Carpal Tunnel Syndrome and Amyloid Cardiomyopathy

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Introduction

- Carpal tunnel syndrome (CTS) can be the initial manifestation of amyloidosis, often presenting years before cardiac involvement becomes apparent.

Hypothesis

- A significant number of older patients undergoing surgery for ‘idiopathic’ CTS may be diagnosed with amyloidosis through tenosynovial biopsy
- In these patients, early detection of amyloid cardiomyopathy may be possible using advanced cardiac imaging techniques

Methods

- Prospective, longitudinal study (ClinicalTrials.gov NCT02792790)
- Consecutive patients from May 2016 – June 2017 undergoing carpal tunnel release by participating physicians in the Cleveland Clinic’s Department of Orthopaedic Surgery
- Inclusion:
  - Males ≥ 50 and females ≥ 60 years
- Exclusion:
  - Known diagnosis of amyloidosis
  - Rheumatoid arthritis
- Tenosynovial biopsy during carpal tunnel release stained by Congo Red to detect amyloid deposits (Figure 1)
- If positive for amyloid:
  - Mass spectrometry for accurate typing
  - Cardiac workup including cardiac biomarkers, electrocardiogram, echocardiography with longitudinal strain, ⁹⁹m-technetium pyrophosphate nuclear scintigraphy (TcPYP)

Results

- 10 of 96 patients (10.4%) were positive for Congo Red on tenosynovial biopsy
  - 2 light chain (AL) subtype
  - 7 transthyretin (ATTR) subtype
  - 1 presumed ATTR
- 2 found to have cardiac involvement
  - 1 AL with IVS 1.5cm and apical sparing on echo with strain
  - 1 ATTR with IVS 1.4cm but no apical sparing on echo with strain, however Grade 2 uptake on TcPYP (Figure 2)

<table>
<thead>
<tr>
<th>Enrolled (n=96)</th>
<th>No Amyloid (n=86)</th>
<th>Amyloid (n=10)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, mean ± SD</td>
<td>68 ± 9.8</td>
<td>67.6 ± 9.8</td>
<td>72 ± 9.4</td>
</tr>
<tr>
<td>Male</td>
<td>49 (51%)</td>
<td>43 (50%)</td>
<td>6 (60%)</td>
</tr>
<tr>
<td>Black race</td>
<td>6 (6%)</td>
<td>5 (6%)</td>
<td>1 (10%)</td>
</tr>
<tr>
<td>Bilateral symptoms</td>
<td>82 (85%)</td>
<td>72 (84%)</td>
<td>10 (100%)</td>
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<tr>
<td>Prior CTS release surgery</td>
<td>36 (38%)</td>
<td>29 (34%)</td>
<td>7 (70%)</td>
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</tbody>
</table>

Conclusion

- Hand surgeons and cardiologists should be aware of the association between carpal tunnel syndrome and amyloidosis.
- Tenosynovial biopsy at the time of carpal tunnel release is a low-risk procedure that may lead to early diagnosis of amyloidosis, thereby allowing for timely intervention in this life-threatening disease.

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