

## Skin-colored nodules in zosteriform pattern

A 33-year-old man presented with painful, skin-colored lesions of 15-year duration on the right half of upper back. He had developed similar lesions on both shoulders over the past 2 years. There were episodes of pain in the lesions, aggravated on exposure to cold, touch, and physical stress.

His general physical and systemic examinations were within normal limits. Dermatological examination

revealed multiple skin-colored erythematous, firm, tender, nodular lesions, varying in size from 2 to 5 cm, arranged in zosteriform pattern over right scapular and interscapular areas (Figure 1). Few lesions were present on the shoulders. An excision biopsy was done from the lesion on the shoulder and histopathological findings are presented in Figure 2.

**WHAT IS YOUR DIAGNOSIS?**



Figure 1: Skin-colored nodules on the right upper back

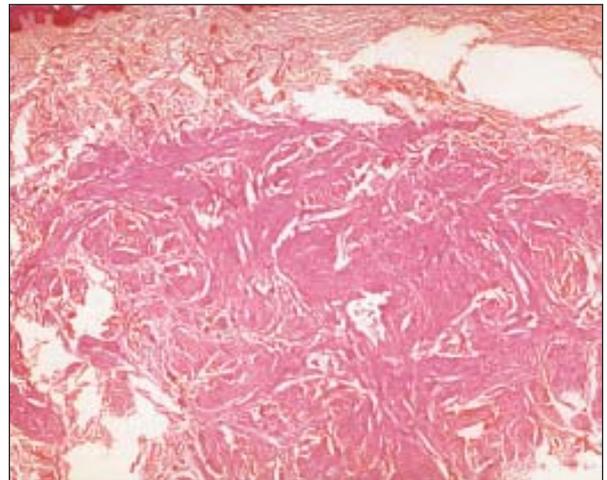


Figure 2: Photomicrograph of skin biopsy (H and E, x100)

**How to cite this article:** Gupta R, Singal A, Pandhi D. Skin colored nodules in zosteriforms pattern. *Indian J Dermatol Venereol Leprol* 2005;72:81-2.

**Received:** October, 2004. **Accepted:** March, 2005. **Source of Support:** Nil.

### Diagnosis: Type-2 segmental leiomyoma cutis

#### Histopathological findings

Biopsy showed a tumor composed of irregularly arranged smooth-muscle fibers, with elongated nuclei and rounded ends, interlaced with variable amounts of collagen in the reticular dermis. Masson's trichrome stained the smooth muscles dark red. The larger lesions were excised and the patient was started on 10 mg nifedipine thrice daily. In the following 2 months, he reported partial relief from pain.

#### DISCUSSION

Cutaneous piloleiomyomas typically present as reddish-brown or skin-colored, firm, often painful nodules, preferentially over the face, neck, trunk, or extremities.<sup>[1]</sup> Lesions on the trunk are typically multiple, also known as leiomyomatosis, and can be disseminated, zosteriform,<sup>[2]</sup> or segmental in distribution. Lesions over the extremities tend to be solitary.

In the pathogenesis of segmental leiomyomas, two theories have recently been suggested.<sup>[3]</sup> In type-1 segmental leiomyoma, heterozygosity of a postzygotic mutation leads to the segmental skin lesions comparable with non-mosaic phenotype. Type-2 segmental leiomyoma reflects postzygotic mutational event in a heterozygous embryo with subsequent loss of heterozygosity, resulting in pronounced pattern of the segmental lesions superimposed on ordinary phenotype of the underlying disease. The index patient had both discrete lesions as well as in a segmental pattern; hence, a diagnosis of type-2 segmental leiomyoma cutis was considered.

The lesions are often sensitive to touch, cold, emotional stress, or spontaneous pain. The pathogenesis of pain associated with the lesions is not clear. It may be owing to local pressure exerted by the tumor on cutaneous nerves; otherwise, the infiltrating mast cells may play some role. Others have suggested that muscle contraction may play a pivotal role in the induction of pain. The excitation of the arrector pili muscle occurs

via sympathetic nervous system. Muscle contraction ensues, triggered by the influx of calcium ions. Hence, nifedipine, a calcium channel blocker has a role in relieving pain associated with cutaneous leiomyoma.

Several conditions are associated with piloleiomyoma,<sup>[4]</sup> for example, Reed's syndrome (associated uterine myomas), erythrocytosis/polycythemia, and visceral involvement (gastrointestinal tract and retroperitoneal area). Therefore, a detailed history, examination, and investigations should be carried out in patients with multiple piloleiomyomas. The important differential diagnoses include neurofibroma, eccrine spiradenoma, dermatofibroma, and angioliipoma.

The treatment of leiomyomas is not satisfactory. In solitary, painful lesions, surgical excision is the therapy of choice. In multiple, painful lesions, chances of recurrence (50%) make surgery impractical. Various systemic therapies have been tried<sup>[5]</sup> with variable results, such as calcium channel blockers (nifedipine),  $\alpha$ -adrenoreceptors blockers (phenoxybenzamine), nitrates, analgesics, antidepressants, and gabapentine. Liquid-nitrogen cryotherapy and CO<sub>2</sub>-laser ablation have also been used with good results.

**Ritika Gupta, Archana Singal,  
Deepika Pandhi**

Department of Dermatology and STD, University College of Medical Sciences and Guru Teg Bahadur Hospital, Shahdara, Delhi, India.

Address for correspondence: Dr. Archana Singal, B-14 Law Apartments, Karkardooma, Delhi-110 092, India.  
E-mail: archanasingal@hotmail.com

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