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Case Report

Mucinous Naevus: A Rare Variant of Connective Tissue Naevus

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Abstract

Mucinous naevus is a rare benign hamartomatous lesion which has been categorized as a variant of both cutaneous mucinosis and connective tissue neavus. These lesions commonly involve the trunk and extremities, appearing as papulonodular or plaque-like lesions. We present a 22 year old boy with congenital mucinous nevus presenting with mole-like lesion on the face.

INTRODUCTION

Mucinous naevus (MN) is a type of benign hamartomatous cutaneous mucinoses characterized by deposition of excessive amounts of mucin in the superficial dermis in a diffuse pattern with absence of collagen and elastic fibres in the mucinous areas. It may be associated with features of connective tissue naevus of proteoglycan type [1]. There is paucity of literature regarding this interesting entity. We report a case of this rare benign hamartoma in a 22 year old boy who presented with a mole-like pigmented lesion on the right cheek since birth. An extensive review of literature is being discussed herewith.

CASE REPORT

A 22 year old boy presented with a single linear raised brown pigmented lesion on the right side of the face since birth. The patient had no relevant systemic abnormalities. Physical examination revealed a single linear brown pigmented plaque measuring 2x1cm over his right cheek. Routine haemotological and biochemical investigations were normal. A clinical diagnosis of mole was proffered. The lesion was excised and sent for histopathological examination. Biopsy revealed a hyperkeratotic epidermis with elongated rete ridges overlying papillary and reticular dermis containing clusters of nevus cells separated by widely spaced collagen fibers and increased mucin deposition (Figure 1). The mucin was positive for Alcian blue at pH 2.5 and toluidine blue (Figure 2a and b) and negative for mucicarmine (Figure 2c) and periodic-acid Schiff (PAS). Van Gieson stain revealed absence of elastic fibres in the dermis (Figure 3). A diagnosis of mucinous naevus was rendered.

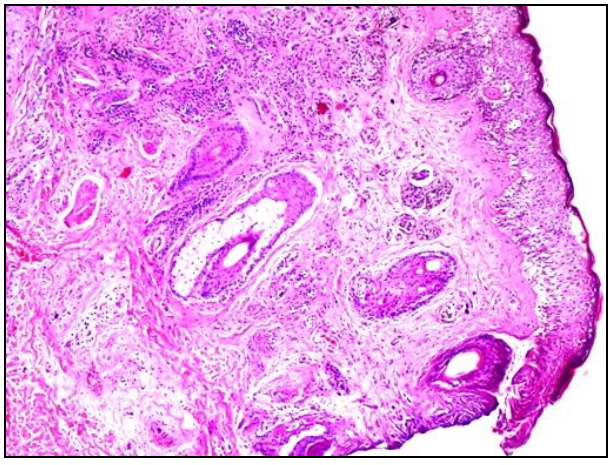

Figure 1. Papillary and reticular dermis showing clusters of nevus cells and mucin deposition. (H&E, x100)

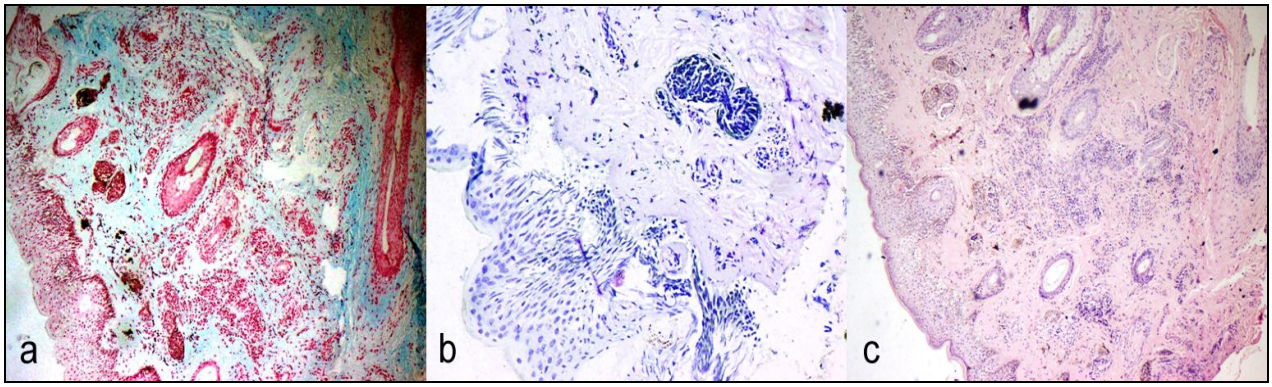

Figure 2. Stains for dermal mucin: a) Alcian blue at pH 2.5 positive b) Toluidine blue positive c) Mucicarmine negative (Histochemistry, x40 for all panel).

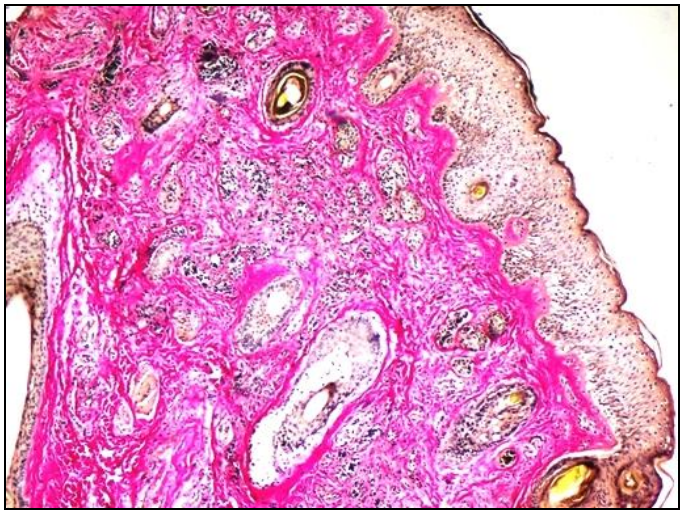

Figure 3. Absence of elastic fibers in the dermis (van Gieson, x200)

DISCUSSION

Mucinous naevus is a neoplastic hamartoma and is a variant of primary cutaneous mucinoses. Recently, it has been classified as a variant of connective tissue nevus (CTN) of the proteoglycan type. The lower back is the most common affected site and these lesions can be present at birth or develop later in early adulthood with a male preponderance [1-3]. A case of familial MN was reported by Chen et al. in a 9 year old child and his younger brother [4].

The source of mucin in these lesions is unclear; however, it has been proposed that abnormal fibroblasts produce excessive mucin in the dermis [5]. Later, Tardio proposed CD34 positive fibroblasts and scarce factor XIIIa-positive dendritic cells in the cellular component of MN [6].

Cutaneous mucinoses (CM) is a heterogenous group of disorder which is characterized by abnormal deposition of mucin (dermal/epithelial) in the skin. The mucin may be dermal or epithelial. Dermal mucin, produced by fibroblasts, consists of acid glycosaminoglycans, which stains with Alcian blue at pH 2.5 and metachromatically with toluidine blue but is PAS negative. Epithelial mucin, on the other hand present in eccrine and apocrine glands consists of both neutral and acid glycosamines and stains positive with Alcian blue at pH 2.5 and PAS but negative for toluidine blue [1].

Mucinous naevus has also been described as a variant of connective tissue naevi (CTN). They are categorized as naevi of reticular connective tissue and naevi of adventitial connective tissue. CTN composed of

predominantly proteoglycans are extremely rare [2,3].

Redondo Bellon et al. were the first to describe MN in 1993, in a 16 year old girl presenting with a congenital plaque-like lesion in the interscapular area demonstrating a naevoid appearance and deposition of mucin in the superficial dermis [7].

Rongioletti et al proposed that congenital or acquired lesions with nevoid features i.e. unilateral and/or linear or dermatomal pattern should be termed as MN. MN without epidermal nevus like changes should be classified as CTN of the proteoglycan type while with epidermal changes, as combined epidermal-CTN of proteoglycan type [8].

All reported cases of MN in the literature have been documented in the trunk or extremities. They typically present as coalescing papules, nodules or plaques arranged in unilateral, linear or zosteriform pattern. They may be mimic epidermal naevus clinically [9]. The present case is the first to present with a pigmented linear plaque on the face which is an unusual site and clinically diagnosed as a mole. Microscopically, Alcian blue and toluidine blue positive mucin deposition was seen in the papillary and reticular dermis similar to those reported in the literature. Depletion of elastic fibres was observed in the dermis.

This case was differentiated from other types of primary CM based on clinicopathologic features and histopathology (Table 1) [1,5,10].

Table 1. Differential diagnosis of Mucinous naevus [1,5,10].

	Age of onset	Pattern and level of mucin deposition	Additional features
Degenerative-inflammatory mucinoses			
Lichen myxedematosus (LM)/papular mucinoses- generalized/ localized	Middle age (30-50 years)	Focal/ diffuse in upper and mid reticular dermis	Marked proliferation of fibroblasts, increased collagen, follicular atrophy, systemic manifestations, monoclonal gammopathy
Reticular erythematous mucinosis	Middle aged	Diffuse in upper reticular dermis	Dermal vascular dilation, perivascular and perifollicular T lymphocytic infiltrate, photoaggravated, direct immunoflouroscence-negative
Self-healing juvenile CM	Children	Diffuse in upper reticular dermis	Acute eruption and spontaneous healing, mild increase in fibroblasts and mast cells, arthritis
Acral persistent papular mucinosis	Adults	Focal in upper reticular dermis	Variant of localized LM, sparse collagen fibres, absence of fibroblastic proliferation
Cutaneous mucinosis of infancy	Congenital/early infancy	Focal in papillary dermis (hugged by epidermis)	Dermal perivascular mononuclear infiltrate, symmetrical/densely grouped papules.
Cutaneous focal mucinosis	Adults	Focal , filling the whole dermis	Solitary lesion, histologic diagnosis, thin collagen fibres, absence of elastic and reticulum fibres.
Follicular mucinosis (FM) – Pinkus FM/ urticaria-like FM	Children and Adults	Focal, confined to hair follicles	Disconnection of keratinocytes in the hair follicular epithelium and sebaceous glands by mucin followed by formation of cystic spaces, perifollicular and perivascular inflammation
Neoplastic/hamartomatous mucinoses			
Angiomyxoma	Adults	Dermis and subcutis	Trichoblastic like-epithelial features, increased vascularity, microhaemorrhage, inflammation, associated with Carneys complex

Secondary mucinoses is divided into epithelial mucinosis, dermal mucinosis and follicular mucinosis. Epithelial mucinosis is seen in skin cancers, mycosis fungoides, warts, seborrheic keratosis and keratoacanthoma. Dermal mucinosis is associated with various conditions such as granuloma annulare, lupus

erythematosus along with epithelial, mesenchymal and neural tumors. Follicular mucinosis is seen in sarcoidosis, lymphomas, leukemia and sometimes in lupus erythematosus and seborrheic keratosis as well [1]. Normal physical examination and routine investigations, diagnostic histologic features with PAS and mucicarmine negative mucin in the dermis in this case ruled out the possibility of secondary mucinoses.

In MN, lesions which are symptomatic or cosmetically disfiguring require surgical excision. Carbon dioxide laser vaporization has emerged as a treatment option for MN of combined epidermal-CTN of the proteoglycan type. These lesions usually do not recur following treatment [5].

To conclude, mucinous naevus is a hamartoma and a form of primary CM and CTN with deposition of dermal type of mucin in the dermis. These lesions may present as papulonodules or plaques, commonly affecting the trunk and can be treated by simple surgical excision. It is imperative to differentiate them from other variants of primary and secondary CM. This case is being reported because of the unusual location and rarity of this lesion.

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