Non Atheromatous Lesions
Fibromuscular Dysplasia

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Presenter Disclosure Information

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Fibromuscular Dysplasia

“String of Beads”

Intimal symmetric narrowing with post stenotic dilatation
Occurrence

- Incidence <1%
- Most commonly occurs in young females
- Presents with:
  - Asymptomatic Carotid Bruit
  - Hypertension (renal ds)
  - Headache
  - Transient Ischemic Attacks
  - Stroke
  - Tinnitus, Vertigo, Arrhythmia, Carotodynia, hearing impairment, Angina …

Olin JW. Cleveland Clinical Journal of Medicine, April 2007.
Pathophysiology

• True etiology remains unknown
  – Congenital defects in media and internal elastic layers may predispose to pathologic change
  – ?? Autosomal Dominant with reduced penetrance in males ??
• Medial fibrodysplasia or intimal fibrodysplasia
  – Non-atherosclerotic
  – Non-inflammatory
• Most often affects the renal and carotid arteries
  – Renal: 85% of cases
  – Carotid: when present, bilateral in 80%
  – Both renal and carotid: 50% of cases

Olin JW. Cleveland Clinical Journal of Medicine, April 2007.
Classification

• Type 1: Medial Fibroplasia
  – 80+% of cases
  – Multiple, irregular concentric narrowings
    (dilations > normal artery)

• Type 2: Intimal Fibroplasia
  – ~7% of cases
  – Focal tubular stenosis

• Type 3: Adventitial Fibroplasia
  – Rare
  – Focal diverticular outpouchings
Associated Conditions

- Patients with FMD have increased risk for:
  - Intracranial aneurysms (20-50%)
  - Intracranial neoplasms
  - Carotid dissection
  - A-V Fistula

- Irritable vessels
  - Vasospasm common

Greenberg 2006.
Fibromuscular Dysplasia (FMD)

Brachiocephalic Location

- Carotid
  - C2: common
  - Proximal ICA: rare
- Vertebral
- Intracranial vessels: rare
Diagnosis

Angiography – Gold Standard
– High-grade stenosis with “string of beads” pattern

Indications for Treatment

- Symptomatic FMD
- Severe flow restriction
- Dissection/ pseudoaneurysm
- Anticoagulation contraindicated
Treatment

- **Carotids**… “string of beads” (Medial FMD)
  - Asymptomatic: antiplatelet medication and sequential imaging
  - Symptomatic: graduated low pressure endoluminal dilation (with EP)
  - **Stent** RARELY necessary

- **Renals**
  - If renal artery stenosis, hypertension or renal impairment, then transluminal angioplasty without stent

Olin JW. Cleveland Clinical Journal of Medicine, April 2007.
FMD: Treatment

• Medial and adventitial
  – Low pressure angioplasty
  – Success rate 80-90%
  – Recurrence 8%
Tubular FMD
segmental tubular narrowing

- High pressure angioplasty
- Success rate variable
- Self-expanding stent
“String Sign”

May be tubular FMD
What is this?

Pseudospasm
What is this?

Stroke in Evolution

String Sign

Progressive, Fluctuating Hemiparesis
ICA & MCA Open Above
CAS
Percusurge & Wallstent
Symptoms Resolved
Carotid “String Sign”

How to Evaluate

microcath
Pseudoocclusion... String Sign

Recurrent Symptoms...

Normal Vessel Above....Consider Reopening

Microcath run
Carotid dissection/ pseudoaneurysm with FMD when symptomatic or flow limiting
- Often track high into cervical segment
- May not be possible to use DEP
  -alternative = flow reversal

Stent
• Xpert – soft, trackable, self-expanding
• **Fibromuscular dysplasia**

FMD is a nonatherosclerotic, noninflammatory arteriopathy of small and medium size arteries that often affects the renal and carotid arteries (22). Angiographically, it is diagnosed by a distinct beading of the artery with alternating areas of dilatation and stenosis. Most often, this finding is of little clinical significance. However, patients can develop secondary dissections and symptomatic stenosis requiring treatment; often tracking high in the cervical segment of the carotid artery where filters are not easily used, but flow reversal might be possible. Soft, trackable self-expanding stents like the Xpert (Abbott Vascular Inc., Redwood City, CA; off-label for this indication) can produce excellent results, and balloon angioplasty alone can often be effective without stenting for pure FMD. Patients with Ehlers-Danlos syndrome, especially type IV, as the underlying condition are a high-risk group for treatment-related morbidity and long-term failure, and the decision to treat such patients should be carefully weighed against the risks of further vessel dissection. One patient treated recently at the University at Buffalo had bilateral, chronic, symptomatic dissections of the carotid artery secondary to FMD (Fig. 2a, left). The left-sided lesion was treated with an Xpert stent without DEP due to the high location and low risk for embolic phenomena; the right side was similarly treated with an Xpert stent (Fig. 2a, middle). Proximal to the stented region, the artery developed significant pseudospasm overlying the preexisting FMD (Fig. 2a, right). Angioplasty of this region was performed with a fair result, and a follow-up angiogram performed the next day showed a remarkably normal artery (Fig. 2b).
**ID:** 49 yo f

**RFR:** Bilateral sx upper cervical carotid stenosis

**PMH:** smoker

**HPI:** L hemisphere stroke (R paresis, dysphasia, mild cognitivei deficit, R eye patchy loss of vision), bilateral transient monocular blindness.

**Imaging MRI:** Multiple bilateral MRI DWI changes in carotid watershed distribution.

**Angio:** bilateral severe distal cervical ICA narrowing associated with FMD-type changes.

**Tx:**

- LICA: 5x30 Xpert 4x20 postdil
- RICA: 4x40 Xpert, 3x30 postdil

**Outcome:** Good clinical outcome
LICA – measurements

A: 4.8mm
B: 3.3mm

CF: 0.1353 mm/p (x1.7)
Abbott Expert Stent
Xpert Nitenol Stent

A stent specifically designed for small vessels

- Low profile
- High flexibility/conformability
- High radial force

Designed to optimize hemodynamics (low porosity)
### Technical details

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*Sheath AV in Beigene*
preop

stent placed
FMD

Three Types

• Intimal fibroplasia (tubular FMD)
• Medial fibroplasia (80%)
  – Fibromuscular hyperplasia
    (Dilated diameter $>$ normal diameter)
• Subadventitial fibroplasia
  – Dilated diameter $=$ normal diameter
FMD: Angiography

- String of beads (80%)
  - Dilated areas wider than normal artery
- Tubular FMD (intimal dysplasia)
  - Long segment = uniform narrowing
“String of Beads”

Intimal symmetric narrowing with post stenotic dilatation