New nomenclature for non-inflammatory ascending aortic disease
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Talk Outline
- Case Vignette 4
- Short history of aortic nomenclature
- Consensus aortic nomenclature
- Simple descriptors for signing out aortic cases

Clinical History
- She has a bicuspid aortic valve and severe stenosis by echocardiogram.
- 35 years earlier she had an operation for subaortic stenosis.
- Frequent follow-up with her primary care physician.
- At surgery a recurrent subaortic membrane was noted and resected.
- The valve & aorta were replaced with a St. Jude valve and graft.

Case Vignette 4
Aortic Specimen

- A gross specimen measuring 5.8 x 4.2 x 2.0 cm (in aggregate) was received and processed.
- 1 section per cm of tissue (6 in total) were taken in two cassettes.
- A Movat Pentachrome stain was obtained on one slide in addition to H&E stains.
Non-inflammatory Ascending Aortic Disease

Is this Marfan Syndrome?

Or is this?
- Bicuspid Aortic Valve with Aneurysm
- Loeys-Dietz Syndrome
- Turner’s Syndrome
- Vascular Ehlers-Danlos Syndrome
- Familial Thoracic Aortic Aneurysms and Dissection (FTAAD)
- Something Else?

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Final Diagnosis
AORTA (ANEURYSM REPAIR): SEVERE MEDIAL DEGENERATION
NOTE: There is extensive elastic fiber fragmentation and loss (movat) and extensive transmamellar mucoid extracellular matrix accumulation.
NOTE: The patient’s history of Turner Syndrome in noted.

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The beginning of aortic disease nomenclature

Medionecrosis aortae idiopathica cystica.

Virchows Arch Path Anat. 1930

Attempts at Consistency (using current terms)

- Homme JL et al. Surgical Pathology of the ascending aorta: a clinicopathologic study of 513 cases. AJSP 2006

Cystic Medial Degeneration
Cystic Necrosis
Medial Necrosis
Mucoid Degeneration
Medionecrosis

Actual Signouts of Aortic Aneurysm Cases

1. AORTA: ANEURYSM (RESECTION): PORTION OF AORTA.
2. AORTA: EXCISION: PORTION OF AORTA WITH HEMORRHAGE.
3. AORTIC ROOT (REPLACEMENT): SEGMENT OF AORTA WITH SEVERAL HEMORRHAGE.

Medionecrosis aortae idiopathica cystica =

Consensus statement on surgical pathology of the aorta from the Society for Cardiovascular Pathology and the Association for European Cardiovascular Pathology: II. Noninflammatory degenerative diseases—nomenclature and diagnostic criteria

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Mucoid Extracellular Matrix Accumulation (MEMA)

- A new descriptor for cystic medial degeneration
- Two flavors – translamellar and intralamellar
- Three levels of severity – mild, moderate, severe
- Three levels of abundance – focal, multifocal, extensive

MEMA - Translamellar

MEMA - Intralamellar

Elastic Fiber Fragmentation and/or Loss

Extent: Focal/Multifocal/Extensive          Severity: Mild/Moderate/Severe

Additional Descriptors

- Smooth Muscle Cell Nuclei Loss
- Laminar Medial Collapse
- Elastic Fiber Thinning
- Elastic Fiber Disorganization
- Smooth Muscle Disorganization
- Medial Fibrosis

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Using the new AECVP/SCVP nomenclature in routine signout

Easy Signout

Aorta

No Inflammation

Extent of Medial Degeneration

Inflammatory

Determine type of inflammation (contact clinician)

Minimal Top Line Diagnosis

• Aorta (Resection): Mild/Moderate/Severe Medial Degeneration
More Involved Diagnosis

Aorta (resection): MILD MEDIAL DEGENERATION
NOTE: There is patchy rare smooth muscle cell nuclei loss. There is no obvious mucoid extracellular matrix accumulation (MEMA), elastic fiber fragmentation and/or loss, or laminar medial collapse (evaluated with Movat's pentachrome stain).

Another Involved Diagnosis

Aorta (resection): SEVERE MEDIAL DEGENERATION.
NOTE: This aorta demonstrates extensive band-like smooth muscle cell nuclei loss and a thin extensive laminar medial collapse (Movat). There is moderate extensive elastic fiber fragmentation and/or loss. There is moderate multifocal intraluminal mucoid extracellular matrix accumulation in the media.

Summation

• Aortic nomenclature dates back to 1930
• Nomenclature revisions in 2016 have given us new and revised terms such as "mucoid extracellular matrix accumulation (MEMA)," "elastic fiber fragmentation and/or loss," "smooth muscle cell nuclei loss" and "laminar medial collapse"  
• Non-inflammatory aorta specimens can be effectively signed out as mild, moderate, or severe medial degeneration on the top line with additional descriptors as one sees fit.

Resource


Figures, Supplemental Figures, Supplemental Routine Pathology Report, Representative Surgical Pathology Reports

Acknowledgements

• AECVP and SCVP Consensus Committee on Aortic Disease

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