



SWEET'S SYNDROME

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Sweet's syndrome, named after the physician who first described the condition, is also known as acute febrile neutrophilic dermatosis. The initial manifestations of this condition are non-specific and often overlooked. These include fever, elevated white blood cell count and flu-like symptoms. As the condition progresses, the patient will abruptly develop numerous tender, red or purple, plaques and papules on the arms, head, neck and trunk.

Sweet's syndrome is a relatively unusual but not rare dermatologic condition. It demonstrates a female prevalence; occurring about four times more commonly in women than men. The exact mechanism underlying Sweet's syndrome is not known at this time. One of the current hypotheses equates the disease to the body developing a hypersensitivity reaction to an underlying infection, autoimmune disease, medication or cancer. The role of the neutrophil, which is a specialized form of white blood cell, in Sweet's syndrome is thought to be an inappropriate response to cellular signals known as cytokines and growth factors. When present in excess amounts, cells like neutrophils will respond to these signals and infiltrate the skin leading the characteristic rash.

The cutaneous manifestations of Sweet's syndrome include tender, non-itchy, red-to-purple papules and plaques that can enlarge as the disease progresses. Sometimes these lesions enlarge and may coalesce to cover large areas of the body. Patients can also develop significant swelling within and around the rash that give them a blister-like appearance. The distribution of skin lesions has a predilection for the head, neck and arms. In cases where Sweet's syndrome is secondary to an underlying disease, the eruption becomes more widespread, occurring anywhere on the body. Furthermore, up to 80% of individuals develop a fever >100.4F and upper respiratory flu-like symptoms.

Criteria for the diagnosis of Sweet's syndrome include 2 major criteria and 4 minor criteria. To confirm the diagnosis, both major and 2 additional minor criteria must be met. They include:

Major

- Sudden onset of classical skin lesions
- Histological examination consistent with Sweet's syndrome

Minor

- Fever or constitutional signs and symptoms
- Elevated number of white cells in the blood
- Marked improvement to the administration of steroids
- Associated with malignancy, inflammatory disorder, drug exposure, vaccination or infection

Sweet's syndrome in and of itself is a benign condition. If left alone, the lesions will resolve spontaneously but may recur in up to 30% of patients. If the disease is secondary to an underlying disease, treatment of the primary condition is recommended. The traditional therapy for Sweet's syndrome is **oral prednisone** at doses of 0.5-1.0mg/kg/day for 4-6 weeks. Alternatives to prednisone include **dapsone**, **potassium iodine** and **colchicine**.

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