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Case Report

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Acute type A aortic intramural haematoma in a hypertensive patient: a case report

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Abstract

Aortic intramural haematoma (IMH) is a less known entity within the spectrum of acute aortic syndromes (AAS), in which a haematoma develops in the media of the aortic wall in the absence of a false lumen and intimal tear. There are many risk conditions for acute aortic syndromes, but the most common predisposing factor is arterial hypertension. We presented a case of acute type A aortic intramural haematoma complicated with cardiac tamponade witch is associated with a 16- fold greater mortality risk. The accuracy of the diagnosis made it possible for the emergency surgery to be successful, saving the patient's life.

Keywords: acute aortic syndrome, intramural haematoma, arterial hypertension

Introduction

Acute aortic syndromes refer to the spectrum of aortic emergencies that include aortic dissection, intramural haematoma, penetrating atherosclerotic ulcer of the aorta, aortic aneurysm leak and rupture and traumatic aortic transection.

Aortic IMH develops in the media of the aortic wall in the absence of a false lumen and intimal tear. It is diagnosed in the presence of a circular or crescent-shaped thickening of 5 mm of the aortic wall in the absence of detectable blood flow. This entity may account for 10- 25% of acute aortic syndromes. [1]

In the IRAD (International Registry of Acute Aortic Dissection) series the in-hospital mortality of Type A IMH was similar to Type A aortic dissection.[2] The therapeutic management in acute IMH should be similar to that for aortic dissection.

Case Report

A 51-year-old female arrived at the emergency department with severe chest and back pain, with abrupt onset about 6 hours ago. The pain was described as

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tearing and it was accompanied by dyspnea at rest and cold sweats.

The patient had multiple conventional cardiovascular risk factors: smoking, obesity, grade III arterial hypertension (up to 240 mmHg) and dyslipidemia. The arterial hypertension was diagnosed when she was 40 years old and it was poorly and intermittently treated with 5 mg of perindoprilum. The patient had no personal history of angina or syncope, also no family history of aortic dissection or sudden cardiac death.

On arrival at the emergency room the patient was hemodynamically unstable: the blood pressure of 110/50 mmHg (before she was a patient with severe hypertension), the heart rate of 110 beats/min (hypophonetic rhythmic heart sounds), respiratory rate of 34 breath/min, the oxygen saturation was 91% breathing ambient air (it increased till 98% with 8 litres of oxygen per minut on mask). The clincal examination was significant for jugular venous distension, paradoxically pulse (no deficit pulse was detected) and cold extremities. The patient was anxious and she had had no diuresis in the last 6 hours.

The electrocardiogram showed sinus tachycardia (106/min) and left ventricular hypertrophy with secondary ST-T changes.

The inital laboratory tests showed severe renal dysfunction (creatinine= 3.27 mg/dl, urea= 98 mg/dl), liver dysfunction (hepatic cytolysis- GOT=1058 U/L, GPT=788 U/L, spontaneous INR=1.3). The levels of D- dimers, lactate and brain natriuretic peptide (BNP) were increased: D-dimers =2,7 mcg/ml (<0.7 mcg/ml), lactate= 3.8 mmol/l, BNP= 304 pg/ml (NR< 100 pg/ml). The cardiac troponin TnI was negative, while creatine kinase levels was mild elevated (CK= 118 U/L, CK-MB= 53 U/L). A complete blood count revealed moderate anemia (Hb= 10.2 g/dl), leukocytosis and neutrophilia (WBC=12,350/mm^3 with 83.4 % neutrophils), platelets were in the normal range. The patient also had mild hyperglicaemia (138 mg/dl) and mild hypercholesterolaemia (237 mg/dl).

The transthoracic echocardiography revealed a hypertrophied left ventricule (IVS 19 mm, PW= 18 mm) with normal global and regional systolic function (LVEF= 55%); mild mitral regurgitation; a normal three leafleats aortic valve , grade II aortic regurgitation, dilated ascending aorta (55 mm) and aortic cross (36 mm); no intimal tear was visualized. Most important, the patient had moderate pericardial effusion with systolic right atrial collapse and diastolic right ventricle collapse (the fluid was circumferentially disposed and it suggested haemoperdicardum) The RV was small (29 mm) with normal systolic function. The inferior vena cava was dilated (27 mm) and without inspiratory collapse.

In this context (back pain, history of uncontrolled arterial hypertension, acute renal failure, positive D-dimers and the ecocardiographic findings- dilatation of the ascending aorta , the aortic regurgitation, peri-

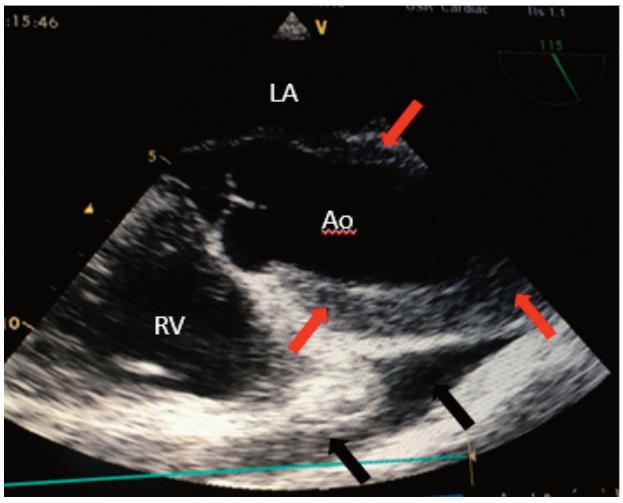

Figure 1. TEE Mid Esophageal LAX- Aortic intramural haematoma which starts at one centimeter above the aortic annulus (red arrows); pericardial fluid (black arrows).

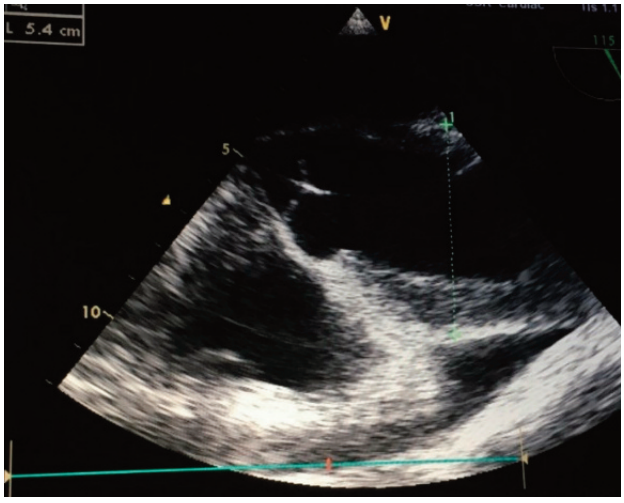

Figure 2. TEE Mid Esophageal LAX- Dilated ascending aorta (54 mm).

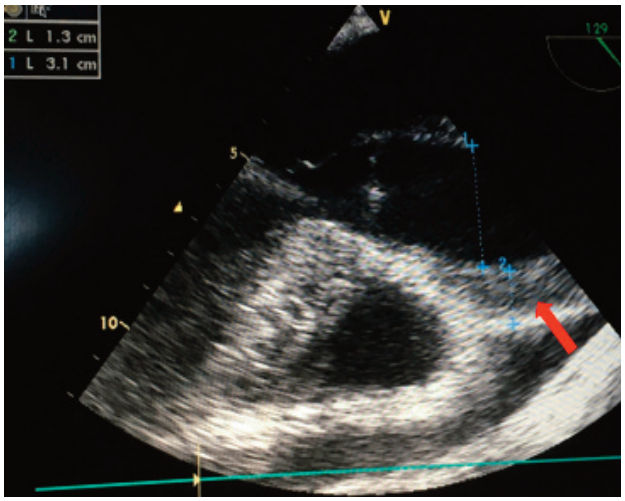

Figure 3. TEE Mid Esophageal LAX- Thickened anterior aortic wall - 13 mm (red arrow); aortic lumen- 31 mm.

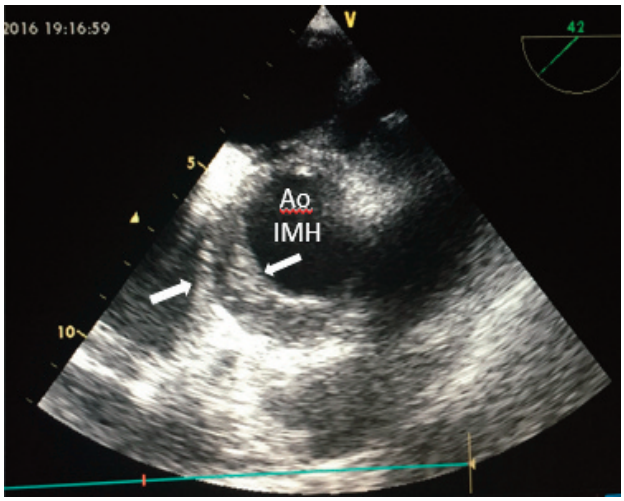

Figure 4. TEE Mid Esophageal Ascending aorta SAX- Intramural haemaoma (white arrows).

cardial effusion- haemopericardum with cardiac tamponade) an acute aortic syndrome was suspected. It was performed a transesophageal echocardiography. It revealed intramural haematoma of the ascending aorta, which starts at one centimeter above the aortic annulus; the anterior aortic wall thickness was 13 mm, the posterior's was 8 mm and the aortic lumen measured 31 mm; the coronary ostia seemed to be unaffected; the aortic regurgitation was mild (probably by dilated aortic annulus- 24 mm); the intramural haematoma affected also the anterior wall of the aortic cross and descending aorta (it was visualized up to 40 cm of the dental arch). The pericardial fluid was circumferentially disposed (maximum of 13 cm around anterior wall of RV). No intimal tear or false lumen were present (Figs. 1–6).

We also performed a carotid artery Doppler ultrasound- the intramural haematoma affected the ostium of the right common carotid artery (Fig. 7).

The following diagnosis was established: Acute type A aortic intramural haematoma. Cardiac tamponade. Mild aortic regurgitation. Severe arterial hypertension. Acute renal failure. The patient needed emergency surgery so she was transferred to the operating room.

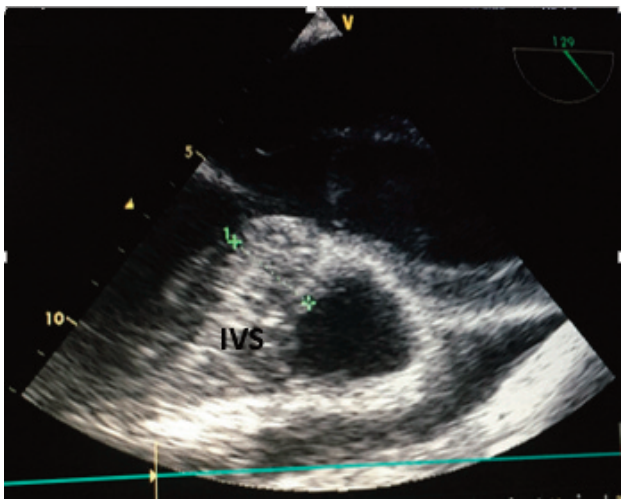

Figure 5. TEE Mid Esophageal LAX- Severe interventricular septum hypertrophy- 19 mm.

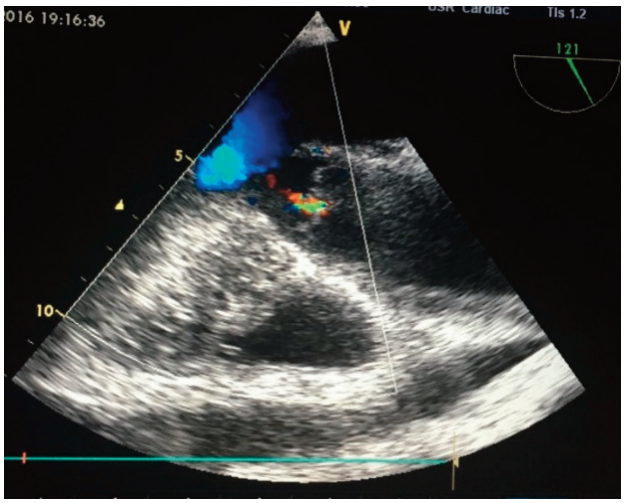

Figure 6. TEE Mid Esophageal LAX- Mild aortic regurgitation.

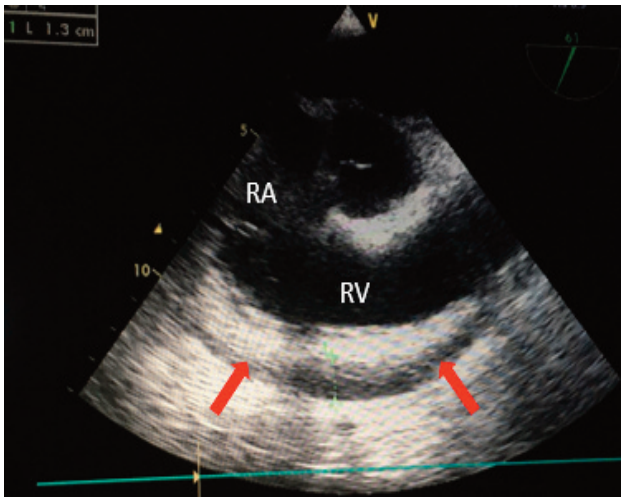

Figure 7. TEE Mid Esophageal SAX- Pericardial fluid around the anterior wall of RV- 13 mm.

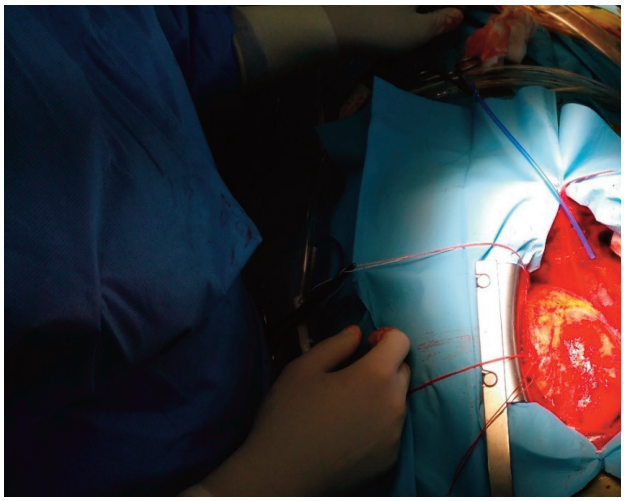

Figure 8: Intraoperative photography- Ascending aorta intramural haematoma.

The left femoral artery was cannulated for cardiopulmonary bypass. Also the carotid arteries were additionally cannulated to allow for antegrade cerebral perfusion during the circulatory arrest period. Intraoperative it was found a blood distented pericardium; 200 ml of blood under pressure were evacuated. The ascending aorta was dilated- about 60 mm. The intramural haematoma started from the right Valsalva sinus but, fortunately, the right coronary artery was unaffected. No intimal tear, flap or false lumen were found. The ascending aorta above Valsalva sinuses and the hemi-cross were resected and a 26 Intergard vascular prosthesis was implanted (Fig. 8).

Remarkably, after surgery the patient had a uneventful recovery; postoperatively she was mechanically ventilated only 82 hours more. She had a transient thrombocytopenia- 75,000/mm^3. Until discharge the renal function was normalized (creatinine 0.95 mg/dl) and the platelets were in the normal range (120,000/mm^3). The renal dysfunction was caused by renal ischemia secondary to arterial hypotension.

The transthoracic echocardiography revealed a normal global and regional LV and RV systolic function, mild aortic regurgitation and a vascular aortic prosthesis with normal function.

After 14 days the patient was discharge. The medical treatment recommended was: antiplatelet (100 mg of aspirin), ACE inhibitors (perindopril 10 mg), calcium channel blockers (amlodipine 10 mg), thiazide diuretic (indapamide 1.5 mg), betablockers (metoprolol 50 mg) and statin (atorvastatin 20 mg).

It was highlighted the importance of lifestyle changes and the cardiovascular risk factors control- in particular the management of arterial hypertension.

Discussion

In 1760, dr Frank Nicholls, physician to King George II, first described on necropsy an acute aortic dissection [3]. New diagnostic imaging modalities, longer life expectancy, longer exposure to elevated blood pressure, and the proliferation of modern noninvasive imaging modalities have all contributed to the growing awareness of acute and chronic aortic syndromes.

Acute aortic syndromes refer to the spectrum of aortic emergencies that include aortic dissection, intramural haematoma, penetrating atherosclerotic ulcer of the aorta, aortic aneurysm leak and rupture and traumatic aortic transection.

It is generally believed that intramural haematoma is caused by rupture of the vasa vasorum, which separates medial wall layers, eventually leading to a secondary tear or to communication to the adventitial space. Some sporadic case reports showing accidental development of typical IMH by percutaneous catheter manipulations, such as coronary angioplasty, insertion of balloon pump, or catheter ablation of left sided bypass tract, support the presence of a "primary intimomedial

tear" in IMH [4]. This hypothesis is also supported by demonstration of a small intimal communication at the time of surgery [5,6].

Aortic IMH develops in the media of the aortic wall in the absence of a false lumen and intimal tear. It is diagnosed in the presence of a circular or crescent-shaped thickening of 5 mm of the aortic wall in the absence of detectable blood flow. This entity may account for 10- 25% of acute aortic syndromes [1].

The involvement of the ascending aorta and aortic arch (Type A) may account for 30% and 10% of cases, respectively, whereas it involves the descending thoracic aorta (Type B) in 60–70% of cases [7,8].

There are many risk conditions for aortic syndrome, but the most common predisposing factor in IRAD was arterial hypertension (72%). Also, the history of atherosclerosis was present in 31% of patients. [9].

Chronic arterial hypertension affects the arterial wall composition, causing intimal thickening, fibrosis, calcification and deposit of extracellular fatty acid. The extracellular matrix undergoes accelerated degradation and apoptosis. Both mechanisms may eventually lead to intimal disruption. Adventitial fibrosis may obstruct vessels feeding the arterial wall as well as small intramural vasa vasorum [10]. Both result in necrosis of smooth muscle cells and fibrosis of elastic structures of the vessel wall, which leads to stiffness and vulnerability to pulsatile forces, creating a substrate for aneurysms and dissections [11].

The studies revealed that the previous blood pressure control on patients with AAS is poor, despite the cases of patients being on combination antihypertensive therapy, particularly in immediately fatal cases [12]. This high rate of treatment-resistant hypertension may reflect increased aortic stiffness or other pathophysiological mechanisms responsible for the development of acute aortic syndromes. However, better control of BP would nevertheless be likely to reduce incidence and case fatality. Improved primary prevention - more aggressive management of hypertension and smoking cessation in particular, may reduce future incidence rates, but treatment-resistant hypertension is likely to remain a challenge.

The presented case confirms the theories above exposed. A femele with history of uncontrolled hypertension (and other cardiovascular risk factors) suffers an acute aortic syndrome- a type A intramural haematoma complicated with cardiac tamponade due to aortic rupture into the pericardial compartment which is associated with a 16- fold greater mortality risk.

For the detection of an acute aortic intramural haematoma, the transthoracic echocardiography (TTE) is inadequate because of its low sensitivity (lower than 40%) [13]. Based on these findings, TTE cannot be used as the sole imaging technique in patients with suspected AAS.

Transesophageal echocardiography (TEE) has a 90%–100% sensitivity and a 91%–100% specificity for the detection of intramural hematoma. However, it does not allow visualization of the entire thoracic aorta, and the quality of the examination is operator dependent. Difficulty in differentiating an intramural haematoma from severe atherosclerosis with focal wall thickening may produce a false-positive or equivocal result [14]. The crucial factors that contribute to confidence in the diagnosis of intramural hematoma at imaging are accurate identification of the intima, which is often echogenic because of calcification, and localization of the abnormality beneath the intima rather than above it (the latter being suggestive of an intraluminal lesion) [14].

Computer tomography (CT) and magnetic resonance imaging (MRI) are the leading techniques for diagnosis and classification of intramural haematoma.

On unenhanced CT, intramural hematoma is hyperdense. The differentiation of intramural haematoma from atherosclerotic thickening of the aorta, thrombus, or thrombosed dissection may be difficult using CT. In those circumstances, MRI can be a valuable problem-solving tool, it may also provide a determination of the age of a haematoma, based on the signal characteristics of different degradation products of haemoglobin [15]. MR imaging has a reported sensitivity of 100% for the detection of aortic intramural haematoma because it provides excellent soft-tissue contrast and characterization of aortic wall thickening. However, this modality receives only limited use for initial diagnosis evaluations because of the lengthy examination time (approximately 30 minutes for evaluation of an intramural hematoma), incompatibility of the magnet with many monitoring devices necessary in critically ill patients, and less availability on an emergent basis. MR imaging can aid in the distinction of slow flow in the false lumen of a dissection from no flow in an intramural haematoma.

In 2010, the ACC/American Heart Association (AHA) guidelines proposed a risk assessment tool based on three groups of information— predisposing conditions, pain features, and clinical examination— and proposed a scoring system that considered the number of these groups that were involved, from 0 (none) to 3. [16] Our patient had a high probability of an acute aortic syndrome (score 2- chest and back pain described as tearing and she developed shock). Initially it was performed a TTE that was inconclusive but it strengthened the initial suspicion (dilatation of the ascending aorta, pericardial effusion). In unstable patients with a suspicion of AAS, TEE and CT it is recommended using imaging modalities (class I, level C by ESC guidelines [5]). In the Emergency Institute for Cadiovascular Diseases "Prof. Dr. C. C. Iliescu", the TEE is 24 hours available; the diagnosis was promptly established. Transoesophageal echocardiography may be of great interest in the highly unstable patient, and can be used to monitor changes in-theatre and in post-operative intensive care.

Acute IMH, when first diagnosed, may be a classic subtle aortic dissection that escapes diagnosis on initial imaging but shows on subsequent imaging or re-review of initial studies. A second scenario is the progression of IMH to classic aortic dissection between an initial diagnosis study and an interim follow-up image. Given these uncertainties, and until further studies provide more predictive data, many experts recommend definitive aortic repair for acute IMH of the ascending aorta similar to type A dissection, and a less aggressive attitude towards haematoma in the descending aorta similar to type B dissection.

The mortality rates of medically treated patients are high. In the IRAD series, the in-hospital mortality of Type A IMH was similar to Type A aortic dissection. Overall, the long-term prognosis of patients with IMH is in favour of patients with AD [2]. On the other hand, several series showed that 30– 40% of Type A IMH evolved into AD, with the greatest risk within the first 8 days after onset of symptoms. [17]

Some authors have identified parameters, such as progressive maximal aortic wall thickness (increasing from 14.5 to 21.7 mm) and aortic diameter (increasing from 55.6 to 63.9 mm) as strong predictors of subsequent dissection with the need of surgical repair within days to weeks after the clinical acute aortic syndrome (CT measurements).[18]

The therapeutic management in acute IMH should be similar to that for aortic dissection.

Based on the ESC guidelines, for the type A intramural haematoma - emergency surgery is indicated in complicated cases with pericardial effusion, periaortic haematoma, or large aneurysms, and urgent surgery (24 hours after diagnosis) is required in most of Type A IMHs. [5]. In elderly patients or those with significant comorbidities, initial medical treatment with a 'wait-and-watch strategy' (optimal medical therapy with blood pressure and pain control and repetitive imaging) may be a reasonable option, particularly in the absence of aortic dilation (<50 mm) and IMH thickness (<11 mm). [5]

Our patient needed emergency surgery, because the intramural haematoma was complicated with cardiac tamponade; this was successfully performed and the patient had an uneventful recovery.

Conclusions

Aortic intramural haematoma is a less known entity within the spectrum of acute aortic syndromes, in which a haematoma develops in the media of the aortic wall in the absence of a false lumen and intimal tear. There are many risk conditions for AAS, but the most common predisposing factor is arterial hypertension. A better control of blood pressure would nevertheless be likely to reduce incidence and casue fatality.

The in-hospital mortality of Type A IMH is similar to Type A AD, so the therapeutic management in acute IMH should be similar to that for AD.

We presented a case of acute type A aortic intramural haematoma complicated with cardiac tamponade which is associated with a 16- fold greater mortality risk. The diagnosis flow chart must combine the pretest probabilities according to clinical data, the laboratory and imaging tests, as should be done in clinical practice in all emergency rooms. The accuracy of the diagnosis made it possible for the emergency surgery to be successful, saving the patient's life.

Conflict of interest

The authors confirm that there are no conflicts of interest.

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