THE AUSTRALASIAN COLLEGE
OF DERMATOLOGISTS

Sweet's Syndrome

Also known as ... Acute Febrile Neutrophilic Dermatosis, Gomm-Button Disease

What is Sweet's syndrome?
Sweet's syndrome is an uncommon condition named after the English dermatologist, Dr Robert Sweet, who first described it in 1964. Sweet's syndrome usually presents with fever, a rise in white blood cells, raised red skin rashes and neutrophils (pus cells) infiltrating the upper layer of the skin.

Fever and a rise in white blood cells are not always present. Sweet's syndrome is sometimes initially mistaken for an infection or medication allergy.

What causes Sweet's syndrome?
Sweet's syndrome is a disorder of neutrophils (pus cells) and although it is not considered hereditary, there is some evidence of a genetic link.

Most experts consider Sweet's syndrome a "reactive condition" - it is driven by an underlying condition in up to 50% of cases. As the appearance of Sweet's syndrome may occur before the underlying condition is recognised, ongoing monitoring is required, especially if the skin eruption is severe, recurrent or treatment-resistant or if blood abnormalities are detected.

What does Sweet's syndrome look like?

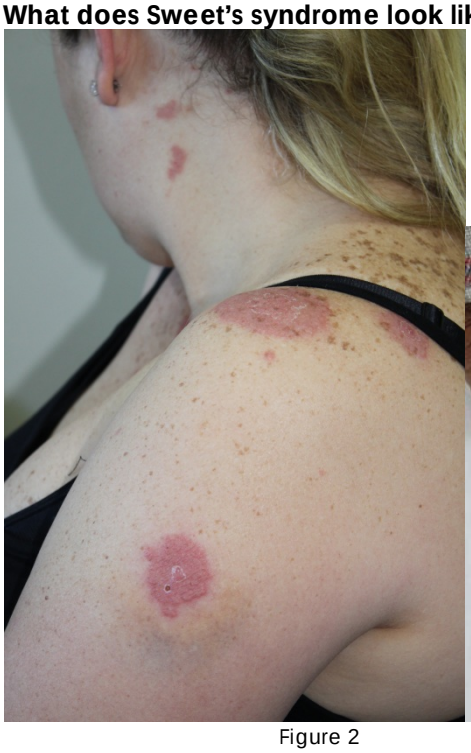

Figure 2

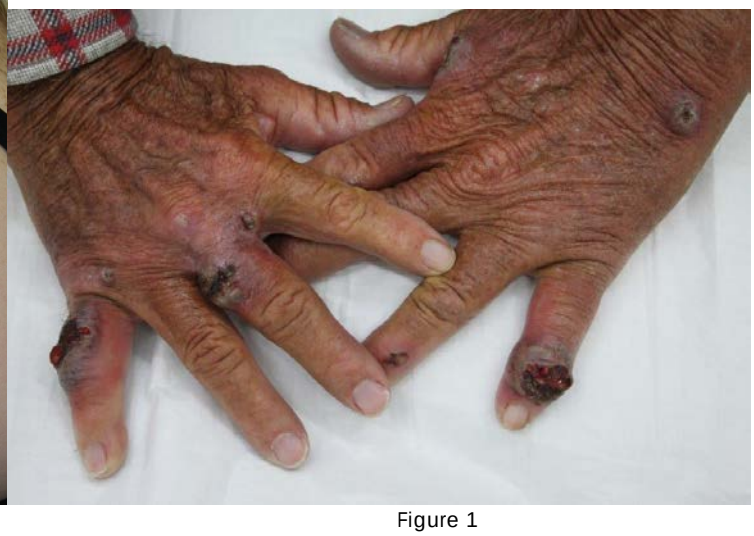

Figure 1

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- Tender red-purple bumps joining into larger raised areas on the skin; often these may look like blisters or may form true blisters or pustules; often round ring-like or arch-shaped; common on the face, neck and upper limbs with one form localised over the back of the hands
- Fever, headache, malaise (feeling unwell) and joint pains with, or prior to, the rash in 50% of cases
- Mouth ulcers
- Sore eyes
- Other organs (eg lungs, kidneys, bones, heart, muscle) are rarely involved.

What are the underlying conditions that can cause Sweet's syndrome?

1. Classical Sweet's syndrome occurs in over 50% of cases, especially in middle-aged women. In these cases, the symptoms of Sweet's syndrome start after a non-specific chest or gastrointestinal infection.
2. Immunologic disease is seen in around 15% of Sweet's syndrome cases. This includes inflammatory bowel disease and connective tissue disorders such as rheumatoid arthritis.
3. Cancer is seen in about 10 to20% of Sweet's syndrome cases. Blood-related disorders such as leukaemia and solid organ tumours (bowel, breast, or kidney cancer) are the most common cancers seen in association with Sweet's syndrome.
4. Drug-induced Sweet's syndrome may be caused by bone-marrow cell growth factors, certain antibiotics, anti-epileptic or anti-inflammatory medications. There are many other medications known to have caused Sweet's syndrome.
5. Pregnancy is known to cause Sweet's syndrome.
6. Other reactive neutrophilic disorders are associated with Sweet's syndrome. These conditions are characterised by lesions occurring in areas of minor skin injury (known as "pathergy") and include conditions such as pyoderma gangrenosum.

How is Sweet's syndrome diagnosed?

A skin biopsy performed by a dermatologist usually confirms the diagnosis. This shows neutrophils, some breaking down, entering the upper layer of the skin, particularly around the blood vessels. The blood vessels may be damaged by chemicals released by the neutrophils.

Blood tests are used to confirm a raised white cell and neutrophil count. Blood tests also show inflammatory markers, such as ESR (erythrocyte sedimentation rate) as being raised.

Other tests such as urine tests, bone marrow testing, cancer screening and imaging studies (x-rays, CT scans) and tests to rule out infection are sometimes needed to look for associated conditions of Sweet's syndrome.

How is Sweet's syndrome treated?

Sweet's syndrome is treated by trying to identify and cure any underlying or associated disorder. In more than 50% of cases no cause or underlying medical association is found and the condition is treated with corticosteroid tablets (prednisone or prednisolone). Generally a rapid improvement is expected and the dosage of corticosteroids is gradually reduced over several weeks or months. Corticosteroids can be applied as a cream or injected into areas of Sweet's syndrome in mild localised cases.

Alternative treatments may be used in cases where corticosteroids are not effective or safe (e.g. the presence of a serious infection). In small studies, a good response to indomethicin and colchicine or potassium iodide has been reported.

People with Sweet's syndrome have responded well to other drugs affecting neutrophil function including: "anti-infective" agents (eg dapsone, clofazimine, metronidazole or doxycycline); retinoids (eg isotretinoin); pentoxifylline and thalidomide; Immunosuppressive and cytotoxic agents (eg methotrexate, cyclosporine, chlorambucil) and biologic agents.

What is the likely outcome of Sweet's syndrome?

The outcome of Sweet's syndrome may be related to that of the underlying disorder if present, e.g. malignancy, rheumatoid arthritis etc.

With treatment, the fever and blood count usually improve within a couple of days and the rash clears within a few weeks. Some cases will clear without treatment over a number of weeks.

Recurrent episodes of Sweet's syndrome affect about one-third of all Sweet's syndrome cases and some will require long-term therapy.
Most areas of Sweet's syndrome fade without permanent marks but the appearance of pigmentation (brown marks) may take several months to fade.

Where can I find more information about Sweet's syndrome?
Information sites:
http://emedicine.medscape.com/article/1122152-overview
Support group:

https://www.facebook.com/search/top/?q=Sweets%20Syndrome%20Australia

If you have a skin rash and fever present you should see your doctor immediately.

This information has been written by Clinical Associate Professor Duncan Stanford