Clinical and Histopathologic Comparison of Generalized Subcutaneous Morphea (GSM) with Eosinophilic Fasciitis (EF)

Fazleomar Mahmood 1, Ann J. Impens 1, Elena Schiopu 1, Kristine Phillips 1, Lori Lowe 2, Stephen Olsen 2 and James R. Seibold 1

1Scleroderma Program, University of Michigan, Ann Arbor, MI, United States
2Division of Dermatopathology, University of Michigan, Ann Arbor, MI, United States

Purpose:
GSM and EF are both characterized by inflammation and fibrosis involving the lower dermis, subcutaneous, fascia and even superficial muscle. Clinical and laboratory findings overlap as well but these entities have been classified as distinct syndromes. We identified 25 patients investigating clinical and histopathologic distinctions as well as response to therapy and outcomes.

Methods:
Retrospective chart review (1998-2008). Cases were selected by ICD9 codes from the database of outpatient clinical services at the University of Michigan Health Center. Reference area: 3200 patients. Patients with systemic sclerosis and other forms of inducative skin disorders were excluded. Full data included demographic, clinical and diagnostic variables. Two dermatopathologists have reviewed 8 out of 24 biopsied specimens to date.

Results: (See Tables)

Conclusions:
Clinical and laboratory features are indistinguishable between GSM and EF. Based on the 8 out 24 biopsy reviews so far, there are some histopathologic differences between GSM and EF, although these data do not support separate terminology, prognosis or approach to therapy.

Background
Deep morphea is a form of localized scleroderma characterized by inflammation and later sclerotic process involving the deep dermis, panniculus, fascia and even superficial muscle. Clinically characterized by morpheaform plaques, the skin appears diffusely indurated and bound down to the underlying fascia and muscle [1]. Flexion contractures of joints are frequently present [2]. Other features include arthralgia, arthritis and myalgia [1].

As opposed to systemic sclerosis there is minimal to no systemic involvement in deep morphea although features such as esophageal dysmotility, abnormal pulmonary function test results and even cardiac and renal disease have occasionally been reported [3]. The systemic involvement is very mild without impact on prognosis. Thus routine detection for systemic involvement in asymptomatic patients is not suggested [3].

Generalized subcutaneous morphea (GSM) and Eosinophilic fascitis (EF) are currently classified under the deep morphea group [4]. Although each of these conditions is prone to involve specific levels of skin, the similarities in clinical features have allowed them to be clustered in the same group.

Research Questions
1. Can generalized subcutaneous morphea and eosinophilic fascitis be differentiated clinically?
2. Can generalized subcutaneous morphea and eosinophilic fascitis be differentiated histopathologically?
3. Are there any long term outcome differences between generalized subcutaneous morphea and eosinophilic fascitis?
4. Are there differences in responses to therapy that might guide design of prospective interventional trials?

Abstract

Design:
• Retrospective cohort study, chart review of potential study subjects by using a combination of ICD-9 codes such as 701.0 (morphea or circumscribed scleroderma) and 728.89 (eosinophilic fasciitis)

Inclusion Criteria:
• Patients with GSM and EF seen at University of Michigan health center clinics in past 10 yrs.

Exclusion Criteria:
• Patients with systemic sclerosis.
• Patients with other forms of morphea such as plaque morphea or linear morphea.
• Patients with other indurated skin disorders such as scleromyxedema or scleroderma adularium.

Variables:
• 19 patients identified as study cohort
• The charts for these patients were reviewed for various demographic, clinical and diagnostic variables.
• The skin histopathology of 8/24 biopsy specimens have been reviewed by 2 dermatopathologists who were blinded to the original biopsy reports.

Patient Characteristics:
• 19 patients with GSM and 6 with EF
• Mean age at diagnosis: 50.7 y (range 18-83) in GSM group and 46.3 y (range 27-62) in EF group.
• Gender: 10/19 (52.6%) males in GSM group and 3/6 (50%) in EF group.

Results:

Table 1: Clinical Variables.

<table>
<thead>
<tr>
<th>Variable</th>
<th>GSM</th>
<th>EF</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (y)</td>
<td>50.7 (range 18-83)</td>
<td>46.3 (range 27-62)</td>
</tr>
<tr>
<td>Gender</td>
<td>10/19 (52.6%) males</td>
<td>3/6 (50%)</td>
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</tbody>
</table>

Table 2: Histopathologic Variables

<table>
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<tr>
<th>Variable</th>
<th>GSM</th>
<th>EF</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dermal Inflammation</td>
<td>50% (5/10)</td>
<td>30% (3/10)</td>
</tr>
<tr>
<td>Subcutaneous Inflammation</td>
<td>60% (6/10)</td>
<td>50% (5/10)</td>
</tr>
</tbody>
</table>

EF tends to have inflammation mostly confined to the fascia. Fascial involvement is quite prevalent in GSM as well. This is greater than has been previously recognized and reported. The presence of dermal and/or subcutaneous inflammation, and in particular plasmacytic inflammation, favors GSM.

Conclusions
Clinical and laboratory features are indistinguishable between GSM and EF. Based on the 8 out 24 biopsy reviews so far, there are some histopathologic differences between GSM and EF, although these data do not support separate terminology, prognosis or approach to therapy.

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