The Face that Launched a Thousand Lab Tests

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Case Presentation

A 69-old-woman with history of hypothyroidism and rosacea conjunctivitis presented to the emergency room with a non-resolving facial rash and new onset of fevers.

- 3 weeks prior to admission (PTA):
  - Nasal redness, dx "nasal vestibulitis" → mupirocin
- 2 weeks PTA:
  - Fevers, headaches, nausea/vomiting
  - Seen by ENT, dx "cellulitis" → trimethoprim-sulfamethoxazole
  - Right nostril improved, continued malaise, mild neck pain developed
- 1 day PTA:
  - Bleeding from nare, neck pain
  - Seen by ENT again, dx "cellulitis" → trimethoprim-sulfamethoxazole
- Day of admit:
  - Severe neck pain, vomiting, fevers, conjunctivitis, photophobia, worsening facial rash, mild confusion

Physical exam:

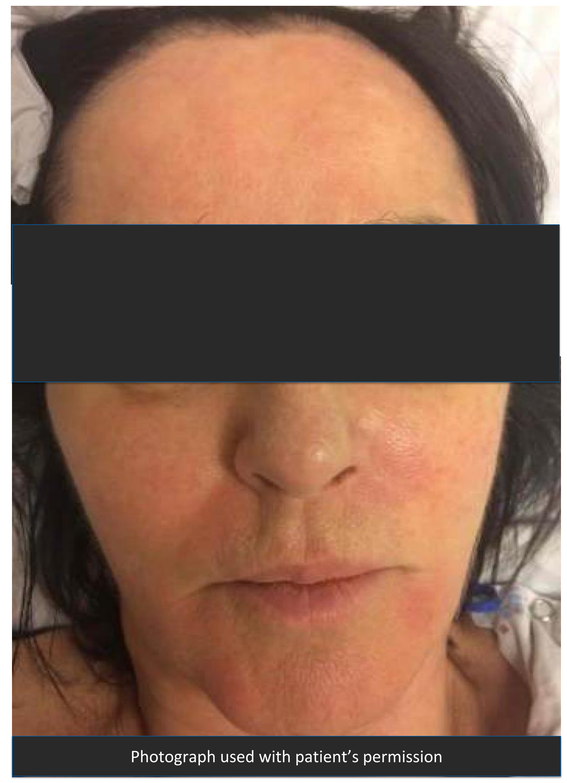
- T 37.6 °C HR 117 RR 16 BP: 93/44 mmHg, SpO2: 94 %
- Bilateral conjunctivitis, scabbing of left nare
- Patchy, tender, erythematous nodular rash on forehead, cheeks, chin
- Cardiac, respiratory, abdominal exams normal
- Neuro: Non-focal, alert and oriented

Labs:

- WBC intially 6.25 K/mm^3 (94.8% neutrophils), otherwise normal CBC
- Normal CMP, lactic acid, TSH
- CRP of 11.1 mg/L (H), ESR 28 mm/hr
- LP with normal opening pressure, 1 WBC, normal protein and glucose

Imaging

- CT head and CT venogram → negative for acute abnormality
- CT maxillo-facial → No abnormal enhancement



Photograph used with patient's permission

Clinical Course:

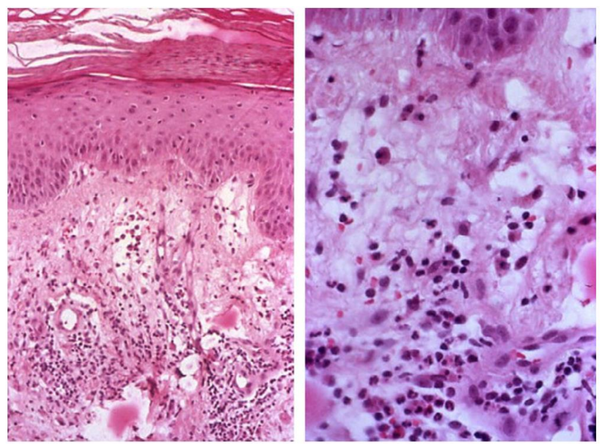
- Resuscitated with IVF, broad spectrum antibiotics started
- Blood pressures decreased to MAPs in 50s, temperature rose to 40 °C
- High dose systemic corticosteroids were initiated for refractory hypotension with rapid resolution of hypotension and fevers.
- Facial rash completely resolved by next day
- Culture data remained negative and a large workup for viral, auto-immune, and rheumatologic causes returned negative

Common medications associated with drug-induced Sweet syndrome

Antibiotics	Minocycline, nitrofurantoin, TMP-SMX, ofloxacin, quinupristin/dalfopristin
Antiepileptics	Carbamazepine, Diazepam
Antiretrovirals	Abacavir
Antihypertensives	Hydralazine
Antipsychotics	Clozapine
Antithyroid drugs	Propylthiouracil
NSAIDs	Celecoxib, diclofenac
Diuretics	Furosemide
Contraceptives	Levonorgesteryl/ethinyl estradiol, levonorgestrel-releasing IUDs
Colony stimulating factors	GCSF, granulocyte-macrophage-colony stimulating factor

Discussion:

- Sweet syndrome (acute febrile neutrophilic dermatosis) is characterized by abrupt onset of tender, erythematous nodular rash on extremities and head/neck, high fevers, elevated inflammatory markers, and neutrophilia
- The syndrome can be drug-induced or secondary to malignancy, inflammatory disease, or upper respiratory/gastrointestinal infection
  - In our case, the most likely culprit was trimethoprim-sulfamethoxazole, a well-documented cause of drug-induced Sweet syndrome
- Biopsy of the nodules is usually performed to confirm the diagnosis, which shows papillary edema, swollen endothelial cells, and a neutrophilic infiltration
- Treatment is with systemic corticosteroids and usually requires a 4-6 week course, though the disease may recur
- Sweet syndrome belongs to a group of conditions called the neutrophilic dermatoses, which also includes abscesses/cellulitis, pyoderma gangrenosum, subcorneal pustular dermatitis, and leukocytoclastic vasculitis
- This case represents one of the many causes of non-responsive nodular facial rash.
- Other diagnostic considerations when faced with this include DRESS (drug reaction with eosinophilia and systemic symptoms), fixed urticaria, angioedema, abscess, erysipelas, or viral illnesses



Representative biopsy of Sweet syndrome

Teaching Points

- Clinicians should consider Sweet syndrome in facial cellulitis type processes, especially with minimal or sluggish response to appropriate antibiotics
- Sweet syndrome can be caused by a number underlying conditions and may require an extensive workup

References

1.Cohen, P.R. Neutrophilic dermatoses: a review of current treatment options. AM J Clin Dermatol (2009) 10: 301.
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