

## Focal mucinosis: clinical and histological features of an unusual condition

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### Abstract

Mucinoses constitute a diverse group of disorders that have in common the deposition of basophilic, finely granular and stringy material in the connective tissues of the dermis (known as dermal mucinoses); this also can occur in the hair follicles (follicular mucinoses) and finally in the epidermis (epithelial mucinoses). We present the clinical and histological findings of a case of focal mucinosis in a 76-year-old man who was otherwise fit and healthy. A literature review revealed a sparse number of such cases none of which was reported in the ENT literature. Focal cutaneous mucinosis is a benign condition but the authors believe it is of local and systemic interest to the Otolaryngologist as an addition to his or her armentarium of diagnoses.

**Key words:** Mucinosis; Nasal Cavity

### Introduction

Mucinoses constitute a diverse group of disorders that have in common the deposition of basophilic, finely granular and stringy material in the connective tissues of the dermis (known as dermal mucinoses); this also can occur in the hair follicles (follicular mucinoses) and finally in the epidermis (epithelial mucinoses).<sup>1</sup> Characteristics of cutaneous focal mucinosis vary from smooth elevated nodules to verrucous lesions. These lesions tend to persist but do not recur following superficial excision.<sup>2</sup> Medline and PubMed literature searches revealed no cases of cutaneous focal mucinosis in the ENT literature. We describe such a case.

### Case report

A 76-year-old gentleman was referred by the Dermatologists to the department of Otolaryngology. He presented with a lesion arising from the midline of the nasal columella. This asymptomatic lesion had been present for six months but recently had increased in size.

On examination there was an obvious non-tender sessile skin-covered mass arising from the columella (Figure 1). There were no further lesions found and no other significant ENT abnormalities noted.

The following week the patient underwent excision of this lesion under local anaesthetic. He subsequently made a good post-operative recovery.

The resected specimen consisted of an ellipse of skin bearing a firm nodule measuring  $0.8 \times 0.5$  cm with a height of 0.6 cm. Microscopic examination revealed the nodule to be composed of an ill-defined dermal collection of acid mucin (hyaluronic acid) in which there were scattered small spindle cells. A few mature fat cells, collagen fibres, blood vessels and eccrine ducts were incorporated in the mucinous area (Figure 2). A diagnosis of focal cutaneous mucinosis was made.

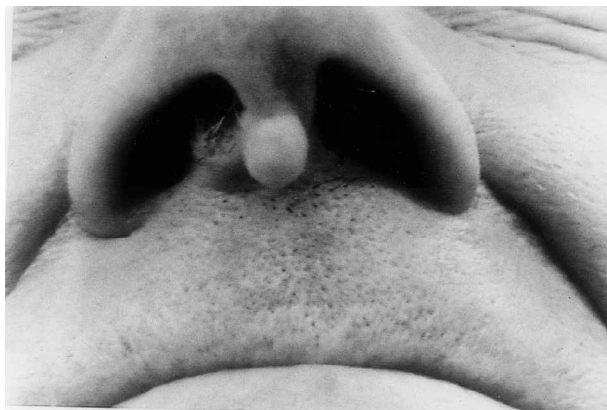


FIG. 1

A single sessile lesion arising from the nasal columella. Such a lesion is typical of focal cutaneous mucinosis.

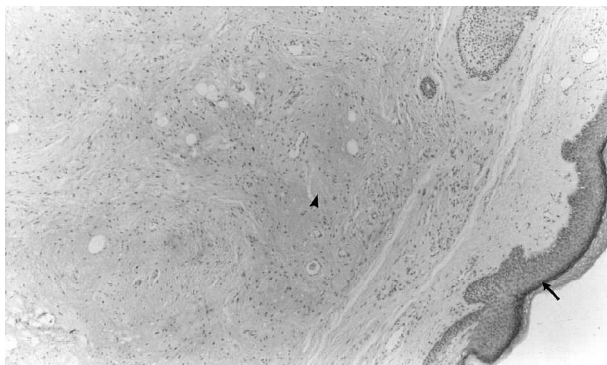


FIG. 2

Photomicrograph of the excised nodule. Normal epidermis (black arrowhead) and dermal mucin (black arrow) can be clearly seen (Staining with Alcian blue pH 8).  $\times 40$ .

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## Discussion

Cutaneous focal mucinosis was first accepted as a distinct clinical entity in 1966.<sup>2</sup> The mucinoses are currently regarded as a very diverse group of disorders divided into three categories: dermal mucinosis, follicular mucinosis and epithelial mucinosis.<sup>3</sup> Some authors believe mucopolysaccharidosis to be another category of cutaneous mucinosis.<sup>1</sup> Dermal mucinoses are of particular interest; they have considerable quantities of glycosaminoglycans (acid mucopolysaccharides) within the dermis. Focal cutaneous mucinosis is included in this group. The pathogenesis of this mucin accumulation in the skin remains poorly understood.

Focal cutaneous mucinosis presents as a solitary flesh-coloured nodule most commonly found on the face.<sup>4</sup> Occurrence on the trunk and mid-extremities has been reported.<sup>5</sup> Multiple lesions have been reported in two cases: the first in a patient with hypothyroidism<sup>6</sup> and the second, an otherwise healthy patient who presented with multiple nodules on the right leg.<sup>7</sup> All of the reported cases occurred in adult patients without gender bias. Malignancy was not identified in any case nor recurrence reported following surgical removal in any case.<sup>2</sup>

In our case the differential diagnosis included:

- (1) cutaneous myxoma (which is usually less cellular with minimal vascularity),
- (2) myxoid neurofibroma,
- (3) dermal nerve sheath myxoma (neurothekeoma) or
- (4) leioma

All of the above were excluded by negativity with appropriate immunocytochemical stains (S100 protein, smooth muscle actin, and desmin). The adipocytic component was insufficient for a diagnosis of myxolipoma.

Focal cutaneous mucinosis therefore appears to be a benign condition with unknown aetiology. Jacobovic described a case associated with hypothyroidism that resolved spontaneously following correction of thyroid hormone levels, the authors therefore suggested monitoring thyroid hormone levels in this condition.<sup>6</sup> The association with thyroid disease was further supported by Schuppli *et al.* when he described a female with normal T<sub>3</sub> and T<sub>4</sub> levels but with high titres of antithyroid antibodies.<sup>8</sup>

Other authors believe focal mucinosis to be solely reactive. Others have suggested that focal mucinosis is the result of a dysfunction of fibroblasts in a localized area.<sup>5</sup> It is thought that increased amounts of hyaluronic acid are produced by fibroblasts at the expense of the connective tissue elements.<sup>1</sup> The resolution of lesions spontaneously and by surgical excision, both total and partial (biopsy), indicated a benign reactive myxomatous alteration of connective tissue elements.<sup>7</sup>

Focal cutaneous mucinosis is a rare benign condition which can be associated with systemic disease. It is successfully treated by surgical excision.

- **This is a single case report of mucinosis presenting on the nasal columella**
- **The authors state that no previous accounts of mucinosis have been reported in the otolaryngology literature**
- **The differential diagnosis and pathophysiology are discussed**

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Miss A.-L. McDermott takes responsibility for the integrity of the content of the paper.  
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