Unexpected cause for subacute cardiac tamponade in a hypertensive patient

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Introduction

Aortic dissection is an uncommon, yet life-threatening condition for which timely diagnosis is paramount. It is frequently associated with risk factors that differ in the younger vs older populations (under vs over 40 years of age). Presentation is variable, often making the clinical diagnosis a challenge.

Case presentation

We present the case of a 39-year-old male patient who arrived at the Emergency Department complaining of fatigue, shortness of breath on mild exertion, and pitting bilateral symmetric edema of the lower extremities.

He reported symptoms first began with sustained effort and worsened progressively for the past month, with dyspnea on minimal exertion for the past few days. He denied having any chest pain and stated he has never experienced similar symptoms. His personal and family history were negative for cardiovascular disease. He worked in constructions and mentioned sporadic diffuse back pain in the thoracic and lumbar areas, recently diagnosed as degenerative disease. He was otherwise unaware of other health problems.

Vitals revealed mild fever (T = 38.4 degrees C), blood pressure (BP) of 186/104 mmHg in his right arm and 178/94 mmHg in his left arm, respiratory rate of 22 bpm, with no cyanosis present. He appeared to be in no distress at rest. His cardiovascular examination was significant for jugular venous distention, distant heart sounds on auscultation, but no gallops, rub or murmurs were heard. Breath sounds were present with fine crackles in the basal lung fields, bilaterally. On inspection, soft, pitting edema of the lower extremities up to the knees was noted.

Laboratory workup revealed thrombocytosis ($532,000/mm^3$), moderate leukocytosis ($20,300/mm^3$), with minimally elevated serum creatinine (1.32 mg/dL, eGF [CKD-EPI] = 67.5 mL/min/1.73m2) and elevated liver enzymes (AST = 102 IU/L, ALT = 97 IU/L). ESR and CRP could not be processed at the time of evaluation, while troponin and CK-MB levels returned negative at a later time.

EKG showed sinus tachycardia and a discrete pattern of electrical alternans, more evident in the inferior

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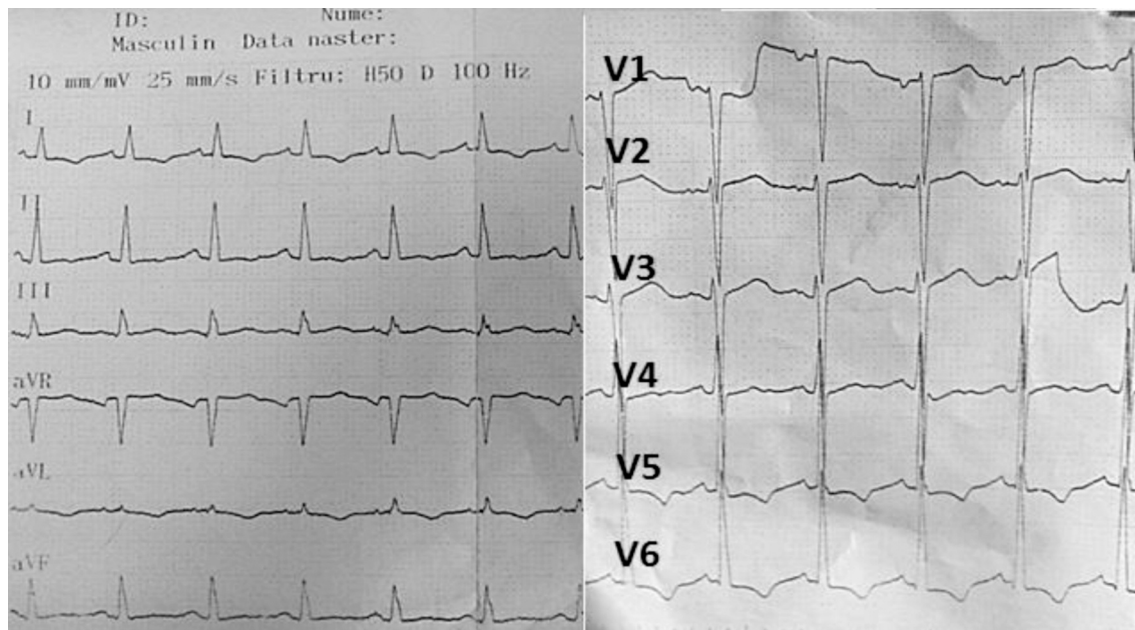e-mail: alexandru_deaconu@yahoo.com

Fig. 1. ECG showing discrete electrical alternans pattern, more evident in the inferior limb leads (specifically aVF and lead II), with signs of left ventricular hypertrophy in the thoracic leads and secondary ST-T changes (also seen in lead I and aVL).

limb leads, suggestive for pericardial effusion (Fig. 1), with signs of left ventricular hypertrophy in the thoracic leads and secondary ST-T changes.

Transthoracic echocardiography (TTE) confirmed the effusion, showing circumferential involvement with diastolic collapse of the right ventricle (RV) and the right atrium (RA) and a mild aortic regurgitation. The maximum width of the effusion was measured at 3.8 cm anterior to the RV and RA. Mild left ventricular hypertrophy was also noted. All echocardiographic

signs indicated cardiac tamponade as the main cause for the patient's heart failure (Fig. 2).

Of note, during monitoring in the Emergency Department, our patient had oscillating blood pressure readings, from 170 mmHg up to 200 mmHg (systolic values), which prompted administration of intravenous beta-blocker and nitroglycerin.

Differential diagnosis of pericardial effusion includes idiopathic effusion, tuberculosis, connective tissue disease, malignancy or aortic dissection.
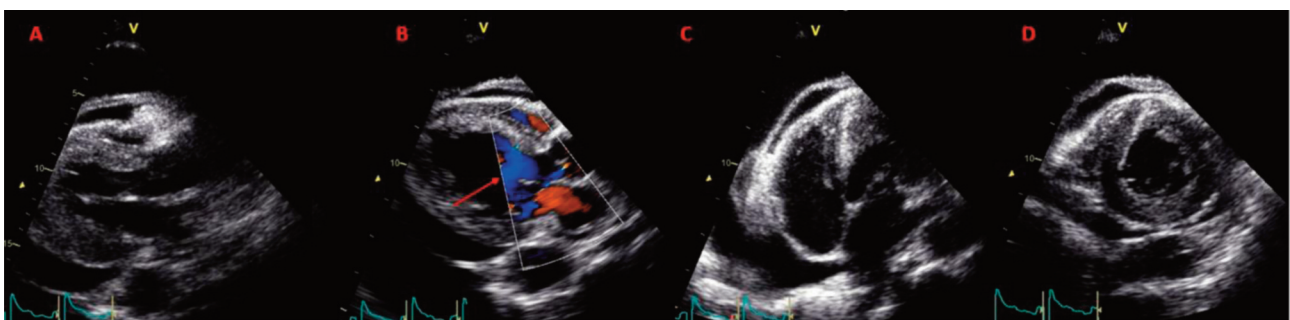

Fig. 2. A: TTE parasternal long axis view, showing a large pericardial effusion; fibrin strand noticeable anterior to the right ventricle, with diastolic collapse also evident. B: TTE color Doppler examination showing mild aortic regurgitation (arrow); C: TTE apical 4-chamber view demonstrating circumferential effusion; D: TTE parasternal short axis view in diastole, emphasizing right ventricle diastolic collapse.

Fig. 3. A: CT scan showing aneurysm of the aortic arch, with hypodensity suggestive of thrombosis; B: Intimal flap evident in the descending aorta; C: Circumferential pericardial effusion, measured at a maximum width of 2.9 cm; in addition, right pleural effusion, left pulmonary atelectasis and dissection of the descending aorta can also be seen.

Because the patient was hemodynamically stable and with no significant renal dysfunction, we decided to perform a CT scan of the chest and abdomen, which unexpectedly revealed an increased diameter of the ascending aorta (5.5 cm), not seen previously on TTE. An area of thrombosis extending into the aortic arch and an intimal flap progressing into the thoracic and abdominal aorta down to the emergence of the renal arteries were also evident (Fig. 3).

Ultimately, the diagnosis was subacute Type A (Stanford) aortic dissection complicated with cardiac tamponade, ascending aortic aneurysm, mild aortic regurgitation and hypertension stage 3 of very high additive risk. The patient was referred to the cardiovascular surgery unit for emergent intervention, with subsequent successful repair of the ascending aorta.

Case particularity and discussion

From a clinical standpoint, aortic dissection has a large spectrum of presentations, therefore a high index of suspicion is required to establish diagnosis. At times, this task proves to be an intricate process that demands a thorough workup including history, physical examination, laboratory and imaging studies. An electrocardiogram may be suggestive, but not specific. Laboratory studies are non-diagnostic, however, recent guidelines recommend testing for D-dimers, if cardiovascular imaging is not available or too costly. In low-risk patients, D-dimer levels <500 ng/mL have a 96% negative predictive value [19]. Imaging studies are required both for diagnosis and follow-up. Contrast-enhanced CT is widely available, particularly in the emergency setting,

is time-effective, the least examiner-dependent, and provides useful information regarding anatomical correlates for surgical and endovascular therapy. The hemodynamic status of the patient and renal dysfunction can limit its use. MR angiography is highly accurate, but not readily available, with limited applicability (patients with claustrophobia, pacemakers or other devices) and cost prohibitive. Transesophageal echocardiography is the imaging study of choice when suspicion is high. It is accurate, can be used in hemodynamically unstable patients, but it is not readily available due to requirement of trained specialists [20].

Our patient presented with symptoms suggestive of right heart failure that warranted a search for the underlying cause. The echocardiogram revealed a large pericardial effusion, with signs of tamponade. Self-limited ascending aortic dissection in a young patient, without known prior risk factors and no symptoms is an exceptional cause of chronic heart failure.

In trying to understand this complex case, we considered two directions of research: (1) the etiology of a previously unknown ascending aortic aneurysm and (2) the atypical clinical presentation and its complications. We will first discuss the latter.

Subacute cardiac tamponade in a young patient, with no relevant medical history should prompt further investigation to find a cause. So far, studies conducted in the United States and Europe have shown different prevalence in the etiology of cardiac tamponade or large pericardial effusion. In a series of 322 cases of moderate-to-large effusions (size of effusion >10mm), Sagrista et al. [1] found that in 29% of cases no cause could be determined (idiopathic pericardial effusion), 13% of the effusions had an underlying neoplastic ori-

gin, 16% were iatrogenic, 6% were of infectious causes, and 5% were established as due to collagen vascular disease. In contrast, in another series of 204 cases of moderate-to-large effusions published three years later, Levy et al. [2] concluded that 48% of the cases were rendered idiopathic, 15% were due to malignancy, 16% due to infectious causes and 10% of the cases were attributed to collagen vascular disease. More recently, in Western countries, the prevalence of infectious causes for pericardial effusions has declined, while an increase in iatrogenic causes has been remarked. In endemic areas or in immigrants from endemic areas, tuberculous pericarditis should be included in the differential diagnosis. In a study conducted in South Africa, Mayosi et al. found that 10% of the patients with confirmed tuberculous pericarditis had presented initially with cardiac tamponade.

Aortic dissection (AD) in association with pericardial tamponade has seen a substantial decline in incidence in the last decades, as other causes for the condition have taken the forefront. This phenomenon has not been closely followed in recent years, however, a single-center study analyzed 150 cases [3] with the diagnosis of cardiac tamponade on admission, out of which 64% had hemopericardium and, of those, only 4% were associated with aortic dissection.

AD is a rather uncommon, yet life-threatening condition that prompts early diagnosis and treatment. Clinical presentation depends on the site of dissection, but pain is the chief complaint in over 90% of cases, and most commonly it is located in the chest or back [4]. Incidence of acute AD in the general population is 2.6-3.5 per 100 000 person-years [5–7]. It affects the elderly more often (60-80 years of age), with a higher incidence in males. Older patients are more likely to have history of hypertension, prior aortic aneurysm, or iatrogenic dissection. Younger patients (under 40 years of age) have a higher likelihood of collagen disorders (50% Marfan syndrome [8], Ehler-Danlos syndrome, annulo-aortic ectasia). Other high-risk conditions include bicuspid aortic valve, aortic instrumentation or surgery, aortic coarctation, Turner syndrome, inflammatory diseases, pregnancy and delivery.

In this particular case, the diagnostic difficulty came from symptoms developing over a longer timeframe than expected. In the ascending type dissection, mortality is approximated at 1-2% each hour for the first 48h and 90% of those who survive this interval

die within 3 weeks [21,22]. Evolution classifies AD as acute in the first 15 days from onset, subacute (15-90 days) and chronic (over 90 days). Moreover, the chief complaint of type A aortic dissection is chest pain, which our patient denied at all times.

Thoracic aortic aneurysm has seen an increase in incidence in the past decades, to an estimate of 10.4 per 100,000 person-years in a Mayo Clinic study, significantly higher than prior to 1980 in the same population [7]. Around 60% of these involve the ascending aorta [9]. It was questioned if the recent epidemiologic data revealed an increase in aortic disease per se, or if it was the direct effect of a wider availability of aortic imaging studies (echocardiography and CT especially). Moreover, aneurysm disease is silent, and is generally detected as an incidental finding during an imaging study [10].

Aneurysms most often result from cystic medial degeneration, with smooth muscle cell dropout, degeneration of elastic fibers, and formation of cystic spaces. These changes cause weakening of the vascular wall, that leads to progressive aneurysm formation [9]. Hypertension is considered an accelerating factor of cystic medial degeneration. In association with advanced age, these two constitute the most important risk factors for aneurysm formation. For younger patients, Marfan syndrome and other connective tissue disorders, aortic valve abnormalities and familial thoracic aortic aneurysm syndrome are considered high-risk conditions [9].

Our patient had not been previously investigated, therefore he was unaware of having a thoracic aortic aneurysm, and he denied any family history of aortic disease.

Regarding potential complications, particularly dissection, among the International Registry of Aortic Dissection (IRAD) population the majority of patients had history of hypertension, 14% had a known thoracic aortic aneurysm (most likely an underestimated value, if considering others with undetected aneurysms prior to dissection). Twenty-five per cent of the young patients (under 40 years of age) with aortic dissection were hypertensive on presentation and 34% had history of hypertension [8].

In most cases, however, aortic dissection occurs in the absence of a preexisting aneurysm. Moreover, aneurysms can later develop as a complication of dissection [11].

Identification of patients at risk for aortic dissection is challenging. In addition to the aforementioned risk factors and predisposing conditions, patients with aortic disease engaging in professional activities that require heavy lifting or strenuous manual labor may be at increased risk for aortic events [12]. It is thought heavy weight-lifting causes abrupt spikes in BP, predisposing to vascular wall abnormalities over time.

Furthermore, in aortic dissection cases, treatment of hypertensive emergency [13] is essential. The objective is to prevent retrograde dissection and occlusion of branch arteries. Careful reduction in systolic BP (SBP) values should be considered in patients with essential hypertension, as they tend to tolerate better higher SBP readings [14].

A hypertensive emergency consists of intense peripheral vasoconstriction (probably from increased catecholamines), an imbalance of the renin-angiotensin system, or altered baroreceptor function [15]. With vasoconstriction, there is a rapid rise in BP, triggering a series of events that uphold and worsen the hypertension. If it remains refractory to optimal medical treatment, renovascular hypertension related to the dissection flap should be considered [16].

In our case, the hypertension was multifactorial (1) possibly preexistent in a heavy weight lifter without a known secondary hypertension, (2) as a consequence of AD, (3) worsened by the renal dysfunction and. In either scenario, the strain on the vascular wall creates the favorable conditions for aortic events.

The echocardiogram showed left ventricular hypertrophy (LVH). Assessing increased wall thickening in the context of tamponade is difficult. Generally, a follow-up echocardiogram is required to establish it as LVH due to preexistent hypertension or 'pseudohypertrophy' [17,18] in the context of tamponade.

Conclusions

Aortic syndromes can be elusive for the physician, with difficulty in establishing etiology, identifying risk factors and predisposition to complications. As emphasized by the European Society of Cardiology in its Guidelines for the Diagnosis and Management of Aortic Diseases, published in 2014, epidemiologic data on the topic are scarce, warranting further research on both incidence and outcomes [27].

There is a 6% estimate of aortic dissection cases presenting with acute congestive heart failure [23]. Precisely due to this uncommon presentation, there is a delay in surgery referral. In addition, our literature research revealed only a handful of cases of insidious cardiac tamponade, all of which were misdiagnosed at first due to atypical presentation [24–26].

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