Diagnosis of Fibromuscular Dysplasia

Chrislyn White, Harvard Medical School Year III
Gillian Lieberman, MD
Agenda

Our Patient:
• Clinical Presentation
• Initial Images
• Classic Appearance

Fibromuscular Dysplasia (FMD):
• Distribution
• Classification

Diagnosis of FMD:
• Algorithm
• Menu of Tests
• Our Patient
• Differential Diagnosis

Summary
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Summary
Our Patient

• 46-year-old woman with a history of migraines presents with headaches, enlarged blind spot, and new-onset hypertension
• Past Medical History: migraines
• Medications: oral contraceptives, Excedrin migraine prn
Our Patient: Clinical Presentation

Headaches:
Change in frequency and severity with unpredictable response to treatment

Blood Pressure:
BP was 210/110. She has no history of hypertension and BP is usually 100/70

Visual Symptoms:
In left eye, centrally-located missing piece of vision when right eye was closed
Our Patient: Dissection on CT

Dissection of Left Internal Carotid Artery

PACS, BIDMC
Head & Neck CT (+C) Axial

PACS, BIDMC
Head & Neck CT (+C) Coronal
Our Patient: Arterial Stenosis and Aneurysm on CT

PACS, BIDMC
Head & Neck CT (+C) Reconstruction
Classic Appearance of FMD

“String of Beads”
(Slovut and Olin, 2004)

Image source:
http://www.ehow.com/how_8192233.tie-between-beads.html
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Summary
**Fibromuscular Dysplasia**

- Nonatherosclerotic, noninflammatory vascular disease
- <2% of all hypertension cases
- Unknown etiology
- **Risk Factors**
  - Cigarette smoking
  - Hypertension
  - Family history
- **Women aged 15 to 50**

(Mounier-Vehier et al., 2002)

(Slovut and Olin, 2004)
Fibromuscular Dysplasia: Distribution of Disease

**Table 1. Arterial Involvement in Fibromuscular Dysplasia.**

<table>
<thead>
<tr>
<th>Arteries Involved</th>
<th>Frequency of Involvement (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Renal arteries</td>
<td>60–75</td>
</tr>
<tr>
<td>Bilateral</td>
<td>35</td>
</tr>
<tr>
<td>Extracranial cerebrovascular circulation (carotid or vertebral arteries)</td>
<td>25–30</td>
</tr>
<tr>
<td>Associated intracranial aneurysm</td>
<td>7–50</td>
</tr>
<tr>
<td>Multiple vascular beds</td>
<td></td>
</tr>
<tr>
<td>Other arterial beds (iliac, popliteal, splanchnic, hepatic, coronary, subclavian,</td>
<td>Uncommon, exact frequency</td>
</tr>
<tr>
<td>brachial, aorta, superficial femoral, tibial, or peroneal)</td>
<td>unknown</td>
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* Fibromuscular dysplasia may be a generalized process; in rare cases, it has also been identified in the venous system.

Fibromuscular Dysplasia: Vessel Involvement

Fibromuscular dysplasia (FMD) mainly affects arteries (rarely veins).

It is classified based on which arterial layer is affected.

(Slovut and Olin, 2004)

Image source:
http://www.as.miami.edu/chemistry/2086/Chapter_21/NEW-Chap21_class_part1.htm
1. Medial dysplasia
   1) Medial fibroplasia (75-80%)
   2) Perimedial fibroplasia (10-15%)
   3) Medial hyperplasia (1-2%)

2. Intimal fibroplasia (<10%)

3. Adventitial (periarterial) fibroplasia (<1%)
Fibromuscular Dysplasia:

1. Medial Dysplasia

1) Medial fibroplasia: “string of beads” appearance
   - Alternating thinned and thickened areas of the media containing collagen
     “Beads” are larger diameter than normal arterial diameter

(Slovut and Olin, 2004)


Image source:
http://www.ehow.com/how_8192233_tie-between-beads.html
2) Perimedial fibroplasia

“Beads” are **smaller** diameter than normal arterial diameter

(Slovut and Olin, 2004)

Marked deposition of collagen in outer half of media

(Nickeleit et al., 1999)


Image source:
http://www.ehow.com/how_8192233_tie-between-beads.html
Fibromuscular Dysplasia:  
1. Medial Dysplasia (Continued)  

3) Medial hyperplasia: 

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<td>Alternating areas of thinned media and thickened fibromuscular ridges containing collagen Internal elastic membrane may be lost in some areas</td>
<td>“String of beads” appearance where the diameter of the “beading” is larger than the diameter of the artery</td>
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<td>Perimedial fibroplasia</td>
<td>10–15%</td>
<td>Extensive collagen deposition in the outer half of the media</td>
<td>“Beading” in which the “beads” are smaller than the diameter of the artery</td>
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<td>Medial hyperplasia</td>
<td>1–2%</td>
<td>True smooth muscle cell hyperplasia without fibrosis</td>
<td>Concentric smooth stenosis (similar to intimal disease)</td>
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<td>Intimal fibroplasia</td>
<td>&lt;10%</td>
<td>Circumferential or eccentric deposition of collagen in the intima No lipid or inflammatory component Internal elastic lamina fragmented or duplicated</td>
<td>Concentric focal band Long smooth narrowing</td>
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<td>Adventitial (periarterial) fibroplasia</td>
<td>&lt;1%</td>
<td>Dense collagen replaces the fibrous tissue of the adventitia and may extend into surrounding tissue</td>
<td></td>
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Fibromuscular Dysplasia: 2. Intimal Fibroplasia

- Focal, concentric stenosis due to deposition of collagen in intima
- Long, smooth narrowing

(Slovut and Olin, 2004)

### Fibromuscular Dysplasia: 3. Adventitial Fibroplasia

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Summary
Algorithm for Diagnosing Suspected Renal Artery FMD

Detect stenosis and kidney height

CTA is superior to MRA due to higher spatial resolution, but neither can accurately estimate degree of stenosis

Perform with simultaneous revascularization procedure due to risk of dissection

Figure 3 Proposed algorithm for establishing the diagnosis of renal artery fibromuscular dysplasia (FMD). PTA, percutaneous transluminal angioplasty.

Diagnosis of FMD: Menu of Tests for Renal Artery Involvement

**Invasive:**
Catheter-based angiography: gold standard

**Noninvasive:**

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<th>Vascular studies to evaluate the renal arteries</th>
<th>Shows the renal arteries and measures flow velocity as a means of assessing the severity of stenosis</th>
<th>Inexpensive; widely available</th>
<th>Heavily dependent on operator’s experience; less useful than invasive angiography for the diagnosis of fibromuscular dysplasia and abnormalities in accessory renal arteries</th>
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<td>Duplex ultrasonography</td>
<td>Shows the renal arteries and perirenal aorta</td>
<td>Not nephrotoxic; useful in patients with renal failure; provides excellent images</td>
<td>Expensive; less useful than invasive angiography for the diagnosis of fibromuscular dysplasia; stents result in imaging artifacts</td>
</tr>
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<td>Magnetic resonance angiography</td>
<td></td>
<td>Provides excellent images; stents do not cause artifacts</td>
<td>Not widely available; the large volume of contrast medium required is potentially nephrotoxic</td>
</tr>
<tr>
<td>Computed tomographic angiography</td>
<td></td>
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Catheter-Based Angiography of Renal Artery: Companion Patient #1

Duplex Ultrasonography: Companion Patient #2

Renal artery peak systolic velocity ≥ 3.5 = 60-90% stenosis

Aorta peak systolic velocity

End-diastolic velocity ≥ 150 cm/s = 80-99% stenosis

(Olin, 2007)
Magnetic Resonance Angiography: Companion Patient #3
Computed Tomographic Angiography: Companion Patient #4

Image source:
http://www.learningradiology.com/archives05/COW%20174-Fibromuscular%20Dysplasia/fmhcorrect.htm
Diagnosis of FMD: Menu of Tests for Cerebrovascular Involvement

- Duplex ultrasonography
- Magnetic resonance angiography
- Computed tomographic angiography
Duplex Ultrasonography of Carotid Artery: Companion Patient #5

Image source:

Magnetic Resonance Angiography of the Carotid and Vertebral Arteries: Companion Patient #6

Computed Tomography Angiography: Our Patient

PACS, BIDMC
Head & Neck CT (+C) Reconstruction
Diagnosis of FMD: Our Patient

- Carotid artery imaging was consistent with fibromuscular dysplasia
- Renal artery involvement was suspected based on new-onset hypertension
- Renal imaging was obtained
Our Patient: Renal Artery Stenosis in Left Kidney

PACS, BIDMC
Abdomen CT (+C)
Maximum Intensity Projection Image
Differential Diagnosis of Renal Artery Stenosis

- Atherosclerosis
- FMD

(Safian and Textor, 2001)
Causes of Renal Artery Stenosis: FMD vs. Atherosclerosis

Patients with FMD:
Young women with no or few cardiovascular risk factors

FMD affects middle or distal arterial segments

Patients with Atherosclerosis:
Older individuals with CV risk factors

Atherosclerosis affects the proximal arterial segment

(Slovut and Olin, 2004)

Image source: http://www.riversideonline.com/health_reference/Heart-Disease/HQ01345.cfm?RenderForPrint=1
Our Patient: Normal Right Kidney on CT

Normal caliber of arteries

PACS, BIDMC
Abdomen CT (+C)
Curved Multi-planar Reformatted Image
Our Patient: Dissected Artery in Left Kidney on CT

Dissected artery: abrupt change in caliber of artery

PACS, BIDMC
Abdomen CT (+C)
Curved Multi-planar Reformatted Image
Our Patient: Areas of Infarct on CT

PACS, BIDMC
Abdomen CT (+C) Coronal

Wedge-shaped lesions (prior Infarcts)
Our Patient: Areas of Atrophy in Left Kidney on CT
Comparison of Our Patient’s Kidneys

Normal Right Kidney

Abnormal Left Kidney

PACS, BIDMC
Abdomen CT (+C)
Curved Multi-planar Reformatted Images
Carotid artery and renal imaging were consistent with fibromuscular dysplasia.

She was diagnosed with fibromuscular dysplasia, hypertension secondary to FMD, and hypertensive retinopathy.

She was medically managed with aspirin, anti-hypertensives, and anti-coagulants.
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• FMD is nonatherosclerotic, noninflammatory vascular disease
• Renal and carotid arteries are commonly affected
• Classic appearance of “string of beads” on radiologic imaging
• Diagnosis usually based on angiographic appearance
References

Acknowledgements

Seth Berkowitz
Gillian Lieberman
Claire Odom