AMSER Case of the Month
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55-year-old female with headache and refractory hypertension

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Patient Presentation

• **HPI:** 55 y/o female presents to outpatient clinic with 1.5 weeks of persistent headache. Has history of refractory hypertension that is poorly controlled with medication. Denies any fevers, chills, palpitations, weakness, or syncope.

• **PMH:** Hypertension

• **Meds:** Losartan, hydrochlorothiazide

• **Allergies:** NKDA

• **Vitals:** T 36.9C; BP 180/120; HR 90; RR 18; SpO2 100%

• **ROS:** Negative except for headache

• **Physical exam:** Unremarkable except right carotid and abdominal bruit noted on exam.
Pertinent Labs

• **CBC:** Within normal limits

• **CMP:**
  - Na+ = 148 (high)
  - K+ = 3.3 (low)
  - Cl- = 99
  - Glu = 130
  - Cr = 1.2
  - BUN = 20 (slightly elevated)

• **Cholesterol:** Normal

• **Triglycerides:** Normal

• **Aldosterone:** Slightly elevated
What Imaging Should We Order?
Select the applicable ACR Appropriateness Criteria

<table>
<thead>
<tr>
<th>Name</th>
<th>Category</th>
<th>Adult RRL</th>
<th>Peds RRL</th>
</tr>
</thead>
<tbody>
<tr>
<td>US duplex Doppler kidneys retroperitoneal</td>
<td>Usually appropriate</td>
<td>0 0 mSv</td>
<td>0 0 mSv [ped]</td>
</tr>
<tr>
<td>MRA abdomen without and with IV contrast</td>
<td>Usually appropriate</td>
<td>0 0 mSv</td>
<td></td>
</tr>
<tr>
<td>CTA abdomen with IV contrast</td>
<td>Usually appropriate</td>
<td>1-10 mSv</td>
<td></td>
</tr>
<tr>
<td>MRA abdomen without IV contrast</td>
<td>May be appropriate</td>
<td>0 0 mSv</td>
<td></td>
</tr>
<tr>
<td>ACE-inhibitor renography</td>
<td>May be appropriate</td>
<td>1-10 mSv</td>
<td></td>
</tr>
<tr>
<td>Arteriography kidney</td>
<td>Usually not appropriate</td>
<td>1-10 mSv</td>
<td></td>
</tr>
<tr>
<td>Venography with renal vein sampling</td>
<td>Usually not appropriate</td>
<td>Varies</td>
<td>Varies</td>
</tr>
</tbody>
</table>

This imaging modality was ordered by the PCP.
Select the applicable ACR Appropriateness Criteria

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<tr>
<td>US duplex Doppler carotid</td>
<td>Usually appropriate</td>
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</tr>
<tr>
<td>MRA neck without and with IV contrast</td>
<td>Usually appropriate</td>
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</tr>
<tr>
<td>MRI head perfusion with IV contrast</td>
<td>May be appropriate</td>
<td>0 0 mSv</td>
<td>0 0 mSv [ped]</td>
</tr>
<tr>
<td>MRI head without and with IV contrast</td>
<td>May be appropriate</td>
<td>0 0 mSv</td>
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</tr>
</tbody>
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This imaging modality was also ordered by the PCP.
Findings (unlabeled)
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Findings (labeled)

- Focal area of alternating stenoses and dilatations at mid portion of right renal artery on coronal CTA Abdomen
- “String of beads” appearance of left renal artery on coronal CTA Abdomen
Thin, linear carotid web noted at lateral wall of right internal carotid artery on CTA.
Final Dx:

Fibromuscular dysplasia of the bilateral renal arteries and right internal carotid artery
Fibromuscular Dysplasia

Definition: Fibromuscular dysplasia (FMD) is a noninflammatory, nonatherosclerotic disorder that can involve any artery, most commonly the renal and internal carotid arteries. FMD can lead to stenosis, aneurysm formation, and dissection of the affected artery.

Epidemiology: FMD most commonly occurs in young women, usually diagnosed between the ages of 30-50.

Clinical features: Patient with FMD may present with renovascular hypertension, abdominal bruit, and flank pain. Patients with cerebrovascular FMD may present with headache, neck pain, pulsatile tinnitus, and carotid bruit. Stroke/TIA are rare but also possible manifestations.
Fibromuscular Dysplasia

**Diagnosis:** Diagnosed by imaging – CT angiography, MR angiography, digital subtraction angiography, and duplex ultrasound.

**Classification:** FMD was previously classified into five categories depending on histology of the affected vessel wall layer. Categories included medial dysplasia, medial hyperplasia, perimedial fibroplasia, intimal fibroplasia, and adventitial fibroplasia. Angiographic correlations have been made from these histologic findings, and FMD is now primarily classified as either being multifocal or focal, based on angiographic appearance. Multifocal FMD corresponds to medial/perimedial histology while focal FMD corresponds to the intimal/adventitial histology.
Fibromuscular Dysplasia

**Radiographic features:** Multifocal FMD has an angiographic appearance of alternating stenoses and dilatations ("string of beads" appearance) at mid-distal portion of affected artery. Focal FMD has an angiographic appearance of concentric/tubular stenosis and, less commonly, carotid web. Other findings include vascular loops, fusiform vascular ectasia, arterial dissection, and aneurysm.

**Differential diagnoses:** Atherosclerosis, vasculitides, vasospasm, vascular injury, and segmental arterial mediolysis.

**Treatment:** Asymptomatic patients are observed. If symptomatic, treatment consists of antihypertensive medication, antiplatelet medication, and revascularization with percutaneous transluminal angioplasty (PTA), typically without stenting.
References:


