Recurrent hyposphagma and ptosis as presenting signs of primary localized orbital amyloidosis with lateral rectus involvement

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Background

Primary localized orbital amyloidosis is rare, with isolated involvement of extraocular muscle being even less common. Among primary localized disease, conjunctival amyloidosis is most commonly reported. Amyloid deposits consist of insoluble β-sheet protein aggregates. The most common histologic stain is Congo red, which yields green, yellow, and orange birefringence.

Case Description

77 year old woman presenting with recurrent left eye irritation, slight ptosis, and subconjunctival hemorrhage for 1 year.

- Visual acuity 20/20 OS
- No propptosis, papillary abnormality, or fundus pathology
- CT: soft tissue mass on anterior globe with left lateral rectus involvement and calcification
- Histopathology of excised mass with cytometry and histopathology consistent with AL amyloidosis
- Systemic workup including EKG, bone marrow biopsy, abdominal fat pad biopsy, and skeletal survey unremarkable
- Patient was treated conservatively with artificial tears and monitoring

Figure 1. Clinical, radiographic, and histologic findings in primary localized amyloidosis. (A) External photo with hyposphagma of left eye. (B) Slit lamp photo showed diffuse red, gelatinous mass over temporal bulbar conjunctiva. (C) H&E stain with revealing pink amorphous material. (D) Congo red stain with green birefringence with polarization. (E) Axial CT scan with thickening and calcification of left lateral rectus (arrows). (F) Slit lamp photo showed diffuse red, gelatinous mass over temporal bulbar conjunctiva. (G) H&E stain with revealing pink amorphous material. (H) Congo red stain with green birefringence with polarization.

Table 1. Review of cases of primary localized amyloidosis with extracocular muscle involvement.

<table>
<thead>
<tr>
<th>Case Description</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Clinical Signs</th>
<th>Location</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recurrent ptosis and hyposphagma</td>
<td>77</td>
<td>F</td>
<td>Left</td>
<td>Lateral rectus</td>
<td>Mass and calcification</td>
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<td>Isolated involvement of extraocular muscle</td>
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Discussion

Isolated extraocular muscle amyloidosis is exceedingly rare. First described by Holmström and Nyman in 1987, there have been few cases reported in the literature. The 3.8% of cases report isolated rectus involvement. Orbital signs are often present, but not in all cases as seen with this patient. CT is useful for mass characterization in adjunct with MRI. Systemic workup should be conducted in all cases. Monitoring is often sufficient, complete excision is typically curative.

References


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