence[8], a finding that was replicated in our cases. Additionally, D-dimers demonstrated a strong prognostic value in our cohort. Patients who either did not survive or were treated conservatively had higher D-dimer levels than survivors or patients treated surgically.

Taking under consideration the noteworthy impact of age, the JRAD registry reported that age ≥80 years was a major risk factor for in-hospital mortality[19]. Nonetheless, the results of our analysis were in line with IRAD data that showed a decrease in the in-hospital mortality in patients <70 years old[20]. Advanced age is reported as the principal risk factor for in-hospital mortality, regardless of the type of management, surgical or conservative[19]. In our case series, the mortality rate was higher in patients transferred to the ED within the first 2 hours after symptom onset. A possible explanation could be that worse clinical status caused alarming symptoms leading to immediate contact with the ED.

The present research was performed on a represen-

tative sample, though distribution may differ in other regions; hence the extrapolation of our results to other populations should be critically evaluated.

Conclusion

AAD type A is an alarming, life-threatening condition with a high in-hospital mortality rate. Current epidemiological data on the occurrence of AADs are inadequate; more information is needed to elucidate the actual incidence, the true mortality rates, and the most appropriate diagnostic and therapeutic approach. Our case series presents, for the first time, data regarding a local Greek cohort. Baseline demographic characteristics of our patient group and risk factors were comparable with those in the existing literature; nevertheless, mortality rates were higher in our cohort. Further research ought to be rendered to shed light on the various aspects of acute management of the disease in the local setting, with an eye on improving survival.

Conflict of interest

Authors declare no conflict of interest.

Acknowledgement

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