CLINICAL VIGNETTE

A Confusing Clinical Picture

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Clinical Scenario

A 67-year-old man with posterior mitral valve prolapse and dyslipidemia presented to the emergency department with sudden onset dyspnea. He was visiting from across the country and was in his hotel when the symptoms began. There was no chest pain, cough, or fever and the dyspnea was constant and profound, associated with nausea and diaphoresis. On examination, he was afebrile with a blood pressure of 96/69 mmHg and in respiratory distress with rate of 28 breaths per minute. Chest exam revealed bilateral rales and wheezing and regular tachycardia to 145 beats per minute with a brief systolic murmur. He was cool on exam without edema and his neck veins were not appreciable due to body habitus. Initial pulse oximetry was 79% on room air and improved to 92% on 100% oxygen with non-invasive positive pressure ventilation (NIPPV). Laboratory testing was notable for a leukocytosis of 25.4 thousand/uL with 75% neutrophils, acute kidney injury with a creatinine of 1.7 mg/dL, and lactic acidosis with a lactate of 7.9 mmol/L and pH of 7.21. B-type natriuretic peptide was mildly elevated at 318.4 pg/mL and the initial troponin was negative. Profound hypoxia was confirmed with an arterial pO2 of 71mmHg despite 100% oxygen and NIPVV. His electrocardiogram showed sinus tachycardia but was otherwise unremarkable and chest radiograph was interpreted as bilateral opacities with the right much worse than the left, consistent with multifocal pneumonia.

The etiology of the patient's presentation was not entirely clear, but the asymmetric infiltrates, significantly worse on the right, leukocytosis with shift, hyperglycemia without a history of diabetes, and tachypnea made the initial diagnosis pneumonia with septic shock. An echocardiogram was later performed but was technically limited due to the patient's respiratory distress and positional issues. There was at least moderate mitral regurgitation, though his tachycardia and poor acoustic windows made the true degree not appreciated. Further while there was at least clear prolapse of the valve, some views suggested a possible flail leaflet. He had some improvement in oxygenation and his lactic acidosis but began to tire from his high work of breathing and was intubated. Shortly after intubation, transesophageal echocardiogram confirmed concerns about a flail segment. There was rupture of the chordae tendinea and flail of the P2 scallop of the posterior leaflet of the mitral valve with severe regurgitation towards the right sided pulmonary veins. The patient was immediately taken for coronary angiogram and bilateral heart catheterization, revealing no coronary artery disease, and confirming cardiogenic shock. An intra-aortic balloon pump (IABP) was placed to reduce the regurgitant volume through the mitral valve and increase forward flow and the patient was taken for emergent cardiac surgery. Due to clinical instability, it was felt he would not tolerate the prolonged time to attempt a mitral valve repair and his valve was replaced with a bio prosthesis. It was difficult to wean him from cardiopulmonary bypass, but with the addition of inhaled nitric oxide, manipulation of the ventilator, and a combination of inotropes and the IABP, he was able to be disconnected from cardiopulmonary bypass. Over several days his renal failure and shock resolved and with diuresis his oxygen requirements progressively improved to the point of extubation. His recovery was prolonged, but ultimately he was discharged to his home with normal cardiac function.

Discussion

This case offers multiple areas for important review and discussion. It took approximately 48 hours to arrive at the correct diagnosis, which is a medical emergency and requires immediate surgical correction. If the severe, acute mitral regurgitation had been recognized earlier his peri-operative and post-operative course may have been simpler and shorter and he might have been able to have a valve repair instead of a replacement. There was a strong focus initially on a suspected pulmonary infection and septic shock. This was due to the patient presenting with three of the four systemic inflammatory response syndrome (SIRS) criteria and the interpretation of the radiograph as bilateral pneumonia. The SIRS criteria have a high sensitivity for sepsis, but a low specificity (approximately 25%) for the condition and simply represent an abnormal physiologic response to a stressor on the body, including things such as trauma or pancreatitis, not necessarily an infection.[1] The limitations of the SIRS criteria in the setting of an infection to indicate the presence of sepsis has been widely recognized and has caused a change in the way that sepsis is conceptualized,[2] focusing on primarily confirming an infection and then focusing on degree of organ dysfunction.

In our patient, the asymmetric pulmonary infiltrates, interpreted as bilateral pneumonia, were felt to represent the infectious source causing sepsis in the patient. However, it is important to always evaluate the radiology findings in the clinical context, and our patient denied any cough or fevers and reported his

symptoms appeared suddenly, all of which go against a diagnosis of pneumonia. One might expect a more insidious onset of symptoms and a significant cough given the extensive radiographic findings. While uncommon, unilateral pulmonary edema does occur, almost always in the setting of severe mitral regurgitation.[3] As only the P2 scallop is flail, the regurgitation jet is primarily focused anteriorly towards the right sided pulmonary veins that drain the right lungs. While the left atrial pressure rises with the acute mitral regurgitation it is more focused to the right, explaining the asymmetric radiography findings. In patients with unilateral pulmonary edema due to severe mitral regurgitation, the edema was more often right sided and as was the case with our patient. A study of patients with unilateral pulmonary edema indicated compared with bilateral pulmonary edema reported delayed diagnosis and increased use of antibiotics[3] which is what happened with our patient.

The only effective treatment for acute, severe mitral regurgitation is to repair the defect. Vasodilating medications, inotropes, and the IABP can help promote forward flow of blood from the left ventricle into the systemic circulation as a temporizing measure, but it cannot be managed without repair. While patients are inevitably critically ill, early valve repair or replacement is superior than initial conservative therapy with delayed surgery. Studies have shown early intervention resulted in reduced mortality in the short and long-term and reduced morbidity including less atrial fibrillation and progressive heart failure.[4] While open surgical repair or replacement has been the standard of care, recently transcatheter edge-to-edge repair (TEER) using a percutaneous placed clip to secure the flail scallop has been employed. Our patient's flail P2 scallop was secured to the structurally normal A2 scallop to prevent it from flailing. This was an appealing option to avoid a major cardiac surgery requiring cardiopulmonary bypass in a critically ill patient. Published series report high success rates with low complications.[5] However, this was not available at the patient's hospital and he was too unstable to transfer. The cardiac surgery team was able to perform an emergent valve replacement without any complications.

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