Giant Cell Arteritis – Large Vessel Vasculitis

Kenneth J. Warrington MD
Associate Professor of Medicine
Mayo Clinic College of Medicine
Rochester, MN, USA

Evidence Based Medicine (EBM)

- Clinical Data Published in a Scientific Journal:

- Review Article & Meta-analysis:

Case

- 66 year old female; HTN
- PMR x 15 months
  - Low dose prednisone, tapered
  - No cranial symptoms or signs
- Presents with
  - Weight loss, malaise, left arm claudication
  - BP: (R) 136/82 (L) 90/
  - Left radial pulse: absent
  - ESR 85 mm/hr

Disclosures

- I have no relevant financial relationships to disclose

PET scan in GCA:
- FDG uptake in large vessels (83%)
- Subclavian arteries (74%)

Isolated PMR
- 31%; subclavian arteries

LV-GCA

Upper Extremity Arterial Involvement

LV-GCA
Upper Extremity Arterial Involvement

- Ultrasound
  - 30%: wall swelling of the axillary, subclavian and/or proximal brachial arteries
  - Bilateral disease common
  - Stenosis (15%)

- CT angiography
  - 42% subclavian arteries

Clinical Features

Older Cranial symptoms Vision loss

Younger Often absent Arm Claudication Vascular findings

Diagnosis

ACR ’90 Criteria (95%)
Earlier Diagnosis TAB

ACR ’90 Criteria (40%)
Delayed Diagnosis Vascular Imaging

Diagnosis: Angiogram

Diagnosis: MRA


Blockmans D, Bley T, Schmidt W. Curr Opin Rheumatol. 2009 Jan;21(1):19-28
Case: TAB negative; CT Angiogram

LV-GCA Treatment Course

- Compared to C-GCA
  - Same disease course\(^1\)
  - More relapses\(^2\)
  - Higher cumulative steroid dose
  - Longer to reach 0 mg

LV-GCA Prognosis

- UE arterial disease (by US)
  - 93% improved/stable
  - No ischemic complications
  - Symptoms resolved gradually
- Retrospective cohort

Revascularization

- Rarely required
- Collateral circulation sufficient to maintain tissue viability

LV-GCA Lower Extremity Arterial Involvement

- Imaging
  - PET scan:
    - Femoral and iliac artery (37%)
  - Ultrasound:
    - LE arteritis (12-50%)
- Clinical manifestations
  - Leg Claudication 1-20%
LV-GCA
Lower Extremity Arterial Involvement

- 19 women
  - 84% LE claudication preceded the diagnosis of GCA
  - Cranial symptoms were absent in 42%
- Proximal arterial disease
  - Superficial femoral and popliteal arteries
  - Bilateral
- Most improve with medical therapy
  - 15-30% critical leg ischemia

LV-GCA
Involvement of the Aorta

LV-GCA
Aortitis at onset of GCA

CT Angio (thickening) PET scan (FDG uptake)

65% 50%


Key Points

- Large-vessel vasculitis may be under-recognized
- Careful history and examination
  - Vascular system: pulses, bruit
  - BP measurement
  - Vascular Laboratory studies
- Vascular Imaging

LV-GCA
Aortitis


CT scan 4 years later

FDG-neg FDG-pos

* P = <0.05

Patients with aortitis at onset may be more prone to develop thoracic aortic dilatation

Printed by "VCRC. J Rheumatol. 2012 Feb;39(2):303-9"
Case

- 71 year-old retired optometrist
  - Asthma
- GCA diagnosis
  - ESR >100 mm/hr
  - Positive TA Biopsy
  - Standard therapy
- 2 ½ years later . . . .
  - Asymptomatic, Normal exam
  - Normal ESR & CRP
  - Prednisone 2 mg daily

Aneurysm of the Ascending Aorta

Active Giant Cell Aortitis

LV-GCA Aortic Aneurysm

- Thoracic Aortic Aneurysms
  - 17.3 – fold increased risk
  - 6.6 – fold increased risk
  - Up to 12% incidence at 10 years of F/U
- No consistent clinical predictors
  - Aortic Regurgitation

Aortic Anurysms - Timing

- At diagnosis
  - 15% of patients had aortic dilation
- As a late complication
  - 3-5 years from diagnosis

Timing of large-vessel involvement after GCA diagnosis: Cohort study 1950-2004

Robson JC. Ann Rheum Dis. 2013 Oct 4
Aneurysm screening

- Systematic screening study
  - 54 GCA pts, at median F/U 5.4 yrs
  - In clinical remission, low inflammatory markers
- Screening: CXR, then CT scan
  - 22% had aortic damage (aneurysm/dilatation)

Prospective F/U study
Median F/U of ~10 years
33% had aortic structural damage

Aortic Dissection

Aneurysm/Dissection is associated with increased mortality

Key Points

- Aortic inflammation common
  - Mostly asymptomatic
- Increased risk of Aortic Aneurysms
  - Late complication, GCA clinically quiet
  - Thoracic aorta
  - May cause premature mortality
  - Screening recommended

LV-GCA

Imaging
PET - CTA / MRA - US
80% . . . . 50% . . . . 30%

Clinical Manifestations
Symptomatic Vascular Stenoses
Aneurysm/Dissection < 30%

Sakurari C et al. Lancet. 2008 Jul 19;372(9634):244-45
Bouquet, Matthieu. Curr Opin Rheumatol 2006;18:10-17


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