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