Rheumatic Valvular Heart Disease

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Case

Ana K is a 45 year old Nepalese woman
She presents with an interpreter hoping to get a refill on a medication she was prescribed back home for an abnormal heart rhythm she has had for several years
This is the only medication she is on
It was started when she had a small stroke effecting her speech 15 years ago. She has no residual problems from the stroke
She does not know what type of abnormal heart rhythm she has
She really has no symptoms, specifically SOB, palpitations, lightheadedness, chest discomfort, weight gain or swelling
She is tired, but no more than she has been
Physical exam

Irregularly irregular pulse
Diastolic murmur (loud)
No residual neurologic deficits
Grading Heart Murmurs

- graded on a 6 point scale
  - Grade 1 = very faint, heard intermittently
  - Grade 2 = quiet but heard immediately
  - Grade 3 = moderately loud
  - Grade 4 = very loud with palpable thrill
  - Grade 5 = very loud and can still hear with edge of stethoscope
  - Grade 6 = heard with stethoscope partly off the chest
- *Note: Thrills are assoc. with murmurs of grades 4 - 6
Ana’s ECG
Echocardiogram
2 D Echocardiogram characteristics

- Thickened and calcified mitral valve leaflets and subvalvular apparatus
- Restricted motion
- Diastolic doming of leaflets (hockey stick appearance)
- Increased LA size
- Fish mouth appearance in short axis view
Domed mitral valve leaflets
Hockey stick appearance of anterior mitral valve leaflet
Stenotic mitral valve end diastole (fish mouth appearance)
Spotted blue puffer fish
Severity by mean gradient across the mitral valve

Severity (by Mean Gradient)

- Mild MS < 5mmHg
- Moderate MS 6-10mmHg
- Severe MS >10mmHg
Epidemiology of Rheumatic Fever

ARF caused by group A beta- hemolytic streptococcus affects 20 million people and is the leading cause of cardiovascular death in the first 5 decade of life.

Mean incidence worldwide is 19 per 100,000.

US 5 per 100,000

- Resurgence in past few decades

The worst affected areas are sub-Saharan Africa, south-central Asia and the Pacific where incidence has been reported as high as 1 percent.
Rheumatic Heart Disease

RF principally involves the heart, joint, CNS, skin and subcutaneous tissues.

Rheumatic Heart Disease refers to the cardiac involvement that develops in 50% of patients with RF.

Can affect the endocardium, myocardium or pericardium.

It may later effect the heart valves causing chronic valve disease leading to CHF and death.

There is a latent period of up to several decades before valve disease becomes severe enough to recognize.
Pathophysiology

Causative agent
Group A Beta-hemolytic streptococci

Untreated strep throat

Rheumatic fever

All layers of the heart and the mitral valve become inflammed

Vegetation forms

Valvular Regurgitation and stenosis

Heart Failure
Key feature of RF is granuloma formation

1. Exudative and degenerative phase
2. The proliferative phase (Granulomatous period)
3. Scar phase (healed phase\Fibrosis phase\Hardening phase)
Key morphologic features of acute rheumatic heart disease.
Acute rheumatic endocarditis: small (diameter 1- to 2-mm) vegetations along the mitral valve margin, insufficient to cause valvular deformation.
Small vegetations (verruca) are visible along the line of closure of the mitral valve leaflet (arrows).
Exudative and degenerative phase
It is characterized by serofibrinous exudate, with deposits of immune precipitate on collagen fibers that lead to fibrinoid necrosis.
Serous pericarditis

Fibrinous pericarditis
pericardial effusion

Can lead to heart sound far, around the heart boundary expanding, serious cardiac X-ray showed a flask.
Adhesive pericarditis is in cardiac surface of patients. From the epicardial surface to the pericardial sac visible fibrinous exudate, which is typical for a fibrinous pericarditis.
The proliferative phase (Granulomatous period)

**Aschoff Body**: pathognomonic for RF

**Structure:**
- **Center**: fibrinoid necrosis
- **Around the center**: Anitschkow cells, lymphocytes, occasional plasma cells

**Distribution:**
Myocardial interstitial, subendocardial and subcutaneous connective tissue.
Anitschkow cells

These distinctive cells have abundant cytoplasm and central round-to-ovoid nuclei in which the chromatin is disposed in a central, slender, wavy ribbon (hence the designation "caterpillar cells" cross section named Owl's eye cells).

Some of the larger macrophages become multinucleated to form Aschoff cells (inflammatory giant cells).
A Rheumatic granuloma

- Collagen fibers
- Fibrinoid collagen necrosis
- Plasma cells
- Lymphocytes
- Artery
- Aschoff cell
- Anitchkov cell (histiocyte)
Rheumatic granuloma
(HE) x 100
Owl's eye cells

Aschoff cells
Early changes to mitral valve on echocardiogram
Valves effected

Mitral valve most common
Then aortic valve
Tricuspid and pulmonary valves effected less than 5 % of the time
Mitral stenosis

Most common cause is RF (up to 99%)
Become symptomatic is 2\textsuperscript{nd} to 4\textsuperscript{th} decade of life
More women than men (2/3rds)
25\% of patients with chronic RF have pure MS
40\% have combined mitral stenosis and mitral regurgitation
Valve changes that lead to stenotic or regurgitant valve

- Commissural adherent and fusion
  - restricted opening of mitral valve
    - “fish mouth” shape of mitral valve orifice
  - Thickening, fusion and shortening of the chordae or papillary muscles
    - funnel-shaped change of valve apparatus
Clinical manifestations of mitral stenosis

Symptoms
- Onset in patients with moderate to severe MS (MVA < 1.5 cm²)

Dyspnea – Principal symptom, often the earliest.
- Precipitated by exertion, fever, a fib or pregnancy
- Orthopnea, weight gain, LE edema, PND

Hemoptysis – blood stained sputum, pink frothy sputum

Cough, especially at night

Hoarsness (Ortner’s syndrome) less common. Compression of left recurrent laryngeal nerve.
Physical exam signs of Mitral Stenosis

Cardiac signs of MS

Diastolic heart murmur

Opening snap and accentuated S1
  – Left sternal boarder or at apex
  – Suggests mitral valve leaflets are still flexible
  – As leaflets become stiffer and more calcified, OS dissapears
Mitral Regurgitation

Etiology – Mitral valve apparatus and or LV structural or functional abnormality

RHD commonly causes MR (one in three) +/- MS and also possibly AV disease

MVP

Ischemic heart disease

Dilated cardiomyopathy

Mitral annular calcification
Systolic murmur of mitral regurgitation

RHD – pan systolic, blowing, high-pitched murmur
  Loudest at apex (axilla, back)
MVP – mid to late systolic murmur
Papillary muscle dysfunction – variable murmur
Rupture of chordae tendonea - musical
Differential diagnosis of systolic murmur (MI.AS.TI.PS)

- Tricuspid regurgitation – systolic murmur along the left sternal border (Erb’s point), Increased with inspiration.
- VSD – loudest at left sternal border and can be accompanied by a parasternal thrill
- Systolic ejection murmur **Aortic Stenosis** vs pulmonic stenosis
- Hypertrophic cardiomyopathy
Aortic stenosis

RHD less common etiology but suspect it when AS is seen with MS and AI

Degenerative calcific AS. Most common cause of severe AS
  – Most common in the elderly
  – Can be associated with mitral annular calcification

Bicuspid AV.
Physical exam in aortic stenosis

Systolic ejection murmur

– Blowing harsh crescendo decrescendo
– Heard best at aortic listening post
– Radiates to neck and apex
– The more severe the AS, the longer the duration of the murmur
– Although, once LV fails (dilates) and cardiac output decreases, murmur will diminish
Aortic regurgitation

RHD is most common cause - 2/3rds originate from RF
– Can be associated with AS and MV disease

 Infective endocarditis

Bicuspid AV

Aortic root dilatation
– Marfan syndrome
– Aortic dissection
– Syphilitic aortitis
Physical examination in AI

Chronic, severe aortic insufficiency

Peripheral arterial signs due to widened pulse pressure

- Water hammer pulse (early rise and fall)
- Pistol shot sounds – loud systolic and diastolic sounds over femoral artery
- Duroziez’s sign – bruit over partially compressed femoral artery
- Quincke’s sign – subunguinal capillary pulsations
- de Musset’s sign
Aortic insufficiency murmur

Diastolic murmur
High-pitched, blowing, decrescendo pattern
When due to primary aortic valve disease
– Diastolic murmur heard best along left sternal border
– 3rd and 4th intercostal space
When due to dilation of the ascending aorta
– Heard best along right upper sternal border
Austin Flint murmur – apical mid or late diastolic low-pitched murmur. Common in severe AI. Due to partial closure of MV by the regurgitant aortic jet.
Final thoughts

Obtaining history of Rheumatic fever as a child
- History of repeated throat infections, fevers, missing school
- Remember half of those later diagnosed with rheumatic valvular heart disease have no recollection of having RF as a child

Atrial fibrillation
- Common consequence of mitral valve disease
- Due to LA dilatation
- Anticoagulate with Coumadin, not one of the new drugs