Sweet’s syndrome is a rare skin condition that is often misdiagnosed and may need long-term management. Patients need to be guided to access accurate information.

In this article...

- How Sweet’s syndrome differs from other skin conditions
- How you can do to support people with Sweet’s syndrome
- How misinformation available online can affect patients

Author
Michelle Holder is a former staff nurse at Cardiff and Vale University Health Board, undertakes voluntary work for the British Skin Foundation and runs the Sweet’s Syndrome UK Facebook group.

Abstract

Sweet’s syndrome (SS) is an uncommon inflammatory condition. The main signs and symptoms are fever, malaise and skin lesions. Misdiagnosis is common and biopsy is needed to detect characteristic physiological changes in the upper dermis. Standard treatment is oral corticosteroids but SS can be difficult to manage, with symptoms recurring in at least 30% of cases. Nurses can offer advice and support for patients and their families, and show them where to find accurate information.

Swee’t syndrome (SS), or acute febrile neutrophilic dermatosis, is an uncommon inflammatory condition, first described in 1964 by Robert Sweet (Cohen, 2007). Several hundred cases have been reported worldwide; most occur in women aged 30-50 years but other age groups and men have been affected (Cohen, 2007). There is no accurate information about the incidence of SS in the UK; there is a lack of awareness among health professionals and it is often misdiagnosed (Paydas, 2012).

The main signs and symptoms are fever, malaise, an elevated white-blood-cell count and a range of non-infectious skin lesions (Sweet’s lesions). One of the most common types of lesion is plaques, where small lesions cluster together before spreading out to form distinctive, tender, raised, red/purple areas. Other common lesion types are papules (circular bumps) and nodules (protruding lumps); pustules, blisters and ulcers also occur. Most lesions appear on the face and neck, but they have been found on the torso, as well as upper and lower extremities (Cohen, 2007; Neoh et al, 2007; Zamanian and Ameri, 2007).

What causes Sweet’s syndrome?
The causes of Sweet’s syndrome can be divided into five distinct groups: classical or idiopathic; paraneoplastic (associated with malignancy); autoimmune or inflammatory disease associated; drug induced; and pregnancy related.

Most cases are classical or idiopathic, especially in women. Despite the cause often being unknown, it can be triggered by gastric or upper respiratory tract infection and vaccination, particularly the BCG and the flu vaccine (Cohen, 2007; 2003; Neoh et al, 2007). The condition can also be caused by malignancy, appearing at the same time as the malignancy or up to 11 years before it is diagnosed; up to 20% of cases are malignancy associated. Other triggers are auto-immune disease, such as rheumatoid arthritis, and inflammatory disease, particularly inflammatory bowel diseases (Cohen, 2003).

Drug-induced SS is a rare adverse drug reaction estimated to occur in fewer than 5% of cases; lesions usually appear 5-7 days after drug administration. Drugs involved include granulocyte monocyte-colony-stimulating factor, all-trans retinoic acid, azathioprine, furosemide, tetracyclines and non-steroidal anti-inflammatory drugs (Kluger et al, 2008; Cohen, 2007; 2003). Pregnancy-related SS is rare, accounting for up to 2% of cases (Cohen, 2007; Burtall, 1999).

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Signs and symptoms
Alongside fever, malaise and skin lesions, patients may experience headaches, arthritis and joint pain, muscular pain and conjunctivitis. Less common signs and symptoms – often associated with lesions spreading to other areas – include:
- Recurrent mouth ulcers and lesions;
- Bleeding gums and gum enlargement;
- Blurred vision and deteriorating vision;
- Impaired hearing due to lesion formation in the ear (Cohen, 2007).

In very rare instances, the spread of lesions can lead to organ malfunction and/or multi-organ failure. Central nervous system involvement can cause problems such as encephalitis, meningitis and neuro-Sweet’s disease (Cohen, 2007).

Investigation and diagnosis
Sweet’s syndrome looks similar to a range of other conditions and misdiagnosis is common. It may be misdiagnosed as acne vulgaris, impetigo, rosacea fulminans,
herpes (simplex and zoster), cellulitis, discoid lupus erythematosus, pyoderma gangrenosum, erythema nodosum and erythema multiforme.

Patients complaining of sudden onset of tender-to-painful skin lesions should be investigated. Lesion biopsy and histology showing lots of closely packed neutrophils in the upper dermis and no primary inflammation of the blood or lymph vessels indicates SS. An absence of inflammation is key to diagnosis and can rule out many other conditions.

Blood results should show three of: elevated erythrocyte sedimentation rate; positive C-reactive protein; elevated white-blood-cell count; and neutrophils of >70%. Patients may have fever of higher than 38°C but this may be absent or intermittent, particularly in malignancy-associated SS.

Management

Lesions can disappear without treatment but this may take several months, so most patients require medication. While some only need one course others need repeat treatments in long-term management.

In drug-induced SS, symptoms usually disappear when the offending drug has been stopped (Kluger et al, 2008); pregnancy-related SS tends to resolve without intervention (Burrall, 1999).

Drug treatment

Oral corticosteroids are the standard treatment but intravenous corticosteroids can be used in more serious cases. If lesions are minor and localised, topical corticosteroids or injections into the lesions can be used.

First-choice alternatives to corticosteroids are oral potassium iodide and colchicine. While colchicine is effective, many patients cannot tolerate its gastrointestinal side-effects. Second-choice alternatives are oral indometacin and clofazimine, which are useful when lesions keep returning.

Patient education

Most patients are treated as outpatients. Alongside medication they need education to prevent lesions and help them manage the condition.

A lack of information has led many to rely on online information and support-group websites. Some of this information is not only inaccurate but also dangerous; patients are distressed by what they read or frightened into not taking medication. Patients should be directed to legitimate sources of information such as the British Association of Dermatologists (www.bad.org.uk/site/792/default.aspx) and the British Skin Foundation (www.communityforpeoplewithskintuations: www.british -skinfoundation.healthunlocked.com).

Medication used can have various side-effects and patients may be tempted to stop treatment. As this can lead to relapse and poor management of the condition, they should be encouraged not to stop taking medication without getting medical advice.

Prevention of skin lesions

Sweet’s lesions return in at least 30% of cases (Cohen, 2007). Minor damage to the skin – such as from scratching, venepuncture, injection, biopsy or insect bites – is a causative factor (Cohen, 2003). Patients should be encouraged to take good care of their skin and avoid or minimise activity that might cause damage.

In 2012 a new variant of the condition – necrotising SS – was reported, which mimics necrotising fasciitis (Kroshinsky et al, 2012). Wound debridement (the usual treatment for necrotising fasciitis) must be avoided in people with SS as it can have significant consequences.

It is not advisable for patients with any form of SS to clean lesions with stringent cleansing solutions that could remove tissue as these may trigger the development of further lesions. Patients should treat lesions as gently as possible and try to avoid using adhesive tapes to secure dressings.

Ultraviolet light has been found to trigger lesions, particularly in areas where the skin has been damaged by sunburn (Meyer et al, 2011). Patients should, therefore, be encouraged to use high-factor sun lotion and advised to avoid sunbeds and overexposure to strong sunlight. They should also be encouraged to keep a diary to establish potential trigger factors to avoid or minimise their exposure.

Scarring and milia

Occasionally, Sweet’s lesions cause scarring, which can be distressing; cosmetic camouflage creams can be used to cover scars and have been shown to improve self-confidence (Deshayes, 2009). Patients should be advised to contact their GP or dermatology clinic for information. Some camouflage creams are available on prescription and the charity Changing Faces (www.changingfaces.org.uk) runs free sessions across the UK to teach people how to apply them.

Patients should be encouraged not to pick lesions as this can lead to secondary infection and scarring. Secondary milia (tiny, white, keratin-filled cysts) can appear where lesions have healed. If these are a problem, patients should be advised to contact their GP or dermatology clinic.