BSH Heart Failure Day for Revalidation and Training 2016

“Catch Me if you Can” - An Unusual Cardiomyopathy

Speaker: Dr Paul M Haydock

Conflicts of interest: None
MA

- 52 y/o Post Office manager
- Admitted 28/06/2013 c/o chest pain & breathlessness
  - Unwell ~4/12
    - First noted after running for a plane
    - Spent journey with left chest pain and SOB
    - Since – SOBE – NYHA II
    - Occasional recurrent chest pain with pleuritic features
    - Nocturnal cough. No PND
- Previously fit and well
PMHx

- Osteoarthritis both hips
  - Bilateral hip resurfacing
    - Right 2004
    - Left 2008
- Not known hypertensive
- No regular meds on admission
- Never smoked
- EtOH – 4 pints beer / week

FHx

- Father heart failure in 50’s
  - Cause uncertain
  - RIP ?MI in late 50’s
Assessment

- Noted to have a large tongue
- Slight JVP + 4cm
- Bibasal crepitations
- BP 155/85
- HS I---II---I + 0
- No peripheral oedema
- No organomegaly

- Hb 126
- WCC 9.6
- Plts 184
- MCV 77.7
- Na+ 138
- K+ 4.0
- Urea 5.7
- Creat 89
- CRP 7
- Trop-I 0.29 (<0.04)
Adult Echo
S5-1
26Hz
19cm

2D
HGen
Gn 31
C 50
3/2/0
75 mm/s
Further Rx

- Pericardiocentesis
  - Serosanguinuous fluid
  - Reactive mesothelial cells w/ background lymphocytes + a few polymorphs. No malignant cells.
  - No organisms / no growth
  - Protein 82g/L (serum total protein 72)
Thoughts?
Further Results

- ESR 13
- Alb 37
- Bili 11
- ALT 18
- ALP 73
- B₁₂ 171
- Folate 6.8
- Ferritin 91
- cCa²⁺ 2.3
- CK 106
- LDH 521

- Normal serum EP strip
- -ve urinary BJP
- ANA / ANCA -ve
- α-galactosidase A 16.1pmol/punc/hr (6.3 – 47)
D/C Home

- Symptomatically well
- Established on
  - Ramipril 2.5mg
  - Bisoprolol 1.25mg
  - Spironolactone 25mg
  - Furosemide 40mg
  - Colchicine 500mcg bd
- Planned early r/v by HFSN for uptitration
- R/V Complex HF Clinic 3/12
Well

Normalization of LV function but persistent LVH
Adult Echo
S5-1
28Hz
19cm

2D
HGen
Gn 36
C 50
3 / 2 / 0
75 mm/s
Colchicine & Furosemide stopped
Referred for genetics re ?HCM
Sister and children advised phenotypic screening
Referred for cardiac Bx given unclear aetiology
Biopsy & Genetics Results

- Negative molecular analysis for 16 HCM genes
- Microscopic examination of 4 fragments of myocardium shows no evidence of amyloid.
- Stains for Congo Red are –ve
- No evidence of inflammation
- No abnormal infiltrate
- No vacuolation of myocytes to indicate a metabolic defect
Clinic Dec 2013

- EF improved to 60%
- Explained all results –ve
- Diagnosis revised to HCM of non-recognized genotype
- ETT
- 24h T
- Presented once in interim with chest pain and borderline troponin (0.37) – reassuring echo.
- Troponin repeated – persistently elevated @ 0.18
Persistent left hip pain since early 2014 – walks with antalgic gait
Never really had a good result from left hip
Seen March 2014
Further Orthopaedic Ix

- MRI hips
  - Left sided complex iliopsoas bursa with probable early loosening and neck erosion – I cannot exclude ALVAL/ARMD

- Whole blood
  - Cobalt 2060 nmol/L (0-20)
  - Chromium 4625 nmol/L (0-17)

- Synovial fluid
  - Cobalt 154700 nmol/L
  - Chromium 2600000 nmol/L
Biopsy
Thoughts...

- **Unifying Dx elusive**
  - Cobalt/Chromium cardiomyopathy
    - ↓EF / pericardial effusion
    - ?Can be associated with hypertrophy
  - Possible dual pathology
    - HCM + Cobalt/Chromium
    - Persistent “troponinitis”

- **Further admission with pericarditis May 2014**
  - Rim of pericardial fluid & mild LVSD
  - HS Troponin 120
Transferred to CCU post-op following further chest pain – HS Troponin 393
Ongoing review

- Doing well – NYHA I – II
- Cobalt / chromium levels reducing over time
- Most recent TTE shows EF now 40%
Cobalt Cardiomyopathy

1965 – ‘Quebec Myocardosis’

‘Beer Drinkers Cardiomyopathy’
- massive pericardial effusion
- low cardiac output, and raised venous pressure
- accompanied in a certain number of cases by polycythemia
- Thyroid dysfunction also common

Mainstay of therapy is removing toxin
- Chelation therapy has been used but benefits unclear
• Impression that pre-existing heart disease or malnutrition predispose to more aggressive forms of disease

• Several case reports of cardiotoxicity related to metal-on-metal hips

<table>
<thead>
<tr>
<th>Authors</th>
<th>Initial</th>
<th>Revision</th>
<th>Peak Co levels (µg/l)</th>
<th>Blood sample acid digestion?</th>
<th>Time to systemic symp-toms following revision</th>
<th>Hip pain</th>
<th>Weakness/ fatigue</th>
<th>Hypothyroidism</th>
<th>Auditory symptoms</th>
<th>Visual symptoms</th>
<th>Cardiac abnormalities</th>
<th>Poly-neuropathy</th>
<th>Other neuro-logical symp-tems</th>
<th>Other findings</th>
<th>Persistent symp-томs following revision</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rizzetti et al17</td>
<td>CoC</td>
<td>MoP</td>
<td>549 (blood)</td>
<td>NR</td>
<td>2 months</td>
<td>NR</td>
<td>NR</td>
<td>Yes</td>
<td>Hearing loss</td>
<td>Blindness</td>
<td>Yes</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>Partial visual loss</td>
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<tr>
<td>Oldenburg et al7</td>
<td>CoP</td>
<td>MoP</td>
<td>625 (blood)</td>
<td>NR</td>
<td>3 months</td>
<td>NR</td>
<td>NR</td>
<td>Yes</td>
<td>Hearing loss</td>
<td>NR</td>
<td>LVH, interstitial fibrosis</td>
<td>Yes</td>
<td>Headaches, convulsions</td>
<td>Weight loss, skin and nail changes</td>
<td>Neurological symptoms</td>
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<tr>
<td>Ikeda et al8</td>
<td>CoC</td>
<td>MoP</td>
<td>&gt; 400 (blood)</td>
<td>NR</td>
<td>2 years</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Hearing loss</td>
<td>NR</td>
<td>Yes</td>
<td>NR</td>
<td>Yes</td>
<td>NR</td>
<td>Mild persistent hearing impairment</td>
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<tr>
<td>Steens et al9</td>
<td>CoC</td>
<td>MoC</td>
<td>398 (serum)</td>
<td>NR</td>
<td>2 years</td>
<td>Yes</td>
<td>NR</td>
<td>Yes</td>
<td>Hearing loss</td>
<td>Partial loss of vision</td>
<td>Yes</td>
<td>NR</td>
<td>Skin changes</td>
<td>NR</td>
<td>Pronounced persistent hearing loss</td>
</tr>
<tr>
<td>Pelcova et al10</td>
<td>CoC</td>
<td>MoP</td>
<td>506 (serum)</td>
<td>NR</td>
<td>14 months</td>
<td>NR</td>
<td>NR</td>
<td>Yes</td>
<td>Hearing loss</td>
<td>NR</td>
<td>LVH, pericardial effusion</td>
<td>Yes</td>
<td>NR</td>
<td>Loss of appetite</td>
<td>Fatal cardiomyopathy</td>
</tr>
</tbody>
</table>

Current study CoC MoP 6521 (serum) Yes 6 months Yes Yes Yes Tinnitus NR Profound cardiomyopathy NR NR Loss of appetite Fatal cardiomyopathy

1 C?, ceramic femoral head, unknown acetabular liner; CoC, ceramic-on-ceramic; MoP, metal-on-polyethylene; MoC, metal femoral head, ceramic acetabular liner
2 Co, cobalt
3 NR, not recorded
4 LVH, left ventricular hypertrophy

Zywiel et al. Bone & Joint 2013
ca. 32 000 metal-on-metal hips in England & Wales