

SWEET'S SYNDROME (ACUTE FEBRILE NEUTROPHILIC DERMATOSIS)

What are the aims of this leaflet?

This leaflet has been written to help you understand more about Sweet's syndrome. It tells you what it is, what causes it, possible treatments and where you can find out more about this condition.

What is Sweet's Syndrome?

Sweet's syndrome (also known as acute febrile neutrophilic dermatosis) is a rare skin disorder characterised by a fever and the appearance of tender red or purple lumps or patches on the skin that may ulcerate. It is not contagious, not hereditary and not a form of skin cancer.

What causes Sweet's Syndrome?

In approximately half of those affected it is not possible to identify a cause for Sweet's syndrome. It is much more common in females between 30-60 years old, although it can occur in males and in other age groups. It can occur in people of any ethnicity and skin type. In the other half of cases, the majority are secondary to an upper respiratory tract or gastrointestinal infection or a drug reaction from medications such as non-steroidal anti-inflammatory medications. In some patients it can be a manifestation of another condition such as inflammatory bowel disease, lupus erythematosus or rheumatoid arthritis. In a few cases, it is a result of hormonal changes such as in pregnancy or thyroid gland disorders and more rarely may be a sign of cancer such as blood, breast or bowel cancer.

What does Sweet's syndrome of the skin look like?

The most common skin changes in Sweet's syndrome are raised red, pink or purplish tender skin lumps. These can be small (known as papules, which can be about 5-10 mm) or larger (known as nodules) and can join together to form bigger areas (known as plaques). They can be single or multiple and can appear anywhere on the skin although the arms, face and neck are affected most often. Sometimes blisters or pustules can be seen and the rash may appear at the site of an injury to the skin.

What are the symptoms of Sweet's syndrome?

Sweet's syndrome may cause some or all of the following symptoms arising together over a period of hours or a few days:

- a rash (described above)
- tiredness, lack of energy and feeling unwell
- high fever (temperature)
- aching joints and muscles
- mouth ulcers
- sore red eyes

How will Sweet's syndrome be diagnosed?

Tests that are useful in Sweet's syndrome include:

- Skin biopsy. This is an important test. A sample of the abnormal skin may be taken by your dermatologist. The skin sample is then examined under a microscope in the laboratory. It takes time for the sample to be processed and analysed by a pathologist (specialist doctor) so you will not get the result on the day.
- 2. **Blood tests** such as high levels of a type of white blood cells called neutrophils, can help to make a diagnosis of Sweet's syndrome.
- 3. Your doctor may recommend other blood tests or imaging studies such as X-rays or CT scans to look for an underlying cause of the Sweet's syndrome, if suspected.
- 4. If a medication is thought to be responsible, your doctor may recommend stopping it for a while to see if this improves your symptoms

Can Sweet's syndrome be treated?

Yes. Treatment is available for Sweet's syndrome and it can improve quite quickly. Treatments can be divided into topical and oral medications.

1. Applications to the skin (topical treatments)

- Strong <u>steroid creams or ointments</u> may help, especially with smaller lumps, and may help reduce the tenderness. Occasionally, steroid injections into the lesions can be used for very swollen or painful lumps.

2. Oral treatments (tablets)

- Steroid tablets (e.g. prednisolone) can be a very effective treatment for Sweet's syndrome. They are usually given at a high dose to start with and then reduced slowly over a number of weeks to prevent the rash coming back.
- Other medications such as <u>dapsone</u>, colchicine, potassium iodide or indomethacin can also be very effective and may be preferred by your doctor if you cannot take steroid tablets because of other medical problems such as diabetes or high blood pressure.
- Immunosuppressive agents such as <u>ciclosporin</u> can reduce the action of the body's own defence system (the immune system) and can also be very useful for treating Sweet's syndrome.

Some of the above medications can have certain side effects and you may require regular blood tests for monitoring while prescribed these treatments.

If an underlying medical problem is found, treating this itself may lead to an improvement in the Sweet's syndrome. Recurrences can occur and therefore treatment may need to be reintroduced until recurrences cease. In rare cases, the condition can persist and treatment would then be given as a maintenance therapy.

Where can I get more information about Sweet's syndrome?

Web links to detailed leaflets and further information:

https://www.dermnetnz.org/topics/acute-febrile-neutrophilic-dermatosis/

http://www.healthline.com/health/sweets-syndrome#Overview1

http://www.pcds.org.uk/clinical-guidance/sweets-syndrome-syn.-acute-febrile-

neutrophilic-dermatosis-gomm-button-dise

http://www.patient.co.uk/doctor/febrile-neutrophilic-dermatosis

https://helpforsweetssyndromeuk.wordpress.com/

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists; individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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