Introduction

Multiple nodular lesions on the skin of human beings create a sense of confusion to the examining dermatologist. He has to rule out many documented nodular skin lesions in an orderly manner before arriving at a final clinically acceptable diagnosis. Even then, he keeps two or more diagnosis for exclusion by the available biochemical and histopathological aids. He will not know till the end what surprise is awaiting him, until when the histopathology shows an entirely different scenario than what he thought of.

One among such nodular skin lesions on the human skin, which many dermatologists will never think of while diagnosing clinically is “Cutaneous Leiomyomas” (CL).

Case Report

A 30 yrs old married woman, came to the dermatology department with skin colored nodules on her right upper back near the shoulder and over the right forearm (Fig. 1-3). They were slowly evolving, one by one adjoining each other for nearly 3 months. The nodules were skin colored, soft, dome shaped, discrete and occasionally painful. There were seven nodules on the right shoulder and multiple over the right forearm. No signs of inflammation or ulceration were seen around the nodules. The nodules over the forearm were slightly hyperpigmented and also discrete. The skin over the nodules was not pinchable, not attached to the underlying structures. Elsewhere her skin, hairs, nails and mucous membranes were normal.

The nodules were skin colored, soft, dome shaped, discrete and occasionally painful. There were seven nodules on the right shoulder and multiple over the right forearm. No signs of inflammation or ulceration were seen around the nodules. The nodules over the forearm were slightly hyperpigmented and also discrete. The skin over the nodules was not pinchable, not attached to the underlying structures. Elsewhere her skin, hairs, nails and mucous membranes were normal. Gynaecological examination was normal.

Clinically the following diagnoses were considered

1. Neurofibroma
2. Neurilemmoma
3. Cutaneous mastocytoma
4. Xanthomas
5. Myxomas

Biological studies were normal. Biopsy of the nodules showed the following features [1].
1. Bundles of smooth muscle interlacing in the dermis (Fig. 4.5).
2. Muscle bundles were straight.
3. Centrally located, thin, very long, blunt edged, “eel-like” nuclei (Fig. 4.6).
4. Intermingling of varying amounts of collagen.
5. No nuclear hyperchromasia, pleomorphism or mitosis. The nodules on the right shoulder were excised and the outcome was excellent. After six months there were no signs of any recurrence on the excised site or elsewhere.

Figure 1. Nodules on right shoulder and back

Figure 2. Nodules on right shoulder and back (closeup view)

Figure 3. Multiple, slightly hyperpigmented nodules – Right forearm

Figure 4. Picture of eel

Figure 5. Histopathology of the nodule showing bundles of smooth muscle interlacing in the dermis

Figure 6. Close up view of Figure 4

1) Eels are long snake like scaleless marine or fresh water fishes.
2) They migrate from fresh water to salt water to spawn.
3) They lack pelvic fins.

Box 1.
Discussion

Cutaneous leiomyomas (CL) are benign tumors originating from the erector pili muscles. They can develop wherever smooth muscle is present. They have equal distribution in both sexes. It has a benign clinical course, and most often presents as multiple cutaneous lesions. Nevertheless, some cases have been described in which single cutaneous leiomyoma appear and even cases in which they appear in families [2-4]. There are reports that associate cutaneous leiomyomatosis to tumors located in other organs, specifically in the uterus and kidneys [5-7]. Pleomorphic adenomas (PA) of the parotid are the most frequently found benign tumors of the major salivary glands [8] and their simultaneous appearance with CL has been reported. First described by Virchow in 1854 [9]. Cutaneous Leiomyomas (CL), are usually not considered while examining any skin coloured nodule with minimal pain, which is sometimes ignored by the patient and missed by the dermatologist. Cutaneous leiomyoma has received little attention in the recent literature. The histopathological features of Cutaneous Leiomyoma is typical and striking to the eyes which are trained to look for it.


Three major types of cutaneous leiomyomas exist:
1. Piloleiomyomas, are believed to arise from the erector pili muscle.
2. Angioleiomyomas, originate from smooth muscle (tunica media) within the walls of arteries and veins.
3. Genital leiomyomas.

- They correspond to the histological or anatomic site.
- Menses or pregnancy, temperature and pressure are supposed to be trigger factors for pain [12,13].
- CL are benign tumors that can be exquisitely painful [12,14-16].
- Pathogenesis of pain associated with these lesions is still a mystery.
- The histological findings do not show that prominent nerve fibers are associated with these tumors.
- Others have theorized that specific infiltrating cells may play a role.
- Yet others have suggested that muscle contraction may be pivotal in the induction of pain.
- Genital leiomyomas tend to be the least common of the 3 types.
- Cutaneous leiomyomas with histopathologic features of uterine symplastic leiomyoma (USL) have also been reported [17,18]. Symplastic leiomyoma is an atypical uterine leiomyoma with cytologic atypia [19].
- Associated morbidity may be due to spontaneous lesional pain, as well as pain evoked by cold and/or tactile hypersensitivity. Additionally, multiple piloleiomyomas have the potential to be cosmetically disfiguring.
- A racial predilection had not been reported.
- The incidence of piloleiomyomas in men and women appear to be equal.

Recent Research:
- The location of the gene for transmission of dominantly inherited, multiple cutaneous piloleiomyomas associated with uterine leiomyomas in female family members [5].
- As reported by Alam et al., the locus is named MCUL1 (Multiple Cutaneous and Uterine Leiomyomata) [5,26].

**Complications:**
1. Erythrocytosis associated with skin leiomyomas [8,25].
2. Pain.

**Distribution patterns:**
- Bilaterally symmetric,
- Grouped,
- Dermatomal,
- Linear patterns,
- Piloleiomyomas develop in the superficial dermis, therefore it is fixed to the skin.

**Problem in contemporary pathology is:**
- The classification and distinction of spindle cell soft tissue tumours of skin.
- Markers such as alpha smooth muscle actin (alpha-SMA) and desmin, considered specific for smooth muscle cell (SMC), have been shown to be expressed in variety of fibroblastic and myofibroblastic processes.

**Conclusion**
1. Cutaneous Leiomyomas are rare, most of the time not thought of by dermatologists till the histopathology confirms it. Problem in contemporary pathology is the classification and distinction of spindle cell soft tissue tumors of the skin.
2. Markers such as alpha-smooth muscle actin (alpha-SMA) and desmin, considered specific for smooth muscle cell (SMC), have been shown to be expressed in a variety of fibroblastic and myofibroblastic processes. High-molecular-weight caldesmon (h-caldesmon), one of two isoforms, is reported to be expressed exclusively by SMC and shown to be a specific marker of SMC tumors.
3. h-caldesmon is a specific marker of fully differentiated smooth muscle and that it can serve to differentiate spindled SMC soft tissue tumors of the skin from tumors of myofibroblastic and/or fibroblastic origin.
1. Characteristic round, thin-walled vessels and the mixed nature of the tumor cells.
2. There is no cytologic atypia or mitotic activity.
3. Thin spindle cells associated with thin, wavy collagen bundles.
4. Loosely spaced in clear or mucinous matrix.

1. Small groups of fibrils surrounded by rows of palisaded nuclei.
2. Nuclei in two parallel rows enclosing between them a space nearly homogenous anucleate material.

Table I. Neurofibroma vs Neuromlemmma (HP showing Verocay body)

REFERENCES