Pathology Image of the Month

Acute Onset of Extreme Shortness of Breath

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A 33-year-old male presented via ambulance to the emergency department (ED) with extreme shortness of breath. A 12-lead electrocardiogram showed a heart rate of 158 beats/minute and atrial fibrillation with frequent runs of right bundle branch block-type aberrant conduction. Notable labs included a brain natriuretic peptide (BNP) level of 1167 pg/ml (<100pg/ml) and Troponin I of 1.23 ng/ml (<0.04ng/ml). Chest X-ray revealed bibasilar interstitial opacities interpreted as pulmonary edema. Chest auscultation was difficult due to obese body habitus, but heart sounds were described as distant and diaphoresis was noted. Bedside echocardiogram revealed severe mitral regurgitation. He became hypoxemic and required emergent endotracheal intubation. During intubation, he became bradycardic and entered pulseless electrical activity. Despite aggressive resuscitative protocols, the patient was declared dead within four hours of his arrival in the ED. Medical history was pertinent for morbid obesity and hypertension, as well as nonischemic cardiomyopathy characterized by recent (five days prior) left and right heart cardiac catheterization without ventriculography, echocardiogram findings of moderate pulmonary hypertension with no coronary artery disease, and a ventricular ejection fraction documented at 20%. Postmortem toxicology was subsequently shown to be negative. Full, unrestricted autopsy permission was granted under coroner authorization.

At autopsy, the heart was 1,020 grams [400±69] with no notable chamber dilatation. The coronary arteries coursed over the heart in the usual right-dominant fashion. All were widely patent with no significant atherosclerotic plaque identified on serial cross sectioning at 2-3 mm intervals. A 4 cm segment of myocardial bridging of the left anterior descending coronary artery was identified, beginning at 5 cm from its vascular origin and coursing at depths ranging from 0.4 to 0.7 cm. Ventricular wall thickness was as follows: left ventricle 2.6 cm, interventricular septum 2.3 cm, and right ventricle 0.4 cm. The heart was opened according to blood flow, beginning with a caval section into the right atrium. Figure 1 reveals the left atrioventricular section across the mitral valve.

Figure 1: Mitral valve at autopsy showing the anterior leaflet and its respective chordae tendinae with complete rupture of the anterolateral papillary muscle at approximately 1.0 cm from its tip.

What is the cause of death?

Diagnosis is on p. 176
DISCUSSION

Acute papillary muscle rupture has long been recognized as an uncommon, often catastrophic, cardiac emergency leading to massive mitral regurgitation, congestive heart failure, cardiogenic shock, and a nearly 80% mortality rate within the first 24 hours.\(^1\) Its stated frequency has most often been estimated by autopsy data such that, in one series, two cases among 6,000 consecutive autopsies were identified.\(^2\) The most common known cause of the rupture is preceding acute myocardial infarct (AMI), two to seven days prior, in the context of significant (>75% stenosis) atherosclerotic coronary artery disease (CAD), a pathogenic association that appears described in scientific literature that dates back to the late 1800s.\(^3\)

In the post-AMI patient population, ruptured papillary muscle is said to occur in less than 1% of all cases but account for up to 5% of all deaths.\(^4\) The posteromedial papillary muscle is preferentially involved five to eight times more often than its anterolateral counterpart, likely due to the fact that its vascular source relies solely on the posterior descending branch of a right-dominant coronary artery. Whereas the anterolateral papillary muscle is dually supplied by branches both of the left anterior descending (first diagonal), as well as the left circumflex (first obtuse marginal) coronary arteries.\(^5\) With the rarity of the condition, no particular constellation of risk factors predisposing AMI/CAD patients to papillary muscle rupture has yet to be described or understood.\(^4\)

With no clinical or pathological evidence either before death or at the time of autopsy for AMI or for CAD, the current case is classifiable as a spontaneous papillary muscle rupture (SPMR). These cases of non-AMI, non-CAD SPMR are exceedingly uncommon with very few described and retrievable in the English medical literature.\(^6-12\) In some cases of SPMR, various mechanisms for rupture have been documented, ranging from endocarditis and syphilis to trauma, as well as small vascular arteritis, sickle cell anemia, and aneurysms.\(^6\) However, in a small subset of SPMR cases, no clearly associated mechanism is evident. To our knowledge, the current case is the fifth reported case wherein no etiologic cause or preceding event is identifiable, and appears to be only the second such case wherein rupture involves the anterolateral papillary muscle. Features from these cases are compared in Table 1, where it is demonstrated that, in all but one case, there is either pathologic evidence and/or a clinical history of hypertension (HTN). Another shared feature illustrated in the table is the histopathologic evidence of ischemic necrosis, or infarction, that is isolated to the papillary muscle bundle itself rather than involving the adjacent left ventricular myocardium (Figure 2a-b). In a 1990 review of 110 consecutive adult hearts retrieved at autopsy, Waters documents evidence of papillary muscle ischemia (PMI) far more frequently (77% of cases) than ischemic changes in the ventricular walls (33% of cases). Acute PMI, specifically, was identified in 23% of post-mortem hearts. Still further, Waters suggests that both cardiomegaly and HTN increase the risk for acute PMI since 42% of hearts weighing more than 500 gm, and >1/3 of hypertensive patients had an acutely infarcted papillary muscle.\(^13\) This causal relationship may be partly explained by the hypothesis that these patients are at a higher risk both for subendocardial hypoperfusion, as well as for PMI, because of an increased resistance to perfusion and a wider separation of a relatively lower volume of perfusing capillaries that results from the greater ventricular mass and wider myocyte fibers.\(^13\) The case presented here is notable and unique, both for the patient’s young age and clinical history of hypertension and for the extreme cardiomegaly (1,020 gm) and left ventricular hypertrophy. Another feature of interest is the long segment (4 cm) of myocardial bridging...
at significant depths of 0.4 - 0.7 cm. What role the bridging may have played on possible transient occlusion of the mid-left anterior descending coronary artery in the region of the origin for the first diagonal branch that serves to supply the anterolateral papillary muscle is unclear. Further reports on cases of SPMR, particularly of the anterolateral muscle where myocardial bridging, marked cardiomegaly, and ventricular hypertrophy are observed, are needed.

ACKNOWLEDGEMENTS

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REFERENCES


Table 1: Clinical and pathologic features of current case compared to reported cases in the literature of non-AMI/non-CAD-associated SPMR with PMI. CTR = chest:thoracic ratio; LVH: left ventricular hypertrophy; HTN: hypertension

<table>
<thead>
<tr>
<th>Age/Gender</th>
<th>Anterior or Posterior Muscle</th>
<th>Clinical Outcome</th>
<th>Pathologic Signs or Clinical History of HTN</th>
<th>Pathology of Ruptured Papillary Muscle</th>
</tr>
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<tr>
<td>Current Case</td>
<td>Male, 33y</td>
<td>Anterior, complete</td>
<td>Fatal</td>
<td>Cardiomegaly, LVH, history of HTN</td>
</tr>
<tr>
<td>Lazar et al.</td>
<td>Female, 76y</td>
<td>Anterior, complete</td>
<td>Mitral valve replacement, survival</td>
<td>None</td>
</tr>
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<td>Tsuboi et al.</td>
<td>Male, 77y</td>
<td>Posterior</td>
<td>Mitral valve replacement, survival</td>
<td>CTR 67%</td>
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<td>Lee et al.</td>
<td>Female, 49y</td>
<td>Posterior, complete</td>
<td>Mitral valve replacement, survival</td>
<td>LVH, history of HTN</td>
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<tr>
<td>Ripoll Vera et al.</td>
<td>Female, 73y</td>
<td>Posterior, complete</td>
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<td>LVH, history of HTN</td>
</tr>
<tr>
<td>Weiss et al.</td>
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<td>Mitral valve replacement, survival</td>
<td>LVH, history of HTN</td>
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