Multiple Cutaneous Focal Mucinosis with Dermatomal Distribution

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We report a case of multiple cutaneous focal mucinosis in a 22 year-old male, who presented with multiple, asymptomatic, nodular lesions on the left upper trunk and left arm which had persisted for about one year. They were 2 to 17 mm in size, firm, yellowish, dome-shaped elevated, slightly movable papules or nodules which were distributed over the area of the left C3-7 and T1-3 sensory dermatomes. The histopathologic findings showed homogeneous mucinous material in the upper dermis. The material was confirmed to be hyaluronic acid by alcian blue stain. Some of the lesions showed improvement with intrallesional injection of triamcinolone acetonide.

Such an entity, to the best of our knowledge, has not yet been reported. (Ann Dermatol 1:46–50, 1989)

Key Words: Cutaneous focal mucinosis, Dermatomal distribution

Cutaneous focal mucinosis usually presents as a solitary, asymptomatic, white to flesh-colored papule or nodule with a predilection for the face, neck, trunk or extremities! There have been some reports of focal mucinosis as a single lesion. But multiple cutaneous focal mucinosis is so rare that we could find only three cases in the English1,2 and Korean literature.3 Herein we report a case of multiple cutaneous focal mucinosis with an unprecedented dermotomal distribution, the like of which have not yet been described.

REPORT OF A CASE

A 22-year-old soldier was first seen at our hospital in June, 1987 with multiple papulonodular lesions on the left upper trunk and left arm of 1 year's duration. The lesions developed almost simultaneously and did not show any change in size and shape with time. His general health was good. Past and family history was noncontributory.

Examination revealed asymptomatic, yellowish or normal skin-colored, dome-shaped papules or nodules distributed over the area of the left C3-7 and T1-3 sensory dermatomes (Fig. 1,2,3). They were 57 in number, ranging from 2 to 17 mm in size, firm and slightly movable on palpation. The clinical diagnosis included nevus lipomatosus cutaneous superficialis, connective tissue nevi, and xanthomas.

Laboratory examinations, including complete blood count, urinalysis, stool test, renal function tests, protein immunoelectrophoresis of serum, thyroid function tests, and T-cell count were all within normal limits. VDRL was negative. The chest X-ray film and electrocardiogram showed no abnormalities. Hepatic aminotransferases were moderately elevated initially (SGOT 66, SGPT 99), but they soon became normalized.

Two scalp biopsies were performed on the nodules of the left upper back and left forearm and revealed the same histologic findings. The hematoxylin-eosin-stained tissue sections showed a localized but not sharply circumscribed area of the dermis in which the collagen was largely replaced by homogeneous mucinous material containing scattered spindle-shaped fibroblasts (Fig. 4,5). The mucinous material stained pale blue with hematoxylin and eosin, and blue with alcian blue at pH 2.5 (Fig.
Fig. 1. Dermatomal distribution of the lesions.

Fig. 2. Multiple nodules on the left upper trunk and left upper arm.

Fig. 3. Dome-shaped nodules on the back.

Fig. 4. Loosely arranged collagen and homogeneous pale-staining matrix (H & E stain ×40).

Fig. 5. Spindle-shaped fibroblasts in myxomatous stroma (H & E stain ×400).

6) But not at pH 1.0. It was metachromatic when stained with toluidine blue, and negative when stained with periodic acid-Schiff (PAS) suggesting the presence of hyaluronic acid. Direct immunofluores-
DISCUSSION

Cutaneous mucinoses are a heterogeneous group of diseases in which accumulation of mucin (acid mucopolysaccharides) in the dermis is a prominent feature. Mucin deposition in these diseases may be a primary (metabolic) or secondary (catabolic) process.

Since Johnson and Helwig's study in 1961, the term cutaneous focal mucinosis, a focal form of primary mucinosis, has been used to describe asymptomatic, white to flesh-colored papules, nodules or plaques which are usually solitary. Sometimes it has the appearance of a cyst. It occurs on the face, neck, trunk or extremities, but never over the joints of the hands or feet. The gross appearance of focal mucinosis is not ordinarily distinctive. According to Johnson and Helwig, the clinical diagnoses in their series of 14 lesions included a cyst in two, nevus in two, tumor in three, and sebaceous cyst, myxoma, keratoses, and hemangioma in one each.

The lesions showed remarkable improvement with intralesional injection of triamcinolone acetonide 10 mg/ml weekly. At the last visit, 9 months after initiation of treatment, we noticed almost complete disappearance of the lesions (Fig. 8).
be seen in the older lesions. Sometimes there may be cleft-like spaces which are similar to those of the early stage of a myxoid cyst. The amorphous mucinous material, predominantly hyaluronic acid, is positive with alcian blue stain at pH 2.5, but not at pH 0.5, negative with PAS, and shows metachromatic staining with toluidine blue. Pretreatment with hyaluronidase eliminates positive staining with alcian blue. In our case, the above-mentioned histologic picture and staining characteristics were demonstrated in the entire dermis.

Johnson and Helwig interpreted that the presence of droplets of mucin within the cytoplasm and/or attached to cytoplasmic membranes showed fibroblasts to be the source of the hyaluronic acid. Recently, electron microscopic studies revealed that fibroblasts in the lesion contained a large amount of a medium electron-dense amorphous substance which is similar to some of the extracellular substance. We also found on electron microscopic examination that the material, both within the endoplasmic reticulum of the fibroblasts and in the extracellular space, was similar.

Differential diagnosis of cutaneous focal mucinosis from some other forms of mucinoses may be difficult. Papular mucinosis (lichen myxedematosus) appears as multiple, discrete to confluent papules occurring commonly on the face and arms without evidence of thyroid dysfunction, and usually shows a paraprotein composed of lambda-type light chains of immunoglobulin G on immunoelctrophoresis. Histopathologically, papular mucinosis differs from focal mucinosis in that it shows a large number of fibroblasts, more collagen, less mucin, and no cleft-like spaces. Myxoid cysts present as soft, smooth, dome-shaped nodules located on the dorsal surface of the distal interphalangeal joints of the fingers and, occasionally, of the toes. Histopathologically, they do not differ from focal mucinosis in the initial stage; but at a later stage, they have a greater tendency for liquefaction and cavity formation than focal mucinosis. Reticular erythematous mucinosis, self-healing juvenile cutaneous mucinosis and acral persistent papular mucinosis must also be differentiated.

Though the pathogenesis of mucinosis remains unclear, certain unknown factors, such as chronic antigenic stimulation, inflammation, trauma, viral infections, and inherited abnormalities may be responsible for stimulation of fibroblasts to produce excessive amounts of hyaluronic acid. The cause of the cutaneous focal mucinosis is likewise obscure. Johnson and Helwig described possible racial differences showing that none of the focal mucinoses occurred in Negroes. They suggested that Negroes may be less susceptible to stimulation of fibroblasts which leads to excessive production of hyaluronic acid. Cheung et al. demonstrated that skin fibroblasts from the lower extremities were more sensitive to the sera of patients with pretibial myxedema than fibroblasts from other areas in stimulation of synthesis of hyaluronic acid. They also suggested that fibroblasts from different regions of the body might possess different characteristics. This idea can be extrapolated to apply to cutaneous focal mucinosis; depending on anatomic locations, the mucin secreting capacity of the fibroblasts may be different. We think that segmental localization of the lesions in our case may be accounted for by the hypothesis that fibroblasts of the involved area are more sensitive to unknown factors.

The treatment of choice for a focal mucinosis is simple local excision, but intralesional injection of steroids may also be used. In our case, multiplicity of the lesions made local excision difficult, and so we tried weekly injections of intralesional triamcinolone. The result was a more pronounced decrease in the size of the injected lesions than the noninjected ones. But the noninjected lesions did show a small, though less marked, decrease in size. Nine months later, we experienced a dramatic response. We do not know the mechanism by which the intralesional steroid injection worked, but the three following possibilities may be thought to be responsible for the improvement: 1) spontaneous regression, 2) direct suppression of proliferation of mucin-secreting fibroblasts by steroid, or 3) absorption of local steroid into the general circulation to exert a systemic effect.

REFERENCES