HOLY C.O.W.!

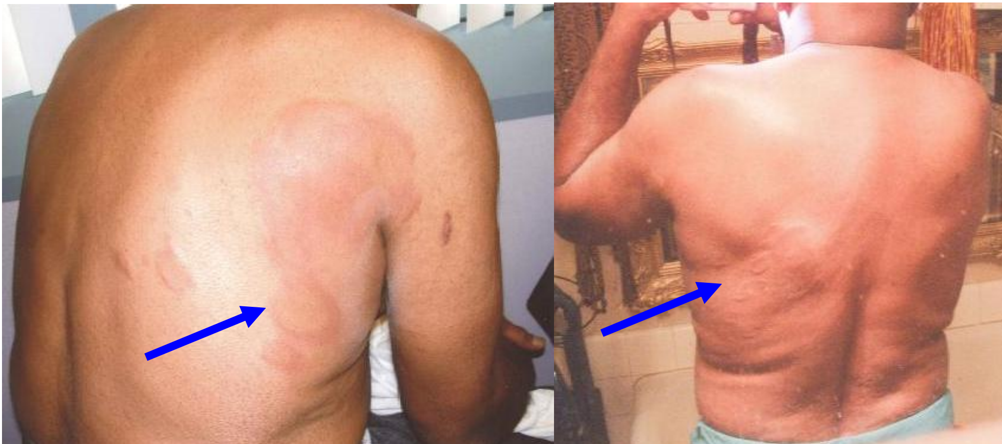
IT'S…

Clinical Question of the Week #29
January 26th, 2009 through February 2nd, 2009

Happy Chinese New Year! Please e-mail your answers to Kuo, Tim, Wendy, and Kevin (klian@mednet.ucla.edu; tprovias@mednet.ucla.edu; wsimon@mednet.ucla.edu; kbreger@mednet.ucla.edu) by 0800 on Monday, February 2nd, 2009. The resident or intern with the most correct answers at the end of each month will receive a prize!

Case: A 55-year-old man presents to urgent care clinic with areas of tender, red, and swollen skin over the past few days. He emails you with pictures that he has taken at home, which are shown below. He recalls an episode in which this happens earlier as well a while back, however his only recent symptoms include a recent head cold with some mild body aches. His past medical history is notable for AML diagnosed two years ago, which has been treated and is currently in remission. Examination reveals tender, erythematous patches of skin, some of which has a little pustular appearance, which developed since the patient took the picture two days ago.

**Special thanks to Dr. Baldeep Singh, who supplied this case.

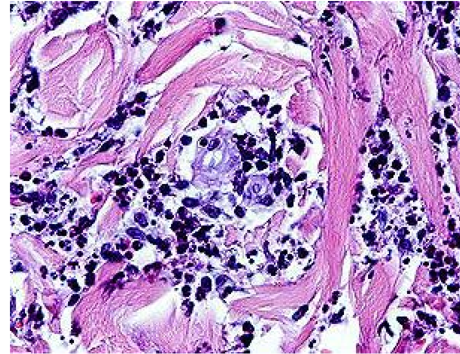

Images taken by the patient of his cutaneous lesions.

Questions:

1. **What is the diagnosis?**
   The prototype for neutrophilic dermatoses, Sweet originally described in 1964 the syndrome of acute onset of fever, leukocytosis, and erythematous plaques infiltrated by neutrophils, which later was named after him as Sweet's disease. Sweet's disease is a rare phenomenon with a female predominance (4:1).

The pathogenesis of Sweet's syndrome is thought to possibly be due to hypersensitivity reaction, with hypothesized causes including immune complex vasculitis, T-cell activation, and altered neutrophil function. Dysregulation of cytokines is thought to be the cause of the clinical manifestations, and include Interleukins (1, 3, 6, and 8), G-CSF (there is an association with Sweet's syndrome and G-CSF therapy), and GM-CSF.

The diagnostic pathology of Sweet's syndrome is dense and nodular perivascular neutrophilic infiltrates, with vasculitis as a rare phenomenon. Neutrophils may also migrate to involve the more superficial dermis, causing pustules or panniculitis. Diagnosis is made on satisfying two major and two minor criteria from the following: 1) typical cutaneous lesions (major); 2) consistent pathophysiology (major); 3) antecedent fever of infection (minor); 4) fever, arthralgia, malignancy or conjunctivitis (minor); 5) leukocytosis (minor), and 6) response to steroids (minor). (0.5)



2. **Name two associated conditions.**
   Sweet's syndrome is associated with a malignancy in 20-25% of cases, most commonly a hematologic malignancy, however in 15% of these cases the association is with a solid tumor. There is also association with G-CSF therapy, so there may be overlap. Sweet's syndrome is also associated with autoimmune processes such as RA, SLE, MCTD, Hashimoto's thyroiditis, SS, and Bechet's, as well as Crohn's and UC.

   Infectious associations include vaccinations, streptococcus, mycobacteria, yersinia, typhus, salmonella, CMV, HIV, and chronic active hepatitis. Multiple drugs may also cause Sweet's syndrome including lithium, lasix, hydralazine, oral contraceptives, minocycline, Bactrim, and imatinib. Finally, rare associations include pregnancy, complement deficiency, subacute necrotizing lymphadenitis, and POEMs syndrome. (1)

3. **Name two clinical features.**
   The primary clinical manifestations include the characteristic inflammatory eruption, a dermal neutrophilic infiltrate without vasculitis on biopsy, fever, and neutrophilia. The skin lesions are usually irregular and may have an area of yellow discoloration in the center. They are frequently painful and nonpruritic, and may progress to blistering or pustule formation. Other symtoms include eye involvement (conjunctivitis, episcleritis, iridocyclitis), arthralgia, myalgia, asymmetrical inflammatory arthritis, neutrophilic alveolitis, sterile osteomyelitis, acute renal failure, involvement of the liver and pancreas, and neurologic or psychiatric changes. (1)

4. **What is the treatment?**
   While little data exists on the treatment of Sweet's syndrome, the usual course of therapy includes oral prednisone taper. Other non-steroid treatments include NSAIDS, dapsone, potassium iodide, colchicine, doxycycline, and clofazimine. (0.5)

Did you catch the clue? Eating **sweets is considered good luck for Chinese New Year, so as to have a "sweet" year.