Incidence and Time to Diagnosis of Pemphigus Foliaceus in a Private Dermatology Practice Over 10 Years

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Abstract
Data on the incidence of pemphigus foliaceus in the community setting are sparse. This retrospective chart review analyzed one decade of patient visits to a private dermatology practice in the greater Tampa Bay area of Florida and identified four new diagnoses of pemphigus foliaceus. Time to diagnosis and factors associated with prompt and delayed diagnosis were noted. This study may allow community dermatologists to recognize this infrequent immunobullous condition more rapidly.

Introduction
Pemphigus foliaceus (PF) is an uncommon autoimmune blistering disorder propagated by antibodies to desmoglein 1, an intercellular adhesion protein found preferentially in the upper epidermis.1,2 Classic cases of PF are characterized by flaccid bullae or crusted erosions in a seborrhoeic distribution on the face, scalp, and upper trunk. Its presentation, however, may be subtle and protein, which can lengthen time to correct diagnosis.

Objective
This study determined the incidence of new diagnoses of PF, established the time to diagnosis (TTD), and examined factors associated with prompt diagnosis and delayed diagnosis.

Methods
This study was designed as a retrospective cross-sectional review using data previously gathered for patient care. The patient data utilized were selected using the following criteria: 1) The patient must have been seen at one of eight private practice dermatology clinics in western Florida from March 2007 to March 2017; and 2) the patient must have a new diagnosis of PF as identified by Florida from March 2007 to March 2017; and 2) the patient must have a new diagnosis of PF as identified by institutional review board exemption. The TTD of PF was substantiated by manual chart review. The TTD of PF during the studied time period, with an incidence of one per 38,558 new patient visits. The median TTD was 240 days (range: 60 to 480), or 3.5 office visits (range: 1 to 8). Fewer office visits were associated with widespread clinical distribution. Additional office visits were associated with limited disease distribution and misdiagnosis by initial histopathology.

Results
Four patients out of 154,230 were newly diagnosed with PF during the studied time period, with an incidence of one per 38,558 new patient visits. The median TTD was 240 days (range: 60 to 480), or 3.5 office visits (range: 1 to 8). Fewer office visits were associated with widespread clinical distribution. Additional office visits were associated with limited disease distribution and misdiagnosis by initial histopathology.

Discussion
While the dermatologic literature is replete with descriptions of PF—its clinical appearance, histology, management, and prognosis—little has been said about its incidence and TTD in the private practice setting. Cited incidence of PF is often combined with pemphigus vulgaris and stated relative to general populations: The incidence of pemphigus ranges from 0.76 to 16.1 new cases per million.3 Our data allow a more specific grasp of the incidence of PF: In a community dermatology clinic, an average of 38,558 new patients must be seen before encountering one new diagnosis of PF. This infrequency leads to unfamiliarity and, possibly, delayed diagnosis.

A retrospective review of seven cases from a tertiary care center reported a mean diagnostic delay of 300 days in PF.4 Our data show a median TTD of 240 days with a sample size of four patients. This demonstrates PF is uncommonly encountered and not immediately diagnosed, regardless of practice setting. Increased office visits were required for cases with disease limited to the head and neck and a biopsy initially diagnosed as lichen simplex chronicus (Figure 1). The limitations of the present study include small sample size and retrospective nature and reflect the rarity of PF.

Conclusion
This report represents unique data on incidence and TTD of PF in the private practice setting. Increased awareness of PF may accelerate its TTD and reduce its morbidity.

Figure 1. Patient 4: PF presenting as erythematous, crusted plaques of the left jawline. Time to diagnosis was prolonged after initial histopathology misdiagnosed lichen simplex chronicus.

Table 1. New diagnoses of PF at western Florida private dermatology practice, Mar. 2007 to Mar. 2017

<table>
<thead>
<tr>
<th>Pt Age (y)</th>
<th>Sex</th>
<th>Lesion Description</th>
<th>Lesion Distribution</th>
<th>Initial Clinical DDx</th>
<th>TTD</th>
<th>Bx Results</th>
<th>TTD-OV</th>
<th>Diagnostic method (H&amp;E, DIF, +/- Abs)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>47</td>
<td>M</td>
<td>Erythematous scaly patch</td>
<td>Left parietal scalp</td>
<td>Seborrheic dermatitis</td>
<td>480</td>
<td>OV 4: PF</td>
<td>4</td>
<td>H&amp;E</td>
<td>Limited disease distribution†</td>
</tr>
<tr>
<td>72</td>
<td>F</td>
<td>Erythematous, crusty, excoriated patches</td>
<td>Upper back, central chest, cheek</td>
<td>Drug eruption with impetigo</td>
<td>270</td>
<td>OV 1: PF</td>
<td>1</td>
<td>H&amp;E, DIF</td>
<td>More distributed at clinical presentation‡</td>
</tr>
<tr>
<td>71</td>
<td>F</td>
<td>Erythematous, scaly papules and plaques</td>
<td>Upper back, left breast, forehead</td>
<td>ACD</td>
<td>60</td>
<td>OV 1: PF</td>
<td>1</td>
<td>H&amp;E, DIF, Abs</td>
<td>More distributed at clinical presentation</td>
</tr>
<tr>
<td>65</td>
<td>M</td>
<td>Erythematous, scaly plaques</td>
<td>Bilateral jawline</td>
<td>LSC, DLE, sarcoidosis</td>
<td>210</td>
<td>OV 4: LSC Ov 8: PF</td>
<td>8</td>
<td>H&amp;E, DIF, Abs</td>
<td>Limited disease distribution Initial Bx results may have increased TTD-OV</td>
</tr>
</tbody>
</table>

Abs, serum antibodies against desmoglein 1; ACD, allergic contact dermatitis; DIF, direct immunofluorescence of perilesional skin; DLE, discoid lupus erythematosus; H&E, histologic analysis of lesional skin with H&E stain; LSC, lichen simplex chronicus; OV, office visit; PF, pemphigus foliaceus; TTD, time to diagnosis, measured in days from onset of symptoms to PF diagnosis; TTD-OV, time to diagnosis, measured in number of office visits until PF diagnosis

†Limited disease distribution: lesions limited to head/neck or trunk
‡More distributed at clinical presentation: lesions on head/neck and trunk
References

