

Asymptomatic nodule over the shin

A 58-year-old male presented with an asymptomatic dome-shaped shiny, slightly scaly indurated nodule over anterior aspect of left leg. Lesion was present since 1 year and gradually increased to the present size. There was no history of trauma, oozing, crusting, or similar lesions elsewhere or family history of similar lesions. The examination revealed single erythematous, indurated, scaly, nontender sessile nodule measuring 1.5×2 cm in size [Figure 1]. Rest of the cutaneous and systemic examination and blood biochemistry were within normal limits. Serum electrophoresis, thyroid

function tests, and venous doppler of bilateral lower limbs showed no abnormality. Antinuclear antibodies and human immunodeficiency virus antibodies were negative.

Hematoxylin and eosin-stained sections of excision biopsy from such a papule are shown in Figure 2, and toluidine blue stain is shown in Figure 3.

WHAT IS YOUR DIAGNOSIS?



Figure 1: Solitary dome-shaped scaly erythematous nodule over anterior aspect of left leg

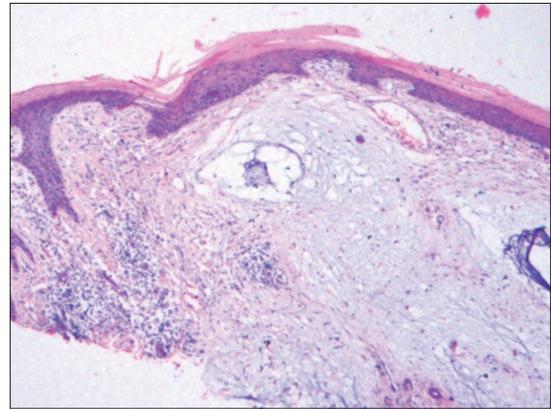


Figure 2: Acanthosis, mild hyperkeratosis, and spares perivascular lymphocytic infiltrate with diffuse dermal mucin deposition (H and E, x200)

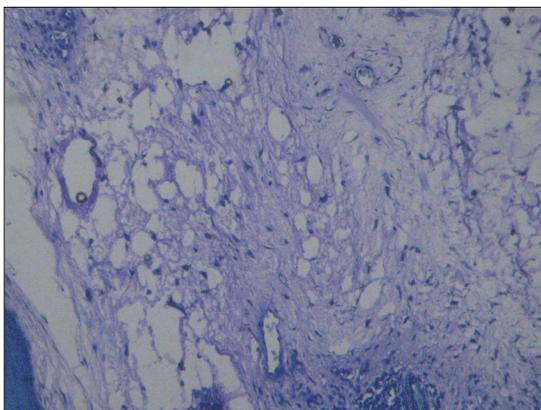


Figure 3: Diffuse dermal mucin stained violet with toluidine blue stain (Toluidine blue, x200)

How to cite this article: Gutte R, Garg G, Kharkar V, Khopkar U. Asymptomatic nodule over the shin. Indian J Dermatol Venereol Leprol 2012;78:123.

Received: April, 2011. **Accepted:** June, 2011. **Source of Support:** Nil. **Conflict of Interest:** None declared.

Answer: Cutaneous focal mucinosis (CFM)

Histological examination of excised specimen revealed mild hyperkeratosis, thinned epidermis, and diffuse mucin deposition in the dermis with bluish myxoid and stringy, and granular appearance of stroma with increased number of mast cells and without folliculotropism or increased vascularity [Figure 2]. Mild superficial perivascular lymphocytic infiltrate without pigment incontinence was also seen. Toluidine blue stain confirmed the presence of diffuse dermal mucin [Figure 3].

DISCUSSION

Cutaneous mucinosis is a heterogenous disorder characterized by abnormal deposition of mucin within the dermis. It could be primary (e.g., CFM) or secondary (e.g., associated with a neoplasm).^[1]

Mucin is an amorphous and gelatinous substance concerned with hydration of ground substance. Although exact source of mucin is not known, fibroblasts are claimed to produce acid mucopolysaccharides that stain with toluidine blue stain. Abnormal deposition of mucin either discrete or diffuse throughout the dermis causes cutaneous mucinosis.^[2]

Mucinosis can be primary, i.e., due to metabolic cause or secondary, i.e., due to catabolic cause.^[2,3] Various causes for secondary mucinosis include thyroid dysfunction, connective tissue disease, granuloma annulare, graft-vs-host disease, follicular mucinosis, Dego's disease, venous insufficiency to post ultraviolet or photochemotherapy treatment, and more recently HIV infection. None of these was present in our patient.^[2,4,5]

CFM was first described as separate entity by Johnson and Helwig in 1966 who also defined histologic criteria for diagnosis of the same.^[3] CFM typically presents usually as an asymptomatic solitary papule or nodule mostly on the face, trunk, or extremities in middle-aged adults affecting both sexes.^[1,3] Multiple lesions are seen with self-healing juvenile and adult form of cutaneous mucinosis and rarely lesions are also described in infants, i.e., cutaneous mucinosis of infancy (CFI).^[1,6]

Exact cause of CFM is obscure but it is thought to be more of reactive than neoplastic lesion, and fibroblasts are claimed to be the major source of hyaluronic acid produced in expense of connective tissue, while tissue

hypoxia has been postulated as possible stimulus for mucin secretion by fibroblast in venous insufficiency and hypoxia-induced hyaluronic acid secretion by articular chondrocytes has been documented.^[1,3,4]

Histopathology is essential for diagnosis of CFM that shows diffuse ill-defined dermal mucinous lesion with normal or hyperplastic hyperkeratotic epidermis and sparing of subcutaneous tissue. Additionally, thinned collagen fibers within mucinous stroma with absent elastic or reticulum fibers without increased vascularity are seen.^[1,3] Follicular induction of epidermis as in dermatofibroma is also seen occasionally in CFM.^[1,7] Mucin in CFM can be stained with Alcian blue at pH 2.5 or with toluidine blue stain.^[1]

CFM should be differentiated from many conditions such as papular and nodular mucinosis, digital mucus cyst, CFI, self-healing cutaneous mucinosis, nodular cutaneous lupus mucinosis and cutaneous myxoma, ganglion, and alopecia mucinosa.^[2,3,8,9]

Excision is the treatment of choice.^[2]

Differentiation from cutaneous myxoma is important which is a progressively growing neoplasm that is composed of mucinous stroma, network of fine reticulum fibers with spindle-shaped fibroblasts entrapped within lesion and increased vascularity with microhemorrhages and occasional inflammation. Multiple cutaneous lesions with a prominent vascular component are recognized feature of Carney's complex consisting of dominantly inherited complex of myxomas (cutaneous, cardiac, and mammary) with spotty pigmentation and endocrine overactivity.^[1,3]

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Access this article online	
Quick Response Code:	Website: www.ijdv.com
	DOI: 10.4103/0378-6323.90978

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