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.
Left coronary artery ostium

Semilunar valve
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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