Archives of the AIRP: Intimal Sarcomas of the Great Vessels

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Disclosures

The authors have nothing to disclose...
Introduction

Aortic and pulmonary artery (PA) intimal sarcomas are rare diagnoses of middle age often missed at initial presentation: this delay in diagnosis impacts outcome.

Clinical symptomatology is frequently related to embolic complications of stroke, pulmonary embolism, end organ or limb ischemia (including claudication and absent pulses).

Metastases and invasion may occur in lung, pleura, pericardium, chest wall, liver, kidney, adrenal glands, brain, and bone.

Treatment is primarily surgical with poor survival rates of 28% at 5 years, 14% at 10 years. Adjuvant radiation and chemotherapy are areas of current investigation.
Spectrum of Initial Imaging Manifestations

- Ventilation/Perfusion Defects
- Mesenteric Ischemia
- Cavitary Lesion
- Pulmonary Embolism
- Distal Embolic Phenomena
- Mediastinal Mass +/- Effusion
Diagnostic Dilemma: Rarity & Low Suspicion

Objective: Learn when to suspect Great Vessel Sarcomas

Common Situation

- Initial Presentation
- Diagnostic Imaging
  - Misdiagnosed as Thrombus
  - months later
- Metastatic Disease
- Disability
- Palliative Treatment
- Death

With Improved Understanding:

- Identify Suspicious Features
- Early Diagnosis
- Treatment Options
- Decreased Disability
- Increased Survival
Gross Appearance of Intimal Sarcoma (Aorta & PA)

Often resembles thrombus or gelatinous tumor mass (some are initially misdiagnosed as embolectomy specimens)

Entirely or predominantly intraluminal, with focal invasion (consistent with origin in the arterial intima, rather than arterial wall)

Lobulated contours correspond to radiologic features

Aortic intimal sarcoma (arrows) shows preferential intraluminal growth

Pulmonary artery sarcoma (arrows) often fills the vessel lumen
Aorta: atypical neoplastic cells on microscopic exam differentiate the lesion from non-malignant, bland emboli

Aorta: frequently the tumor grows as a layer of neoplastic cells overlying a core of thrombus/necrosis (less commonly observed in pulmonary artery sarcomas)

Pulmonary Artery: intimal sarcomas often appear more cellular, with nearly total occlusion of vessel lumen (heterogeneous mixture of tumor & thrombus evident throughout the lesion)
Two most common aortic intimal subtypes: epithelioid angiosarcoma and undifferentiated angiosarcoma.

Differentiation between subtypes frequently requires immunohistochemistry.

Pulmonary artery intimal sarcomas do not appear to have a predominant subtype.

Compared to aorta, pulmonary artery intimal sarcomas more often contain heterologous elements: osteosarcoma and chondrosarcoma.

Histology of Intimal Sarcoma (Aorta vs. PA)

Low power view shows tumor cells coating thrombus on intimal surface of aorta.

High power view shows marked cellular atypia, plentiful mitoses, myxoid background.
Great Vessel Intimal Sarcoma: Review Methodology

82 Literature Articles Reviewed

Of these, 14 articles describe suspicious imaging features in 57 Cases: 8 Aortic & 49 PA Intimal Sarcomas

AIRP Archives & University of Maryland School of Medicine provide

24 Cases: 7 Aortic & 17 PA Intimal Sarcomas

Total Analyzed: 15 Aortic & 66 PA Intimal Sarcomas
Suspicious Features: Reporting Method

- Imaging features reviewed: combined AIRP, UMMC, and Literature Cases
- Feature occurrence quantified: combined total from available material
- Some features were not described for all cases reviewed
- Intimal sarcoma features quantified as arising in **PA, Aorta, or Both**

<table>
<thead>
<tr>
<th>Feature</th>
<th>Cases</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Name of suspicious feature</td>
<td>Ratio of total cases demonstrating feature</td>
<td>Percent of total cases demonstrating feature</td>
</tr>
</tbody>
</table>

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Exemplary Images and annotations
# PA Intimal Sarcoma

## Feature #1, *Characteristic*

<table>
<thead>
<tr>
<th>Lobulated Margin</th>
<th>24/33</th>
<th>73%</th>
</tr>
</thead>
</table>

[CTA images of PA Intimal Sarcoma]
# PA Intimal Sarcoma

## Feature #2, *Characteristic*

<table>
<thead>
<tr>
<th>Wall Eclipsing Sign</th>
<th>32/66</th>
<th>62%</th>
</tr>
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</table>

Lumen obliterated on both sides (“eclipsed”)

Appearance on axial imaging

![MRA T1 Black Blood](CTA.png)

![CTA](CTA.png)
## PA Intimal Sarcoma

### Feature #3, Characteristic

<table>
<thead>
<tr>
<th>Contrast Enhancement</th>
<th>8/13</th>
<th>62%</th>
</tr>
</thead>
</table>

**CTA**
- Heterogeneous Attenuation

**Fine Linear**

**VIBE post gad**
- More conspicuous on MRI

**VIBE post gad**
## PA Intimal Sarcoma

### Feature #4, *Characteristic*

<table>
<thead>
<tr>
<th>Unilateral Mass</th>
<th>22/66</th>
<th>33%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right PA Mass</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left PA Mass</td>
<td></td>
<td></td>
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<tr>
<td>CTA</td>
<td></td>
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<tr>
<td>CT-post</td>
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<tr>
<td>CT-post</td>
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<tr>
<td>Non-contrast</td>
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<tr>
<td>MRA T1 Black Blood</td>
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</tbody>
</table>
PA Intimal Sarcoma

Late Feature, *Characteristic*

<table>
<thead>
<tr>
<th>Interval Enlargement</th>
<th>30/48</th>
<th>63%</th>
</tr>
</thead>
</table>

Prior

Follow up

Prior CTA

Follow up CT

HIGHLY Suspicious, however our goal is to diagnose earlier
# PA Intimal Sarcoma

**Late Feature, **Characteristic**

<table>
<thead>
<tr>
<th>Metastasis or Invasion</th>
<th>10/32</th>
<th>31%</th>
</tr>
</thead>
</table>

- Vena cava
- Paracardiac lymph node
- Tumor thrombus
- Pericardium
- T1 post
- T1 post subtraction

**HIGHLY Suspicious,** however due to earlier onset of symptoms, most are diagnosed earlier.
## Aortic Intimal Sarcoma

**Feature #1, Characteristic**

<table>
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<tr>
<th>Lobulated Margin</th>
<th>13/15</th>
<th>87%</th>
</tr>
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</table>

- **CTA**

![CTA Image 1](image1.jpg)

![CTA Image 2](image2.jpg)

![CTA Image 3](image3.jpg)

![CTA Image 4](image4.jpg)
Aortic Intimal Sarcoma

Feature #2, *Our Observation*

<table>
<thead>
<tr>
<th>Atherosclerosis</th>
<th>13/15</th>
<th>87%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absent to Minimal</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

CTA CTA CTA CTA
## Aortic Intimal Sarcoma

### Feature #3, *Characteristic*

<table>
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<tr>
<th>Thoracic Location</th>
<th>12/15</th>
<th>80%</th>
</tr>
</thead>
</table>

Most sarcomas occur in the thoracic aorta, whereas most benign intimal thrombus occurs in the abdominal aorta.
### Aortic Intimal Sarcoma

#### Feature #4, Characteristic

<table>
<thead>
<tr>
<th>Contrast Enhancement</th>
<th>1/8</th>
<th>13%</th>
</tr>
</thead>
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Enhancement is unusual in aortic intimal sarcomas, which are largely comprised of central necrosis/thrombus coated with a superficial layer of tumor. (Enhancement appears more common in PA sarcomas, perhaps due to the more heterogeneous composition of tumor and thrombus.)

Contrast enhancement = HIGHLY SUSPICIOUS for malignancy (on CT and MRI)
Aortic Intimal Sarcoma

Late Feature, *Characteristic*

<table>
<thead>
<tr>
<th>Metastasis or Embolism</th>
<th>7/8</th>
<th>97%</th>
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</table>

HIGHLY suspicious, however our goal is to diagnose earlier.

Left Adrenal

Right Adrenal

Left Temporal Lobe
Both PA & Aortic Intimal Sarcoma

Common Feature, *Shared Characteristic*

<table>
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<tr>
<th>Lobulated Margin</th>
<th>37/48</th>
<th>77%</th>
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<table>
<thead>
<tr>
<th>PA Intimal Sarcoma</th>
<th>Aortic Intimal Sarcoma</th>
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<tbody>
<tr>
<td><img src="image1.png" alt="PA Intimal Sarcoma" /></td>
<td><img src="image2.png" alt="Aortic Intimal Sarcoma" /></td>
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CTA
Both PA & Aortic Intimal Sarcoma

Common Feature, *Our Observation*

<table>
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<tr>
<th>Eccentric Endoluminal Mass</th>
<th>27/32</th>
<th>84%</th>
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</table>

- **PA Intimal Sarcoma**
  - CTA
  - VIBE post gad

- **Aortic Intimal Sarcoma**
  - CTA
  - VIBE post gad

Both PA & Aortic Intimal Sarcoma

Most Diagnostic Feature

| 18F-FDG PET Avidity | 8/8 | 100% |

![Scout](image1)
![Non-contrast CT](image2)
![FDG PET/CT Fused](image3)
Intimal Sarcoma on Chest Radiography

PA (conspicuous)

Aortic (inconspicuous)
Intimal Sarcoma: Rad/Path Correlation

Pulmonary Artery Sarcoma (Right)
Intimal Sarcoma: Rad/Path Correlation

Pulmonary Artery Sarcoma (Left)
Intimal Sarcoma: Rad/Path Correlation

Aortic Intimal Sarcoma
Imaging Findings That Suggest Intimal Sarcoma

- Eccentric endoluminal filling defect (often low attenuation on CT: easily interpreted as thrombus)
- Lobulated margins
- Contrast enhancement
- Complete vessel occlusion (more typical of PA sarcoma)
Imaging Findings That Suggest Intimal Sarcoma

- Pulmonary embolism without risk factors and absence of deep vein thrombus on ultrasound Doppler (PA sarcoma)
- Central or peripheral embolism without risk factors for thromboembolic disease (Aortic intimal sarcoma)
- Absence of significant atherosclerotic disease (Aortic intimal sarcoma)
- Metastases
Imaging Findings That Suggest Intimal Sarcoma

- 18F-FDG PET avidity*

(*strongly recommended when suspicion for intimal sarcoma is high, based upon clinical and/or radiologic findings)
Proposed Diagnostic Algorithm

Identify suspicious features on CT or MRI which suggest great vessel Intimal sarcoma, then recommend 18F-FDG PET/CT.

18F-FDG PET/CT

- Early Diagnosis
- Treatment Options
- Decreased Disability
- Increased Survival

CTA or MRA

Identify Suspicious Features

- Misdiagnosed as Thrombus

Months later

Metastatic Disease

Disability

Palliative Treatment

Death

Initial Presentation
### Suspicious Features Quantification Summary

#### Intimal Sarcoma Features by Total Combined Occurrence: Literature, AIRP, & UMMC

<table>
<thead>
<tr>
<th>Feature</th>
<th>Pulmonary Artery</th>
<th>Aorta</th>
<th>Both</th>
<th>Late Feature</th>
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<tr>
<td>Lobulated Margin</td>
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**Pulmonary Artery**
- Lobulated Margin: 24/33 (73%)

**Aorta**
- Lobulated Margin: 13/15 (87%)

**Both**
- 18F-FDG PET Avidity: 8/8 (100%)

**Late Feature**
- Interval Enlargement: 30/48 (63%)

**Thoracic Location**
- 12/15 (80%)

Most sarcomas occur in the thoracic aorta, whereas most benign intimal thrombus occurs in the abdominal aorta.

**Contrast Enhancement**
- 8/13 (62%)
- 1/8 (13%)

Enhancement is common in aortic intimal sarcomas, which are largely comprised of central necrotic/intimal thrombus covered with a superficial layer of tumor. Enhancement appears more common in PA sarcomas, perhaps due to the more heterogeneous composition of tumor and thrombus.

**Unilateral Mass**
- 22/66 (33%)

**Gross Pathology**
- PA Sarcoma
- Aortic Sarcoma
References


Thank you for learning with us!

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