Renal Artery Fibromuscular Dysplasia

Michael Meuse, M.D.
Vascular and Interventional Radiology
Case

- 54 yo Caucasian woman
- Severe hypertension despite 3 meds
- 2 admissions for hypertensive crisis
- What are the considerations?
- What should be the next step in evaluating this patient?
Fibromuscular Dysplasia

- Middle-age Caucasian woman
- More common amongst 1st degree relative
- May be autosomal dominant with incomplete penetrance and variable expression
- Most patients have no family history
- Prevalence < 0.5% general population
Fibromuscular Dysplasia

- Non-atherosclerotic
- Non-inflammatory
- Pathogenesis – not fully known
  - Hormonal effects
  - Alpha-1-antitrypsin deficiency
  - Mechanical stresses (Rt > Lt)
  - Peculiar distribution of vasa vasorum
  - Tissue hypoxia
  - Genetic factors
FMD

Distribution

- Renal $\rightarrow$ 60-75%, distal main and branches (30-40% bilateral)
- Carotid/Vertebral $\rightarrow$ 25-30%, involvement associated with intracranial aneurysms in 7-51% (intracranial FMD is rare)
- Mesenteric – usually coexistent renal
- Other arteries – almost every artery (rare)
- Multiple arteries involved – 20-30%, almost always with renal
Case

- 54 yo caucasian woman
- Severe hypertension despite 3 meds
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- What are the considerations?
- What should be the next step in evaluating this patient?
Fibromuscular Dysplasia

• Challenging because of subtlety of lesions
• Requires high resolution imaging
• Entity demonstrated with all modalities (Duplex, CTA & MRA)
• DSA “gold standard”
• MDCTA also very accurate

53 yo female with severe hypertension
ceMR Angiography - RAS

- Sensitivity 88-100%; Specificity 71-100%
- Detection of ARAS is excellent, accessory renal arteries good, but FMD problematic
- Unsuitable after stent placement
- Claustrophobia & metal implants: 2 - 4%
Right FMD & Abdominal Coarctation
53 yo female with severe hypertension
## CTA vs ceMRA: Renal Arteries

<table>
<thead>
<tr>
<th></th>
<th>ceMRA</th>
<th>CTA</th>
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<tbody>
<tr>
<td>Sensitivity</td>
<td>62-100%</td>
<td>64-99%</td>
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<tr>
<td>Specificity</td>
<td>77-100%</td>
<td>89-98%</td>
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<tr>
<td>Occlusion</td>
<td>+++</td>
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<tr>
<td>Stenosis quantification</td>
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<tr>
<td>FMD</td>
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<tr>
<td>Hemodynamic significance</td>
<td>+++</td>
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<tr>
<td>Accessory renal arteries</td>
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<tr>
<td>Aneurysms, Dissections</td>
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<tr>
<td>After stent placement</td>
<td>-</td>
<td>+++</td>
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16 channel MDCTA
CT Angiogram

Smaller Rt kidney with associated renal artery FMD
Digital Subtraction Aortogram
Selective Rt Renal Angiogram
Fibromuscular Dysplasia

• What type of FMD is present?
  - Medial fibroplasia
  - Subadventitial fibroplasia
  - Intimal fibroplasia
FMD

- ~ 75-80% cases of FMD
- Classic “string of beads” where beads > diameter of native artery
- Media has areas of disruption with fibrous tissue accumulation or medial thinning → aneurysmal dilatation
- EEL may be fragmented
- Intima, IEL and adventitia are relatively uninvolved
- Can be progressive → rarely to occlusion
FMD

Medial fibroplasia
Perimedial (Subadventitial) Fibroplasia

- ~ 10% cases of FMD
- May coexist with medial fibroplasia
- “String of beads”, but beads are smaller than native artery diameter
- Excessive tissue in the outer media at its junction with the adventitia
FMD

Perimedial (Subadventitial) Fibroplasia

Accumulation of elastic tissue at the medial-adventitial junction
Fibromuscular Dysplasia

• What is the best treatment option in this patient?
  - PTRA
  - Stenting
  - Cutting balloon
  - Other endovascular therapy
  - Surgery
  - Medical therapy
FMD - PTRA

UVA Experience

- N = 66 Patients (85 Lesions)
- Tech Success = 100% (66/66)
- BP Cured 41% (27/66), Improved 57% (38/66)
- Renal Insufficiency (N = 14)
  - 86% (12/14) Improved
  - 14% (2/14) Stable
- Complications = 15% (10/66)

PTRA for FMD

F-U RANGE: 1-121 MONTHS
F-U MEAN: 39 MONTHS
F-U MEDIAN: 32 MONTHS

Fibromuscular Dysplasia

- What tools are used to perform PTRA?
- How does one size the balloon?
- What is the endpoint of the angioplasty?
RAST – GUIDE SHEATHS
PTRA – Guide Catheter

FMD
PTRA – Guide Catheter

POST-PTRA
Fibromuscular Dysplasia

- What tools are used to perform PTRA?
- How does one size the balloon?
- What is the endpoint of the angioplasty?
How do you determine balloon diameter?
Fibromuscular Dysplasia

• What tools are used to perform PTRA?

• How does one size the balloon?

• What is the endpoint of the angioplasty?
PTRA

51 y.o. female with ↑ BP

MEDIAL FIBROPLASIA
POST - PTRA
12 MOS F/U

FMD
PTRA

27 y.o. FEMALE WITH 4 yr HX ↑ BP

PRE - PTRA

27 MOS F/U

FMD
PTRA for FMD
Post-PTRA for FMD
FMD - Summary

• Noninflammatory, nonatherosclerotic
• Young, caucasian females
• 3 main histologic types
  – **Intimal fibroplasia** (overlaps with medial hyperplasia and adventitial fibroplasia)
    • Pediatric patients (adult patients – variants)
    • Long, smooth stenoses or focal resistant webs
  – **Medial fibroplasia** (most common type seen)
    • Classic “string of beads”
  – **Perimedial (subadventitial) fibroplasia**
    • Beads smaller than native artery
FMD - Summary

• May progress, especially in pediatric and fibrous lesion subtypes
• Appears to arrest at menopause and rarely leads to renal insufficiency
• If symptomatic, PTRA without stenting is the first treatment option
• ? consensus to screen carotids for FMD if any FMD is present
• Intracranial aneurysms associated with carotid/vertebral FMD
REFERENCES

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63 yo female with worsening hypertension; renal function normal; CTA performed

Extra case
57 yo female with difficult to control hypertension and normal renal function; no cardiovascular risk factors

Case 4
PTRA with a safety wire in the side branch
Post- PTRA
PTRA of main renal artery using an 0.014” system & 6F guide catheter
Post- PTRA