

Skin problems in patients with Sjögren's Syndrome

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Sjögren's Syndrome affects many different parts of the body, including the skin. Studies have shown that over half of patients with Sjögren's Syndrome suffer from skin problems related to their Sjögren's Syndrome.

Dry Skin

Dryness is a common problem, often resulting in flaking and itching. Dry skin tends to be worse in cold

weather. Measures to improve symptoms include avoiding contact with detergents, using rubber gloves when washing up or washing clothes, avoiding soap and using pH balanced gels or washes instead. Moisturising the skin is important, non perfumed preparations are sometimes better tolerated by sensitive dry skin.

Rashes

Sensitivity to sunlight

Some patients with Sjögren's Syndrome develop rashes in response to sunlight, these 'photosensitive' rashes are more common in patients who have a certain antibody in their blood (anti-Ro). Often the rash develops on the upper chest, arms and sometimes the face, sometimes it is itchy.

If you suffer from photosensitive rashes it is worth showing your rash to your doctor, either your GP or your Rheumatologist. It may be useful to get someone to take a photograph of the rash in case it fades before the doctor sees it. This may help your doctor to diagnose the rash. Sometimes it is necessary to involve Dermatologists in diagnosing and treating rashes.

There is a characteristic rash associated with Sjögren's Syndrome called 'sub acute cutaneous lupus'. It usually develops in sun exposed areas of skin such as arms and chest, and can appear in circles (the 'annular' form) or form a more diffuse rash ('papulo-squamous' type). Although this rash is called a lupus-type rash, and it can occur in patients with lupus, it is also seen in patients who have Sjögren's Syndrome.

Photosensitive rashes can be avoided by protecting skin from sun exposure by avoiding sunbathing, covering up in the sun, wearing long sleeved clothes and a hat. Using a high factor sunblock lotion is also effective, although these tend to be expensive, it may be possible to obtain sun block on prescription. The medication hydroxychloroquine ('plaquenil') is sometimes used to treat/prevent photosensitive rashes. Troublesome photosensitive rashes sometimes require treatment with cortisone based creams or prednisolone (steroid) tablets.

Rashes due to problems with blood vessels

Another rash which can commonly occur in patients with Sjögren's Syndrome is called 'purpura' or purpuric rash. This usually affects the shins and ankles, small purplish spots appear, sometimes after standing. There are different causes of purpuric rashes in Sjögren's Syndrome. One of the commonest is due to high protein levels (gamma globulins) in the blood (this rash has a long name, Waldenström's hypergammaglobulinaemic

purpura). This rash doesn't usually have any long term complications and usually doesn't require any specific treatment. Treatment with the tablet hydroxychloroquine is sometimes useful, to try and reduce the protein level in the blood, and thus reduce the rash.

Rarely purpuric rashes can be a sign of inflammation in the blood vessels, which is called 'vasculitis'. Sometimes it is necessary to take a sample of skin (a 'biopsy') affected by the rash, to find out if vasculitis is present.



If vasculitis is diagnosed, there is a risk of damage to nerves and kidneys. Therefore treatment with prednisolone (a 'steroid') is used, usually by tablet but sometimes it is given intravenously. Occasionally stronger treatment is needed with 'chemotherapy' treatments such as cyclophosphamide, which is usually given intravenously. Cyclophosphamide is usually given for six months after which treatment with azathioprine is sometimes used.

In summary mild skin problems are frequent and recurrent in patients with Sjögren's Syndrome. Rashes should be diagnosed correctly to ensure that the correct treatment is offered. Serious skin problems occur more rarely and may need treatment with prednisolone or other medications.

Glossary

- Anti Ro antibodies:** A protein (antibody) found in the blood of some patients with Sjögren's Syndrome. The presence of these antibodies may be related to photosensitive rashes.
- Azathioprine:** A treatment (tablet) used to suppress the immune system.
- Cyclophosphamide:** A treatment (tablet or intravenous) used to suppress the immune system.
- Dermatologist:** A doctor who is a specialist in diseases of the skin.
- Gamma globulins:** Proteins in the blood, present in healthy individuals, increased levels detected in some patients with Sjögren's Syndrome.
- Lupus:** An autoimmune disease.
- Photosensitivity:** A reaction in the skin due to exposure to sunlight. Photosensitive rashes are different to sunburn.
- Prednisolone:** A treatment (tablet) used to suppress the immune system.
- Sub acute cutaneous lupus:** A rash, which has two forms, annular, or papulosquamous, found in patients with Sjögren's Syndrome and in patients with lupus.
- Vasculitis:** Inflammation of the blood vessels, usually arteries and small vessels (capillaries).
- Waldenström's hypergammaglobulinaemic purpura:** A rash due to high protein levels (gamma globulins) in the blood, seen sometimes in patients with Sjögren's Syndrome.