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Reticulohistiocytosis

Reticulohistiocytosis is a rare form of [histiocytosis](#) that can affect the skin and other organs.

Reticulohistiocytosis has a wide range of severity. It can be a limited disease with a single yellowish–red skin lump through to multiple skin lesions with internal organ involvement. It usually occurs in adults.

What is solitary reticulohistiocytoma?

Solitary reticulohistiocytoma refers to a single skin lesion (also known as ‘solitary reticulohistiocytosis’ or ‘solitary histiocytoma’). It tends to spontaneously resolve over a period of months to years and affected individuals remain healthy.

Surgical removal of solitary skin lesions usually results in a cure.

Multiple skin lesions

Reticulocytosis may present with multiple skin lesions, in which case there is a high chance of other organs being involved. These may include:

- Heart
- Eyes
- Lungs
- Thyroid
- Liver
- Kidney
- Muscle
- Salivary gland
- Bone marrow

Symptoms may include weight loss and fever. Blood tests may reveal anaemia, and increased cholesterol is present in one third of patients. Heart involvement may be fatal.

What is multicentric reticulohistiocytosis?

Multicentric reticulohistiocytosis is sometimes also referred to as ‘lipoid dermatoarthritis’, ‘lipoid rheumatism’, and ‘giant cell reticulohistiocytosis’.

Multicentric reticulohistiocytosis is a very rare and more aggressive condition characterised by skin lesions, mucosal lesions and arthritis. It usually arises in middle–aged women. In one half of patients the first sign of the disease is arthritis, whilst one quarter first develop skin papules (small bumps) and nodules (larger lumps), and the remainder develop skin and joint manifestations at the same time. It is classified with other proliferative disorders of histiocytes (a type of immune cell).

Feature of skin lesions include:

- Skin–coloured or reddish brown papules and nodules that are 1–2 mm to several centimetres in diameter
- Lesions may occur in isolation or in clusters or crops with a cobblestone appearance

- They most commonly occur on the upper half of the body, especially the face, ears, mucosal surfaces (lips, tongue, gums, nostrils, throat, eyelids), hands and forearms
- Skin lesions may cause destruction of cartilage around the ears and nose resulting in a disfigured appearance
- Multiple tiny lumps around the nails may occur. This has been described as 'coral-beading'.
- Lesions are usually symptomless but about one third of patients complain of pruritus (itching).

Arthritis most commonly involves the fingers, hands, knees and shoulders but any joint can be affected. The symptoms of arthritis may wax and wane, but can rapidly become severe and lead to joint destruction and deformity. About 45% of patients suffer severe arthritis.

Multicentric reticulohistiocytosis



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What is the cause of multicentric reticulohistiocytosis?

The cause of multicentric reticulohistiocytosis is unknown but in about 20–30% of cases, the disease is associated with an underlying malignancy. If there are multiple skin lesions but no arthritis then there is no association with cancer. Controversy exists about whether multicentric reticulohistiocytosis is a true paraneoplastic disorder (a paraneoplastic disorder is one that occurs as a result of circulating factor(s) produced by an underlying cancer). Reasons for this include:

- No consistent type of cancer has been associated with multicentric reticulohistiocytosis;
- Because multicentric reticulohistiocytosis is very rare, this raises the question whether associations are merely coincidental
- A correlation between removal of the cancer and disappearance and/or improvement of arthritic and skin symptoms of multicentric reticulohistiocytosis has not been established.

Multicentric Reticulohistiocytosis is also associated with:

- [Vasculitis](#) (inflammation of the blood vessels)
- Autoimmune diseases

What is the treatment for multicentric reticulohistiocytosis?

There is no specific treatment for multicentric reticulohistiocytosis. In many patients, after an average course of 8 years the disease can go into remission, however, by this time considerable joint destruction may have occurred. Patients are left with crippling, deformed joints and disfigured facial appearance.

Because approximately 30% of cases may stem from an underlying malignancy, evaluation for a possible malignancy is essential. Thorough history taking and physical examination with age-appropriate cancer screening is recommended if symptoms of multicentric reticulohistiocytosis are present.

The following treatments may be used:

- [Methotrexate](#)
- Short courses of [oral corticosteroids](#) (prednisolone/prednisone)
- [Ciclosporin](#)
- [Azathioprine](#)
- [Cyclophosphamide](#)

Related information

References:

- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- [Cutaneous markers of internal malignancy](#)
- [Histiocytoses](#)

Other websites:

- emedicine dermatology, the online textbook
 - [Paraneoplastic Diseases](#)
 - [Muticentric Reticulohistiocytosis](#)

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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