Clinical vignette: An atypical case of Sweet's syndrome presenting as facial cellulitis

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An atypical case of Sweet’s syndrome presenting as facial cellulitis

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CASE PRESENTATION
A 73 year old female presented with acute onset fever and right sided facial swelling, erythema and pain. She was initially diagnosed with facial cellulitis. The patient was placed on a course of cephalexin; however, her symptoms worsened, and she developed bilateral facial edema. Routine laboratory data was not indicative of leukocytosis. An incision and drainage was performed and minimal aspirate was retrieved; aspirate cultures were negative for bacterial and fungal growth. The patient’s facial swelling continued to progress; she developed bilateral peri-orbital edema, and she was intubated for airway protection. The patient’s antibiotics were then broadened to vancomycin and piperacillin-tazobactam, but after Infectious disease consultation, they were changed to ampicillin-subactam and fluconazole. Repeat cultures performed were again negative for any microorganisms. However, her symptoms did not improve with antimicrobial therapy, so it was discontinued and a non-infectious etiology was considered more likely. Rheumatology was consulted, but the rheumatologic workup was negative. Then, Dermatology was consulted; they performed a left cheek biopsy which showed pervascular and interstitial acute and chronic inflammation of the dermis, and a diagnosis of febrile neutrophilic dermatosis was made. Consequently, she was started on high-dose corticosteroids, and her symptoms dramatically and rapidly improved.

DISCUSSION

History and Introduction
Febrile neutrophilic dermatosis, or Sweet’s syndrome, was first characterized by Robert Sweet in 1964. The syndrome is characterized by fever, neutrophilia and asymmetric skin lesions. The pathogenesis remains unclear; however, it is believed to be a hypersensitivity reaction that leads to cytokine, auto-antibody or immune complex formation which results in neutrophil activation and infiltration.

Classification
1. Classical (idiopathic) Sweet’s syndrome
   - Occurs typically in women in the age range of 30 to 60
   - Typically follows an upper respiratory tract infection or gastrointestinal. Other associated conditions include inflammatory bowel disease or pregnancy.
2. Malignancy-associated Sweet’s syndrome
   - Occurs equally in men and women
   - Most common malignancy: AML
3. Drug-induced Sweet’s syndrome
   - Most common drug: Granulocyte colony stimulating factor

Clinical Manifestations
- Fever, neutrophilia
  - Asymmetric, tender erythematous skin lesions (papules, nodules or plaques).
  - Papules may have a clear vesicle-like appearance but on palpation are solid.
  - Lesions may be studded with pustules. Papules often coalesce into circinate or annular plaques.
  - Most frequent locations: upper extremities, face and neck
- Additionally, there may be extracutaneous manifestations involving bone, CNS, ears, eyes, mouth, heart, lung, kidneys, intestines, liver, spleen, and muscles.

Diagnosis

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Classic or Malignancy-associated</th>
<th>Drug induced</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (Major)</td>
<td>Sudden onset of painful erythematous plaques or nodules</td>
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<tr>
<td>2 (Major)</td>
<td>Bizarre consistent with a neutrophilic infiltrate without evidence of leukocytoclastic vasculitis</td>
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<tr>
<td>3 (Minor)</td>
<td>Temperature &gt; 38°C</td>
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<tr>
<td>4 (Minor)</td>
<td>Association with underlying malignancy, inflammatory disease, pregnancy, or preceded by upper respiratory or gastrointestinal infection or vaccination</td>
<td>Relationship with drug ingestion and clinical presentation, or recurrence after oral challenge</td>
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<tr>
<td>5 (Minor)</td>
<td>Excellent response to treatment with systemic steroids or potassium iodide</td>
<td>Resolution of lesions after drug withdrawal or treatment with systemic steroids</td>
</tr>
<tr>
<td>6 (Minor)</td>
<td>Three of the following items: ESR &gt; 20mm/hr, CRP &gt; 10mg/L, C reactive protein, WBC &gt; 8000, neutrophils &gt; 70 percent</td>
<td></td>
</tr>
<tr>
<td>Diagnosis</td>
<td>Both major criteria and two more criteria</td>
<td>All 5 criteria</td>
</tr>
</tbody>
</table>

Pathology
- Typically shows inflammation and edema of dermis, with neutrophilic infiltrate.

Treatment
- First-line: systemic steroids; prednisone (1 mg/kg/day) for 4 weeks up to 6 months. Other agents: potassium iodide, colchicine
- Second-line: indomethacin, clofazimine, cyclosporine, dapsone

SUMMARY POINTS
1. Sweet’s syndrome is characterized by fever, neutrophilia and asymmetric skin lesions. It is classified into either classical, malignancy-associated, or drug-induced based on associated factors.
2. It is diagnosed based on different major and minor criteria; however, diagnosis is based primarily on tissue biopsy.
3. Treatment depends on the underlying cause, but systemic glucocorticoids are the mainstay of therapy.
4. This patient had no malignancy, prior viral illness, and had not been taking any medications that are typically associated with Sweet’s syndrome, which makes this presentation unique.

CONCLUSIONS
This case not only exemplifies the common diagnostic dilemma of limiting a differential diagnosis but is also an unusual presentation of an already rare syndrome.

REFERENCES