LICHEN MYXEDEMATOSUS

An Autoimmune Dermatological Condition

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Lichen myxedematosus is a chronic autoimmune skin disorder characterized by flesh-colored papules and nodules.

What is Lichen Myxedematosus?

Lichen myxedematosus, which is also called papular mucinosis, is an autoimmune skin disorder that may be confused with lichen planus or localized myxedema. However, although lichen myxedematosus may accompany other autoimmune conditions, it is not associated with autoimmune thyroid disease. Lichen myxedematosus may cause localized eruptions involving small areas of the body or disseminated cases involving multiple body sites. When confluent papules are accompanied by sclerosis or skin hardening the condition is known as scleromyxedema, a condition sometimes referred to as a subtype of lichen myxedematosus.

Who is affected?

Lichen myxedematosus is a rare condition first reported in 1953 that affects males and females equally. It typically occurs in adults between the ages of 30 and 70 years of age. Rarely, it is seen in infants and children.

Symptoms and Disease Course

The primary lesions of lichen myxedematosus are flesh-colored or reddened dome-shaped or flat-topped papules ranging from 2-4 mm in diameter. Often, the lesions coalesce or merge together, forming plaques that may appear linear. Less often, urticarial (hive-like), nodular, or annular lesions are formed.

The skin of the hands, face, elbows, fingers, ears, and toes are most often affected. The mucosa, including the inside of the mouth and nose, are not affected. The coalescence or patchy grouping of papules on the faces, particularly in the area between the eyes above the nose, results in a longitudinal folding, causing a leonine expression. Ophthalmologic symptoms include ectropion and corneal opacities.

Lichen myxedematosus usually persists as a chronic condition and over time its symptoms may become progressively worse. The prognosis is generally worse in patients who develop cardiac disease or myeloma.

Subtypes
The subtypes of localized lichen myxedematosus include:

- A discrete papular form involving any site
- Acral persistent papular mucinosis involving only the extensor surface of the hands and wrists
- Self-healing papular mucinosis in juveniles and adults
- Papular mucinosis of infancy
- Nodular form

Variants are also present that do not meet the criteria for localized lichen myxedematosus or scleromyxedema but fit some of the criteria for each condition.

**Scleromyxedema**

Scleromyxedema, which is sometimes reported as the same disorder as lichen myxedematosus, tends to involve large parts of the body. The skin in scleromyxedema develops reddened, toughened indurations that can reduce mobility of the affected areas, usually the lips, hands, arms, and legs. Systemic manifestations may also occur, including proximal myopathy (muscle inflammation), inflammatory polyarthritis, central nervous system symptoms, esophageal spasms, and hoarseness. Involvement of the internal organs in scleromyxedema may be fatal.

**Diagnosis**

Tissue studies of lesions in lichen myxedematosus show an abnormal paraprotein, usually containing immunoglobulin G-8 (IgG-8) in 90 percent of cases. Bone marrow studies may be normal or they may show increased numbers of plasma cells. Histology studies show a horizontal band of mucinous glycosaminoglycan material situated between the collagen bundles of the upper dermal layer. Increased fibroblasts and dermal fibrosis may also be observed.

The diagnostic criteria for localized lichen myxedematosus includes:

- The localized forms of lichen myxedematosus include:
  - Papular or nodular/plaque eruption
  - Mucin deposition with variable proliferation of fibroblasts
  - The absence of both monoclonal gammopathy and thyroid disease

**Treatment**

Topical ointments are of little value in lichen myxedematosus. Immunosuppressants such as cyclophosphamide and melphalan, either alone or in combination with corticosteroids, have been used successfully. Other medications used for lichen myxedematosus include interferon-alpha, cyclosporine, PUVA photochemotherapy, electron-beam therapy, intravenous immunoglobulin therapy, and dermabrasion.
Resources:

Elizabeth Liotta, Lichen Myxedematosus, eMedicine, Sept 2006, accessed June 1, 2007,


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