31st Annual State of the Art Echocardiography | San Diego, CA

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Diseases of Aorta: Marfan, Dissection, Atheroma

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Disclosures

Speakers Bureau (Philips, Medtronic)
Advisory Board (Siemens)

ARNOLD SCHWARZENEGGER SAYS: ‘IT’S NOT A TUMOR!’

KID: ‘What’s the matter?’
ARNOLD: ‘I have a headache’
KID: ‘It might be a tumor.’
ARNOLD: ‘It’s not a tumor. Not a tumor. At all!’
**CASE PRESENTATION**

80-year-old man presents with severe headache and nonexertional chest pain in the setting of severe hypertension (BP 210/90 mm Hg)

<table>
<thead>
<tr>
<th>PAST MEDICAL HISTORY</th>
<th>WORKING DIAGNOSES</th>
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<tbody>
<tr>
<td>Hypertension</td>
<td>Hypertensive urgency</td>
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<tr>
<td>Coronary artery disease (s/p CABG &amp; PCIs)</td>
<td>Acute coronary syndrome</td>
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<td>Abdominal aortic aneurysm</td>
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<td>Bradycardia (s/p Permanent pacemaker placement)</td>
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*Transthoracic echocardiogram was ordered...*

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**TRANSTHORACIC ECHOCARDIOGRAM**

Hypertensive heart disease with paced rhythm. Otherwise, no wall motion abnormalities; no valvular disease.
CLINICAL COURSE

Headache resolved after normalization of blood pressure
He was ruled out for acute coronary syndrome

YET...

Severe nonexertional chest pain continued...

MAYBE IT’S AORTIC DISSECTION

Let’s order a chest CT...
**CHEST CT**

The good news is there is no aortic dissection...

...but, there is something very concerning on **noncontrast** portion of CT.

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**NONCONTRAST CHEST CT**

*Crescentic hyperdense lesion* involving distal arch & descending aorta
CT Diagnosis

Acute intramural hematoma (IMH) of distal aortic arch and descending thoracic aorta

First described on autopsy in 1760 of king George II of England

Classic Aortic Dissection

Intramural Hematoma (IMH)

First described in 1920 in Germany on autopsy.

Penetrating Atherosclerotic Ulcer (PAU)

First described in 1986 on aortography & CT at Mayo Clinic.
INTRAMURAL HEMATOMA | FIRST DESCRIPTION

Krukenberg E. Beiträge zur Frage des Aneurysma dissecans
[Contributions to the question of dissecting aneurysm].
Beitr Pathol Anat Allg Pathol. 1920;67:329-351.

Krukenberg correctly deduced that IMH results from rupture of vasa vasorum.

INALMAL HEMATOMA | FIRST DESCRIPTION

ERNST KRUKENBERG
(1871-1946)
German pathologist

Although he was first to describe intramural hematoma (IMH) of the aorta....

...he is actually better known for describing ‘Krukenberg tumors’ – transperitoneal ovarian metastases from stomach and colon cancers.
Back to our patient

Six weeks later, severe chest pain recurred...
...and repeat chest CT was ordered

Chest CT #2 | 6 weeks after initial CT

Increase in the thickness of IMH with appearance of contrast inside IMH.
**CHEST CT #2 | 6 Weeks After Initial CT**

Increase in the thickness of IMH with appearance of contrast inside IMH.

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**CHEST CT #3**

Done 1 day after chest CT #2
Conversion of IMH into aortic dissection

Chest CT #3 | One Day After CT #2

Conversion of IMH into aortic dissection

Chest CT #3 | One Day After CT #2
CONVERSION OF IMH INTO AORTIC DISSECTION

TEACHING POINTS

- Intramural hematoma (IMH) was first described by Ernst Krukenberg of Germany in 1920
- IMH is likely the result of vasa vasorum rupture in the aortic media
- IMH is visualized as crescentic thickening of the aortic wall
- IMH may be a precursor to aortic dissection
INTRAMURAL HEMATOMA | ONE OF CAUSES OF ACUTE AORTIC SYNDROME

The term **ACUTE AORTIC SYNDROME** was first proposed in **2001** by Vilacosta & San Román of Spain

**Editorial**

*Heart* 2001;85:365–368

**Acute aortic syndrome**

Although the chest pain of acute aortic dissection is widely recognised, less consideration has been given to pain associated with other aortic pathologies. In light of contemporary concepts in aortic pathology we would like to present the pathology of a new cardiovascular syndrome—acute aortic syndrome (AAS). 1

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ACUTE AORTIC SYNDROME | INTRAMURAL HEMATOMA

Crescentic thickening of the aortic arch wall
**ACUTE AORTIC SYNDROME | PENETRATING ATHEROSCLEROTIC ULCER (PAU)**

PAU = Erosion of atherosclerotic plaque into the media

Note the intramural hematoma underneath the PAU

**ACUTE AORTIC SYNDROME | PAU LEADING TO PSEUDOANEURYSM**

PAU >> Pseudoaneurysm (Focal aortic rupture)
ACUTE AORTIC SYNDROME ON TEE | CLASSIC AORTIC DISSECTION

Noncontrast TEE

TEE with Microbubble Contrast

Michael Ellis DeBakey (originally: دباغي: Dabaghi) 1908-2008
Lebanese-American surgeon


Stanford Type A & B classification is now used for IMH, PAU & dissection.
Chapter 159: Acute aortic syndrome
Pages 671-679

159 Acute Aortic Syndrome
Muhamed Saric, MD, PhD
Itzhak Kronzon, MD

Acute aortic syndrome (AAS) encompasses several life-threatening clinical entities with overlapping features including acute onset of chest pain, disruption of the aortic wall media, and a need for urgent medical care (Fig. 159.1). The term “acute aortic syndrome” was first proposed in 2001 by the Spanish physicians Vilacosta and San Roman. The following three entities were originally included in the spectrum of acute aortic syndromes: aortic dissection, intramural hematoma (IMH), and penetrating atherosclerotic ulcer (PAU). Traumatic aortic rupture (TAR; transection) due to blunt deceleration trauma as well as aortic aneurysm leak and rupture may also be included in the spectrum of AAS.

Thank You!

New York University Langone Medical Center