A 27-Year-Old Man with Multiple, Skin-Colored Nodules in a Segmental Distribution on the Right Side of the Face

A 27-year-old male patient presented with a 4-year history of multiple reddish brown to skin-colored, raised lesions on the right cheek [Figure 1a]. He did not give any history of itching, burning sensation, or pain associated with the lesions. His personal and medical history was unremarkable. There was no family history of any neoplasm. Physical examination showed four to five skin-colored to erythematous papules and nodules of size ranging approx. from 2 mm × 2 mm to 4 mm × 5 mm present on the right cheek in segmental distribution with a smooth surface and no associated secondary changes [Figure 1b]. Lesions were firm in consistency and nontender on palpation. A 3-mm punch biopsy was taken.

Biopsy showed non-epithelial neoplasm involving the reticular dermis with features of differentiation toward smooth muscles [Figure 2a]. The neoplastic cells were arranged in small and long fascicles that resembled normal arrector pili muscle [Figure 2b]. The neoplastic cells showed abundant pink, vacuolated cytoplasm and elongated, wavy, blunt-ended nuclei [Figure 2c and d]. Surrounding dermis showed dense collagen. The special stain Masson's trichrome (MTS) confirmed the positivity for smooth muscle fibers [Figure 3a]. Immunohistochemistry study revealed positivity of α-smooth muscle actin (α-SMA) [Figure 3b].

What is Your Diagnosis?

Multiple pilar leiomyomas.

Discussion

Leiomyoma is a benign tumor of smooth muscle origin, most commonly found in uterus, gastrointestinal tract, skin, and subcutaneous tissue.[1] Less than 1% of all leiomyomas are found in the head and neck.[2] Malhotra et al.[3] described a clinicopathological series of 37 cases of leiomysa cutis, in which four were present in head and neck (one solitary and three multiple), including a case of leiomyoma on the face of a young male with aceneiform distribution. Leiomyoma of the skin is divided into three main subtypes based on the origin of the smooth muscle: pilar leiomyoma originates from the arrector pili muscle, angioleiomyoma originates from the tunica media of superficial blood vessel, and genital leiomyomas are derived from the darts, vulva, or areolar smooth muscle associated with genitals or breasts.[4,5]

Pilar leiomyoma is the most common type of cutaneous leiomyoma. It may be solitary or multiple; when multiple, leiomyomas may develop in a grouped, linear, or segmental distribution.[4,5] These tumors are usually fixed to the undersurface of the skin, but are movable over underlying structures, and the overlying skin often acquires a reddish-brown discoloration. Leiomyomas of the skin and subcutaneous tissues may be innervated and produce paroxysms of pain, but those occurring in the head regions are usually not painful.

Multiple cutaneous leiomyomas may be associated with uterine leiomyomas, also known as multiple cutaneous and uterine leiomymatosis (MCUL), familial leiomymatosis cutis et uteri, Reed syndrome, or multiple leiomymatosis. They may also be associated with renal cell carcinoma. Therefore, patients with multiple cutaneous leiomyomas should be enquired about the presence of personal and family history of uterine fibroids and renal cell carcinoma, and if found positive, should be...
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adequately managed and followed up. Patient was advised surgical excision, and no recurrence was observed during the 3 months follow-up.

Because of nature of skin findings and their rarity, leiomyomas are usually misdiagnosed clinically and their presence is identified only after histopathologic examination. In a study of 53 lesions, only three were accurately diagnosed prior to biopsy.\[4\] Staining with α-SMA, MTS, and desmin can be used to confirm the presence of smooth muscle. The differential diagnosis includes dermatofibroma, histiocytoma, angiolipoma, neurofibroma, adnexal tumors, and smooth muscle hamartoma. The treatment is complete surgical excision, and recurrence is extremely rare if the excision is complete.\[1\]

Our patient had taken treatment in the form of topical steroids, antibiotics, and topical and oral ayurvedic medications on and off without any improvement. The lesions were asymptomatic without any pain or tenderness, or any change in size with itching or cold exposure. This unusual presentation accompanied with rarity of this disease on the cheek makes the clinical diagnosis difficult, and it can be easily missed if histopathologic examination is omitted. Although extremely rare in head and neck, leiomyomas should be included in the differential diagnosis of well-circumscribed and encapsulated lesions.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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