Severe Esophageal Involvement in Cicatricial Pemphigoid

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INTRODUCTION

- Cicatricial Pemphigoid (CP): is a rare, scarring, vesiculobullous, autoimmune disease.
- Presenting symptoms: typically present with painful, shallow erosions & ulcers from fragile vesicles & bullae of varying size.
- Oral mucosa is often the most involved region (~90%) with the pharynx, larynx and esophagus being the least involved (~10%).
- Esophageal & anogenital involvement ~4% of the cases.
- Esophageal involvement: indicator of aggressive course.
- Few case reports of CP present as isolated esophageal disease.
- Hallmark of the disease:
  - Lesions at any anatomical site to heal with scar formation.
  - Loss of function, most commonly blindness, is a consequence of the disease.
  - This is a major cause of morbidity & mortality.  
- Minimal evidence on interventions to limit scarring.
- No recommendations to limit scarring sequelae. Clinical remission is required prior to intervention as trauma can exacerbate disease.

CASE PRESENTATION

PATIENT

- 89-year-old Caucasian male
- Multiple dermatologic issues of 12 months duration
- 60-pound weight loss from painful oral ulcers [Figure 1]
- Referred by wound care for back ulcer that had not responded to standard care
- He had been evaluated by multiple medical specialties

On exam
- A 8cm friable, bleeding, tender ulcerated plaque on his lower back [Figure 2 below], 6cm friable ulcerated plaque on his right forearm, several 1cm oral erosions, 1cm ulcer on scalp; fused penis to scrotum secondary to an active, fibrotic lesion [Figure 3 below] with anal erosions, bleeding from both nares

Initial Treatment
- Liquid prednisone 20mg QD, triamcinolone 0.1% dental paste, betamethasone dipropionate 0.05%

Pathology
- Multiple biopsies sent for direct immunofluorescence along with indirect immunofluorescence with salt-split skin results
- Pathology report showed CP, IgA variant

Course
- Hospitalized because of weakness & falls, received a PEG tube
- Inhaled budesonide 180mcg, dapsone 50mg QD & triamcinolone 1% ointment were added
- 1 month: resolution of nasal ulcers & Mycoprophate 2500mg added
- 3 months: oral mucosa & skin ulcerations persisted
- Perianal & anal canal: intralesional injection of 1cc 10mg/cc triamcinolone, Cloobetasol 0.05% ointment BID
- Budesonide & dapsone increased to BID.
- Mild improvement with continued dysphagia & globus sensation. EGD: Severe esophageal strictures. GI recommended against dilatation due to risks of perforation. He elected for hospice.
- 6 months: Passed away due to the complications

DISCUSSION

- Case reports indicate that esophageal involvement is typically asymptomatic, however there are cases with asymptomatic, yet severe, involvement. This could potentially lead to under-diagnosis and inadequate treatment.

Current Recommendations

Based on disease severity, determined by rate of progression, & areas of involvement

<table>
<thead>
<tr>
<th>Low risk:</th>
<th>High risk:</th>
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<tbody>
<tr>
<td>Lesions restricted to oral mucosa &amp; skin</td>
<td>Ocular, anogenital, laryngeal, nasopharyngeal or esophageal involvement</td>
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<tr>
<td>Numerous treatments with varying results have been used</td>
<td>Severe:</td>
</tr>
<tr>
<td>Topical treatments (corticosteroids or tacrolimus)</td>
<td>Systemic treatments (corticosteroids, dapsone, cyclophosphamide, mycophenolate mofetil or intravenous immunoglobulin)</td>
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- Our gentleman presented with a several months history of undiagnosed CP with severe & extensive oral, nasopharyngeal, anogenital & skin involvement with subsequent fibrosis.
- Involvement of his esophagus wasn’t diagnosed until late in his course.
- His course was fatal secondary to complications of malnutrition & severe esophageal strictures.
- This case is useful to raise awareness of this condition & the need for early, aggressive treatment & monitoring for debilitating sequelae of CP

REFERENCES

1. Trimarchi, T., Bellini, C., Fabiano, B., Gerevini, S., Bussi, M. Multiple mucosal involvement in cicatricial pemphigoid. Brasileiros de Dermatologia 2018;98;660-666
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