Cardiomyopathies
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- Cardiomyopathy is a disease of the heart muscle
- reduces its ability to pump blood to the rest of the body
- is a leading cause of heart failure
- is the common reason for heart transplantation
- is so dangerous because: - often goes unrecognized and untreated
  - frequently affects younger people
- **Cardiomyopathy: is a group of diseases that primarily involve the myocardium and produce myocardial dysfunction**
- usually present with heart failure and arrhythmias
- there are 3 main types of cardiomyopathy:
  1- dilated cardiomyopathy
  2- hypertrophic
  3- restrictive

1- Dilated cardiomyopathy: (DCM)

- is characterized by:
  1- four-chamber dilation
  2- myocardial hypertrophy
  3- impairment of contractility (systolic dysfunction)
- can occur at any age
- only 25% of patients survive more than 5 years (after diagnosis)

Pathogenesis:

- the cause is frequently unknown (idiopathic) but certain pathological conditions may contribute:

1- genetic defect: i- mutations in sarcomere (actin, myosin, troponin)

ii- mutations in cytoskeleton (desmin, dystrophin)

2- alcohol toxicity : due to direct alcohol toxicity or its metabolite (acetaldehyde) on myocardium

3- peripartum: - disease is discovered within months before or after delivery

- mechanism is uncertain, the association with pregnancy suggests: 1- volume overload

2- nutritional deficiency contribution

4- postviral myocarditis: myocarditis can progress to DCM

Morphology:

grossly: - cardiomegaly, chamber dilation, myocardial hypertrophy

- mural thrombi (stasis, poor contractile function)

microscopically: - myocyte hypertrophy and interstitial fibrosis

Clinical manifestation:

- heart failure
- arrhythmias

- stroke
- sudden death

DCM: grossly: - cardiomegaly, chamber dilation, myocardial hypertrophy
- mural thrombi (arrow-head)

microscopically: - myocyte hypertrophy and interstitial fibrosis

2- Hypertrophic cardiomyopathy: (HCM)

- is characterized by:
  1- *myocardial hypertrophy*
  2- abnormal diastolic filling
  3- ventricular outflow obstruction (in one third of cases)

Pathogenesis: - idiopathic or genetic defect may contribute

1- familial form:
- autosomal dominant
- occurs in young individuals
- due to mutation in genes coding for proteins of cardiac muscle sacromere (myosin
Troponin)

2- sporadic form: - occurs in elderly

Morphology:

grossly: - marked cardiomegaly
- myocardial hypertrophy
- asymmetrical ventricular septal hypertrophy leading to left ventricular outflow obstruction
microscopically: - myocytes hypertrophy
- myocyte and myofiber disarray
- interstitial fibrosis

Clinical manifestation:

- HCM can be: - asymptomatic or
- symptomatic (presents in young adults, with dyspnea, angina, near-syncope and CHF)

- complications: 1- atrial fibrillation with mural thrombus and embolization

2- infective endocarditis

3- left ventricular outflow obstruction

4- CHF

5- sudden death (more common than in other forms)

HCM: **A,** marked myocardial hypertrophy, septal hypertrophy. **B,** microscopically: myocyte hypertrophy and disarray. **C,** Sarcomere of cardiac muscle, showing proteins in which mutations cause defective contraction

3- Restrictive cardiomyopathy:

- rare
- *characterized by:*
  - *1- reduced ventricular compliance resulting in*
  - *2- impaired ventricular filling during diastole*
  - *3- leading to reduced cardiac output*

Pathogenesis:

1- idiopathic

2- secondary to: amyloidosis, radiation-induced fibrosis, hemochromatosis, sarcoidosis

- there is infiltrative process within myocardium result in stiffening of heart muscle which interferes with pumping action

Morphology:

Grossly: - ventricles are of approximately normal size

- the cavities are not dilated
- firm myocardium (fibrosis)

Microscopically: - there is interstitial fibrosis

C/F:

- HF, arrhythmias

**** Arrhythmogenic right ventricular cardiomyopathy (dysplasia):**

- is a recently recognized cardiomyopathy
- it is typically familial disorder
- characterized by:

1- right-sided failure

2- rhythm disturbances (ventricular tachycardia, sudden death)

Morphology: - thinned Rt ventricular wall

- myocyte loss and fatty infiltration

Clinical features:

- death occurs secondary to: CHF

embolism or mural thrombi
fatal arrhythmias

Thank you