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CASE REPORT

# "ASYMPTOMATIC CUTANEOUS PILAR LEIOMYOMA: A RARE AND DISTINCT ENTITY"

**Priyanka Anand[1], Namrata Sarin[2] and V.K Khurana[3]**

[1,2]Department, Pathology department, Hindurao hospital and NDMC Medical College, New Delhi, India

[3]Department, Dermatology department, Hindurao hospital and NDMC Medical College, New Delhi, India

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**ABSTRACT**

Leiomyoma is a rare benign tumor that occurs in solitary or multifocal forms. Most common location is uterus. Skin is the second commonest site comprising of about 3-5% of all leiomyomas. Cutaneous leiomyomas arise from smooth muscles in the skin; from arrector pilar muscles associated with the hair follicles (pilar leiomyoma), media in the blood vessel (angioleiomyoma) and smooth muscle in the scrotum or labia (genital leiomyoma). Piloleiomyoma is the most common type of cutaneous leiomyoma Clinically they mimic other cutaneous lesions and pose a diagnostic challenge. Therefore, histopathological examination is mandatory to differentiate them from spindle cell lesions and is the gold standard for an accurate diagnosis. Herein, we report a case of cutaneous pilar leiomyoma in a 50 year old female who presented with multiple papules over shoulder and back.



## INTRODUCTION

Leiomyoma is a rare benign tumor that occurs in solitary or multifocal patterns. They are benign soft tissue tumors that arise from smooth muscle. The most common location is uterus (about 95% of all leiomyomas). Skin is the second commonest site comprising of about 3-5% of all leiomyomas.[1] Cutaneous leiomyomas arise from smooth muscles in the skin; from arrector pilar muscles associated with the hair follicles (pilar leiomyoma), media in the blood vessel (angioleiomyoma) and smooth muscle in the scrotum or labia (genital leiomyoma). Cutaneous leiomyomas account for about 75% of extra-uterine leiomyomas (2-4%). Piloleiomyoma is the most common type of cutaneous leiomyoma.[2] It usually originate as reddish- brown papules <15mm in diameter. Clinically, they mimic other cutaneous lesions and pose a diagnostic challenge. Therefore, histopathological examination is the gold standard for a precise diagnosis. Herein, we present a case of cutaneous pilar leiomyoma in a 50 year old female.

### *Case presentation*

A 50 year old female presented with multiple skin colored to yellowish colored papules over bilateral shoulder and bilateral flank, these papules were present since six years. These lesions were asymptomatic; was not associated with pain and pruritus. There was no history of any trauma. There was a history of uterine fibroid and renal complicated cyst and the patient got operated for this two years back. Family history was insignificant. On local examination, a single 2-2.5 cm, reddish brown colored papule on both shoulders and flank region was noted. A punch biopsy was performed from the lesion and tissue was sent for histopathological examination. We received a single skin covered soft tissue bit measuring 0.6x0.5x0.3cm. On microscopic examination, multiple serial sections showed mild atrophy of the epidermis. A clear grenz zone was evident. In the dermis a well circumscribed, unencapsulated tumor composed of interlacing fascicles and whorled bundles of smooth muscle cells was seen. These cells were spindle shaped with indistinct cell borders, nucleus was elongated, cigar shaped, with blunt ends, finely dispersed chromatin and abundant fibrillary eosinophilic cytoplasm. No mitoses or cellular atypia was noted. Smooth muscle cells were positive for alpha- smooth muscle actin and negative for S100 on immunohistochemistry. A diagnosis of cutaneous leiomyoma, piloleiomyoma was rendered. Patient is currently under follow up period of six months.

## DISCUSSION

Leiomyoma arises from smooth muscles. It is classified into various subtypes based on origin of the smooth muscle. It can arise from arrector pili muscle of the pilosebaceous unit (piloleiomyoma), tunica media within the walls of arteries and veins (angioleiomyoma) and from the dartos muscle of the

scrotum, labia majora, erectile muscle of nipple (genital leiomyoma)[3] and leiomyoma with mesenchymal elements. Cutaneous leiomyoma comprise approximately 3-5% of all leiomyomas,[4] is a rare benign tumor of skin. Piloleiomyoma is the most common variant of cutaneous leiomyoma.[5] It occurs over the face, neck, trunk and extremities. They are more common in adults than in children. It can present in a solitary or multiple form. Multiple piloleiomyoma is 1-2 times more frequent than solitary pattern. It emerges as small, firm, reddish brown intradermal nodules arranged in a group or linear arrangement most commonly involving the extremities and trunk but may affect face and neck and rarely mouth.[6] Multiple cutaneous leiomyomas can have an association with uterine leiomyoma, also known as multiple cutaneous and uterine leiomyomatosis (MCUL), familial leiomyomatosis cutis et uteri, reed syndrome or multiple leiomyomatosis.[7] A subset of these families has a predilection to develop renal cell carcinoma. The gene localized to chromosome 1q42.3-43 and that encoding fumarate hydratase has been proposed to be a predisposition gene for reeds syndrome.

## Figures

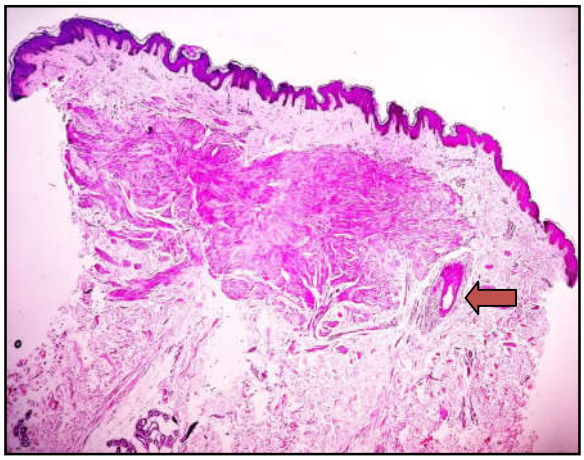

**Fig 1** Multiple serial sections showed a well encapsulated tumor composed of proliferation of spindle cells in the upper dermis. Adjacent to the lesion, a hair follicle is seen (red arrow). (H&E,4x).

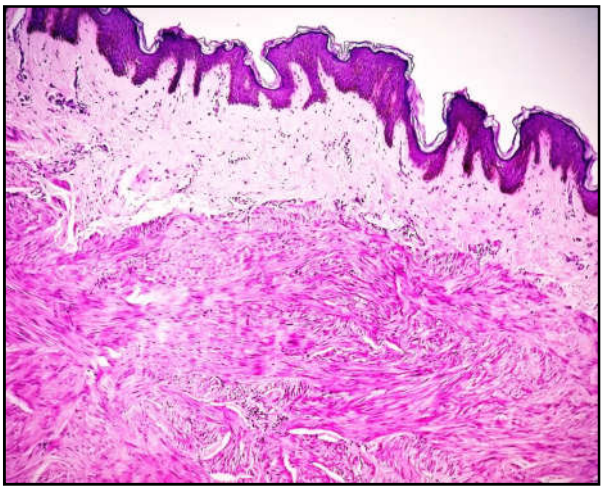

**Fig 2** Microphotograph showing interlacing spindle shaped cells arranged in whorls and fascicles. (H&E, 10x).

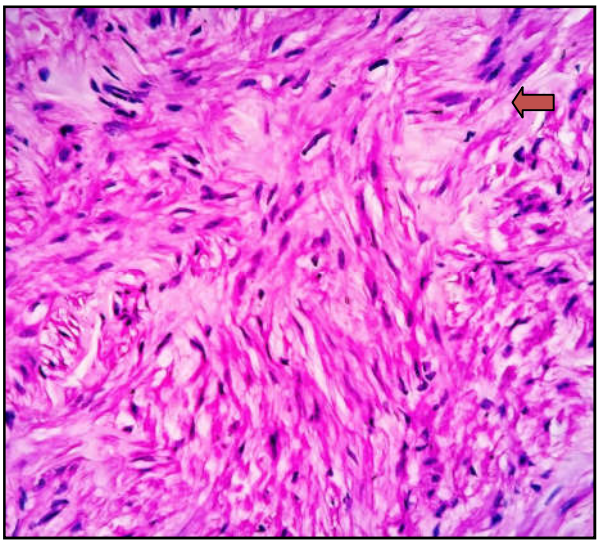

**Fig 3** Microphotograph show spindle cells having blunt-ended cigar shaped nuclei, vesicular chromatin and abundant eosinophilic cytoplasm as shown by an arrow. No cytological atypia was observed. (H&E, 40x).

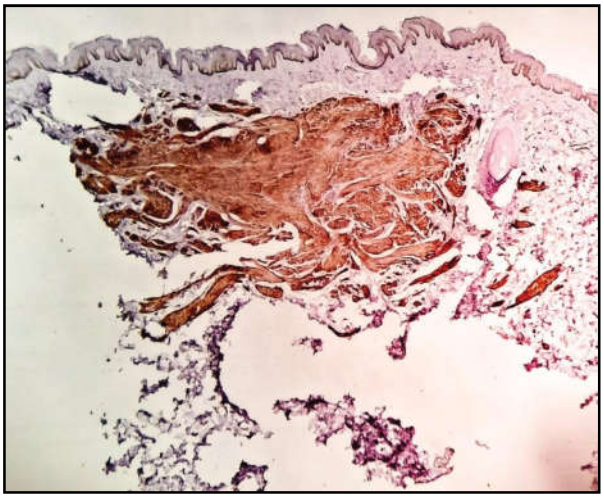

**Fig 4** Microphotograph shows Alpha- smooth muscle actin positivity thereby confirming the diagnosis of cutaneous leiomyoma. (Immunostain alpha-SMA, 4x).

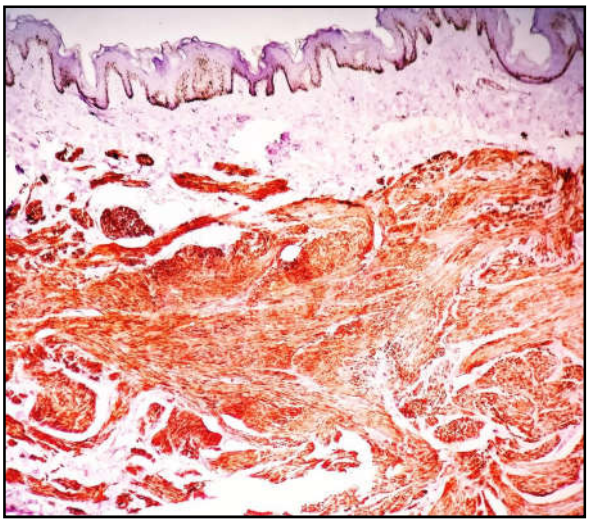

**Fig 5** Immunohistochemical stain highlighting the tumor. (Alpha-SMA immunostain, 10x)

The lesions gradually increase in size and new lesions form.[8,9] The characteristic symptom is pain that is elicited upon light touch. Patient generally shows hypersensitivity to light touch

and cold temperature that is spontaneous or induced by trauma. The reason behind its occurrence is due to localised compression of the tumor itself or by contraction of the muscle fibres of the tumor[10,11] or by infiltration of tumor mast cells. Solitary leiomyomas are infrequently painful in contrast to multiple papules with no symptoms of associated pain. It is believed that the painful sensation is usually an itching sensation with evidence of central sensitization.[12] It can be misdiagnosed as keloid because of the itching sensation. Cutaneous leiomyomas do not undergo malignant transformation.[13] The etiology and pathogenesis is still unknown. Apoptotic and antiapoptotic factors may have a role. An increase in antiapoptotic BCL-X with a decrease in proapoptotic BCL-2 was reported by Wortham *et al*.[14] Clinically, they are often wrongly diagnosed as lipomas, glomus tumor, eccrine spiradenoma, neurofibroma, sebaceous cysts and fibroepithelial polyps.

Therefore histopathological examination is significant in making an accurate diagnosis. The arrector pili muscle, from which piloleiomyoma arises, attaches proximally to the hair follicle and to multiple attachment sites within the papillary and reticular dermis distally as well as to the basement membrane. Histologically, tumor consists of interlacing bundles of smooth muscle cells that stains red with masson trichome and on immunohistochemistry, is positive for smooth muscle actin. Electron microscopic examination has demonstrated that piloleiomyomas consists of normal appearing smooth muscle cells having a central nucleus surrounded by an area composed of endoplasmic reticulum and mitochondria and in the periphery, numerous myofilaments arranged in bundles are present. Dense bodies, both marginal and cytoplasmic are also seen.[13]

## CONCLUSION

Cutaneous leiomyomas are rare benign tumors. It is one of the differentials of papulonodular skin lesions and is frequently misdiagnosed thus performing a biopsy for the confirmation of diagnosis by histopathological examination is mandatory to differentiate them from spindle cell lesions like dermatofibroma, neurofibroma and myofibroblastic lesions like nodular fasciitis, fibromyoma and smooth muscle hamartoma. Complete excision of the lesion is the only definitive treatment.

### *Declarations*

Consent for publication- Written informed consent was obtained from the patient for publication of this case report and accompanying images.
Competing interests- None.
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