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Case report and review of solitary cutaneous focal mucinosis: a unique primary cutaneous mucinosis unrelated to mucinosis-associated systemic diseases

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Abstract

Localized deposition of mucin in the upper dermis is referred to as cutaneous focal mucinosis. Patients with this condition either present with a single skin lesion (solitary cutaneous focal mucinosis) or numerous skin lesions (multiple cutaneous focal mucinosis). A man with solitary cutaneous focal mucinosis is described and the features of this condition are reviewed. Solitary cutaneous focal mucinosis has a slight male predominance and typically presents in adults, ranging in age from 29 years to 60 years, as a nodule or papule that is flesh-colored or white and most commonly located on an extremity or the trunk. Microscopic examination shows deposition of mucin in the upper dermis; the overlying epidermis can be normal, atrophic or hyperplastic. The skin lesion is often removed at the time of biopsy. However, recurrence has not been observed when the mucin deposition is present at the edge of the biopsy or excision specimen. Although the pathogenesis of this condition remains to be established, in contrast to individuals with multiple cutaneous focal mucinosis, solitary cutaneous focal mucinosis is a unique primary cutaneous mucinosis unrelated to mucinosis-associated systemic diseases.

Keywords: cutaneous, disease, focal, mucinosis, multiple, primary, solitary, systemic

Introduction

Cutaneous mucinosis are conditions characterized by increased mucin deposition within the skin. They have been classified as either primary (in which the distinctive clinical lesions that occur result from the abnormal amount of mucin that is present) or secondary (in which the mucin is an additional feature associated with another systemic condition or tumor). In addition, primary cutaneous mucinosis have been further subtyped as neoplastic and hamartomatous or degenerative and inflammatory; the latter subtype is further differentiated depending on whether the location of the mucin is dermal or follicular [1].

Cutaneous focal mucinosis is a dermal degenerative primary mucinosis. The term was coined by Johnson and Helwig in 1966 to describe the solitary lesion. However, since 1961, the authors had used the designation focal mucinosis when referring to this entity [2]. Chen et al. subsequently suggested that the term cutaneous focal mucinosis be changed to a solitary soft fibroma-like polypoid mucinosis in 2004. Although their proposed name for this condition provided an excellent descriptive term, it was too redundant for practical use and did not achieve general acceptance [3]. More recently, in 2016, Kuo et al. suggested that this skin lesion be referred to as solitary cutaneous focal mucinosis. They emphasized that the new nomenclature would differentiate patients with a single skin lesion from those individuals with more than one skin lesion who were



Figure 1. Clinical presentation of solitary cutaneous focal mucinosis appearing as a flesh-colored papule on the back. **A)** Distant, and **B)** closer views of an asymptomatic flesh-colored papule (circled with purple ink) on the mid back of a 63-year-old man.

also being described to have cutaneous focal mucinosis [4].

Solitary cutaneous focal mucinosis is a reactive benign process that has not typically been associated with systemic conditions [2-11]. In contrast, various medical conditions have been found in patients with diffuse mucinosis or multiple cutaneous focal mucinosis [12-16]. A man with solitary cutaneous focal mucinosis—without any mucinosis-related disease—is described and the features of this lesion are reviewed. In contrast to individuals with cutaneous focal mucinosis who have multiple skin lesions, laboratory evaluation for

mucinosis-associated systemic conditions is usually not necessary for patients with only a solitary lesion.

Case Synopsis

A 63-year-old healthy man presented for evaluation of pruritic rashes on his right upper eyelid, chest, and abdomen of four months' duration. There had been no improvement with twice daily topical application of nystatin cream (100,000 units per gram). He had no history of skin cancer.

A complete, head to toe, cutaneous examination was performed. There was lichenification with erythema and scaling on his right upper eyelid. Erythematous dermal plaques were present on his right chest and lower abdomen. An asymptomatic, 2×2 millimeter flesh-colored papule was noted on his mid back. He had not been aware of the papule and there were no additional, similar-appearing, lesions on his body (**Figure 1**).

Systemic treatment with oral prednisone (60 milligrams daily for seven days, followed by 40 milligrams daily for five days and then 20 milligrams for three days) was given to treat his dermatitis. In addition, twice daily topical application for seven days was started with 2.5 percent hydrocortisone cream for his upper eyelid and 0.5 percent betamethasone cream for his chest and abdomen.

The suspected differential diagnosis of the mid back lesion included a basal cell carcinoma, nevus, or seborrheic keratosis. The lesion was circled with purple ink (**Figure 1**). An excisional shave biopsy was performed.

Microscopic examination of the hematoxylin and eosin stained biopsy specimen showed a focal, homogenous and basophilic appearing, deposition, consistent with mucin, in the upper dermis; it was associated with a loose stroma and stellate fibroblasts. The overlying epidermis showed compact orthokeratosis, normal thickness, and regular elongation of the rete ridges. The dermal deposition appeared dark blue after colloidal staining of the specimen, confirming that it was mucin. The lesion did not extend to the margins of the specimen (**Figure 2**).

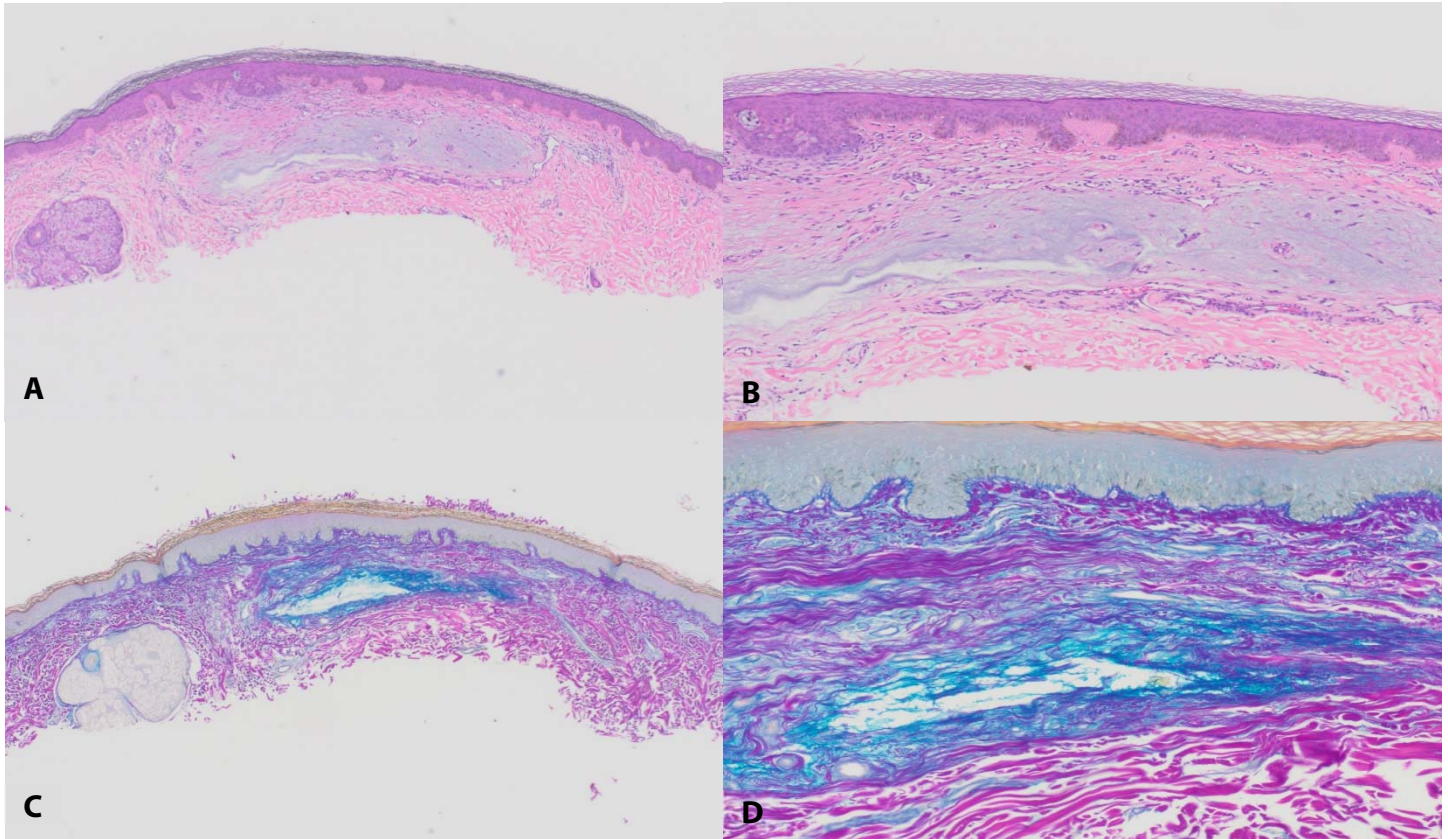


Figure 2. Microscopic examination of solitary cutaneous focal mucinosis after staining with hematoxylin and eosin or colloidal iron. **A)** Distant, and **B)** closer views of the hematoxylin and eosin stained tissue show homogenous slightly basophilic-appearing collection of mucin that extends from the upper dermis to the mid dermis between the collagen bundles. **C)** Distant, and **D)** closer views of the colloidal iron-stained tissue shows strongly positive blue staining of the amorphous material in the dermis thereby confirming that it is composed of mucin. H&E, **A)** 4x; **B)** 10x; colloidal iron, **C)** 4x, **D)** 20x.

Correlation of the clinical morphology and the pathologic findings established the diagnosis of solitary cutaneous focal mucinosis. He returned for follow-up after two weeks. The biopsy site had healed and the dermatitis on his eyelid, chest, and abdomen had completely resolved.

Additional laboratory investigation was done. The following studies were either normal or negative: complete blood cell counts and platelet count, serum chemistries (including a fasting blood sugar), hemoglobin A1c, thyroid stimulating hormone, total thyroxine, total triiodothyronine, thyroid peroxidase antibodies, thyroglobulin antibodies, serum protein electrophoresis, antinuclear antibody, double strand deoxyribonucleic acid antibody, Sjogren syndrome A antibody, Sjogren syndrome B antibody, Smith antibody, ribonucleic protein antibody, and scleroderma 70 antibody. He was again seen for a

follow-up skin check six months later; there was neither recurrence of the solitary cutaneous focal mucinosis on his mid back nor the appearance of any new skin lesions.

Case Discussion

Solitary cutaneous focal mucinosis has been described in case reports of individual patients [3,5-8]. It has also been reported in large studies of individuals with this lesion (**Table 1**), [2,4,9-11]. Many of these latter investigations originate from retrospective review of patient features provided with the pathology requisitions; in some studies additional follow up information was also secured [2,4,9]. One of the studies summarized the observations from not only patients with solitary cutaneous focal mucinosis but also several

Table 1. Characteristics of patients from retrospective studies with solitary cutaneous focal mucinosis.

| Pub Year; Reference ^a | 1966; [2] | 1994; [9] | 2017; [4] | 2018; [10] | Total |
|----------------------------------|----------------------------------|------------------------|-------------------------|-------------------------|--------------------------|
| Cases ^b | 14 | 11 | 11 | 98 | 134 |
| Race | Caucasian | German | Taiwanese | American | Variable |
| Men:Women | 9:5 | 6:5 | 7:4 | 56:42 | 78:56 |
| Age range; Mn/Md | 6-49 yrs ^c ; 36yrs/NS | 39-61 yrs; 51yrs/50yrs | 36-66 yrs; 52yrs/50yrs | 25-86 yrs; 55yrs/NS | 6-86 yrs; 52yrs/50yrs |
| Duration | 8mon-20yrs | Not stated | Mon-yrs | Not stated | Mon-yrs |
| # of lesions | 1 | 1 | 1 | 1 | 1 |
| Symptoms | None | None | None | None | None |
| Size | 4-8 mm | Not stated | 2-18 mm | Not stated | 2-18 mm |
| Color | Flesh-color; white | Not stated | Flesh-color; red; white | Not stated | Flesh-color; red; white |
| Morphology | Nodule; Papule | Not stated | Nodule; Poly papule | Not stated | Nodule; Papule |
| Site | Ext=6; Trunk=3; H&N=5 | Ext=4; Trunk=3; H&N=4 | Ext=5; Trunk=4; H&N=2 | Ext=59; Trunk=33; H&N=6 | Ext=74; Trunk=43; H&N=17 |
| AssocDz | HypoT=1 ^d | None | None | None | None |
| Treatment | Ex=12; Bx & curet; Ed&c, bx=1 | Not stated | Ex=11 | Not stated | Ex=23; Other=2 |
| Recurrence ^e | 0% (0/11) | 0% (0/8) | 0% (0/11) | Not stated | 0% (0/30) |

Abbreviations: AssocDz, associated disease; Bx, biopsy; curet, curettage; Ed&c, electrodesiccation and curettage; Ex, excision; Ext, extremities; HypoT, hypothyroid; H&N, head and neck; Md, median; mm, millimeters; Mn, mean; Mon, months; NS, not stated; Poly, polypoid; Pub, publication; yrs, years; #, number; % percent; -, to.

^aThe information is summarized from retrospective studies of patients with solitary cutaneous focal mucinosis.

^bA report of a 50-year-old Japanese man with solitary cutaneous focal mucinosis and review of an additional 54 patients with cutaneous focal mucinosis from the Japanese literature from 1982 to 2004. The patients either had a solitary lesion (40 patients), or two or three lesions (3 patients) or multiple lesions (12 patients). The characteristics of the patients are all combined without differentiating which features were associated with the individuals with solitary lesions versus those in patients with more than one cutaneous focal mucinosis lesion [11].

^cThe study included one girl who was 6 years old; the other patients were adults ranging in age from 29 years to 49 years; their mean age was 38 years [2].

^dOne of the patients had a history of hypothyroidism; however, no additional details were provided regarding the temporal relationship between the onset of thyroid disease and the diagnosis of solitary cutaneous focal mucinosis [2].

^eRecurrence refers to the percentage of recurrence and the number of recurrences per number of patients with follow-up.

individuals with multiple cutaneous focal mucinosis lesions [11].

Solitary cutaneous focal mucinosis has been reported in at least 182 individuals. The ratio of men to women with solitary cutaneous focal mucinosis is 1.4 to 1.0 [2-11]. Most patients (82 percent, 36 of 44) range in age from 29 years to 60 years at the time of diagnosis [2-9]. However, one Caucasian girl was six years old and one Chinese girl was 12 years old [2,5].

All the patients with solitary cutaneous focal mucinosis only had a single skin lesion. Johnson and Helwig mentioned that one of their patients had a similar-appearing lesion [2]. However, the diagnosis

of the additional lesion was not confirmed by biopsy; hence, it may not have actually been a cutaneous focal mucinosis lesion [2].

Solitary cutaneous focal mucinosis usually presents as an asymptomatic papule [2,4]. Indeed, similar to the reported patient, other individuals were also not aware that the lesion was present [4]. However, a 30-year-old woman with trauma-induced solitary cutaneous focal mucinosis that developed after blunt injury to the site and had been present for three years observed recurring mucinous discharge from the lesion for two months prior to biopsy diagnosis and subsequent planned excision [6].

Solitary cutaneous focal mucinosis typically appears flesh-colored [2-4,7,8]. It has also been observed to be white or red [2,4]. However, albeit less common, the color of the lesion has appeared to exhibit a slight erythema at the base, hyperpigmentation with a light brownish center, erythema with hyperpigmentation, or white color with a blueish hue [2,5-7].

The size of the lesion has ranged from two-by-two millimeters to two-by-four centimeters [4,6]. However, most lesions are smaller than ten-by-ten millimeters. An increase in the size of the lesion was noticed by one patient, a 12-year-old Chinese girl whose right chin solitary cutaneous focal mucinosis began as a small papule and enlarged to a one-by-one-centimeter plaque during a period of one year [5].

The entity usually appears as a papule or nodule [2,4,5,7,8]. However, it has presented as a firm plaque or a soft polypoid fibroma-like lesion [3,4,6]. Rarely, the surface of the lesion was verrucous [2,11].

The extremities were the most common location of solitary cutaneous focal mucinosis [8,11]. More than half the lesions (55 percent, 74 of 134) were located on the extremities of solitary cutaneous focal mucinosis patients described in studies; the arm (59 percent, 44 of 74) was more common than the leg (41 percent, 30 of 74), [2,4,9,10]. The trunk (32 percent, 43 of 134) and the head and neck (13 percent, 17 of 134) were other solitary cutaneous focal mucinosis sites [2,4,9,10]. Post-traumatic solitary cutaneous focal mucinosis was observed on the areola of two men following either folliculitis and laser epilation or removal of nipple ring (implanted three years previously by piercing) at this location [7].

Dermoscopic observations of solitary cutaneous focal mucinosis have recently been described in a 55-year-old man with an asymptomatic flesh-colored eight-millimeter nodule of long duration on his back and a 61-year-old woman with an asymptomatic flesh-colored six-millimeter nodule on her left arm. Both lesions showed a non-specific homogenous whitish pattern. In addition, the lesion on the man's back exhibited a sharply demarcated yellow border. The investigators postulated that the nonspecific

homogenous whitish pattern that they observed under polarized dermoscopy of the solitary cutaneous focal mucinosis lesion was caused by either a decreased number of melanocytes in the epidermis above the dermal mucin or the birefringent properties of mucous, or both [8].

The principal pathology feature of solitary cutaneous focal mucinosis is the deposition of mucin in the upper dermis. The mucinous change, which appears lightly basophilic-staining on hematoxylin and eosin-stained sections, can extend into the deeper dermis and rarely into the superficial subcutaneous fat [1-4,7-9]. Several stains (including alcian blue, colloidal iron, and toluidine blue) have been used to confirm that the homogenous-appearing mucinous material in the dermis is hyaluronic acid; glycogen is absent since the material does not stain with periodic acid-Schiff (PAS) stain [1-9,11].

The mucin is localized in the dermis but it is not encapsulated. Indeed, the borders are not well delineated and its peripheral edges blend gradually into the surrounding normal connective tissue. Scattered fibroblasts are present within the mucin. However, other connective tissue elements such as collagen fibers, elastic fibers, and reticulum fibers are diminished [1-9,11].

The overlying epidermis may be normal, atrophic, or hyperplastic [1,2,4,5,8]. Rarely, an epidermal collarette extends from the epidermis to surround the dermal mucin [4,10]. The epidermal rete ridges may be regularly elongated into the dermis or may be flattened [1,2,4,5,8].

The induction of follicular changes extending from the overlying epidermis into upper dermis has also been observed by several groups of investigators [10]. In one study, 11 percent (11 of 98) of solitary cutaneous focal mucinosis lesions not only had these changes but also demonstrated positive cytokeratin 20 (CK20) staining of Merkel cells within the basaloid follicular epithelial proliferations [10]. Rarely, the coexistence of cutaneous focal mucinosis and a basal cell carcinoma has been observed [11]. However, the presence of CK20 positive staining cells in the basaloid follicular epithelial proliferations of solitary cutaneous focal mucinosis permit it to be

differentiated from pathologically similar appearing aggregates of basal cell carcinoma tumor cells with surrounding mucinous stroma, which do not demonstrate CK20 staining [10].

Additional immunoperoxidase studies have also been performed on solitary cutaneous focal mucinosis specimens to characterize the spindle-shaped or stellate-shaped cells in the dermal mucin. Vimentin staining was regularly observed; its presence correlated with the number of fibroblast-like cells. Approximately 30 percent of cells stained positive for factor thirteen a (FXIIIa) and less than five percent of the cells demonstrated positive staining for CD34. The investigators interpreted these findings to represent that the dermal dendrocytes was passively incorporated into the solitary cutaneous focal mucinosis and did not represent a major cell component of the lesion. The fibroblast-like cells did not stain with antibody to alpha-smooth muscle actin (alpha-SMA), desmin, Leu 7, and S-100 [7,9].

Electron microscopy of the cutaneous focal mucinosis lesion confirms the presence of fibroblasts embedded in the fibromyxoid dermal stroma [11]. The cytoplasm of the fibroblasts contains vacuoles or secretory granules [11]. In addition to fibroblasts, electron microscopy of a cutaneous focal mucinosis lesion from patients with multiple lesions also show enlarged or peculiar macrophages in the myxomatous area with a variable number of intracellular vacuoles or phagolysosomes [16,17].

The clinical differential diagnosis of solitary cutaneous focal mucinosis is diverse (**Table 2**), [2-5,9,10]. Similar to the patient in this report, the most commonly considered diagnoses by the clinician submitting the specimen were basal cell carcinoma (31 percent) and nevus (26 percent). Other frequently entertained diagnoses included cyst (12 percent), seborrheic keratosis (8.5 percent), and fibroma (seven percent). The possibility of cutaneous focal mucinosis was never entertained by the clinicians who submitted the specimen to pathology.

Table 2. Clinical differential diagnosis of solitary cutaneous focal mucinosis.

| Diagnosis | Number | Percentage | References ^a |
|-----------------------------------|--------|------------|-------------------------|
| Basal cell carcinoma ^b | 51 | 31.0 | [9,10] |
| Nevus | 43 | 26.0 | [2,4,5,10] |
| Cyst ^c | 19 | 12.0 | [2,4,9,10] |
| Seborrheic keratosis ^d | 14 | 8.5 | [2,10] |
| Fibroma ^e | 11 | 7.0 | [3,4,9,10] |
| Skin appendage tumor ^f | 6 | 3.7 | [4] |
| Neurofibroma | 5 | 3.0 | [10] |
| Dermatofibroma ^g | 3 | 1.8 | [4,9] |
| Myxoma | 3 | 1.8 | [2,4,9] |
| Angioma ^h | 2 | 1.0 | [2,9] |
| Miscellaneous ⁱ | 7 | 4.2 | [2,4,5,9] |
| Total ^j | 163 | 100.0 | [2-5,9,10] |

^aThe differential diagnoses of the patient in the current report included basal cell carcinoma, nevus and seborrheic keratosis; he is also included in the total category.

^bThis category includes basal cell carcinoma (45) and nonmelanoma skin cancer (six).

^cThis category includes sebaceous cyst (three), follicular cyst (two) and cyst not otherwise specified (14).

^dThis category includes seborrheic keratosis (13) and keratosis not otherwise specified (1).

^eThis category includes fibroma (eight), soft fibroma (two) and acrochordon (one).

^fThis category includes eccrine poroma (two) hidroacanthoma simples (one), nodular hidradenoma (one), and skin appendage tumor not otherwise specified (two).

^gThis category includes bullous dermatofibroma (one), histiocytoma (one) and sclerofibroma (one).

^hThis category includes angioma (one) and hemangioma (one).

ⁱThis category includes tumor-not otherwise specified (three, 1.8 percent) and one of each of the following (0.6 percent each): granuloma annulare, leukemia cutis, Rosai Dorfman disease, and xanthoma.

^jThere were 163 diagnoses from 137 patients; some of the patients had at least two or three submitted clinical diagnoses.

However, a myxoma was suggested in two percent (three of 137) of the patients [2,4,9].

Several conditions characterized by mucin deposition in the dermis are in the pathologic differential diagnosis. Some of these include mucinoses that can have a focal appearance. They include lupus erythematosus (tumid variant), mucinous nevus, mucinous variation of a soft fibroma, myxoid cyst, and self-healing juvenile cutaneous mucinosis [1,4,18].

However, the main condition in the differential diagnosis is a myxoma (which is also referred to as an angiomyxoma), which is a neoplastic primary cutaneous mucinosis. When multiple myxomas are present, they may be a component of Carney complex: cutaneous myxomas, cardiac myxomas, spotty pigmentation, and endocrine overactivity [1,4,18]. A myxoma is clinically larger than solitary cutaneous focal mucinosis and the mucin is located in the deep dermis and subcutaneous fat. Increased dilated capillaries and epithelial strands within the mucinous matrix are very suggestive of myxoma. However, rarely, epithelial proliferation within the mucin can be observed in solitary cutaneous focal mucinosis [1,9]. Also, in contrast to solitary cutaneous focal mucinosis, the spindle-shaped cells in myxoma are usually smooth muscle actin positive and FXIIIa negative [1].

The pathogenesis of solitary cutaneous focal mucinosis remains to be determined. It is considered to be a benign local reactive mucinosis [4]. Although some of the individuals with solitary cutaneous focal mucinosis, including the reported patient, have been evaluated for laboratory abnormalities that can be found in persons having conditions associated with either secondary cutaneous mucinoses or with other primary (degenerative or inflammatory) mucinoses, patients with a single cutaneous focal mucinosis lesion typically do not have any mucinosis-associated systemic diseases [5]. Yet, one of the solitary cutaneous focal mucinosis patients included in Johnson and Helwig's series had a history of treatment for hypothyroidism. However, the temporal relationship between the patient's diagnosis of thyroid disease and the diagnosis of

solitary cutaneous focal mucinosis was not provided [2].

Trauma localized to the site of solitary cutaneous focal mucinosis has also been hypothesized as an etiologic factor in the development of the lesion. A 30-year-old woman developed a solitary lesion of cutaneous focal mucinosis that presented as a four-by-two centimeter erythematous to pigmented nodular plaque on her upper back at the site of blunt trauma six months earlier [6]. Two men also developed a nodular lesion of trauma-induced solitary cutaneous focal mucinosis on the right areola. These patients were a 47-year-old man with a history of folliculitis and laser epilation to the site and a 30-year-old man whose lesion appeared at the site of piercing within three months after he removed his nipple ring that had been present for three years and associated with relapsing episodes of topical antibiotic-treated infectious dermatitis [7].

The cutaneous management of patients with solitary cutaneous focal mucinosis is usually partial or complete removal. Most of the individuals have had their lesion removed at the time of their initial evaluation and biopsy. Some of the patients, whose solitary cutaneous focal mucinosis were only partially removed during the biopsy, had the residual lesion excised [2]. However, lesion recurrence was not observed in several patients in whom the dermal mucin reached the edge of the biopsy or excision specimen [2]. One patient's residual lesion reduced in size after it was injected with ten milligrams per milliliter of triamcinolone acetonide [5].

Patients with cutaneous focal mucinosis may have more than a solitary lesion. Some patients only have two or three lesions [11,19]. However, there are several individuals who have multiple lesions [12-18,20].

Schneider et al., in 1991, suggested a new classification for cutaneous focal mucinosis and defined two subtypes: (1) digital mucous or myxoid cyst and (2) extradigital cutaneous focal mucinoses [17]. The latter subtype was further divided into solitary nodular mucinosis and multiple nodular mucinosis [17]. However, their classification for cutaneous focal mucinosis was not incorporated into Rongioletti and Rebora's subsequent classification

which considers digital mucous cyst and cutaneous focal mucinosis as separate subtypes of dermal degenerative-inflammatory mucinoses [1]. Therefore, it might be reasonable to designate patients having numerous cutaneous focal mucinosis as individuals with multiple cutaneous focal mucinosis, in contrast to the patients who only have a single lesion and are now classified as individuals with solitary cutaneous focal mucinosis.

The individual lesions of a patient with multiple cutaneous focal mucinosis usually have the same clinical morphology and pathologic features as those that develop in a patient with solitary cutaneous focal mucinosis. Also, similar to solitary cutaneous focal mucinosis patients, some of the patients with multiple cutaneous focal mucinosis do not have any mucinosis-associated systemic diseases [17]. However, many of the patients with multiple cutaneous focal mucinosis have systemic conditions which might be associated with their skin condition.

Several of the patients with more than one cutaneous focal mucinosis lesion had thyroid disease (which has been associated with mucin-related myxedema), or positive titers to thyroid antibodies [11,14,15,17,19]. Also, some of the patients with multiple cutaneous focal mucinosis lesions had either received treatment with tumor necrosis factor inhibitors (which has been speculated to induced cutaneous focal mucinosis lesions) or had other systemic diseases such as Birt-Hogg-Dube syndrome with associated renal cell carcinoma, diabetes mellitus, hyperlipidemia, hypertension, lung cancer, or psoriasis vulgaris [12-16]. Additional conditions in the cutaneous focal mucinosis patients summarized by Takemura et al. included acromegaly, Cushing syndrome, dermatitis (atopic or asteototic), left ventricular myxoma, nerve sheath myxoma, psoriasis vulgaris, rheumatoid arthritis, and a history of burn [11].

The dermal mucin-associated systemic diseases include not only thyroid disease, but also papular mucinosis, scleromyxedema (which can be associated with an underlying gammopathy), and lupus erythematosus [1]. Hence, similar to individuals with solitary cutaneous focal mucinosis, some of the cutaneous focal mucinosis patients with

multiple lesions have had laboratory studies [13,14,17,18,20]. These have included some or all of the following: thyroid stimulating hormone (TSH), thyroxine (T4), triiodothyronine T3), thyroid antibodies (antimicrosomal, antiperoxidase and antithyroglobulin), serum protein electrophoresis, antinuclear antibody (ANA), double strand deoxyribonucleic acid (dsDNA) antibody, Sjogren syndrome A (SSA)/Ro antibody, Sjogren syndrome B (SSB)/La antibody, Smith antibody, ribonucleic protein (RNP) antibody, and scleroderma 70 (scl70) antibody.

Several treatment options have been attempted to resolve the skin lesions in patients with multiple cutaneous focal mucinosis. Unsuccessful topical treatments have included corticosteroid ointment and 0.1 percent tacrolimus ointment [15,18]. Indeed, resolution of the lesions is occasionally observed without any additional intervention [12,17].

Cutaneous focal mucinosis has also been observed following joint replacement. A 73-year-old man, one year following joint replacement, developed multiple, asymptomatic, bluish, two-to-three millimeters, cystic papules along the surgical scar on his left knee. The cutaneous focal mucinosis lesions were associated with a prepatellar ganglion cyst that had dissected, via multiple channels, to the skin. All the lesions resolved without recurrence at two year follow-up, after two treatments sessions, three weeks apart, in which the cystic lesions were aspirated and then injected with one percent povidocanol solution [20].

Spontaneous involution of the right leg cutaneous focal mucinosis lesions occurred in a 57-year-old man with diabetes mellitus within four months of diagnosis [16]. Another patient, a 54-year-old woman with multiple cutaneous focal mucinosis lesions on her trunk and extremities of five years' duration, whose diagnosis of goitrogenic hypothyroidism was established during the hospitalization for the resection of her right upper lobe lung adenocarcinoma, also experienced spontaneous resolution of her cutaneous focal mucinosis lesions within four months after starting L-thyroxine, which resulted in her becoming euthyroid (clinically and biochemically) after two months of

treatment [20]. Similarly, a 62-year-old patient also with adenocarcinoma of the lung had a three month history of developing flesh-colored infiltrative plaques of cutaneous focal mucinosis on the fronto-occipital scalp. Slight improvement of the existing lesions was noted and no new skin lesions developed following surgery and chemotherapy for the lung cancer [13].

Conclusion

Solitary cutaneous focal mucinosis is a variant of cutaneous focal mucinosis in which there is only a single lesion. It typically presents as an asymptomatic, less than one centimeter, flesh-colored papule on either the extremities, trunk, or head and neck of middle-aged men or women. In contrast to patients with multiple cutaneous focal mucinosis, individuals with solitary cutaneous focal

mucinosis do not have any dermal mucin-associated systemic diseases and additional laboratory studies to evaluate the patient for these conditions is therefore not necessary. The diagnosis of solitary cutaneous focal mucinosis is rarely considered prior to receiving the pathology report and the etiology of this benign condition remains to be established. If the cutaneous mucinosis has not been removed during the biopsy, the residual skin lesion can either be observed or conservatively excised.

Potential conflicts of interest

The authors (Christof P. Erickson MD and Antoanella Calame MD) declare no conflicts of interests. The author (Philip R. Cohen MD) declares the following: Dr. Cohen is a paid consultant for ParaPRO; however, this activity has no influence as a potential conflict of interest with regards to the manuscript.

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