Background and Objective

Amyloid myopathy is very rare. The objective of this study is to investigate the clinical, pathological, and radiological features of systemic immunoglobulin light-chain (AL) amyloidosis-associated myopathy (AL amyloid myopathy).

Methods

The patients with AL amyloid myopathy were selected from a total of 240 systemic AL amyloidosis patients who were diagnosed at our department between Sep 2001 and Feb 2017. Their clinical presentation and histopathological findings were retrospectively investigated. The diagnosis of amyloid myopathy was made based on the clinical presentation of apparent muscle weakness with increased level of creatine kinase (CK) and histological findings of muscle biopsy that were not compatible with the other established etiologies. The patients were also examined by Carbon-11 labeled Pittsburgh compound B ($^{11}$C-PiB)-positron emission tomography (PET) scan.

Results

Table. Clinical features of patients with AL amyloid myopathy

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Initial symptom</th>
<th>Age of initial symptom</th>
<th>Initial symptom of myopathy</th>
<th>Age of myopathy onset</th>
<th>Dominant clinical symptom due to myopathy</th>
<th>Most dominantly involved muscles</th>
<th>Serum CK level at the time of myopathy onset</th>
<th>Initial diagnosis of muscle biopsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>F</td>
<td>Proteinuria</td>
<td>55</td>
<td>Myalgia in the hip</td>
<td>56</td>
<td>Muscle weakness in the legs</td>
<td>Quadriceps femoris</td>
<td>800~1200 IU/L</td>
<td>Unchanged</td>
</tr>
<tr>
<td>Patient 2</td>
<td>F</td>
<td>Dropped head</td>
<td>80</td>
<td>Dropped head</td>
<td>80</td>
<td>None</td>
<td>Nuchal muscles</td>
<td>300~800 IU/L</td>
<td>Unspecific myopathy</td>
</tr>
</tbody>
</table>


Table 1. Continued

<table>
<thead>
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<td>Patient 1</td>
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<tr>
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<td>F</td>
<td>Dropped head</td>
<td>80</td>
<td>Dropped head</td>
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<td>None</td>
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Figure 1A. Patient 1. Left quadriceps femoris muscle biopsy

Figure 1B. Patient 2. Left biceps brachii muscle biopsy

Figure 2A. Patient 1

Figure 2B. Patient 2

Figure 2. $^{11}$C-PiB-PET images

PiB-PET scan suggested very slight tracer uptake in the involved thigh (2A) and showed obvious abnormal uptake in the involved neck (2B), respectively.

Discussion

Our histopathological findings were comparable with the reported cases that have documented minimal myopathic change with mild amyloid deposition in perimysium and endomysium and dominantly within intramuscular blood vessel walls but not in muscle fibers (Gertz et al., Mayo Clin Proc 2016). It is needed to be investigated how these trivial histological changes cause clinical symptoms.

Conclusion

- AL amyloid myopathy involves proximal muscles.
- Careful attention to the intramuscular blood vessel walls is needed to make appropriate pathological diagnosis.
- PiB-PET might be useful to detect muscle involvement among AL amyloidosis patients.