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