

## Review Article

# Cutaneous mucinoses: an overview

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**Abstract** The cutaneous mucinoses are a group of connective tissue disorders characterized by the deposition of mucin, either focally or diffusely, in the interstices of the dermis. The diseases may occur as a primary (metabolic) event or as a secondary (catabolic) process in certain dermatoses such as lupus erythematosus and dermatomyositis. Systemic abnormalities are seen with most of these disorders. A brief review and classification of the disorders included in this group is given here.

**Key words**

Cutaneous mucinoses, dermal mucinoses, mucin, connective tissue disorders.

The cutaneous mucinoses are a heterogeneous group of diseases so named because of the variable amount of mucin accumulation in the skin or within the hair follicle. Mucin is jelly-like acid mucopolysaccharide (glycosaminoglycan) of the ground substances and probably plays a role in extravascular exchange of metabolites. It is produced by fibroblasts and is composed of hyaluronic acid bound to heparin and chondroitin sulphate.<sup>1,2</sup> We divide the cutaneous mucinoses into two groups: the distinctive cutaneous mucinoses in which the mucin deposit is a distinctive histopathologic feature that manifests as a clinically specific lesion (also called the metabolic or primary group), and the diseases associated with histopathologic mucin deposition as an additional finding (also called the catabolic or secondary group). The former are further divided into degenerative-inflammatory mucinoses,

which may be either dermal or follicular, and into neoplastic-hamartomatous mucinoses. Histopathologic diagnosis is particularly difficult for dermal mucinoses and requires clinicopathologic correlation. Three histologic clues, namely the pattern of mucin distribution (diffuse or focal), the level of mucin deposit in the dermis and some additional findings may help diagnosis. Follicular mucinoses have the easiest pattern to recognize histologically. Neoplastic-hamartomatous cutaneous mucinoses include mucinous nevus, a benign hamartoma, and myxoma, which is a benign tumor to be differentiated from reactive cutaneous focal mucinosis.<sup>1-4</sup> Histologically, mucins are stained with alcian blue at pH 2.5 (blue), mucicarmine (red), colloidal iron (blue green) and the colouration depends on the number and nature of the acid groups. They are also stained metachromatically with toluidine blue, methylene blue and thionine.<sup>5,6</sup> Periodic acid-schiff (PAS) stains heparin but not hyaluronic acid. On the basis of histological site and pattern of mucin deposition and extent of involvement, cutaneous mucinoses may also be divided into focal, follicular and

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diffuse. Focal mucinoses are localized benign forms that are usually not associated with other organ involvement. Follicular mucinosis is generally considered with cutaneous lymphomas. Diffuse cutaneous mucinoses differ from local mucinoses by greater cutaneous involvement, systemic-organ dysfunction from mucin deposition and associated extra-cutaneous disease not related to mucin deposition. The full description of clinicopathological and diagnostic features in case of individual disorders is beyond the scope of this article, therefore, only the major mucinoses are listed and classified below and only treatment aspect has been touched briefly in the end.

### **Clinicopathological classification**<sup>3,4,7-10</sup>

#### **A. Primary (metabolic) mucinoses**

These include cutaneous mucinoses in which the mucin deposit is a distinctive histopathologic feature that manifests as a clinically specific lesion and occurring as a primary pathology. This group is further divided into diffuse, focal and follicular on the basis of histological pattern and extent of involvement.

(i) *Diffuse* These generally comprise of disorders with diffuse cutaneous involvement associated with variable systemic organ involvement such as endocrine system.

- Generalized myxedema
- Pretibial myxedema
- Reticular erythematous mucinosis
- Scleredema
- Scleromyxedema
- Diffuse papular mucinosis
- Lichen myxedematosus

- Self-healing juvenile cutaneous mucinosis
- Cutaneous mucinosis of infancy
- Papular and nodular mucinosis associated with lupus erythematosus (tumid LE)
- Papular mucinosis of the toxic oil syndrome

#### **(ii) Focal**

Focal mucinoses are localized benign forms and generally, are not associated with other organ involvement

- Acral persistent papular mucinosis
- Cutaneous focal mucinosis
- Oral focal mucinosis
- Digital mucous cyst
- Mucocele
- Cutaneous myxoma
- Nevus mucinosis (Hunter syndrome)

#### **(iii) Follicular forms**

These are characterized clinically by infiltrated scaly plaques with loss of hair and histologically by mucin deposition in sebaceous glands and the outer root sheath of hair follicles. The cases may or may not be associated with lymphomas.

- Alopecia mucinosa (follicular mucinosis)
- Urticaria-like follicular mucinosis

#### **B. Secondary (catabolic) mucinoses**

This group includes the diseases in which dermal mucinosis may, to a greater or lesser extent, be associated with otherwise typical histopathological features of that disease.

- Lupus erythematosus
- Dermatomyositis
- Granuloma annulare
- Jessner's lymphocytic infiltrate

- Degos disease (malignant atrophic papulosis)
- Hereditary progressive mucinous histiocytosis
- Papular mucinosis in L-tryptophan-induced eosinophilia-myalgia syndrome
- Mucinosis accompanying mesenchymal and neural tumours
- Mucopolysaccharidoses

### Treatment<sup>11-16</sup>

Treatment of most of the cutaneous mucinoses remains unsatisfactory and no uniformly effective therapy exists, although several treatments are routinely used. Topical therapy is generally of no benefit. There have been occasional reports of spontaneous resolution of localized lesions. Clearance of lesions has been reported with melphalan and cyclophosphamide alone or in combination with prednisone. Both isotretinoin and etretinate have been associated with improvement. Interferon-alpha, cyclosporine, PUVA photochemotherapy, electron-beam therapy, IVIg, and dermabrasion have also been attempted with variable success. Isolated cases document the beneficial responses of dapsone, indomethacin, and interferons. Pulsed dye laser has been found successful in treating reticular erythematous mucinosis in one report. The overall prognosis for extensive disease is poor. Because of the variable course of the disease and the likelihood of spontaneous resolution, therapeutic efficacy is difficult to prove.

### Conclusion

The cutaneous mucinoses are a complex group of dermatologic diseases with local, follicular, or diffuse disease. The diffuse cutaneous mucinoses are remarkable not only for their dermal disease, but also for the numerous systemic manifestations. Because of the variability of associated systemic manifestations, some with substantial morbidity and mortality, it is important for the clinical dermatologist to be able accurately to diagnose and differentiate various diffuse cutaneous mucinoses.

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