SkIndia Quiz 19

Soft tumors in segmental fashion

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A 25-year-old male presented with multiple skin-colored to hyperpigmented, soft to firm raised lesions of varying sizes on the right side of the chest, back, and right upper extremity since six years. Lesions were tender to touch and had gradually increased in number and size to the present status. Family and personal history were noncontributory. Examination revealed multiple grouped, soft to firm, tender, skin-colored papules, and nodules ranging from 2 mm to 10 mm distributed on the right side of the chest extending from midline to anterior axillary line (T2-T6 dermatome) [Figure 1] and on the extensor aspect of the right upper extremity (C5-6 dermatome). [Figure 2]. Button-hole sign was negative and cold sensitivity was absent. Systemic clinical examination, urine microscopy, abdominal and pelvic sonography was normal. Histopathology showed a poorly demarcated tumor in the dermis around the hair follicle composed of interlacing bundles of spindle-shaped cells with elongated rounded nuclei and moderate cytoplasm [Figure 3].

WHAT IS THE DIAGNOSIS?

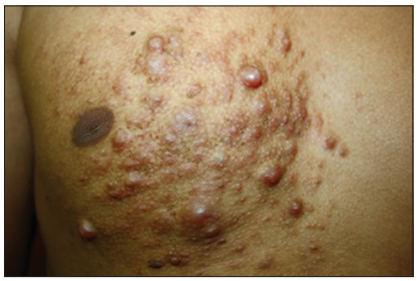

Figure 1: Cluster of skin-colored papules and nodules ranging from 2 mm to 1 cm, grouped on the right side of the chest in T2-T6 dermatome

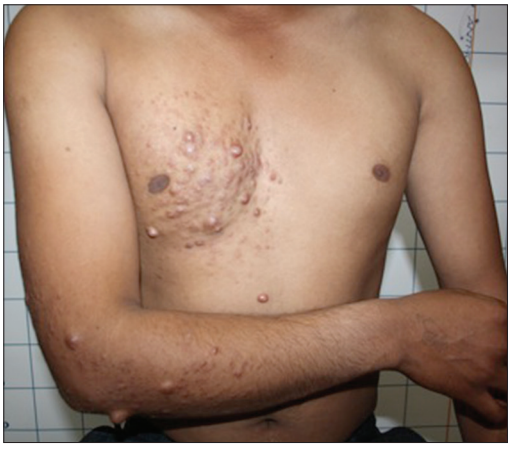

Figure 2: Similar multiple papules and nodules present on the extensor aspect of the right upper extremity, ranging from few millimeters to 2 cm

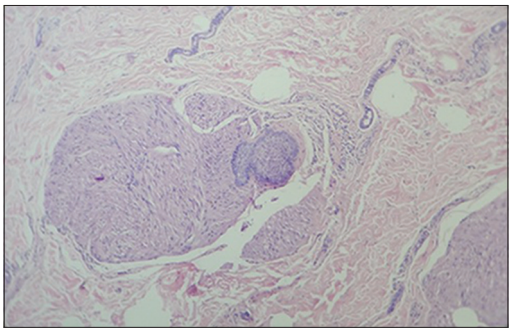

Figure 3: A poorly demarcated tumor in the dermis around the hair follicle composed of interlacing bundles of spindle-shaped cells with elongated blunt nuclei and moderate cytoplasm suggestive of smooth muscle. (H and E, x40)

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ANSWER

Segmental piloleiomyoma

DISCUSSION

Leiomyoma is a benign tumor of smooth muscles. Cutaneous leiomyoma comprises 5% of all leiomyomas and has no sex predilection.[1] These lesions are classified as:

Pilar leiomyoma (leiomyoma cutis)

Tumor arising from pilomotor muscle of skin. It is the most common among the three types. The age of presentation is early adulthood, that is, 10-30 years.

Genital leiomyoma (dartoic myoma)

Arises from the smooth muscle of the genitalia and areola of the nipple. It can occur at any age.

Angioleiomyoma

Arises from the tunica media of the arteries and veins, and is seen mainly in middle age or later.

This classification reflects the origin of the smooth muscle tumor and corresponds to the histologic and anatomic site where they are found.[2]

Pilar leiomyoma generally presents as a collection of pink, red or dusky brown, firm dermal nodules of varying size. The clinical differential diagnosis to be considered includes segmental neurofibromatosis. However, the nodules of leiomyomas are often painful. The pain can be provoked by touching or chilling the skin, or by emotional disturbance. The characteristic distribution of leiomyomas is in a group; but they can occur as linear, segmental, and zosteriform variants. A total of 80% of the lesions are multiple-the condition is termed ‘‘leiomyomatosis.’’ The occurrence of large number of leiomyomas is termed as ‘’Myomatosis Cutis Miliaris.”[1,9]

Recent literature mentions that the leiomyomas can be hereditary and the mode of inheritance is autosomal dominant, with incomplete penetrance and variable expressivity.

These dominantly inherited leiomyomas are distributed in a segmental fashion. The exact etiology is not known but the proposed hypothesis is a mutation in fumarate hydratase gene located on 1q42.3-43. Two types have been mentioned. In type 1, heterozygosity of a postzygotic mutation leads to segmental skin lesions similar to that in nonmosaic phenotype. Type 2 reflects a postzygotic mutational event with subsequent loss of heterozygosity resulting in a pronounced pattern of segmental lesions. Segmental distribution is more often seen in type 2 variant.[3-5]

Multiple cutaneous leiomyomas with inherited predisposition are linked to uterine leiomyomas and an increased incidence of renal cell carcinoma, mostly type 2 papillary. These entities have been described as multiple cutaneous and uterine leiomyomatosis (MCUL) and hereditary leiomyomatosis and renal cell cancer (HLRCC).[6] A broad range of other benign and malignant tumors has also been observed in MCUL/HLRCC families. The anecdotal reports include breast, prostate, bladder, testis, ovarian and kidney cysts, cerebral, cavernous, adrenal gland adenomas.[1-4,10]

Treatment

It consists of surgical excision of solitary lesions but in case of multiple lesions, it is elusive; recurrence is common. For this, medical management is preferred which consists of use of calcium channel blockers, phenoxybenzamine, doxazocine, gabapentin, topical 9% hyoscine hydrobromide.[8] All these medications reduce the pain and provide symptomatic relief. There are two proposed hypotheses regarding the mechanism of pain in leiomyomas. One is compression of nerves in the tumor and the other is abnormal muscle contraction leading to pain. Still, the treatment of leiomyomas is unsatisfactory.[1,7]

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