

**Fatal spontaneous aortic
dissection of ascending
aorta having cystic
medial degeneration:**

An autopsy case

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Background

Anatomy of the AORTA

- Two main parts:

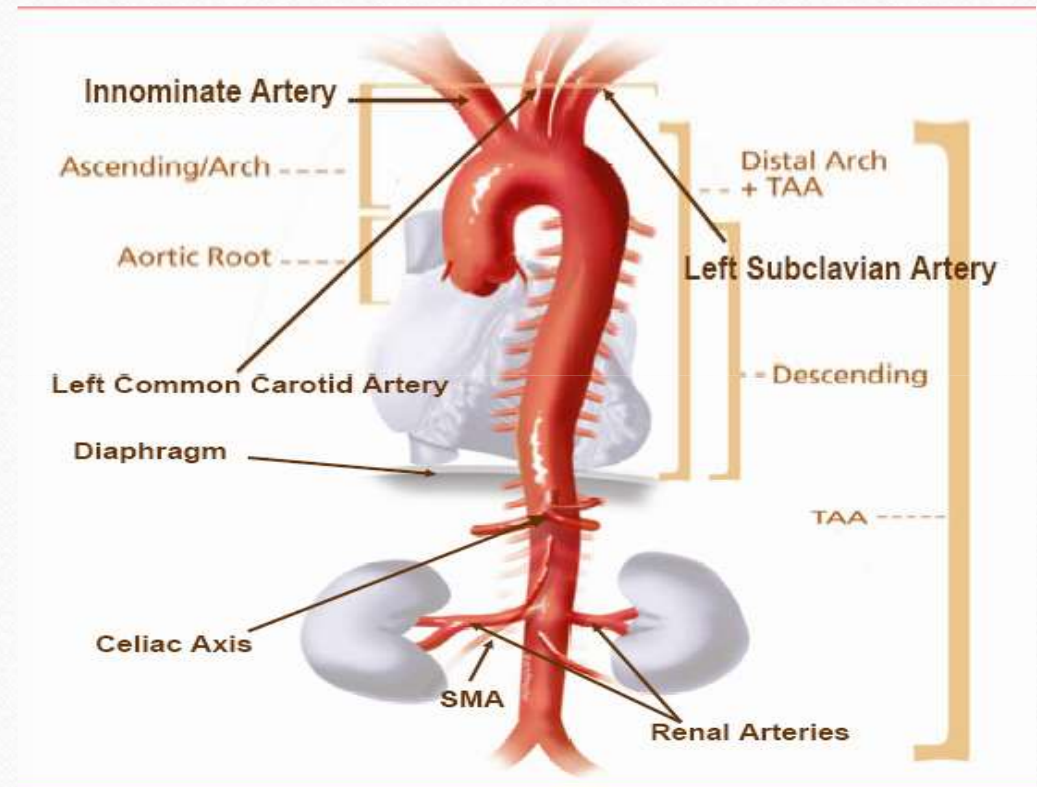
Proximal.

Distal.

- Proximal divided into:

Ascending.

Transverse.



Histology of the AORTA

- **Tunica Intima:** endothelium.

- **Tunica Media:**

Contains lamellae of : Elastin, collagen, smooth muscle cells.

Aorta derives elasticity & tensile strength from T Media.

- **Tunica Adventitia:** collagen, vasa vasorum.

Aortic Dissection

- Most catastrophic event involving the aorta.
- If left untreated: 33% of patients die within the first 24 hr & 50% die within 48 hr.
- **It is either :**

Acute (< 2 weeks)

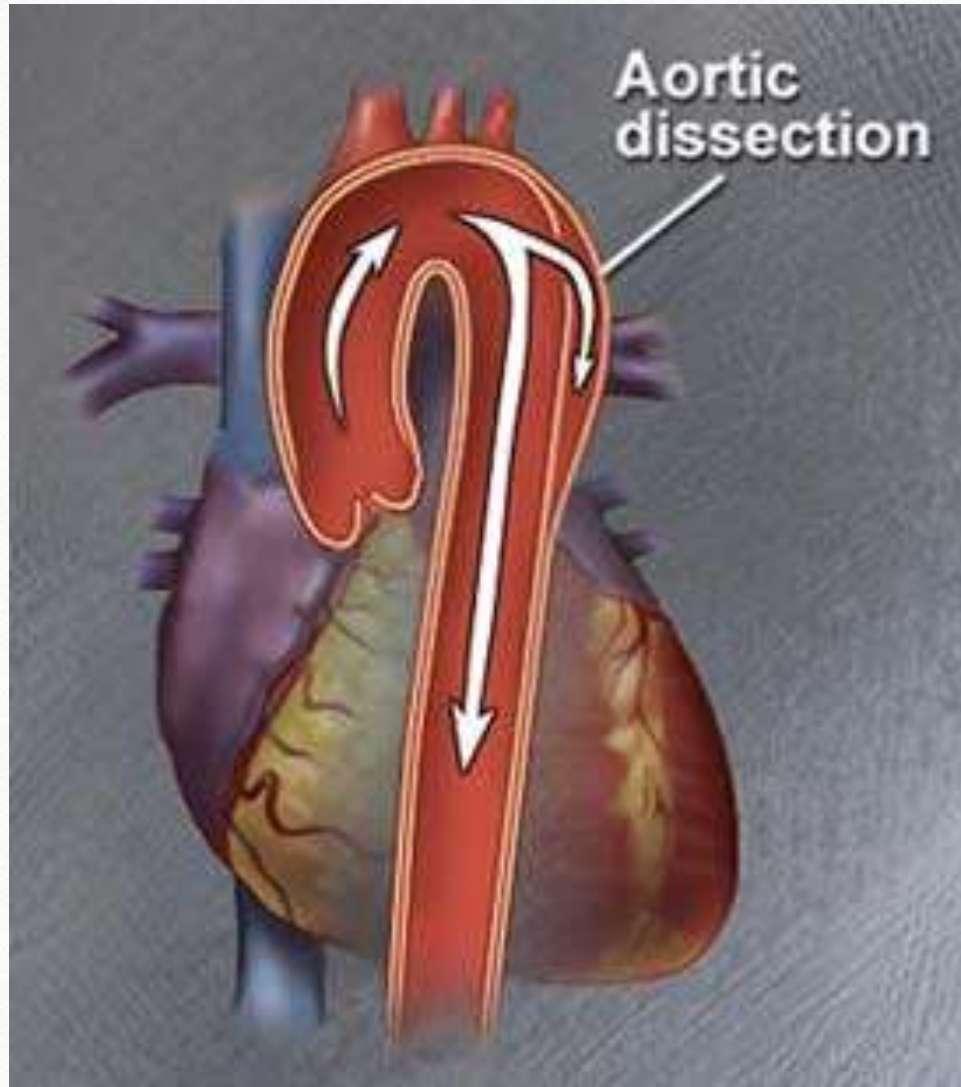
Chronic (> 2 weeks)

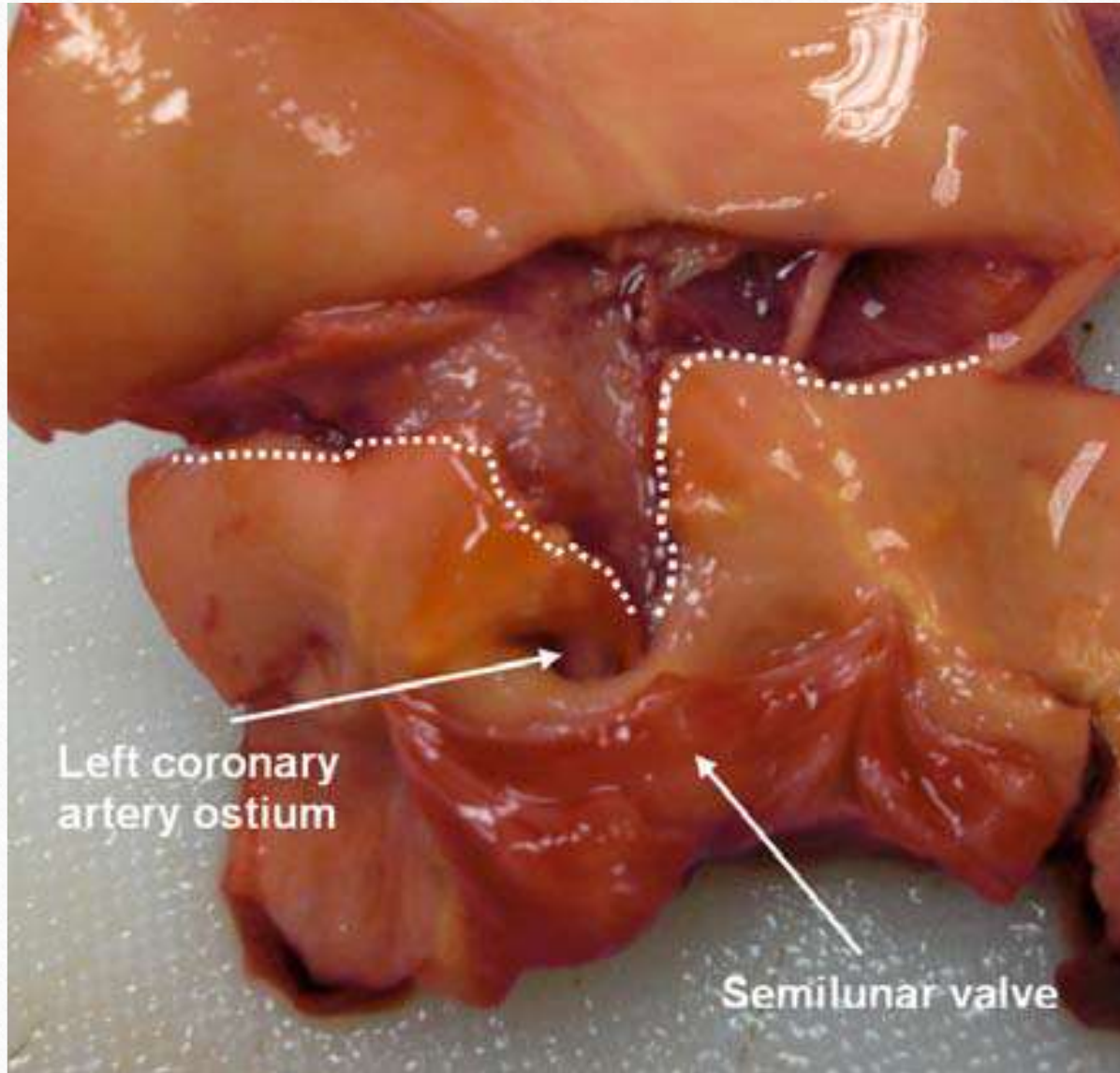
Aortic Dissection

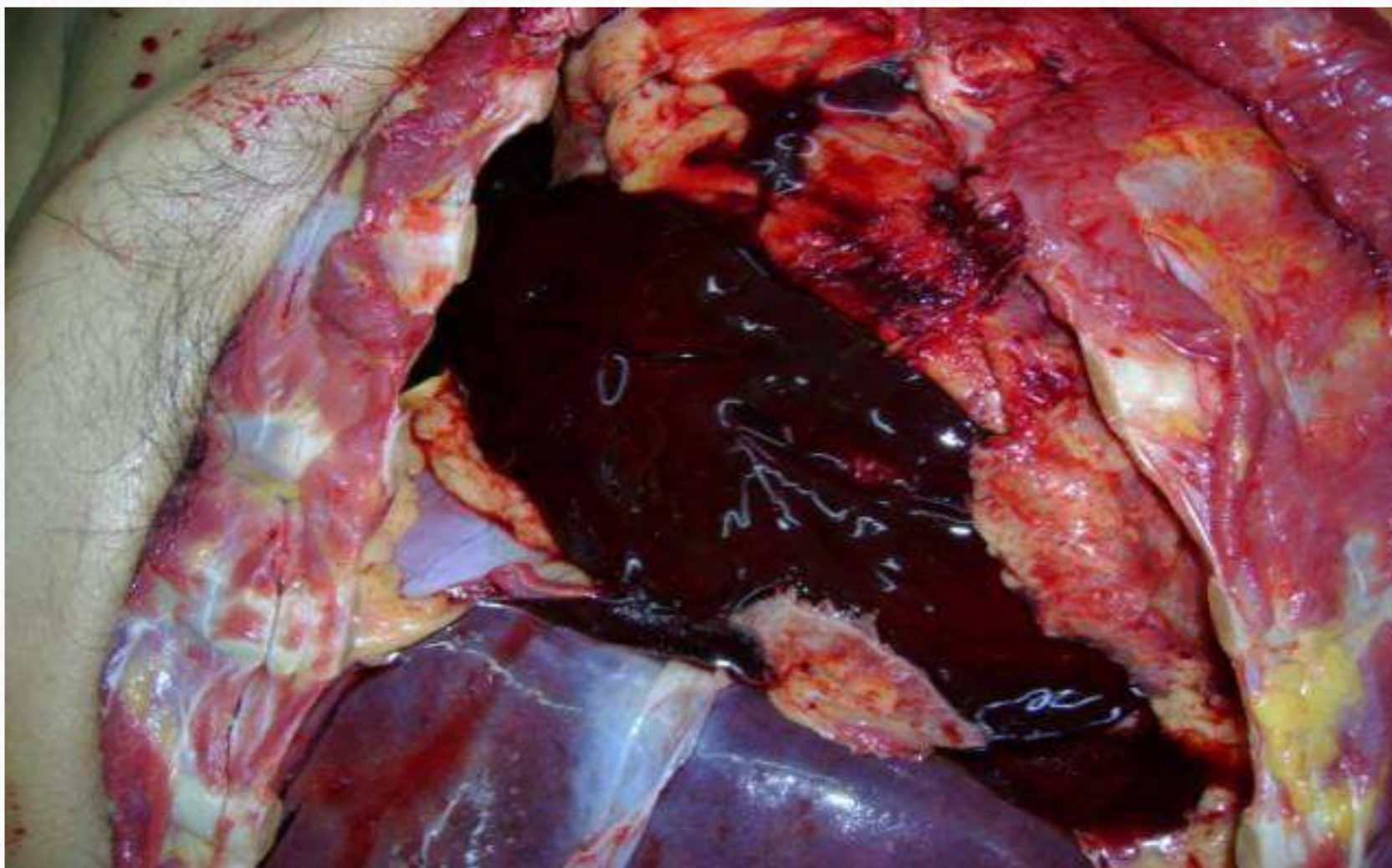
- *Defined as* intimal tear that allows blood to enter the media causing split (dissection) creating a double-barreled aorta.
- Split is frequently horizontal or diagonal.

Aortic Dissection

- *Most common site* of dissection is the 1st few centimeters of the ascending aorta, with 90% occurring within 10 cm of the aortic valve.
- Ascending aortic involvement may *results in* death from wall rupture, hemopericardium and tamponade, occlusion of the coronary ostia with myocardial infarction, or severe aortic insufficiency.







Aortic Dissection

Two aortic channels/lumen are formed:

- **True lumen:**

Smaller, lined by intima.

- **False lumen:**

Within the media, slower blood flow than true lumen.

Often becomes aneurysmal when subjected to systemic hypertension.

Aortic Dissection

- The **first** well-documented case of aortic dissection occurred in **1760**, when *king George II of England* died while straining on the commode.
- In **1761**, the Italian anatomist *Morgagni* provided the 1st detailed pathologic description of aortic dissection.

Aortic Dissection

- In **1955**, Dr. *DeBakey* performed the 1st successful operative aortic **repair** and construction.
- Ironically, Dr. DeBakey himself **developed** aortic dissection at the age 97, and at the age 98 became the **oldest** patient to survive the surgical procedure he pioneered.

Stanford classification of Aortic dissection

- **Type A :**

Acute , in the ascending aorta.

- **Type B :**

Acute / Chronic , in the descending aorta.

DeBakey classification of Aortic dissection

- **Type I :**

Involves ascending aorta, aortic arch & descending aorta.

- **Type II :**

Confined to the ascending aorta.

- **Type III :**

Confined to the descending aorta to the left subclavian artery.

Etiology of aortic dissection

- *Congenital* and *acquired* factors, alone or in combination.
- *More common in* patients with hypertension, connective tissue disorders, congenital aortic valve lesions.

Etiology of aortic dissection

Congenital causes:

- Marfan syndrome.
- Ehlers-Danlos syndrome.
- Bicuspid aortic valve.
- Connective tissue disorders.
- Familial aortic dissections.

Etiology of aortic dissection

Acquired conditions:

- Arterial hypertension: 70% of cases.
- Pregnancy: 50% of cases <40 yr were associated with pregnancy.
- Others: Syphilitic aortitis, Cocaine use.

Epidemiology of aortic dissection

- The true frequency is *difficult* to estimate.
- Most estimates are based on *autopsy* studies.
- It is found in 1-3% of all autopsies in USA.
- Majority in *males*.
- More in *blacks* than in whites.

Signs & symptoms of Aortic Dissection

- **No** one sign and symptom can positively identify Acute Aortic Dissection.
- *Clinical manifestations include:*

Sudden severe chest / jaw pain.

Syncope.

Cerebrovascular accident symptoms.

Dyspnea, hemoptysis, dysphagia.

Aortic Dissection

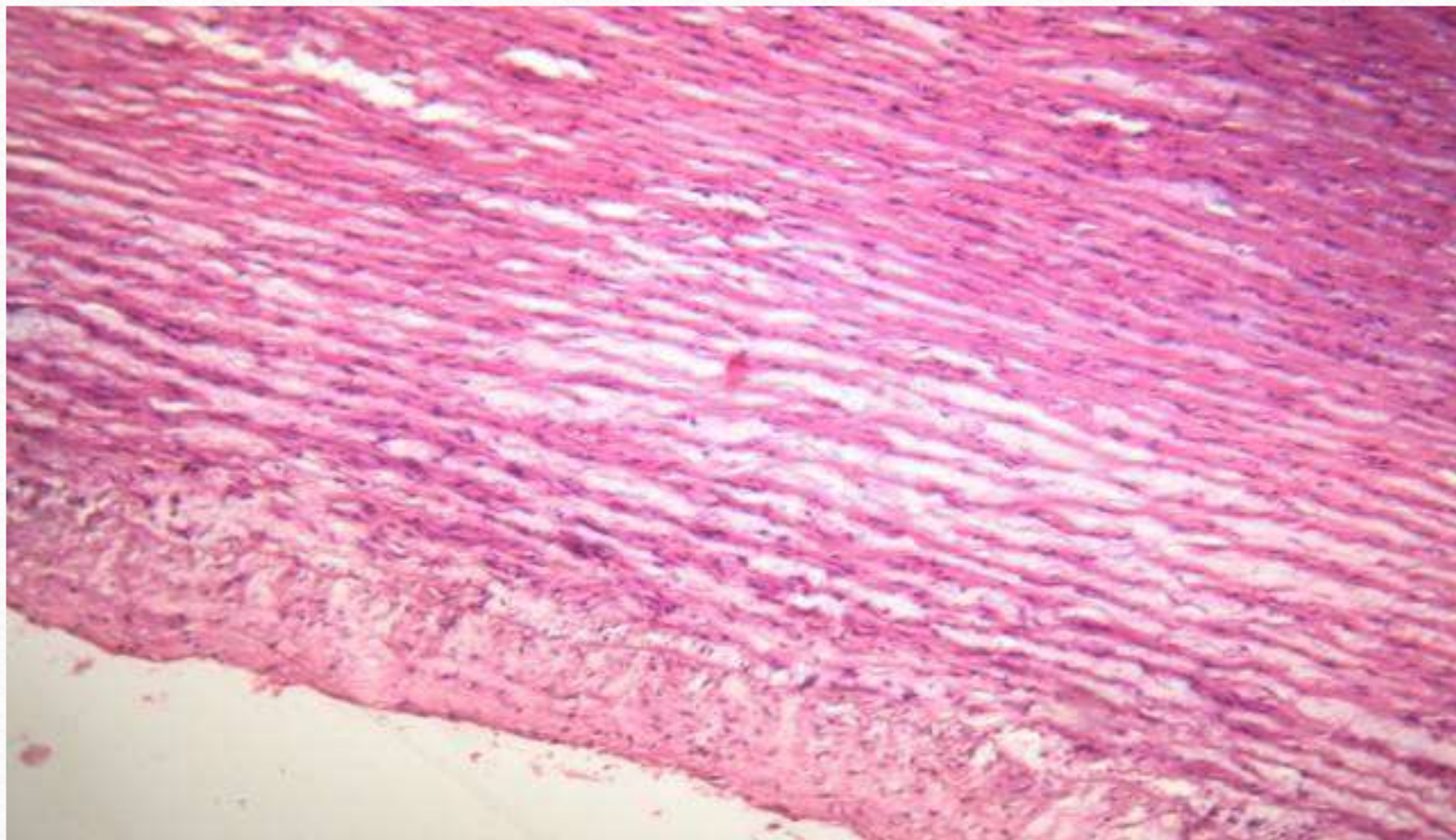
- Modern techniques of diagnosing and repairing aortic dissection transformed the condition from a **death sentence** to a **treatable disorder**.
- However, mortality remains high, as illustrated by the death of actor *John Ritter* and diplomat *Richard Holbrooke*.

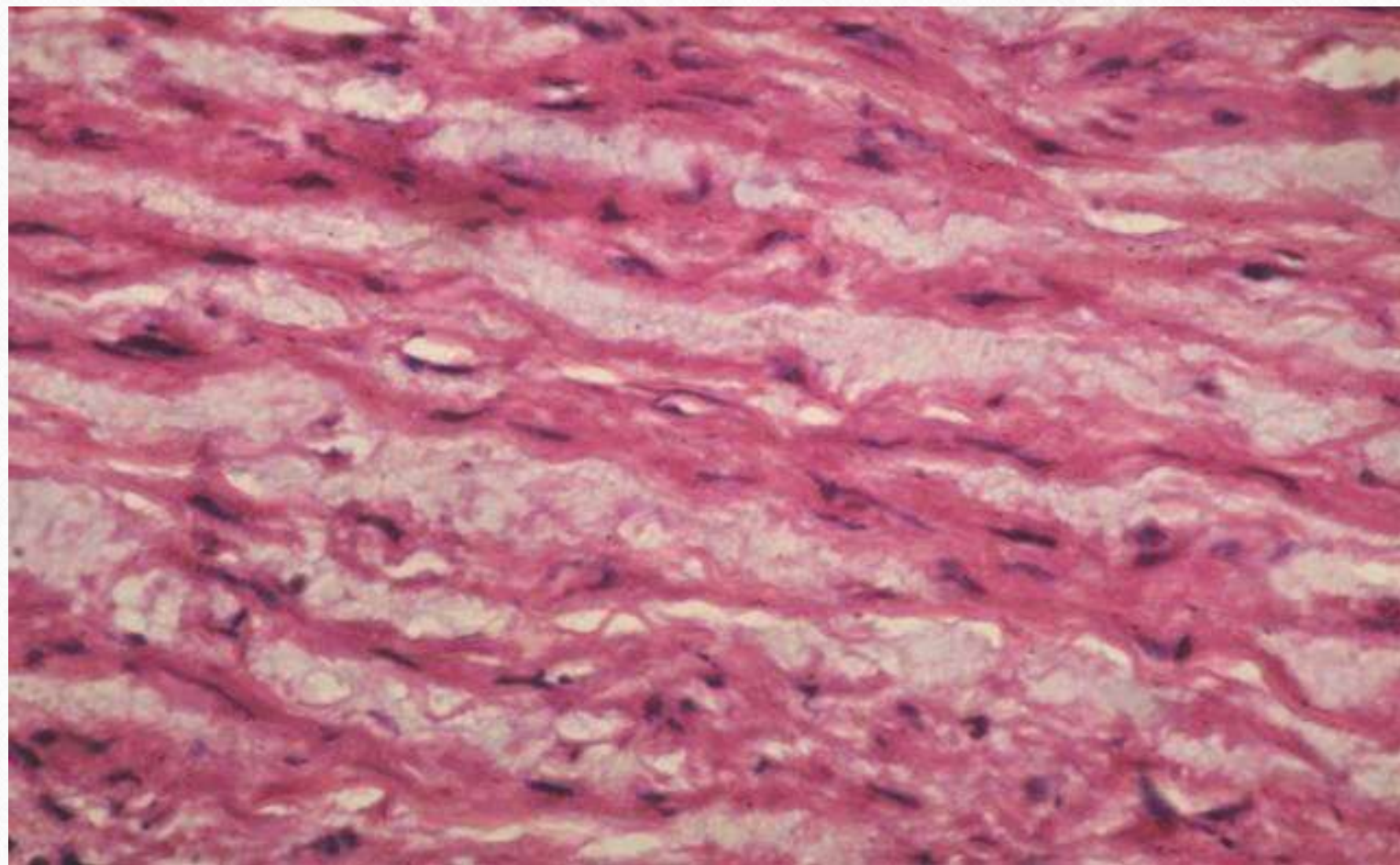
Cystic Medial Degeneration

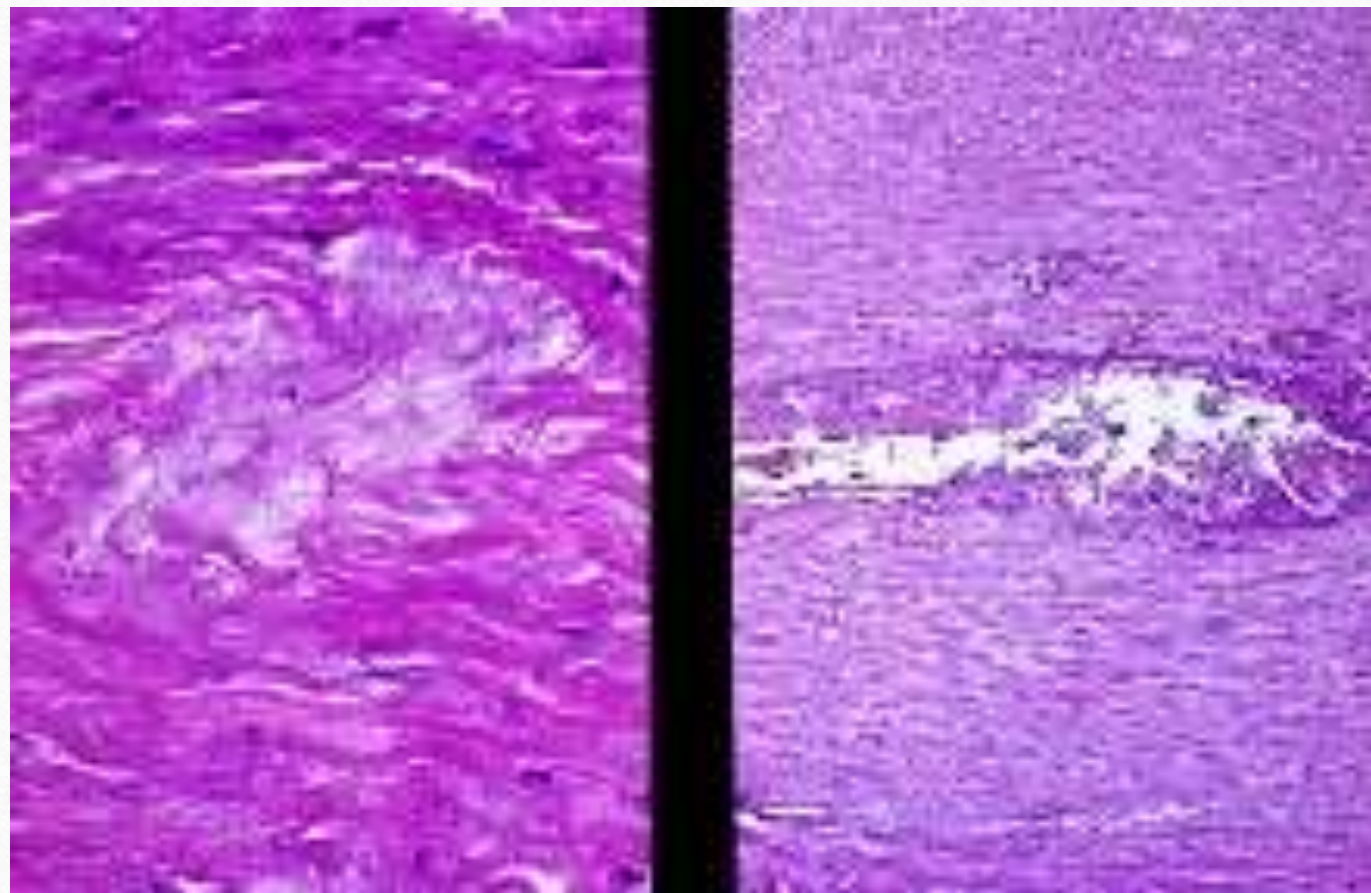
- *With aging*, degenerative changes lead to breakdown of the collagen, elastin, and smooth muscle and an increase in basophilic ground substance.
- It is a *hallmark* histologic change associated with dissection in those with *Marfan syndrome*.

Cystic Medial Degeneration

- It was first described by *Erdheim* in 1929.
- It was **described as** accumulation of basophilic ground substance in the media with formation of cyst-like pools.







Our Autopsy Case

Case history

- This report describes an autopsy case with *sudden death*.
- A *female* in the *3rd* decade *collapsed* at home after a *quarrel* with her husband.
- She arrived to the hospital *dead*.

Autopsy findings

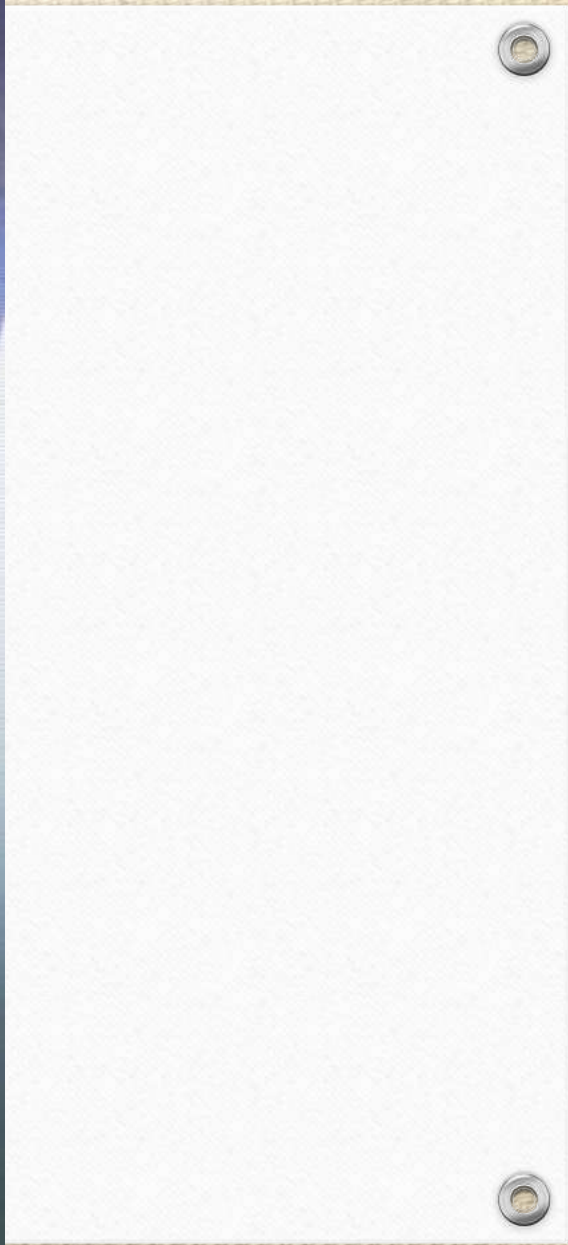
- Autopsy revealed *no* evident external injuries over the body.
- Internally, moderate *hemo-pericardium* was found in the chest cavity.

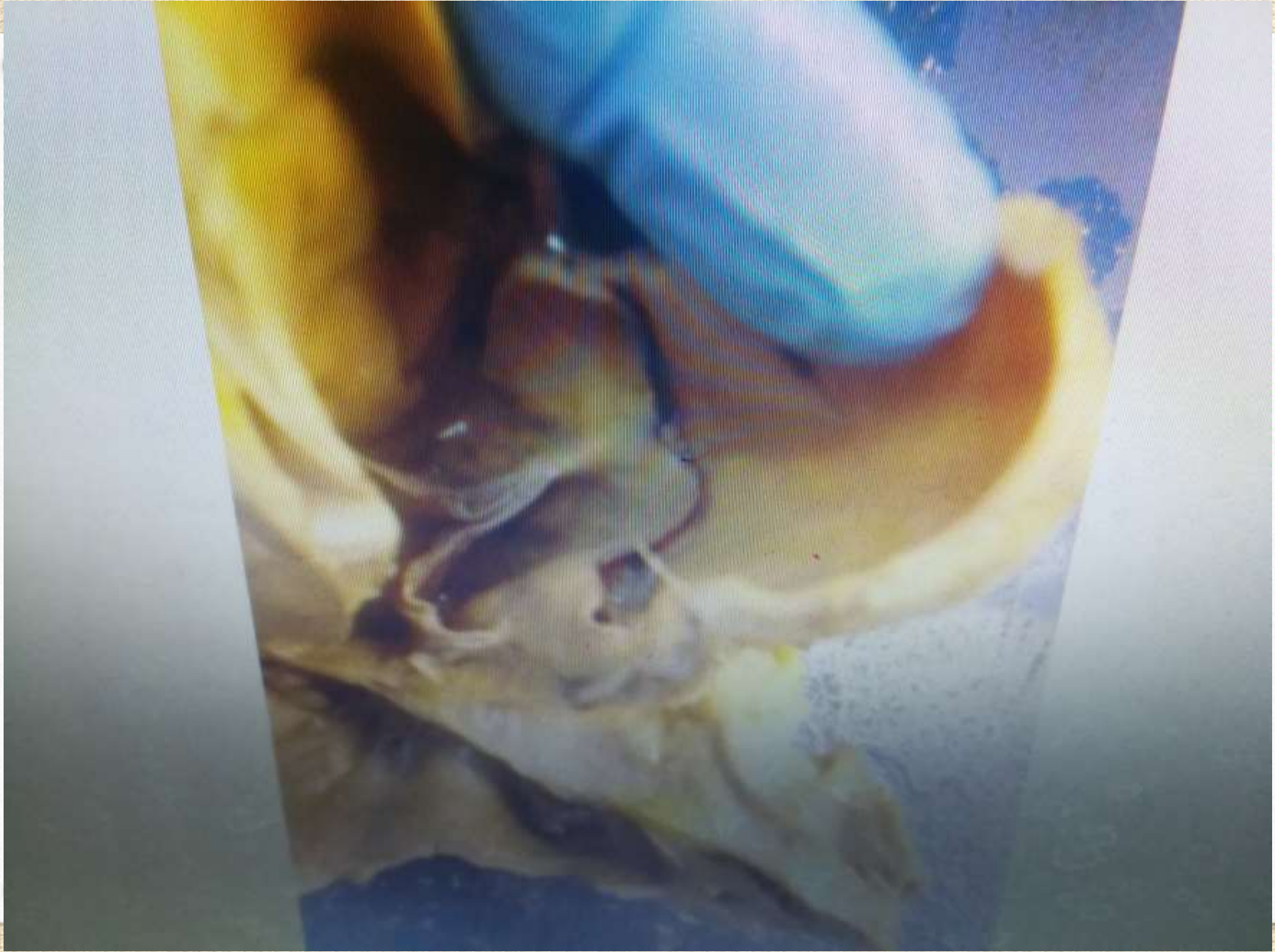
Gross Heart Examination

- There was an external 1 cm *tear* in the ascending aorta above its base by 2 cm. dark area and *aneurysmal dilatation* about 3x2 cm.
- By opening the aorta, a *dissection* in the wall was seen with a 3 cm intimal tear about 1 cm above the aortic valve.





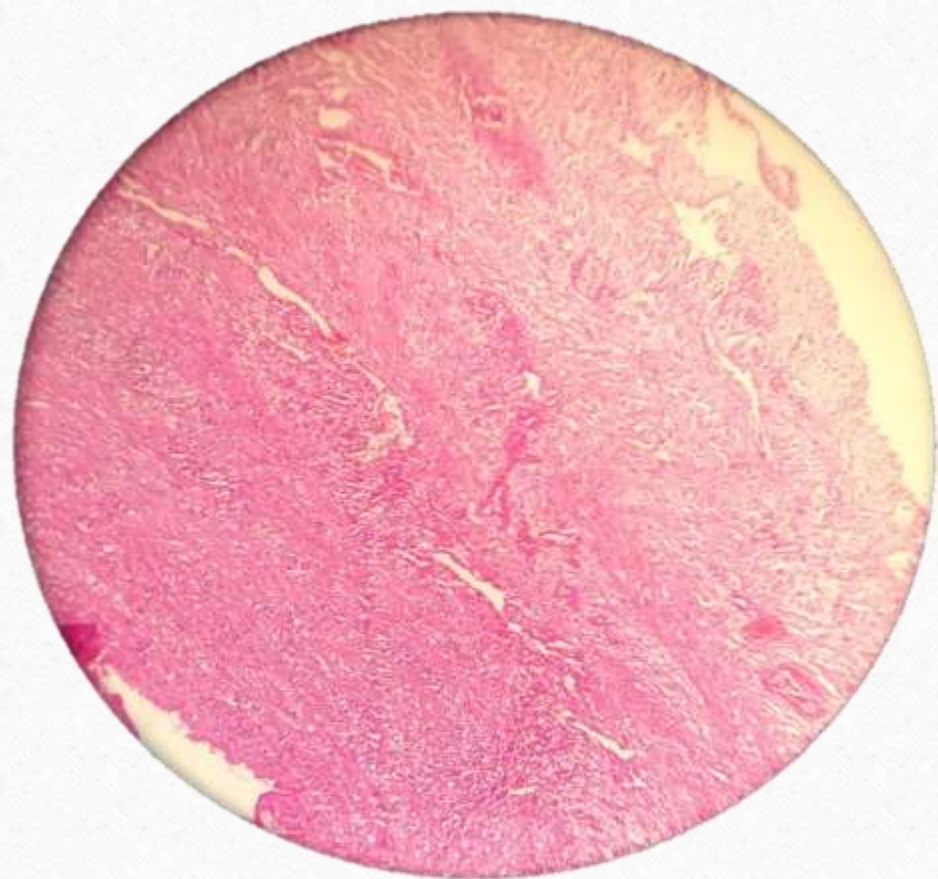




Histopathological examination

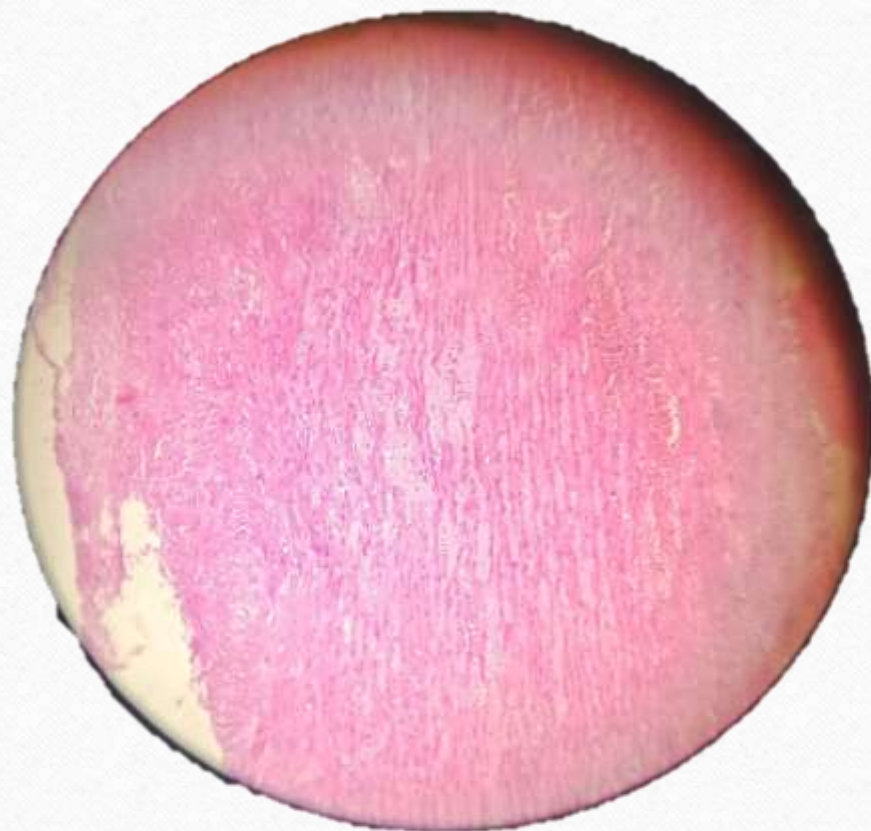
Sections from the wall of the ascending aorta showed:

- *Myxoid* degeneration in the media.
- Elastic tissue *fragmentation*.
- *Separation* of the fibromuscular and elastic elements of the media by numerous ***cystic cleft- like spaces*** containing *basophilic* amorphous extracellular matrix/ground substance.











Histopathological examination

- These findings are consistent with *Cystic Medial Degeneration*.
- In addition to collection of red blood cells in the dissected tunica media in suspected gross aortic dissection.

Cause of Death

- Death was certified as “ due to ***cardiac tamponade*** consequent to rupture of the *dilated dissected aorta* having *cystic medial degeneration*”.

Discussion

Discussion

- In the instant case, there was **no** definite physical characteristics suggesting that the victim was suffering from either Marfan's syndrome or Ehler Danlos syndrome.
- Hence, it appears most likely that the case is one of *idiopathic cystic medial degeneration*.

Discussion

- In the instant case, the ascending aorta showed an aortic aneurysm mostly as a result from the process of cystic medial degeneration.
- A common complication of aortic aneurysm is “*Dissection*”. The blood penetrates into the diseased media through an intimal tear.

Discussion

- Such a dissecting aneurysm frequently *ruptures* spontaneously or following a bout of *hypertension*, resulting in ***Hemopericardium***.
- ***Cardiac tamponade*** is a clinical syndrome caused by the accumulation of fluid in the pericardial space, resulting in reduced ventricular filling and subsequent hemodynamic compromise.
- If *unrelieved*, ***death*** follows.

Take home message

- ***Aortic Cystic Medial Degeneration*** disorder was first thought to be a degenerative process associated with **old** age, but subsequent reports have shown that it occurs not infrequently in **young** people.
- A ***genetic study*** in the instant case would have been helpful in arriving at a diagnosis, to prove or disprove that the victim may have been suffering from a variant of Marfan's syndrome.

Thank you

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