A 41-year-old Woman With Rheumatic Mitral Stenosis, Atrial Fibrillation, and Right-sided Heart Failure

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Case Presentation

A 41 year old Southeast Asian woman presented to the Emergency Department (ED) complaining of acute onset of nausea and abdominal pain as well as a two weeks of palpitations and two months of cough productive of white sputum. The patient also complained of fever, shortness of breath and dyspnea on exertion over the previous week. Patient history revealed a past medical history of asthma. The patient had no prior surgical or psychiatric history, had no known drug allergies, and consumed no chronic medications. The patient reported that she had immigrated to the United States from Laos in 1986. She denied any tobacco, alcohol or illicit substance abuse.

The patient was afebrile and demonstrated hemodynamic stability in face of her EKG demonstrating atrial fibrillation at a ventricular response rate of 80 bpm. Lung auscultation revealed bilateral crackles at the lower bases, but no rales or wheezing. Cardiac examination demonstrated and an irregularly irregular pulse with a mid-diastolic rumbling murmur heard best at the apex without radiation. She demonstrated elevated jugular venous distention to ~12 cm H_2O and demonstrated 1+ pitting lower extremity edema.

Laboratory data was significant for a beta-natriuretic peptide (BNP) of 972 pg/mL. The patient had a microcytic anemia with a hemoglobin 12.6 mg/dL. Troponins were all within normal limits and the EKG did not reveal any ischemic changes. X-ray findings, later corroborated by chest CT, demonstrated bilateral pleural effusions, bilateral lower lobe infiltrates with mediastinal and hilar lymphadenopathy. The patient was admitted to the critical care unit for transesophageal echocardiogram (TEE) which demonstrated:

The left atrium is severely dilated. The right atrium is moderately dilated. There is moderate to severe tricuspid regurgitation. Right ventricular systolic pressure is elevated at >60mmHg. Left ventricular systolic function is borderline reduced. Flattened septum is consistent with RV pressure/volume overload. The right ventricle is mildly dilated. The right ventricular systolic function is hypokinetic. There is severe mitral stenosis. The mean gradient was 11 mm Hg and the peak gradient was 20 mm Hg. The valve area is 1.1cm^2.

The results of the TEE suggested that the most likely etiology for the patient's valvular and clinical findings was that of rheumatic mitral stenosis. The patient's age, immigration status, and relative lack of medical history also fit the prototypical model of a patient with complications of prior acute rheumatic fever. Possibly superimposed on this disease process is a bacterial pneumonia causing the bibasilar infiltrates or these radiologic findings which may simply have been an extension of the sequelae of the rheumatic heart disease.

Discussion

Rheumatic fever is a delayed consequence of pharyngeal infection with *group A streptococcus* (GAS). The disease manifestations can affect several areas of the body, namely the cardiovascular, musculoskeletal, neurological and dermatological systems. These are believed to be a result of a diffuse inflammatory process that results in an autoimmune attack on the body due to the phenomenon of molecular mimicry. The GAS cell wall contains M proteins that are antigenically similar to proteins found in the human body. When an immune response is mounted to the initial GAS infection, antibodies are formed against this M protein, which then circulate throughout the body and bind to normal protein epitopes in human tissue. The complexes formed then induce a T-cell mediated attack on normal tissues, causing the long term sequelae of the disease.[1]

Diagnosis of acute rheumatic fever has remained relatively unchanged since it was first described in 1889 by William Cheadle in London. In 1965, the American Heart Association released the Jones guidelines, laying out a specific set of criteria to be used for diagnosis.[2] In 1992, the Jones criteria were revisited and revised, and these guidelines are currently used in clinical practice:

Table 1. For diagnosis, two major criteria OR one major criterion with two minor criteria is needed, in addition to evidence of prior GAS infection

5 major manifestations:	4 minor manifestations
Carditis	Arthalgia
Migratory polyarthritis	Fever
Sydenham's chorea	Elevated ESR or CRP
Erythema marginatum	Prolonged PR interval
Subcutaneous nodules	
Evidence of preceding streptococcal infection:	
Positive throat culture for GAS or positive rapid streptococcal antigen test	

Rheumatic carditis is considered to be the most serious of the manifestation of GAS infection. It is very difficult to detect before the disease is severe enough to become symptomatic. The disease can manifest in any of the three layers of the myocardium, but most often affects the cardiac valves, especially the mitral valve (85% of affected patients). Small nodules known as Aschoff bodies initially form on the valve leaflets and slowly enlarge due to increased fibrin deposition. These deposits slowly decrease the functional ability of the valve leaflets causing mitral stenosis or mitral regurgitation, and functionally limit normal

blood flow. Chronically, this leads to left atrial enlargement (which can lead to atrial fibrillation), pulmonary hypertension, and eventually right-sided heart failure.[4] Natural history studies suggest that the average amount of time from the onset of rheumatic fever to onset of mitral valve symptoms is 16.3 years. Further progression to severe mitral disability from the onset of symptoms is 9.2 ± 4.3 years.

Treatment of rheumatic carditis is dependent on the extent of the disease on presentation. If the cardiac damage is not extensive, prophylactic treatment includes aspirin for mild cases and steroids for more severe cases.[4] In patients with severe mitral regurgitation or stenosis, surgery is indicated to improve blood flow through the cardiac system.[5] It has been shown that percutaneous balloon valvuloplasty and mitral valve repair have shown similar initial results and efficacy at three years; therefore, percutaneous balloon valvuloplasty is the preferred surgical treatment due to better long term patency, lower cost, and decreased need for open thoracotomy.[6]

Patient Course

The patient's atrial fibrillation was rate controlled with a combination of beta-blockers and digoxin. She was bridged from intravenous heparin to oral warfarin for anticoagulation and her bibasilar pneumonia was treated with broad spectrum antibiotics. She was transferred to another institution for cardiothoracic evaluation of potential percutaneous balloon valvuloplasty of the mitral valve.

References

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