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Case Report

Cardiac Amyloidosis Causing Free Wall Rupture During an Exercise Stress Test: A Case Report and Review of the Literature

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Abstract

Background: There are three major mechanical complications from acute myocardial infarction: left ventricular free-wall rupture, ventricular septum rupture and acute mitral valve regurgitation. The left ventricular free-wall rupture is a serious and often lethal complication, typically following an ST elevation myocardial infarction. Free wall rupture outside the setting of a recent ST elevation myocardial infarction is very unusual. Furthermore, rupture during stress testing is extremely uncommon with very few cases in the literature. In addition to causing restrictive cardiomyopathies, cardiac amyloidosis can also cause severe ischemia with potentially catastrophic complications including free wall rupture.

Case Presentation: We present a 75-year-old white male with dyslipidemia who presented to his internist with intermittent chest pressure for several days which was initially exertional. The night prior to seeing his internist, the discomfort was present while lying in bed. His EKG was unchanged from prior with no acute ischemic changes and an old RBBB. A stress echo was ordered which was obtained six days later. His baseline echocardiogram revealed normal wall thickness, an ejection fraction of 60% with very mild hypokinesia of the lateral wall. He exercised for a total of 7 minutes and 30 seconds and stopped exercise due to light headedness. He developed 4 mm of ST depression in the right precordial leads in early recovery. He subsequently went into PEA arrest and was noted to have a new pericardial effusion with tamponade physiology. Despite ACLS protocol and attempt at pericardiocentesis, he died in the stress lab before he could be taken for emergent surgery. Autopsy conveyed acute and subacute infarction of the posterior and lateral wall with left ventricular rupture. The degree of coronary disease was only moderate. Further evaluation of the myocardium conveyed diffuse transthyretin amyloidosis (ATTR).

Conclusion: Underlying cardiac amyloidosis can directly cause ischemia and infarction due to microvascular disease. If severe ischemia develops before significant left ventricular hypertrophy occurs, myocardial rupture may be more likely due to the already compromised myocardial substrate. Early treatment of subclinical amyloidosis, especially ATTR, should be considered to reduce complications such as this.

Keywords: Myocardial Rupture; Free Wall Rupute; Stress Test; Cardiac Amyloidosis; Transthyretin Amyloidosis (ATTR).

Abbreviations: RBBB: right bundle branch block; ATTR: Transthyretin amyloidosis; PEA: Pulseless electrical activity; ACLS: Advanced Cardiac Life Support; STEMI: ST elevation myocardial infarction; NSVT: Non sustained ventricular tachycardia.

Background

Cardiac rupture during stress testing is uncommon and rarely reported, with less than ten cases described in the scientific literature [1]. In general, the incidence of left ventricular free-wall rupture post-acute myocardial infarction is less than 1%. That being said, ventricular free wall rupture is ten-fold more common than septum or papillary muscle rupture and caries a high mortality. It has a bimodal presentation with greater incidence in the first 24 h and another peak at 3-6 days [1]. As scar tissue has time to develop, the incidence goes down significantly with time. Late fibrinolysis or late percutaneous intervention, advanced age, female sex, hypertension, absence of prior history of ischemic heart disease, first transmural myocardial infarction, marked ST segment elevation, anterior location, obstruction of the culprit artery, and use of corticosteroids or nonsteroidal anti-inflammatories increase the risk of free wall rupture [2-3]. During exercise, the increased intracardiac pressure can overwhelm weakened tissue and cause this devastating occurrence.

Amyloidosis is a metabolic disease that results in organ dysfunction because of the deposition of amyloid proteins in various tissues with the heart being a common place for involvement. Myocardial infarction as a result of narrowing of the epicardial coronary arteries and more commonly as a result of amyloid in the intramural (microvascular) coronary arteries has previously described in patients with cardiac amyloidosis at autopsy. Deposition of amyloid in the heart reduces effective muscle mass and can theoretically reduce space for collagen deposition post myocardial infarction [4]. Both of these factors would contribute to rupture in the setting of vulnerable ischemic tissue.

Case Presentation

We present a 75-year-old man with dyslipidemia who presented to his internist with intermittent chest pressure for several days which was initially exertional. The night prior to seeing his internist the discomfort was present while lying in bed. The symptom seemed to partially resolve with TUMS and a non-steroidal medication. On examination by his internist, he was noted to have a blood pressure of 140/82 with a pulse of 78 and an oxygen saturation of 98%. His cardiopulmonary exam was normal with a normal S1 and S2, no murmurs, rubs or gallops, clear lung sounds and no peripheral edema. His EKG was unchanged from his prior EKG and was only significant for a RBBB see (Figure 1). A proton pump inhibitor was prescribed and a stress echo was ordered which was obtained six days later.

When he arrived for his stress test, his baseline EKG was identical to his EKG at his internist's office. His baseline blood pressure was 128/70 with a pulse of 72 beats per minute. The baseline echocardiogram revealed normal left ventricular wall thickness, an ejection fraction of 60% and very mild hypokinesia of the lateral wall. The left atrial size and aortic root was normal. The aortic valve was trileaflet and opened normally. The mitral valve was normal in structure and function. The right ventricle was normal in structure and function. The right atrial size was normal. The tricuspid valve was normal in structure and function. No pericardial effusion was demonstrated (Figure 2).

He exercised for 7 minutes and 30 seconds, stopping due to light headedness. His heart rate reached 140 beat per minute and his maximum blood pressure was 150/70. He developed 4 mm of ST depression in early recovery followed by 16 beat run of non-sustained ventricular tachycardia at 2:03 into recovery (Figures 3-4). At approximately 4 minutes into recovery, he lost his pulse and was noted to have atrial fibrillation with slow ventricular response (Figure 5). In addition, the voltage on the ECG was noted to be significantly lower compared to baseline. At this time, he was also noted to have a rapidly enlarging pericardial effusion (Figure 6). A crash cart was brought to the room and an interventional cardiologist was called to attempt pericardiocentesis. Unfortunately, no significant pericardial fluid could be drained secondary to clots of blood in the pericardial space. Attempts at resuscitation lasted over 20 minutes with persistent PEA with three salvos of ventricular fibrillation which were successfully defibrillated with 360 J of energy. At 25 minutes into the code, he was noted to be in asystole and the code was called (Figure 7). The family was made aware of the situation and came to the hospital to see the patient one last time. An autopsy was performed.

At the time of autopsy, the heart weighed 340 grams. There were 275 grams of clotted blood with 50 ml of liquid blood in the pericardial space, as well as a 1.5 cm defect/rupture site on the posterior-lateral side of the left ventricle (Figures 8-9). The epicardium surrounding the site of rupture was hemorrhagic and mottled. The mottled area spanned from the defect posteriorly and laterally. The maximal thickness of the left ventricle was at the base (1.5 cm) which is mildly thickened. The right ventricle measured 0.5 cm (normal). The atrial bilaterally were normal. The valves were all normal. The left anterior descending artery conveyed a 40% stenosis, the left circumflex a 50% stenosis and the right coronary artery conveyed a 40% stenosis proximally and 60% distally. All of which are considered non flow limiting lesions.

Microscopic examination of the heart revealed multifocal acute infarctions of the posterior and lateral wall of the left ventricle with coagulation necrosis and neutrophils (Figure 10). There were adjacent areas with granulation tissue and early fibrosis. However, what came as a surprise were multiple focal areas of amorphous material surrounding the myocytes throughout the left ventricle (Figure 11). There was chronic inflammation present within the adventitia of the coronary arteries, likely related to amyloidosis. A congo red stain was positive with apple-green birefringence on polarized light. Kappa and lambda stains were negative. Transthyretin stain was strongly positive. There was no amyloid detected in any other organ system supporting age related, single organ, ATTR cardiac amyloidosis.

Discussion

Myocardial rupture in the setting of ischemia/infarction has become less common with the wide spread availability of cardiac catheterization labs which can limit the degree of infarction in the setting of a STEMI. In this case, the patient clearly did not have a STEMI prior to his visit with the internist or prior to his stress test based on his benign resting EKG. It was during the stress test when significant ECG changes were seen, most notably in early recovery. The ST depression seen in the right sided precordial leads likely represented "reciprocal changes," and actual posterior wall ST elevation with acute infarction of the posterior and lateral wall of the left ventricle. The transmural ischemia during exercise, in the setting of increased intracavitary pressures, was able to precipitate a complete tear in the myocardium leading to acute hemorrhagic cardiac tamponade. What makes this case extremely unique in that there was only moderate epicardial atherosclerosis seen at autopsy without evidence of recent plaque rupture or thrombus in the left circumflex or right coronary arteries which could typically cause this type of presentation. The most likely explanation is that the cardiac amyloidosis was a major factor in his sudden death.

Amyloidosis is a metabolic disease that results in organ dysfunction because of the deposition of amyloid proteins in body tissues. Cardiac amyloid typically presents as a restrictive cardiomyopathy secondary to interstitial amyloid deposition; however, it can uncommonly present with ischemic symptoms [5]. The pathophysiology behind this presentation is the progressive luminal narrowing of small intramural coronary arteries caused by amyloid deposition in the arterial vessel wall resulting in myocardial ischemia [6]. Coronary angiography is typically unremarkable, as would have been the case here based on the autopsy, as the infiltration is predominantly microvascular [7].

In this case, the combination of moderate atherosclerotic epicardial disease in conjunction with amyloid induced microvascular disease caused this man to start developing anginal symptoms approximately 8-10 days prior to his death. Based on his EKG, which failed to demonstrated Q waves, he did not have a significant infarction prior to the stress test. He likely had a subacute infarction prior to the stress test based on the baseline echocardiogram showing a very mild posterolateral wall motion abnormality and fibroblasts/early fibrosis noted on autopsy which typically takes several days to form after an ischemic insult. The ischemia during the stress test was profound with transmural ischemia of the posterior and lateral walls in a myocardial segment which was already compromised by recent subacute infarction and amyloid infiltration. This constellation of findings led to the catastrophic result. As his cardiac amyloidosis was in its earliest stage based on the absence of significant left ventricular hypertrophy, this may have put him at higher risk for a rupture. In a more advanced staged with a thickened wall, he may have been more protected against an event like this.

In addition to myocardial perforation, bowel perforation in the setting of amyloidosis has also been described with a similar proposed mechanism of microvascular ischemia with compromised bowel tissue [8]. In this case, there were no findings on autopsy of organ involvement outside of the heart. This is not unique, as ATTR amyloidosis, in contrast to light chain amyloidosis, has the strongest propensity to involve the myocardium, peripheral nerves and tendons. The latter were not examined at the time of the autopsy.

In conclusion, we describe an unfortunate case of myocardial rupture during exercise with cardiac amyloidosis as a major contributor. In this case, it does not seem that the outcome could have been altered as a moderate degree of exercise at home would have likely led to the same outcome. This case does remind us that ATTR amyloidosis is not benign and begs the question if screening and treatment before overt heart failure symptoms develop is warranted to avoid complications such as this.

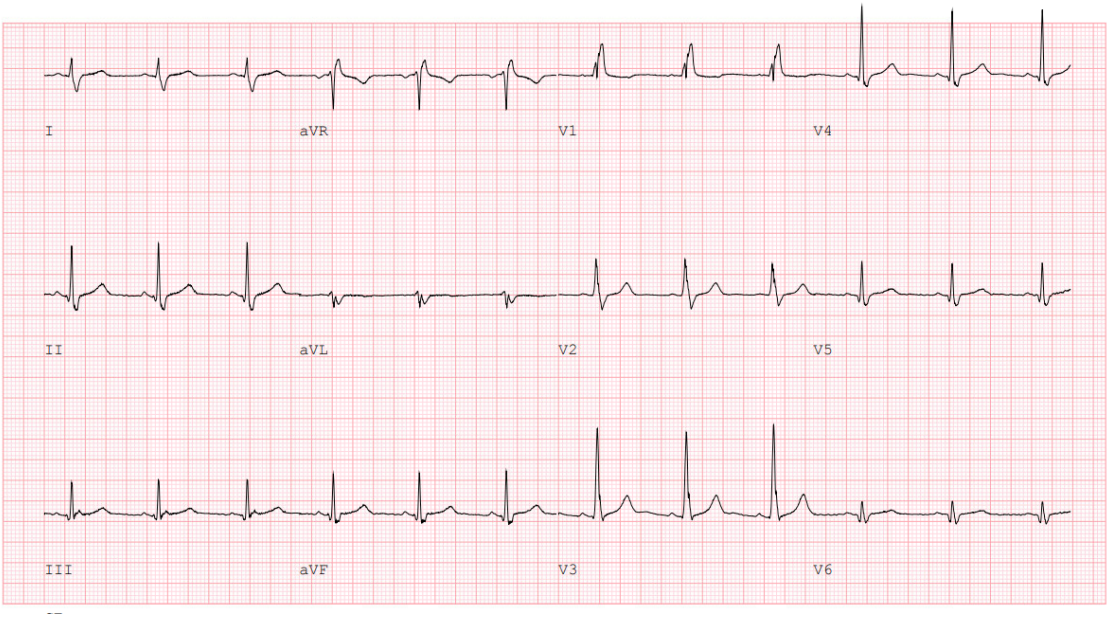

Figure 1: ECG showing normal sinus rhythm with RBBB

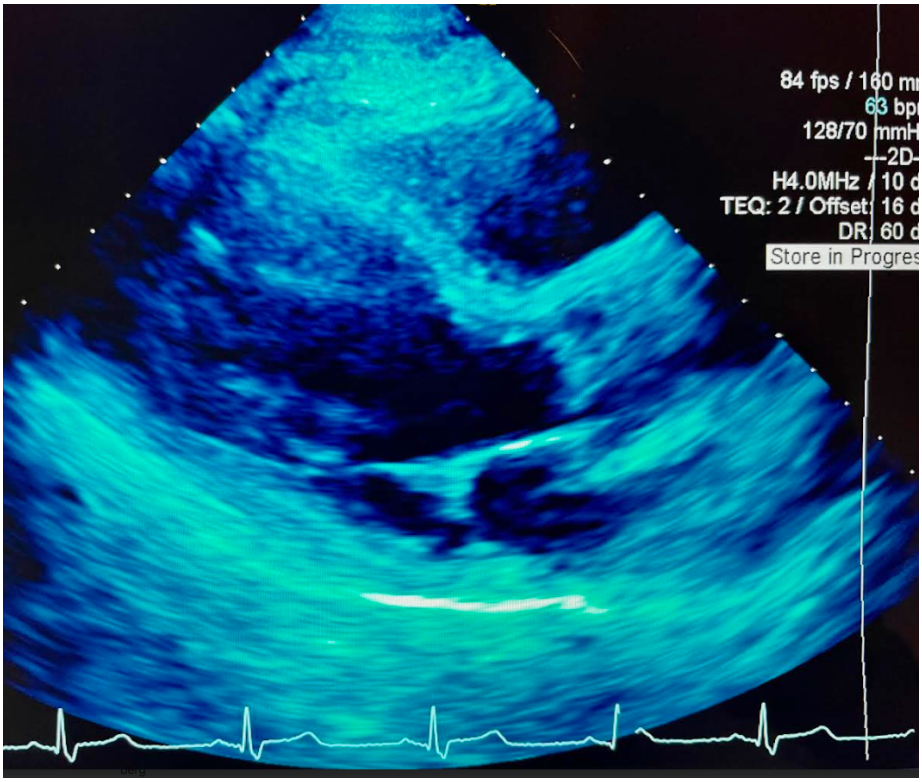

Figure 2: Parasternal long axis view showing normal LV size and thickness without pericardial effusion

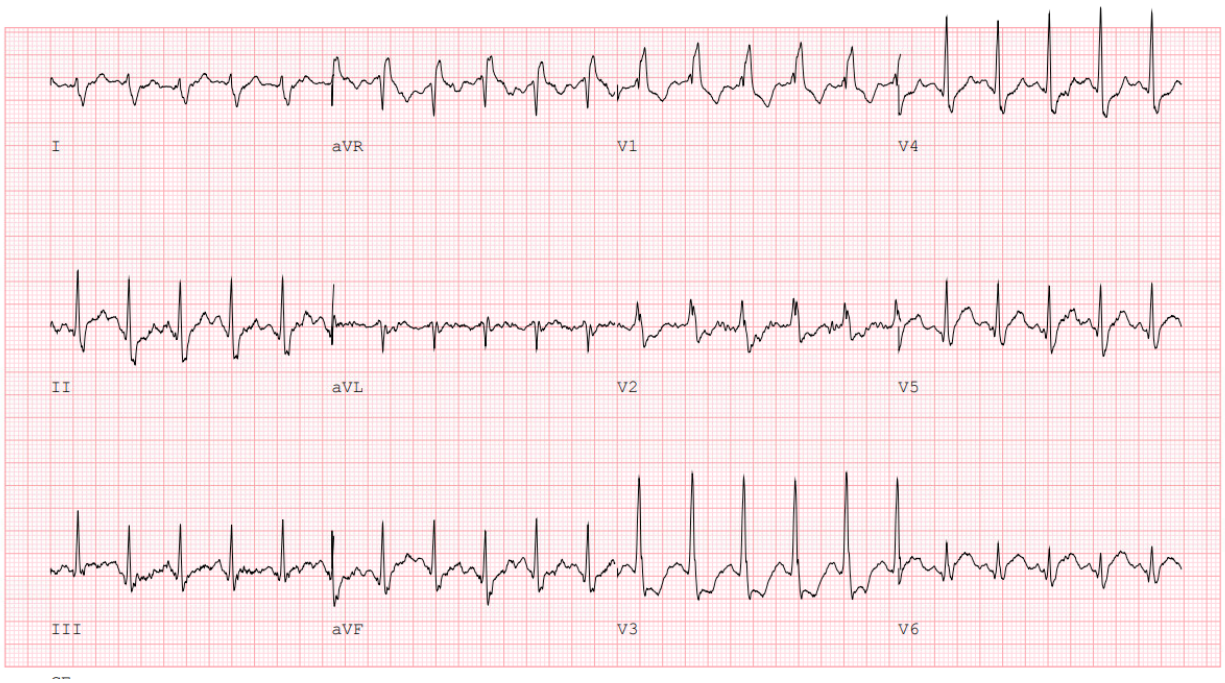

Figure 3: At peak exercise and immediately into recovery, up to 4 mm of ST depression in V2-V3

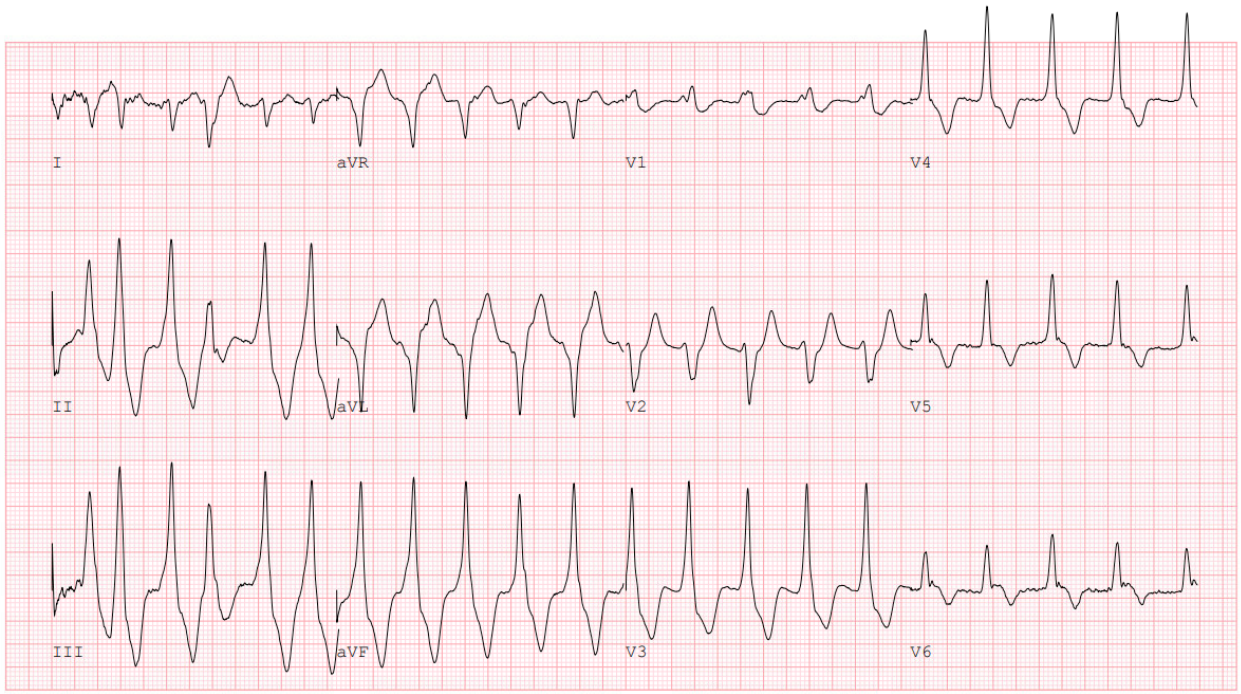

Figure 4: 16 beat runs of NSVT

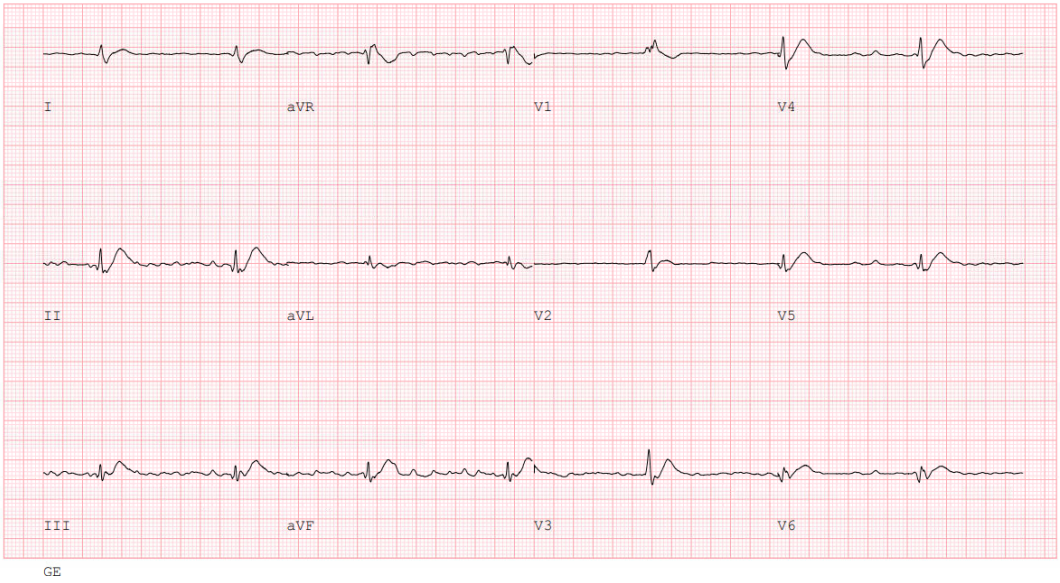

Figure 5: Atrial fibrillation with slow ventricular response with dramatic change in QRS voltage

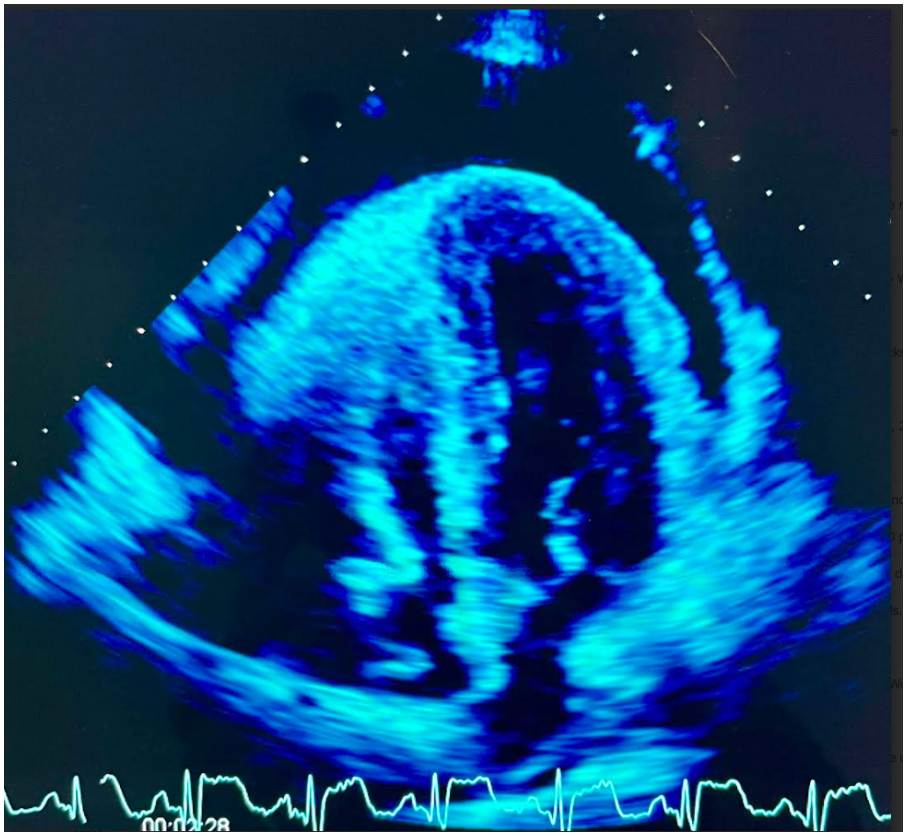

Figure 6: Apical 4 chamber view showing a large pericardial effusion with compression of the right-side chambers as well as the left atrium.

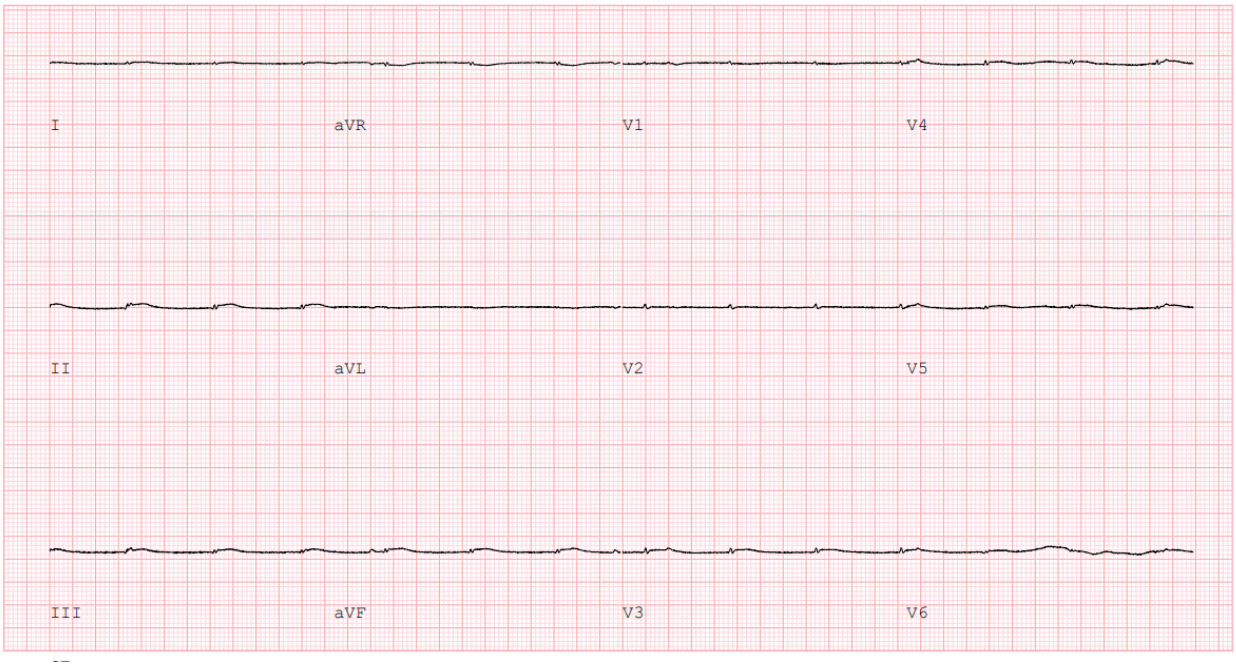

Figure 7: Asystole

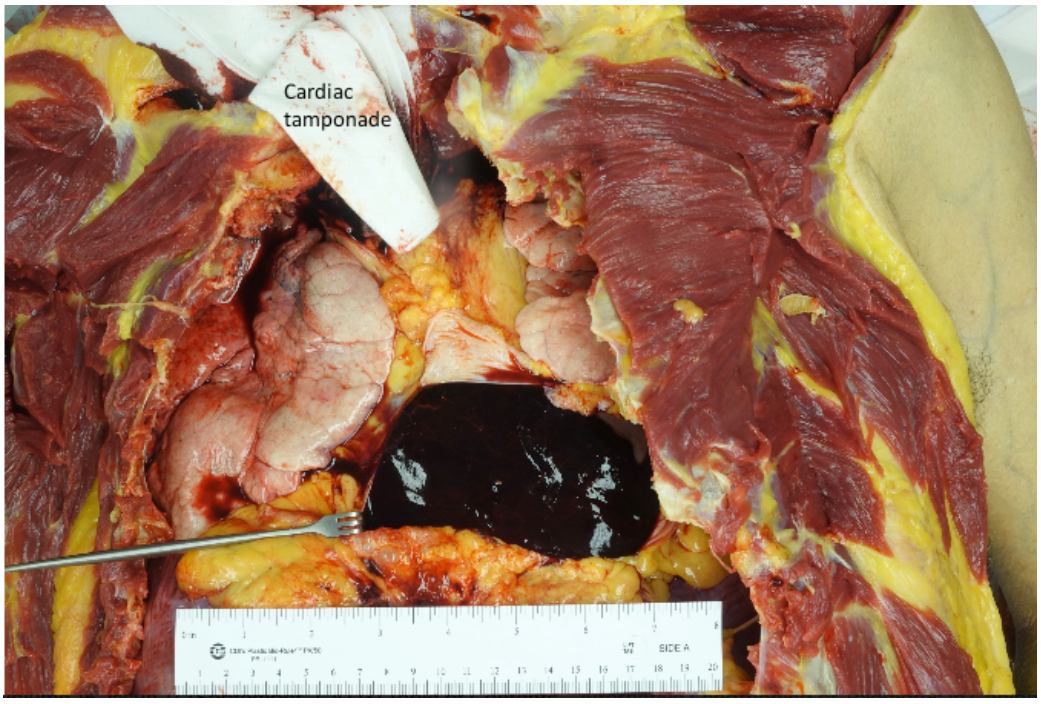

Figure 8: Clots of blood in the pericardial space

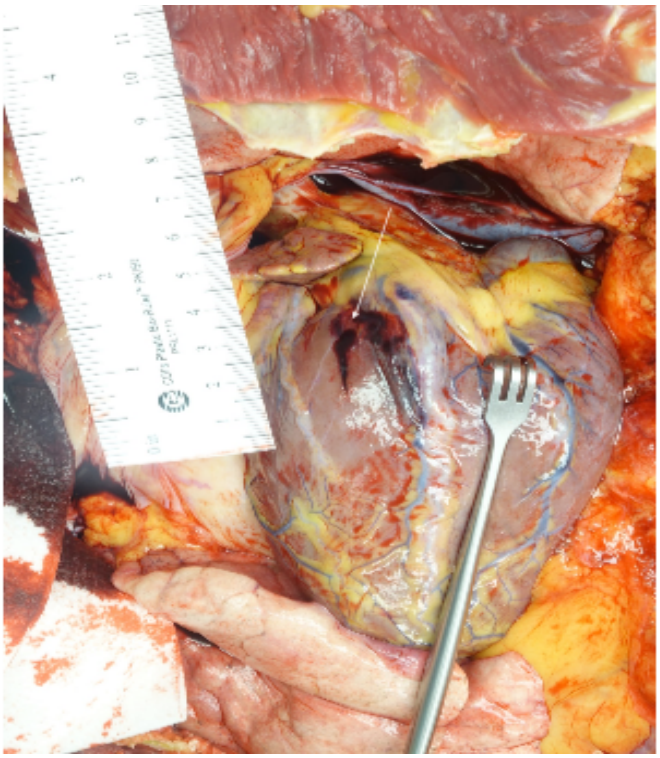

Figure 9: Slit like laceration in the posterior and lateral wall of the left ventricle

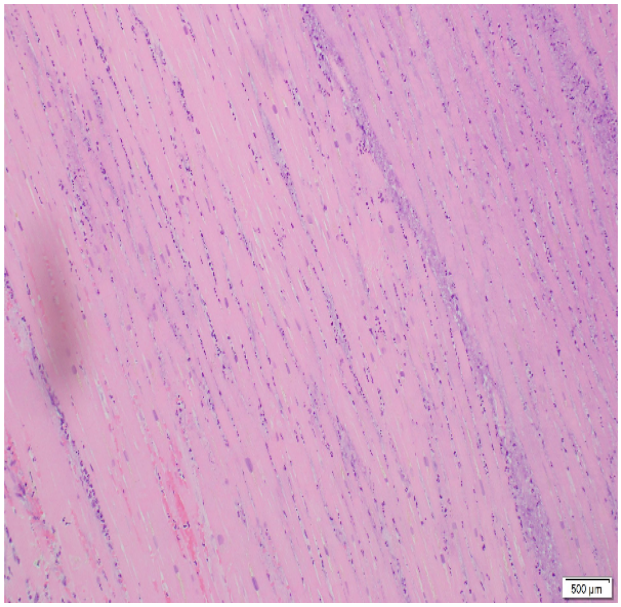

Figure 10: Myocardium at the defect site showing acute MI (loss of myocyte nuclei with interstitial neutrophilic infiltrates

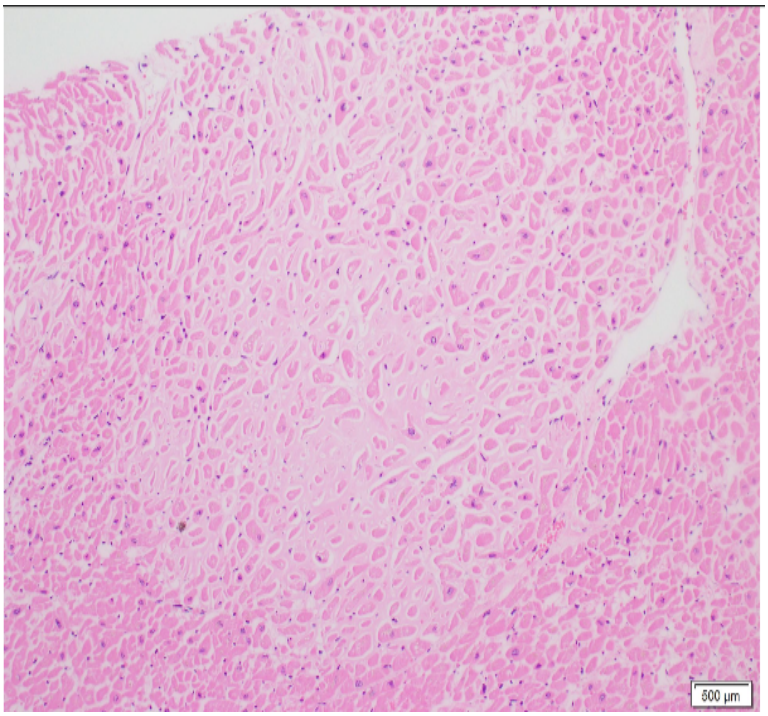

Figure 11: Myocardium adjacent to defect site showing cardiac amyloidosis with amorphous pink material between myocytes

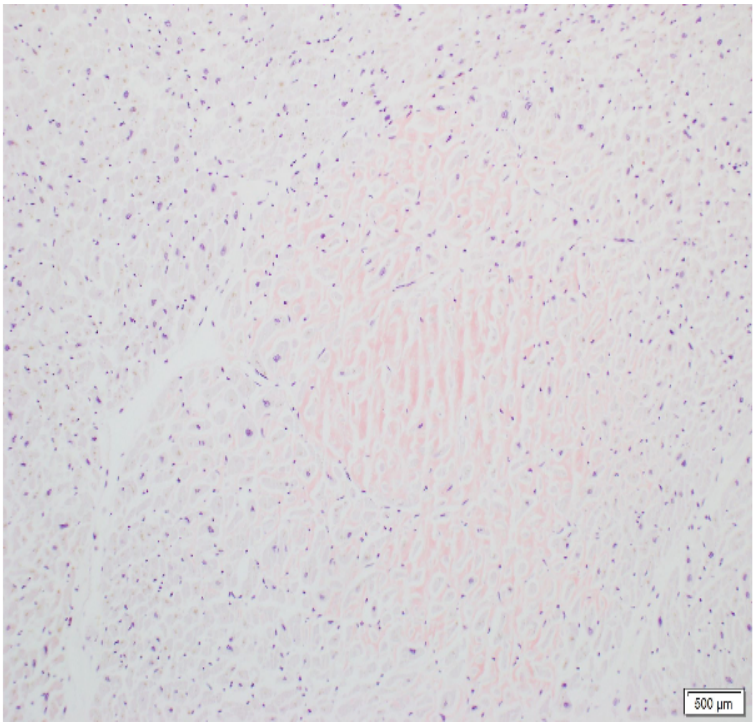

Figure 12: Congo red stain showing salmon pink staining of amyloid

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